

JAO CR

Official Journal of the American Osteopathic College of Radiology

PEDIATRIC INTERVENTIONAL RADIOLOGY



Guest Editor: Tammam Beydoun, D.O.

Editor-in-Chief: Daniel J. Wale, D.O.

July 2021, Vol. 10, Issue 3

JAOCR

Official Journal of the American Osteopathic College of Radiology

Aims and Scope

The Journal of the American Osteopathic College of Radiology (JAOCR) is designed to provide practical up-to-date reviews of critical topics in radiology for practicing radiologists and radiology trainees. Each quarterly issue covers a particular radiology subspecialty and is composed of high-quality review articles and case reports that highlight differential diagnoses and important teaching points.

Access to Articles

All articles published in the JAOCR are open access online. Subscriptions to the journal are not required to view or download articles. Reprints are not available.

Copyrights

Materials published in the JAOCR are protected by copyright. No part of this publication may be reproduced without written permission from the AOCR.

Guide for Authors

Submissions for the JAOCR are by invitation only. If you were invited to submit an article and have questions regarding the content or format, please contact the appropriate Guest Editor for that particular issue. Although contributions are invited, they are subject to peer review and final acceptance.

Editor-in-Chief

Daniel J. Wale, D.O., Ann Arbor, MI

Editor Emeritus

William T. O'Brien, Sr., D.O., Cincinnati, OH

Editorial Board

Abdominal/Body Radiology

Sharon A. Kreuer, D.O., Monroeville, PA

Rocky C. Saenz, D.O., F.A.O.C.R., Farmington Hills, MI

Breast Radiology

Stacy J. Ries, D.O., Detroit, MI

Matthew Tommack, D.O., Eugene, OR

Michelle C. Walters, D.O., Dallas, TX

Chest and Cardiac Radiology

Mark Guelfguat, D.O., Bronx, NY

Douglas Johnson, D.O., Charlotte, NC

Musculoskeletal Radiology

Christopher Cerniglia, D.O., M.Eng., Westborough, MA

Matthew Tommack, D.O., Eugene, OR

Donald von Borstel, D.O., Tulsa, OK

Neuroradiology

Robert Koenigsberg, D.O., F.A.O.C.R., Philadelphia, PA

Alysha Vartevan, D.O., Scottsdale, AZ

Nuclear Medicine

Timothy McKnight, D.O., Farmington Hills, MI

Daniel J. Wale, D.O., Ann Arbor, MI

Pediatric Radiology

Brooke S. Lampl, D.O., Cleveland, OH

Emily Janitz, D.O., Akron, OH

Vascular and Interventional Radiology

Aaron T. Rucks, D.O., M.S., Erie, PA



JAOCR

PEDIATRIC INTERVENTIONAL RADIOLOGY

Guest Editor: Tammam Beydoun, D.O.

From the Editor

In this Issue 4

Tammam Beydoun, D.O.

Review Articles

Vascular Anomalies: Basics of Imaging and Treatment.....5

Tammam Beydoun, D.O., Azam Eghbal, M.D., William N. Holmes, M.D.

Review of Image-Defined Risk Factors in Neuroblastoma 15

Andrew Sung, M.D., Elyssa M. Rubin, M.D., Hollie Lai, M.D.

Differential-Based Case Reviews

Abdominal Mass in a Female Pediatric Patient 21

Tuan Dao, M.D., Hollie Lai, M.D.

Patellar Bone Lesion 24

Danny Nahl, D.O., Andrew Sung, M.D.

JAOCR at the Viewbox

Congenital Extrahepatic Portosystemic Shunt (Abernathy Malformation) 28

Tammam Beydoun, D.O., Daniel Nahl, D.O.

Parkes Weber Syndrome.....30

Tammam Beydoun, D.O., Tuan Dao, D.O.



"The only way to make sense out of change is to plunge into it, move with it, and join the dance."
—Alan Watts

In this Issue

Tammam Beydoun, D.O.

Director, Pediatric Interventional Radiology, Department of Radiology,
Children's Hospital of Orange County, Orange, CA

In July 2012, William E. Shiels, D.O., served as guest editor to the JAOCR in its first year. That same year, I attended his lectures at an AOCC conference and discovered that I too wanted to serve as a pediatric interventional radiologist. As a small token of gratitude to his legacy, I dedicate this issue to his memory.

I would like to thank Editor-in-Chief Dr. Wale for the opportunity to serve as guest editor for this issue of JAOCC and for his commitment to moving this journal forward. I would also like to recognize the heavy lift that Editor Emeritus Dr. O'Brien made to get the journal off the ground in the early years. Thanks also goes to the editorial board and AOCC staff, who support this outlet for our college and colleagues.

In this issue, we visit various topics in pediatric radiology and hopefully leave insights for our radiology colleagues to reference in their encounters with the pediatric population. The first review article covers vascular malformations in an effort to recondition the ubiquitous "hemangioma." Keeping in mind that our reports often trigger reactionary responses from our clinical colleagues, our "impression" should add value and support clinicians in making the best decision for *our* patient.

In our second review article, Drs. Sung and Lai describe neuroblastoma and provide a detailed approach to staging this common

pediatric malignancy. This review serves as a reminder that our knowledge of pathology and, therefore, approach to treating patients, is ever-changing. What was, may no longer be, and it should be our goal to give families the most current care possible.

Imaging is one of many tools to deliver a diagnosis, and often narrowing the differential is the best we can do. With that in mind, we present short, high-yield, case-based reviews of an abdominal mass in the female pediatric abdomen by Dr. Dao, et al, and a patellar bone lesion by Dr. Nahl, et al. Finally, we round out the issue with two rare cases of portosystemic shunt and Parkes Weber syndrome, for which an early imaging diagnosis can change the course of patient care and outcome.

I finish with the same sentiment Dr. Shiels left us with nine years ago: We hope these small examples of pediatric radiology encourage you toward the never-ending pursuit of knowledge and to always keep focus on the patient behind the image.

Vascular Anomalies: Basics of Imaging and Treatment

Tammam Beydoun D.O., Azam Eghbal M.D., William N. Holmes M.D.

Departments of Diagnostic and Interventional Radiology – Children’s Hospital of Orange County

Vascular anomalies is a broad term used to encompass congenital errors in vascular development. This comprises a large group of separate entities including all vessel types, locales, and sizes, as well as syndromic presentations. In the day-to-day setting, these lesions can be frustrating for patients and their families, adversely affecting quality of life.

The clinical presentation of vascular anomalies is varied and may be particularly troublesome, requiring unique considerations in management. These can include life-threatening risks such as airway compromise, congestive heart failure and severe bleeding.¹⁻⁴ There are also functional risks involving the eyes, lips, ears and nose, which can affect vision, speech, feeding and hygiene.^{5,6} Although the majority of vascular anomalies are benign, some can present with sequelae of ulceration or leaking, leading to chronic infections.

As a group, these lesions carry a varied amount of morbidity, but just as important is their psychosocial effect on patients. All in all, swift recognition and appropriate multidisciplinary management become paramount in treating this group of diseases.

Old Terminology

The initial description of a vascular anomaly, namely the arteriovenous

malformation, is credited to Dr. William Hunter who described the “particular species of aneurysm” in 1761.⁷ Since then, nonspecific and nondescript terms applied to vascular anomalies have grown, unfortunately without concrete pathologic or imaging characteristics. An example of these antiquated and misleading terms is listed in **Table 1**. Standardizing classification and characterization of these lesions was needed to facilitate proper diagnosis and subsequent treatment.

The seminal paper introducing a method to classify vascular anomalies based on histopathological and clinical characteristic-based classification was published by Mulliken and Glowacki in 1982,⁸ and has since been cited over 4300 times. Mulliken and Glowacki’s observations led them to separate vascular anomalies into two main categories: tumors and malformations. The former is characterized by vasoproliferative activity and the second by a congenital lesion that grows in proportion with the patient. This classification system has been refined over the years with adoption by the International Society for the Study of Vascular Anomalies (ISSVA),⁹ but the core principle remains the same.

Numerous subsequent publications have been released through a myriad of journals across multiple disciplines, yet the wastebasket use of hemangioma to

Table 1. Common Nonspecific Terms for Different Vascular Anomalies

Old Terminology

Angioma
Nevus angiectoides
Varice aneurysm
Cirroid aneurysm
Hemangioma cavernosum
Arteriovenous angioma
Red angioma telangiectaticum
Cystic hygroma
Lymphangioma

describe all vascular anomalies persists. Although true hemangiomas are a type of vascular anomaly, most vascular anomalies, particularly malformations, cannot be described by hemangioma. For example, the suffix “-oma” alone implies neoplasia, which is often not the case. Lacking specificity, it can generate confusion, misdiagnosis and inadequate or improper management.

Initial Considerations and Evaluation

A multidisciplinary approach has largely become standard of care with dedicated clinics for the diagnosis and treatment of vascular anomalies.¹⁰⁻¹² These clinics have proven vital given the range of pathology presented and treatments offered. These clinics also provide continuity of care with re-evaluation of

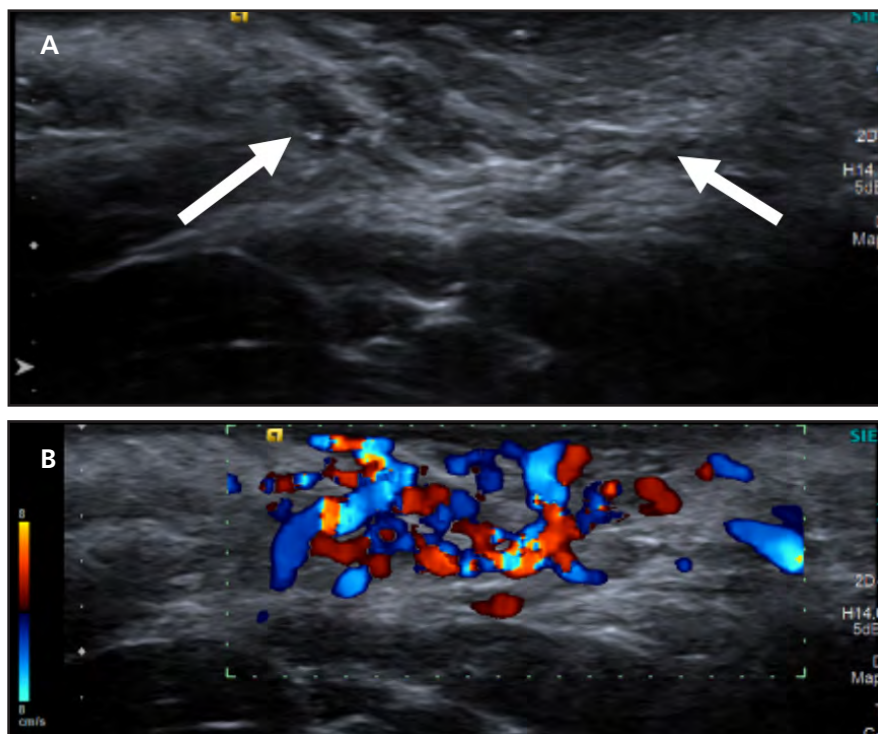


FIGURE 1. Ultrasound imaging of an infantile hemangioma demonstrating a heterogeneous hypoechoic lesion (A) with hypervascularity lacking a distinct feeding vessel (B). Note the low velocity scale.

disease progression and management of comorbidities over the patient's lifetime. At our institution, this team consists of hematology, diagnostic and interventional radiology, plastic surgery and otolaryngology, and meets monthly to discuss and coordinate each patient's care.

For many of these anomalies, an initial history and physical examination can often lead to a diagnosis without the need for diagnostic imaging. That said, in a modern practice environment, it is recommended that these lesions be imaged, most often with MRI and ultrasound. Less often, CT can be used to troubleshoot the angioarchitecture of complex lesions, evaluating bony involvement or used for logistical purposes in which MRI may not be feasible or is contraindicated.

"True" Hemangiomas

The term hemangioma should be reserved for true neoplasms characterized by proliferative tissue. Most hemangiomas are diagnosed clinically, although nonsuperficial or complex hemangiomas

may require imaging, which can detail the extent of disease and clarify the diagnosis. The infrequent biopsy may also be necessary in lesions that exhibit more aggressive characteristics, discussed later in this article.

Most vascular anomalies are true hemangiomas,¹⁴ which are split into two types: infantile and congenital. Both exhibit similar imaging characteristics but are distinct entities differentiated by clinical history and immunohistochemistry, specifically the expression of the GLUT-1 glucose transport protein. Only infantile type hemangiomas express GLUT-1.^{13,14}

Infantile Hemangioma

Infantile hemangiomas represent the most common head and neck vascular anomaly and tumor in infants with a preponderance for the head and neck.^{6,15} Incidence is relatively high, identified in up to 2.6% of term infants, with 10% to 12% presenting by 1 year of age. There is a female predilection in addition to being more common in low birth weight and Caucasian patients.¹⁵⁻¹⁷ Reportedly, there

is also increased incidence of infantile hemangiomas correlating with chorionic villus sampling at 9 to 12 weeks' gestation.

Clinically, these lesions present as lumps/bumps palpated by a parent, which may not be visually obvious. They exhibit progression through four phases: nascent, proliferative, involutional and involuted. Classically, infantile hemangiomas are not visualized at birth, although premonitory or precursor patches of skin discoloration may be identified by the scrutinizing physician in up to 50% of patients.¹⁵

Depending on the depth and character of the lesion, the proliferative phase may be detected as early as a few weeks of age.¹⁸ Most pronounced growth occurs during 3 to 6 months of age, with a minority continuing to grow beyond 1 year. After a period of quiescence, involution takes place lasting 5 to 6 years.² Hemangiomas reach involution at a classically described rate of 50% by 5 years and 70% by 7 years.

Imaging

In the evaluation of infantile hemangioma, ultrasound is often an ideal first choice. On ultrasound, hyperechoic and/or hypoechoic lesions are seen demonstrating uniformly scattered punctate vascularity, which are high velocity/low resistance on Doppler interrogation^{11,19} (Figure 1). Involved lesions generally demonstrate an isoechoic appearance and may not be distinguishable from surrounding soft tissue.²⁰

MR findings typically demonstrate a well-circumscribed lesion with non-aggressive features. Hemangiomas generally exhibit increased T2 and intermediate T1 intensity with strong enhancement on postcontrast sequences. Classically, flow voids depicting faster flow vessels are present^{21,22} and there should be no adjacent soft-tissue edema (Figure 2).

Treatment

Infantile hemangiomas are predominantly self-limiting in that nearly all involute without complication. Medical intervention is warranted in

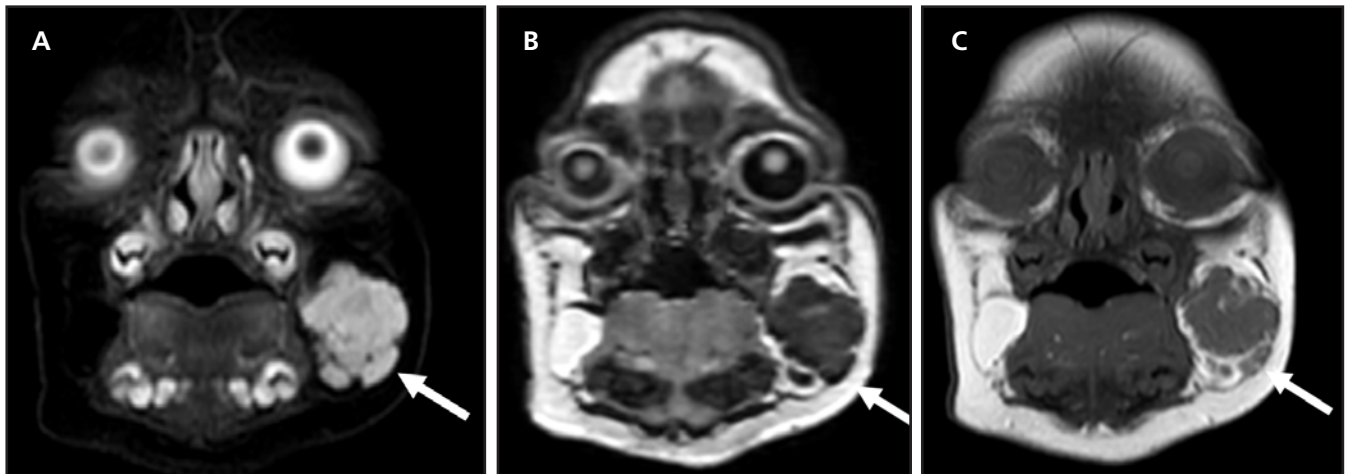


FIGURE 2. MRI of the face demonstrating a well-circumscribed infantile hemangioma (arrows) as T2 hyperintense (A) and T1 hypointense (B) with moderate relative enhancement (C).

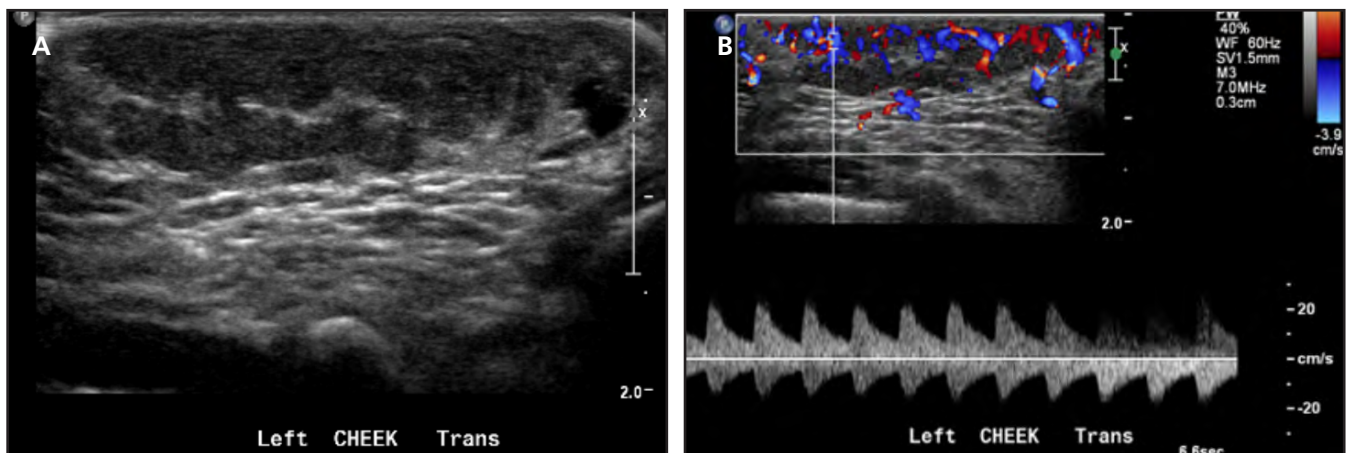


FIGURE 3. Ultrasound of a soft-tissue mass on the left demonstrating heterogeneous hypoechoic appearance (A), which is not very distinguishable from hemangiomas in general. On Doppler interrogation (B), we see the same scattered punctate vascularity with a slight difference exhibited by increased vascular density and a high resistive waveform pattern raising concern for a more aggressive lesion. Punch biopsy proved lesion to be a rapidly involuting congenital hemangioma.

approximately 10% to 20% of cases where lesions generate complications including high output heart failure, airway compromise, vision impairment, ulceration, or facial lesions whose unabated growth can lead to more permanent disfigurement.

First line medical therapy includes weight-based dosing of propranolol,¹⁸ followed by the addition of corticosteroids and/or vincristine in resistant lesions. Lesions responding poorly to medical therapy or with significant comorbidities (eg, Kasabach Merritt syndrome with high output cardiac failure) are candidates for either surgery, endovascular embolization, or a combination of both coordinated with input from the multidisciplinary team.

Finally, in select cases where there are residual remnants of fibrofatty tissue after complete lesion involution, surgical resection may be warranted or elected.

Congenital Hemangiomas

The distinction between congenital and infantile hemangiomas is based on two main features. Congenital hemangiomas are fully formed at birth having completed their proliferative phase. Secondly, they do not express GLUT-1 on immunohistochemical staining. Two forms of congenital hemangiomas exist: noninvoluting congenital hemangiomas (NICH) and rapidly involuting congenital hemangiomas (RICH).^{8,22,23}

As their names suggest, NICHs do not involute, whereas RICHs usually

involute by 14 months of age.²⁴ As is the case with infantile hemangiomas, uncomplicated congenital hemangiomas are well identified clinically. Nonetheless, ubiquitous availability of imaging allows for easily accessible assistance in generating a more complete diagnosis, especially determining the extent of deeper lesions.²⁰ In some instances, a biopsy may be performed to confirm diagnosis.

Imaging

Imaging of congenital hemangiomas is similar in approach and appearance to that of infantile hemangiomas. They are generally well-circumscribed masses with variable echogenicity and high vessel density. Where they differ

from infantile hemangiomas is gray-scale visibility of tubular vessels, echogenic heterogeneity, and the occasional calcification (RICH > NICH). Also, NICH lesions may demonstrate increased vascular velocities compared with RICH and involving infantile hemangiomas.^{14,24}

The MR imaging characteristics of NICH and RICH are also similar to that of infantile hemangiomas although congenital hemangiomas may present with ill-defined margins. Adjacent fat-stranding may also be present, more commonly with NICH. Soft-tissue masses with low vessel density, adjacent edema or high resistive indices should prompt further interrogation to rule out other soft-tissue tumors^{22,25} (**Figure 3**). Aggressive features such as infiltration or bony involvement should raise suspicion for kaposiform hemangioendothelioma.²⁶ Additional differentials are listed in **Table 2**.

Treatment

RICH lesions require no treatment given their self-limiting rapid involution, although sequelae such as remnant excess cutaneous tissue or discoloration may require cosmetic correction. Smaller NICH lesions are generally treated conservatively while larger lesions require laser therapy and/or surgical resection. In very large lesions, endovascular embolization prior to resection may have a role.

Vascular Malformations

To reiterate, vascular malformations represent a morphogenic abnormality of various vessels present at birth. They are nonproliferative and demonstrate normal cellular turnover, growing in proportion to the child and may not be distinctly visible at birth. Although they maintain a nonmitotic “mature” endothelium, they can still grow in response to trauma (iatrogenic or otherwise), hormonal changes, or infection.

Vascular malformations are subdivided initially by the dominant vessel type (eg, venous, capillary, lymphatic, arterial) or a combination thereof with further subdivision based on flow veloc-

ity. In theory, these lesions can have overlapping characteristics manifesting as a gradient of lesions that may require a balance of therapies including: intralesional or interstitial sclerotherapy, surgical resection, laser therapy and chemotherapy. From a treatment perspective, we will focus on the classic subtypes.

Capillary Malformations

These slow-flow vascular anomaly malformations are generally restricted to usually involve the capillaries of the superficial dermal layers and present as discolored macules or patches. Prior antiquated terminology included terms like angel's kiss, stork bite or salmon patch (nevus simplex) or port-wine stain (nevus flammeus). They occur in approximately 0.3% of infants²⁷ and are usually noticed at birth, although initially they may be dismissed as erythema or bruising from birth trauma. Single or multiple lesions may be present as small patches and involve a portion of the face or an entire limb. Facial involvement may herald additional syndromic associations and, for this reason, children with craniofacial involvement should undergo additional workup in the form of a brain MRI (or, less preferred, CT). This specifically pertains to capillary malformations in the trigeminal nerve distribution, which is highly associated with anomalies of the ipsilateral leptomeninges, ie, Sturge-Weber syndrome.¹¹

Craniofacial capillary malformations are diagnosed on a clinical basis and often require no imaging outside of excluding associated syndromic disease. Sonographic features are non-specific. MR imaging may demonstrate skin thickening or abnormality of the subcutaneous tissue, if anything.^{22,25} The primary treatment option of capillary malformations is laser therapy, initially pulsed-dye laser (PDL), given the superficial involvement.²⁷

Venous Malformation

Congenital venous malformations are characterized by a collection of abnormal, ectatic veins with irregular channels present at birth. The smooth muscle

Table 2. Basic Differential for Soft-Tissue Mass Mimicking Atypical Appearance of Hemangioma

Hemangioma Differential

Rhabdomyosarcoma
Tufted angioma
Sarcoma
Hemangiopericytoma
Myofibromatosis
Neuroblastoma
Metastatic neuroblastoma
Fibrosarcoma
Kaposiform hemangioendothelioma

layer is discontinuous and thin, resulting in inability of veins to constrict normally after distention from increased volume. Associated absence of valves aggravates swelling, and dysplastic veins become progressively enlarged. Stagnation of blood leads to further thrombosis, swelling and pain. Chronic thrombi calcify forming phleboliths, which are pathognomonic for venous malformations.^{10,12}

Detection is usually early in superficial lesions with dermal involvement showing varying levels of bluish discoloration. Deeper lesions (ie, intraosseous, intramuscular) may not be identified until later in life when significant swelling or pain manifests. Characteristic enlargement with heavy activity, Valsalva, or crying, as well as dependent position can often assist with clinical diagnosis. Rapid enlargement may also occur with trauma, hemorrhage, or hormonal influences (ie, puberty, pregnancy). Superficial lesions are compressible without audible bruit or pulsatility and may have palpable phleboliths within them.

Imaging

Venous malformations are most commonly described as cystic, dysplastic or spongiform in morphology. In practice, they are a combination of these characteristics with associated, usually dysplastic, narrow draining vessels,²⁸ which eventually often communicate with the normal venous system.

As with most vascular anomalies, initial evaluation with ultrasound and

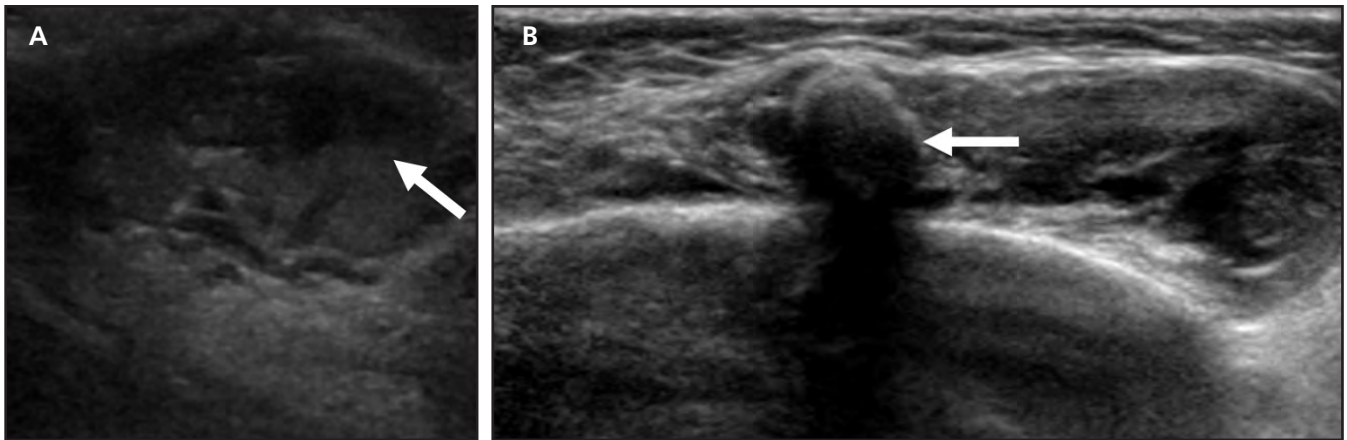


FIGURE 4. Gray-scale sonographic image (A) demonstrates a hypoechoic area with subtle layering internal echoes (arrow) consistent with the hematocrit effect, a common finding in venous malformations. Well-circumscribed, heterogeneously hypoechoic area (B) with internal crescentic echogenicity shadowing posteriorly is consistent with phlebolith, which makes the diagnosis for venous malformation pathognomonic.

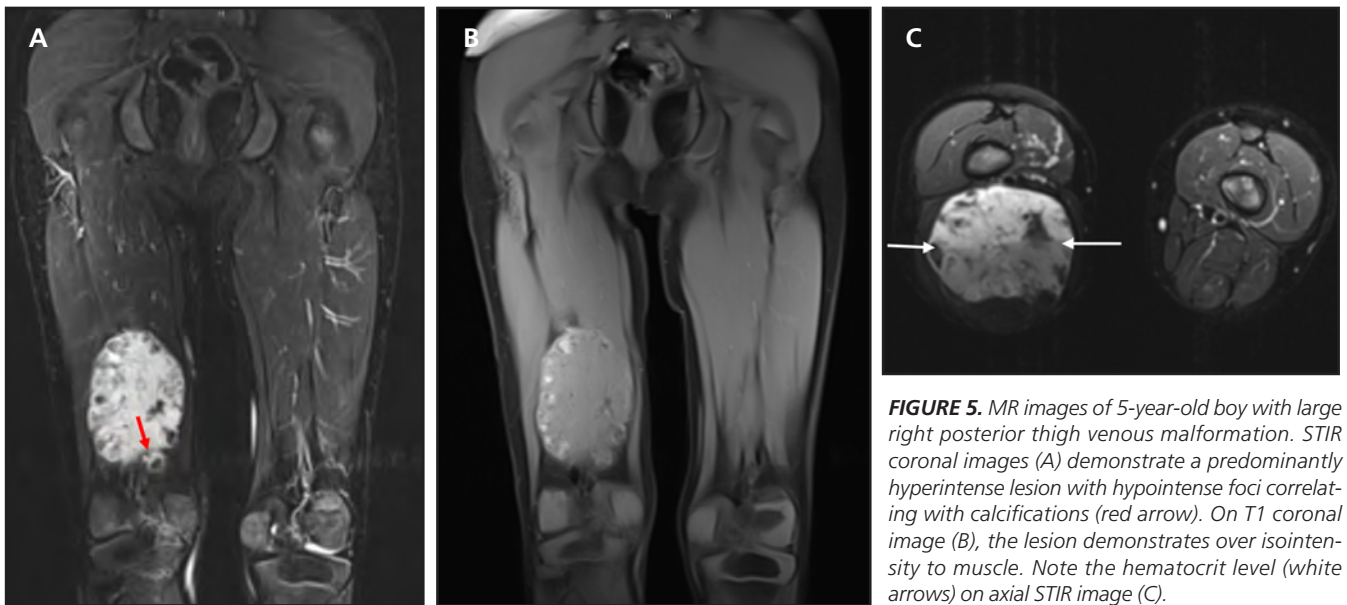


FIGURE 5. MR images of 5-year-old boy with large right posterior thigh venous malformation. STIR coronal images (A) demonstrate a predominantly hyperintense lesion with hypointense foci correlating with calcifications (red arrow). On T1 coronal image (B), the lesion demonstrates over isointensity to muscle. Note the hematocrit level (white arrows) on axial STIR image (C).

MR imaging are the mainstays. Ultrasound allows for evaluation of flow velocity, compressibility, and visualization of thrombus/phleboliths but is limited to relatively superficial areas (**Figure 4**). Multiple-region MRI can also demonstrate secondary characteristics of flow velocities such as fluid-fluid levels (hematocrit effect) and phleboliths (signal voids) but adds the ability for large field-of-view imaging, evaluating the deeper extent and multiplicity of lesions.^{11,21,28} It should be mentioned that phleboliths can be detected radiographically and are usually pathognomonic for venous malformations.

Gray-scale sonography typically

shows a compressible hypoechoic lesion, which can be well circumscribed, but are more often ill-defined or infiltrative. Mobile internal echoes can be visualized sonographically with compression maneuvers while tourniquets can engorge lesions. Tubular structures may be visualized in larger malformations consistent with ectatic veins.

Venous malformations are hyperintense on fluid-sensitive MR sequences. Well-circumscribed hypointense T1 and T2 foci would represent phleboliths. Fluid-fluid levels are also visualized representing a hematocrit effect. Venous malformations are generally considered to enhance diffusely.^{22,25,29}

In our practice, however, we have found enhancement to be variable, including minimal enhancement, enhancement with layering, and avid enhancement (**Figure 5**). Reporting on venous malformations should include lesion size and extent, tissue compartments involved, flow velocities if any, compressibility, and presence of phleboliths.

Venography is often performed during the treatment of venous malformations. This is facilitated by direct ultrasound-guided puncture of anechoic spaces followed by fluoroscopy with contrast injection (**Figure 6**). Venography depicts the individual morphology – cystic, lobulated or dysplastic – of

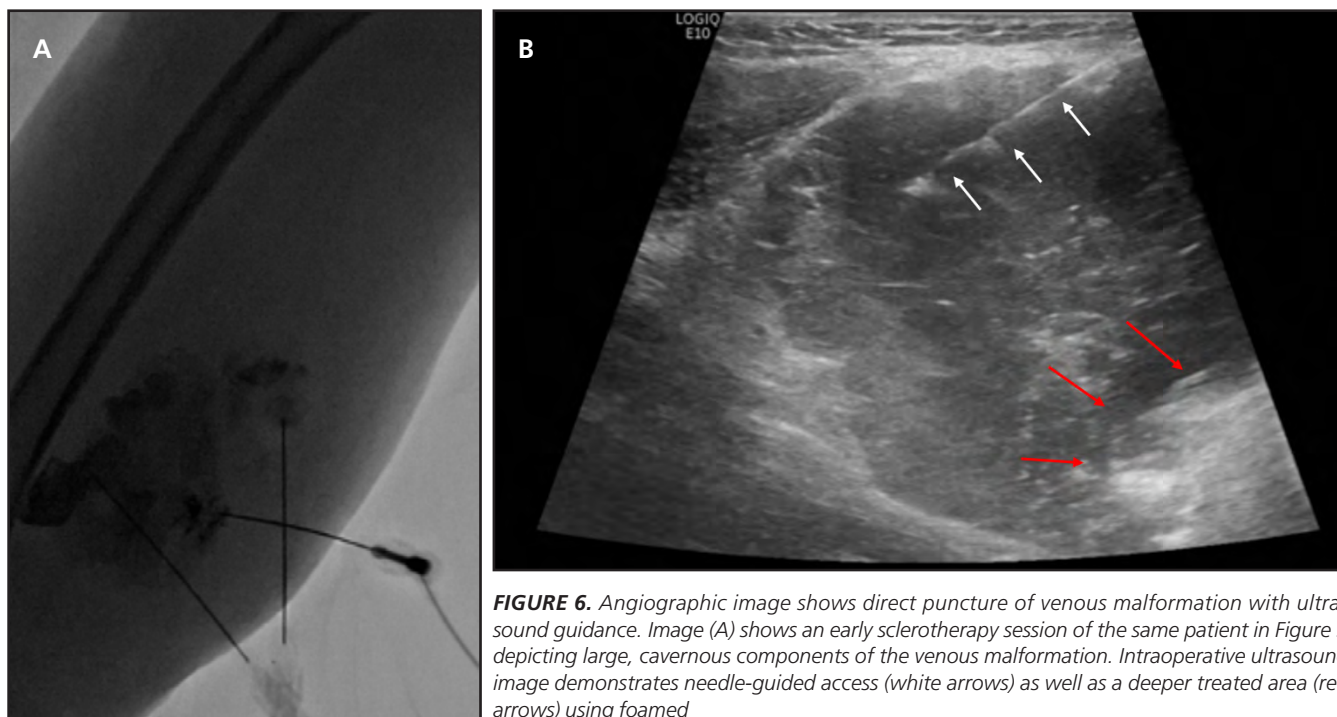


FIGURE 6. Angiographic image shows direct puncture of venous malformation with ultrasound guidance. Image (A) shows an early sclerotherapy session of the same patient in Figure 5 depicting large, cavernous components of the venous malformation. Intraoperative ultrasound image demonstrates needle-guided access (white arrows) as well as a deeper treated area (red arrows) using foamed



FIGURE 7. Another example of intraoperative direct puncture angiogram demonstrating a cavitory/spongiform component of a left venous malformation. Manual extrinsic compression occludes caudal draining vessel using a gauze-covered hemostat focalizing sclerosant within the malformation.

each component within the venous malformation and identifies any prominent drainage of the malformation, which is necessary in guiding therapy.

Treatment

Treatment depends on the location and extent of the venous malformation. Multiple options exist including compression, laser therapy, surgery and sclerotherapy, or a combination.^{5,29}

Local compression may be of benefit in isolated locations such as extremities. However, unlike extremities, compression garments cannot be practically applied to venous malformations in the head and neck. Surgical excision is reserved for accessible and localized lesions; otherwise, these anomalies are managed most often with sclerotherapy.

With sclerotherapy, patient and family expectations must be tempered

during the initial consultation, especially in extensive lesions involving large areas and multiple components. The goal of sclerotherapy is not curative, but rather management of the disease for reducing the comorbidities, which range from pain to disfigurement. It is often unclear initially how many treatments will be required, ranging from a single therapy to numerous repeat sessions over several years. This

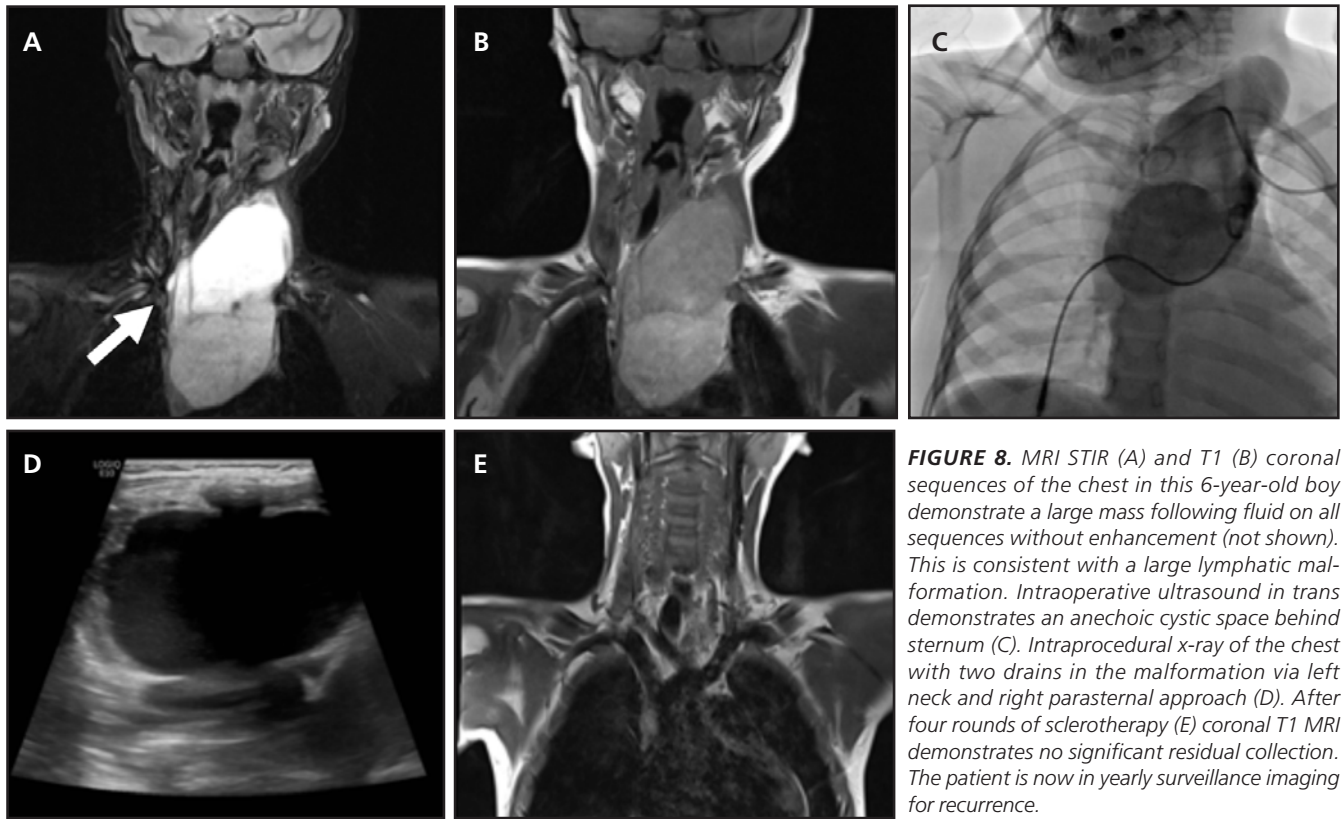


FIGURE 8. MRI STIR (A) and T1 (B) coronal sequences of the chest in this 6-year-old boy demonstrate a large mass following fluid on all sequences without enhancement (not shown). This is consistent with a large lymphatic malformation. Intraoperative ultrasound in trans demonstrates an anechoic cystic space behind sternum (C). Intraoperative x-ray of the chest with two drains in the malformation via left neck and right parasternal approach (D). After four rounds of sclerotherapy (E) coronal T1 MRI demonstrates no significant residual collection. The patient is now in yearly surveillance imaging for recurrence.

depends not only on disease extent but on variables such as patient satisfaction with cosmesis, pain and relapsing disease as the child grows, and external forces such as trauma or infection.

Sclerotherapy begins with contrast venography to depict lesion size and draining veins, and to assess deep veins and sclerosant volume needed (**Figures 8, 9**). The goal of sclerotherapy is localized destruction of endothelial lining, inflammation, and thrombosis of the malformation. Most commonly used sclerosants include 98% ethanol, 1% or 3% sodium tetradecyl sulphate foam, doxycycline, Ethibloc, polidocanol and bleomycin. Ethanol and sodium tetradecyl sulphate are generally considered the most common, effective and available sclerosants. Manual compression may be used to reduce outflow during treatment. In scenarios where there is “rapid” or significant outflow, coils or glue can be used to occlude outflow prior to sclerosant injection. Coils can also be utilized to protect nontarget veins. Complications of sclerotherapy include systemic toxicity of sclerosant,

nontarget embolization, temporary nerve injury, fibrosis and skin necrosis in superficial lesions.

Lymphatic Malformation

Lymphatic malformations represent vascular anomalies proceeding from disorganized lymphangiogenesis and are estimated to account for approximately 6% of all benign childhood head and neck lesions.²¹ Similar to venous malformations, lymphatic malformations are characterized by cystic spaces lined by “mature” endothelial cells, which do not exhibit neoplasia.⁸ Larger lymphatic malformations are still often referred to by old terminology such as cystic hygromas or lymphangiomas, which should be discouraged. These lesions may be visualized in utero, as early as the second trimester and are present at birth. Between 65% and 75% are diagnosed at the time of birth – as they are not always visible on physical examination – and 80% to 90% by 2 years of age. They can present at any age with sudden onset rapid growth from incidental trauma, infection (eg, upper respiratory) or hormonal

change (puberty, pregnancy, etc.), which is often disconcerting to parents and some primary care physicians.

Lymphatic malformations have a greater propensity to insinuate between soft tissues as well as a greater likelihood of demonstrating communication between cystic spaces compared with venous malformations. Lymphatic malformations are characterized as either microcystic (< 1 cm), macrocystic (≥ 1 cm), or a combination of both. This classification helps determine treatment plans.

Imaging

Lymphatic malformations are very slow flow structures, which is well depicted on imaging, particularly ultrasound. Macrocysts are usually well-defined, loculated fluid collections, which can have few internal septae (**Figure 8**). They are hypoechoic or anechoic in character, although debris or hemorrhage can be seen within. Microcystic morphology is hyperechoic in character due to the closely packed cysts/septae (**Figure 9A**), which demonstrate increased vascularity.

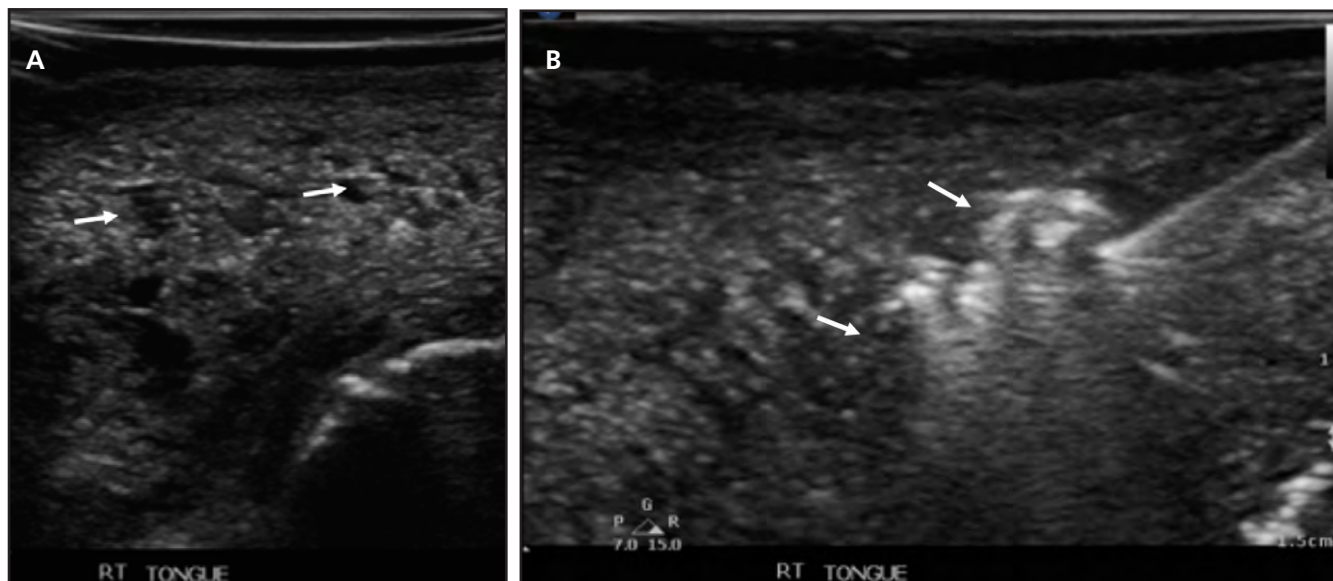


FIGURE 9. Sonographic grayscale image of the tongue belonging to the same patient as in Figure 8 demonstrating a microcystic component of the lymphatic malformation. (A) This is evidenced by numerous punctate anechoic cysts (arrowheads) interspersed throughout the tongue without internal vascularity (not shown). Image (B) demonstrates injection of Doxycycline emulsion (arrow) while retracting needle (arrowheads) 'threaded' through these cysts under sonographic guidance.

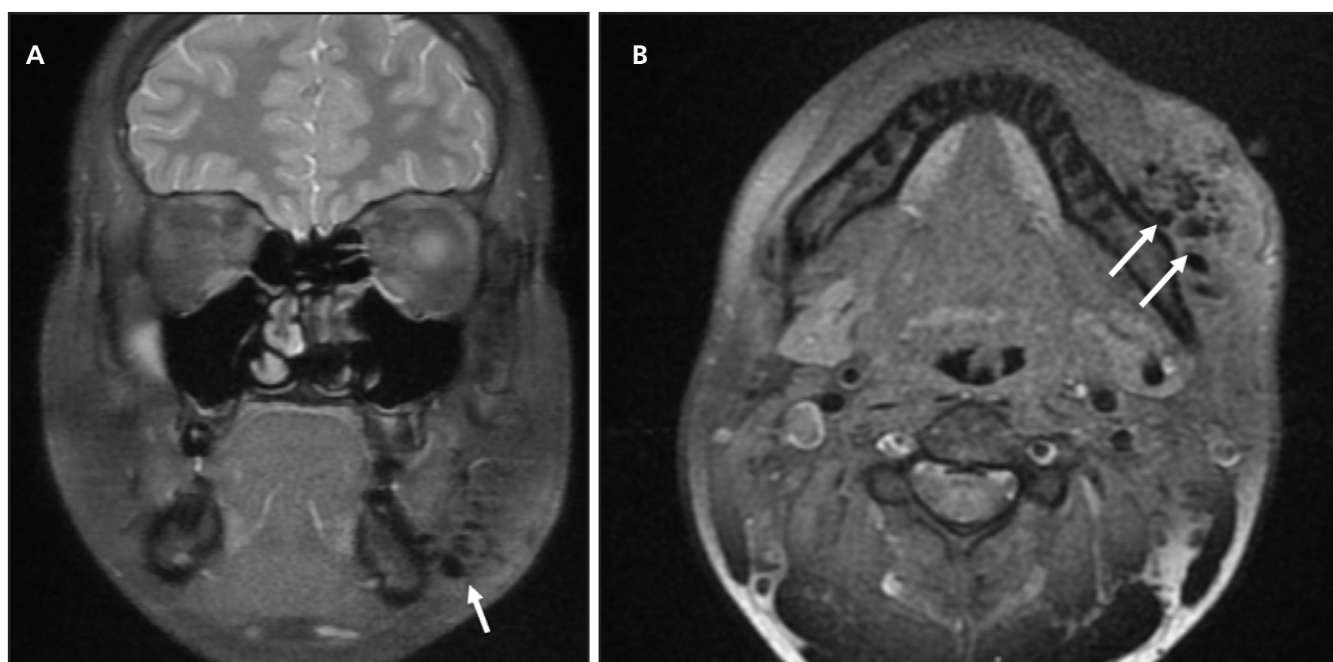


FIGURE 10. Axial T1 (A) and T2 (B) MR images of the face demonstrate intertwining flow voids (arrows) in the left cheek. There is tissue hypertrophy without discrete associated soft-tissue mass interdigitating the lesion.

On MRI, lymphatic malformations are septated cystic structures usually following fluid signal (high T2/low T1) internally, the caveat being that proteinaceous or hemorrhagic content can increase the T1 signal. Increased confidence in identifying the presence of enhancement can be aided by performing subtraction images (subtract-

ing the unenhanced from the enhanced T1-weighted images in postprocessing). When present, septations may demonstrate enhancement, while there should be no enhancement of the cystic components. This is an important distinction in differentiating lymphatic from venous malformations and translates to CT imaging. With CT, cystic spaces are low

attenuation with usually thin walls. The exception is infection, which generates thickening of the cyst walls and adjacent inflammatory changes.^{21,22,25}

Conventional angiographic imaging is key in the treatment of macrocystic lymphatic malformations. Although diagnostic value is relatively limited, morphology and presence of draining

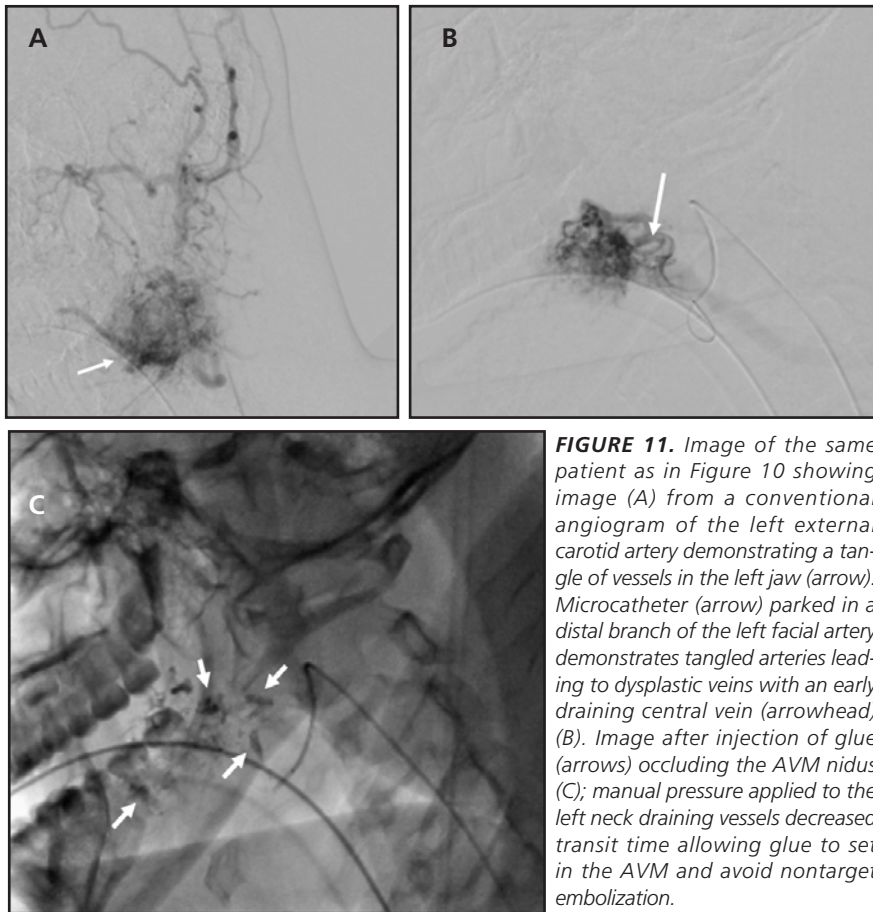


FIGURE 11. Image of the same patient as in Figure 10 showing image (A) from a conventional angiogram of the left external carotid artery demonstrating a tangle of vessels in the left jaw (arrow). Microcatheter (arrow) parked in a distal branch of the left facial artery demonstrates tangled arteries leading to dysplastic veins with an early draining central vein (arrowhead) (B). Image after injection of glue (arrows) occluding the AVM nidus (C); manual pressure applied to the left neck draining vessels decreased transit time allowing glue to set in the AVM and avoid nontarget embolization.

vessels/venous can aid in developing a more complete diagnosis, especially with complex or combined venous and lymphatic malformations.

Treatment

Regarding management, the large majority of lymphatic malformations should not be treated conservatively as there is only an approximately 3% chance of spontaneous regression.³⁰ Conservative therapy is, therefore, a relatively poor option as there is progression of disease with susceptibility for infection, ulceration, bleeding and sequelae of mass effect (eg, airway compromise). Sclerotherapy is generally the first-line treatment for macrocysts with high long-term success rates and low rates of complication. Microcystic lesions can also be treated with sclerosant injection under ultrasound guidance, most commonly using a doxycycline emulsion (**Figure 9B**) and, alternatively, bleomycin.

In well-circumscribed smaller and superficial lesions, surgical resection may be considered. Staged surgical debulking in conjunction with sclerotherapy may be effective in massive or life-threatening lesions. However, surgical involvement can lead to complication of macrocysts requiring a significant increase in efforts for subsequent percutaneous therapy. For extensive disease, as in diffuse superficial microcystic disease where sclerotherapy or surgical debulking is not feasible, pharmacotherapy may be considered. Specifically, sirolimus has shown promise.³¹ This echoes the need for the involvement of a multispecialty team familiar with the natural history and management of the disease.

Arteriovenous Malformation

Arteriovenous malformations (AVMs) are the prototypical congenital high-flow vascular malformations. AVMs typically have a central nidus with feeding artery/arteries and draining

vein(s) meeting at a central location. There is no intervening capillary bed between the arterial and venous components generating variable amounts of shunting.^{4,8} Like other vascular malformations, arteriovenous malformations are structural abnormalities with normal mature endothelial lining. The most common locations are intracranial followed by extracranial head and neck.³²

Superficial lesions present as a mass, which can be pulsatile and warm on palpation. Deeper lesions tend to manifest later in life as areas of pain. Skin discoloration is noted in most patients presenting with superficial, extracranial AVMs as infants, whereas less than 50% of postpubertal patients exhibit skin discoloration.³² Bruits may also be heard on auscultation. Deeper lesions tend to manifest later in life and can present due to pain. Generally, AVMs do not exhibit rapid growth but, like other vascular malformations, growth can be triggered by trauma, surgical/endovascular intervention and hormonal changes (pregnancy/puberty). The natural history is progression of the disease with growth and recruitment of additional vasculature increasing the complexity of the lesion. Untreated lesions can have an aggressive clinical course including life-threatening bleeding, ischemia, ulceration, high-output heart failure and significant pain.

Imaging

AVMs are typically a tangle of intertwinning vessels best demonstrated with conventional angiography or MR/CT angiography. There is a soft-tissue component interdigitating the hypertrophied vessels, which has been shown to have angiogenic effects. Initial MR or contrast-enhanced CT assist in obtaining the overall picture and are paramount for planning treatment. Ultrasound may be the first modality used for superficial lesions on initial presentation demonstrating an overall hypoechoic mass. Doppler shows a low resistance pattern in feeding arteries and arterial waveforms in draining veins (arterialization). Aliasing or turbulence

is seen in the nidus, and abnormal pulsatility is present in draining veins.^{4,11}

Similar to other malformations, CT and MR are excellent for demonstrating extent of lesion including intramuscular and intraosseous involvement. Depiction of multiple hypertrophied arteries and dilated veins associated with the lesion and their relation are necessary for classification and treatment planning. MR exhibits flow voids on T1 and T2 sequences and an overall lack of identifiable soft-tissue components (**Figure 10**). Presence of a soft-tissue mass should prompt consideration for neoplasm such as sarcoma. Temporal imaging with or without contrast enhancement is helpful to separate arteries from veins in complex lesions.¹¹ Time-resolved MRA techniques are especially helpful (TRICKS – GE Healthcare and TWIST – Siemens Healthineers). Finally, traditional angiography exhibits the highest image and temporal resolution facilitating visualization of early draining veins (**Figure 11**). Angiography also allows for concurrent intervention minimizing the number of examinations.

Treatment

Small, asymptomatic lesions may be conservatively managed initially with observation and repeat imaging. However, the expected natural course is progression toward expansion of the lesion with associated sequelae of pain, bleeding, ulceration, etc.

Symptomatic lesions are treated most commonly with endovascular therapy, classically a transluminal intra-arterial approach with coil or glue embolization as well as absolute ethanol (**Figure 11**). Particles are usually not a recommended embolic agent as shunting will likely result in a nontarget injury. An almost expected progression of arterial recruitment often complicates subsequent treatments; therefore, a more comprehensive initial embolic therapy from combined arterial and venous approaches is critical. Postembolization surgical intervention may also be combined in certain accessible lesions.

Complex lesions require a multidisciplinary team with diagnostic and interventional radiology, plastic surgery and/or vascular surgery. Each lesion is unique, requiring a combination of therapy and follow-up. There is a high recurrence rate in the long term.⁵

Conclusion

Vascular anomalies are a group of life-altering and life-long diseases with far-reaching physical and psychosocial consequences. Understanding the basic presentation, pathology and imaging characteristics allows for accurate diagnosis with swift and appropriate treatment. Multidisciplinary teams are an essential component in improving the long-term outlook for this patient population, significantly improving quality of care and prognosis.

REFERENCES

- Enjolras O, Riche MC, Merland JJ, Escande JP. Management of alarming hemangiomas in infancy: a review of 25 cases. *Pediatrics* 1990;85(4):491-498.
- Adams DM, Lucky AW. Cervicofacial vascular anomalies. I. Hemangiomas and other benign vascular tumors. *Semin Pediatr Surg* 2006;15(2):124-132.
- Orlow SJ, Isakoff MS, Blei F. Increased risk of symptomatic hemangiomas of the airway in association with cutaneous hemangiomas in a "beard" distribution. *J Pediatr* 1997;131(4):643-646.
- Kohout MP, Hansen M, Pribaz JJ, Mulliken JB. Arteriovenous malformations of the head and neck: natural history and management. *Plast Reconstr Surg* 1998;102(3):643-654.
- Buckmiller LM, Richter GT, Suen JY. Diagnosis and management of hemangiomas and vascular malformations of the head and neck. *Oral Dis* 2010;16(5):405-418.
- Haggstrom AN, Drolet BA, Baselga E, et al. Prospective study of infantile hemangiomas: clinical characteristics predicting complications and treatment. *Pediatrics* 2006;118(3):882-887.
- Fox RH. William Hunter, Anatomist, Physician, Obstetrician, (1718-1783). H. K. Lewis; 1901.
- Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. *Plast Reconstr Surg* 1982;69(3):412-422.
- Monroe EJ. Brief description of ISSVA classification for radiologists. *Tech Vasc Interv Radiol* 2019;22(4):100628.
- Donnelly LF, Adams DM, George S, Bisset I. Vascular malformations and hemangiomas. *Am J Roentgenol* 2000;174(3):597-608.
- Dubois J, Alison M. Vascular anomalies: what a radiologist needs to know. *Pediatr Radiol* 2010;40(6):895-905.
- Domp Martin A, Vikkula M, Boon LM. Venous malformation: update on aetiopathogenesis, diagnosis and management. *Phlebology* 2010;25(5):224-235.

- North PE, Waner M, Mizeracki A, Mihm MC Jr. GLUT1: a newly discovered immunohistochemical marker for juvenile hemangiomas. *Hum Pathol* 2000;31(1):11-22.
- Mulliken JB, Enjolras O. Congenital hemangiomas and infantile hemangioma: missing links. *J Am Acad Dermatol* 2004;50(6):875-882.
- Bruckner AL, Frieden IJ. Hemangiomas of infancy. *J Am Acad Dermatol* 2003;48(4):477-493; quiz 494-496.
- Drolet BA, Esterly NB, Frieden IJ. Hemangiomas in children. *N Engl J Med* 1999;341(3):173-181.
- Lowe LH, Marchant TC, Rivard DC, Scherbel AJ. Vascular malformations: classification and terminology the radiologist needs to know. *Semin Roentgenol* 2012;47(2):106-117.
- Léauté-Labrèze C, Hoeger P, Mazereeuw-Hautier J, et al. A randomized, controlled trial of oral propranolol in infantile hemangioma. *N Engl J Med* 2015;372(8):735-746.
- Dubois J, Patriquin HB, Garel L, et al. Soft-tissue hemangiomas in infants and children: diagnosis using Doppler sonography. *Am J Roentgenol* 1998;171(1):247-252.
- Pattiel HJ, Burrows PE, Kozakewich HP, Zurakowski D, Mulliken JB. Soft-tissue vascular anomalies: utility of US for diagnosis. *Radiology* 2000;214(3):747-754.
- Baer AH, Parmar HA, DiPietro MA, Kasten SJ, Mukherji SK. Hemangiomas and vascular malformations of the head and neck: a simplified approach. *Neuroimaging Clin N Am* 2011;21(3):641-658, viii.
- Navarro OM, Laffan EE, Ngan B-Y. Pediatric soft-tissue tumors and pseudo-tumors: MR imaging features with pathologic correlation: part 1. Imaging approach, pseudotumors, vascular lesions, and adipocytic tumors. *RadioGraphics* 2009;29(3):887-906.
- Berenguer B, Mulliken JB, Enjolras O, et al. Rapidly involuting congenital hemangioma: clinical and histopathologic features. *Pediatr Dev Pathol* 2003;6(6):495-510.
- Gorincour G, Kokta V, Rypens F, Garel L, Powell J, Dubois J. Imaging characteristics of two subtypes of congenital hemangiomas: rapidly involuting congenital hemangiomas and non-involuting congenital hemangiomas. *Pediatr Radiol* 2005;35(12):1178-1185.
- Laor T. MR imaging of soft tissue tumors and tumor-like lesions. *Pediatr Radiol* 2004;34(1):24-37.
- Ji Y, Chen S, Yang K, Xia C, Li L. Kaposiform hemangioendothelioma: current knowledge and future perspectives. *Orphanet J Rare Dis* 2020;15(1):39.
- Garzon MC, Huang JT, Enjolras O, Frieden IJ. Vascular malformations. *J Am Acad Dermatol* 2007;56(3):353-370.
- Flors L, Leiva-Salinas C, Maged IM, et al. MR imaging of soft-tissue vascular malformations: diagnosis, classification, and therapy follow-up. *RadioGraphics* 2011;31(5):1321-1340.
- Zheng JW, Mai HM, Zhang L, et al. Guidelines for the treatment of head and neck venous malformations. *Int J Clin Exp Med* 2013;6(5):377-389.
- Smith RJH. Lymphatic malformations. *Lymphat Res Biol* 2004;2(1):25-31.
- Laforgia N, Schettini F, De Mattia D, Martinelli D, Ladisa G, Favia V. Lymphatic malformation in newborns as the first sign of diffuse lymphangiomatosis: successful treatment with sirolimus. *Neonatology* 2016;109(1):52-55.
- Marler JJ, Mulliken JB. Current management of hemangiomas and vascular malformations. *Clin Plast Surg* 2005;32(1):99-116, ix.

Review of Image-Defined Risk Factors in Neuroblastoma

Andrew Sung, M.D., Elyssa M. Rubin, M.D., Hollie Lai, M.D.

Departments of Radiology and Oncology, Children's Hospital of Orange County, Orange, CA

Neuroblastoma is the most common extracranial solid tumor in infancy, accounting for approximately 15% of pediatric deaths from oncologic causes.^{1,2} Neuroblastoma is an umbrella term encompassing neuroblastic tumors including neuroblastoma, ganglioneuroblastoma, and ganglioneuroma. These tumors are comprised of neuroblasts, primitive undifferentiated sympathetic cells, and stroma, and can arise anywhere along the sympathetic chain.³ The median age at diagnosis is approximately 16 months, with 95% of neuroblastomas diagnosed by 7 years of age.⁴ Common tumor sites are: the adrenal glands (35%), extra-adrenal retroperitoneum (30% to 35%), and posterior mediastinum (20%).⁵ Given the wide range of histopathology, the natural progression of neuroblastoma is quite variable, with some tumors undergoing spontaneous regression, and others progressing to death. Even with recent advanced treatment regimens, patients with metastatic neuroblastoma have a poor 3-year,

event-free survival of about 60%, despite multimodal therapy.⁶

A few staging strategies have been developed to stratify patients and help deliver appropriate therapy based on individual risk. The International Neuroblastoma Staging System (INSS), developed in 1988 and modified in 1993, was the first widely used staging system to standardize the extent of disease in neuroblastoma patients.⁷ The INSS is a surgical staging system, with stages 1-3 defined by the extent of surgical excision of tumor, and stage 4 representing metastatic disease. However, pretreatment risk assessments and comparisons across clinical trials were difficult given its reliance on surgical outcomes, which can vary across surgeons. In 2009, the International Neuroblastoma Risk Group established the INRG Staging System (**Table 1**), which standardized neuroblastoma staging in a presurgical manner.^{4,8} The INRG staging system incorporates image-defined risk factors (IDRFs) (**Tables 2, 3**) as well as other

genetic and biomolecular markers to risk stratify patients. IDRFs are tumor imaging features, such as involvement of vasculature or other critical organs that have been associated with risk of resecting neuroblastoma. Tumors without an IDRF are considered L1, whereas a tumor with an IDRF is L2. Metastatic disease is stage M, as in the INSS. Stage MS applies to patients with metastatic disease confined to the skin, liver and/or bone marrow in patients 18 months or younger. This differs from the INSS stage 4S, which has an age cutoff of 12 months. In both staging systems, metastatic disease must be iodine-123 metaiodobenzylguanidine (MIBG) negative with less than 10% of total nucleated cells identified as malignant on bone marrow smears or biopsy.^{7,8}

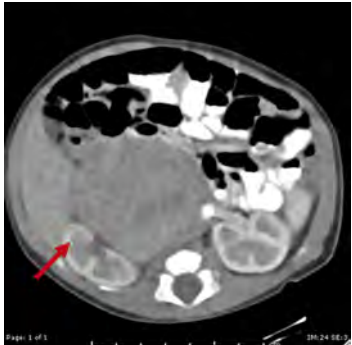
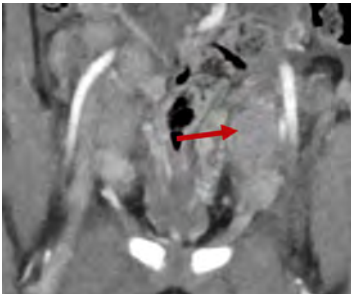
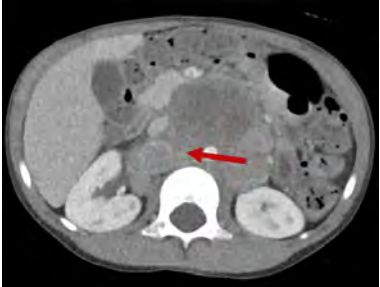
A variety of imaging modalities are employed during the initial diagnosis of neuroblastoma.⁹ Ultrasound is often the first imaging study ordered when there is suspicion of a mass. Ultrasound is

Table 1. International Neuroblastoma Risk Group Staging System*

Stage	Definition
L1	Localized tumor not involving vital structures as defined by the list of image-defined risk factors and confined to one body compartment
L2	Locoregional tumor with one or more image-defined risk factors
M	Distant metastatic disease (except stage MS)
MS	Metastatic disease in children younger than 18 months with metastases confined to skin, liver, and/or bone marrow

*Adapted from Monclair et al⁸

Table 2. Summary of Image-Defined Risk Factors (IDRFs)¹³ with Examples

Term	Description	IDRF?	Examples
Separation	"Visible layer, usually fat, is present between the tumor and the neighboring structure."	No	 The right kidney is displaced but maintains a fat plane (arrow) between it and the mass.
Contact	"No visible layer is present between the tumor and the neighboring structure." For an artery, contact occurs when less than 50% of the vessel circumference is enveloped by tumor.	No*	 Tumor surrounds aorta (arrow) by a little less than 50%.
Flattened	Used to describe when a vein is decreased in diameter, but the lumen remains partly visible.	No*	 The left external iliac vein is flattened by the adjacent tumor with contrast identified in the lumen (arrow). The left external iliac artery is immediately lateral to the vein.
Encasement	"Neighboring structure is surrounded by the tumor." More than 50% of the vessel circumference is in contact with the tumor. 100% of the vessel circumference in contact with the tumor indicates 100% encasement. A flattened vein with no visible lumen is also considered encasement.	Yes	 The tumor completely surrounds the aorta (arrow) with no visible layer of normal tissue between the tumor and aorta.

*A tumor in contact with the renal vessels is said to be invading the kidney and is considered IDRF-positive.

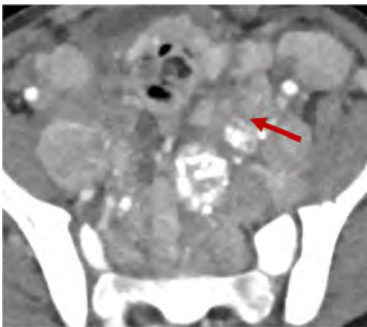
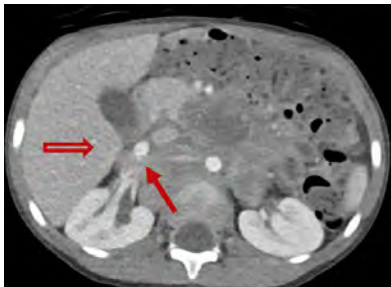
Term	Description	IDRF?	Examples
			 The left iliac vein is flattened by the adjacent tumor with loss of lumen (arrow). The iliac artery is immediately anterolateral to the vein.
Compression	"Tumor is in contact with the airways and causes the short axis of the airway to be reduced." This only applies to airways.	Yes	 Tumor is displacing and reducing the diameter of the left mainstem bronchus (arrow). There is also complete encasement of the aorta.
Infiltration	"Extension into a neighboring organ, causing the margins between the tumor and the infiltrated structure to be lacking or ill defined."	Yes	 There is an ill-defined margin between the right suprarenal mass and right kidney (arrow). Infiltration was confirmed at surgery.
Invasion	Contact or encasement of the renal vessels.	Yes	 The tumor is in contact with the right renal vein (open arrow) and encases the right renal artery (solid arrow). Both findings meet the criteria for invasion. There is also encasement of the aorta and invasion of the left renal pedicle (not entirely shown).

Table 3. Image-Defined Risk Factors in Neuroblastic Tumors***Ipsilateral tumor extension within two body compartments**

Neck-chest, chest-abdomen, abdomen-pelvis

Neck

Tumor encasing carotid and/or vertebral artery and/or internal jugular vein
 Tumor extending to base of skull
 Tumor compressing the trachea

Cervico-thoracic junction

Tumor encasing brachial plexus roots
 Tumor encasing subclavian vessels and/or vertebral and/or carotid artery
 Tumor compressing the trachea

Thorax

Tumor encasing the aorta and/or major branches
 Tumor compressing the trachea and/or principal bronchi
 Lower mediastinal tumor, infiltrating the costo-vertebral junction between T9 and T12

Thoraco-abdominal

Tumor encasing the aorta and/or vena cava

Abdomen/pelvis

Tumor infiltrating the porta hepatis (Figure 3) and/or the hepatoduodenal ligament
 Tumor encasing branches of the superior mesenteric artery at the mesenteric root
 Tumor encasing the origin of the celiac axis, and/or of the superior mesenteric artery
 Tumor invading one or both renal pedicles
 Tumor encasing the aorta and/or vena cava
 Tumor encasing the iliac vessels
 Pelvic tumor crossing the sciatic notch (Figure 4)

Intraspinal tumor extension

More than one-third of the spinal canal in the axial plane is invaded and/or the perimedullary leptomeningeal spaces are not visible and/or the spinal cord signal is abnormal

Infiltration of adjacent organs/structures

Pericardium, diaphragm, kidney, liver, duodeno-pancreatic block, and mesentery

Conditions to record but are not considered IDRFs

Multifocal primary tumors
 Pleural effusion, with or without malignant cells
 Ascites, with or without malignant cells

*Adapted from adapted from Park, et al⁶

defined. Some studies have shown higher sensitivity than MIBG for soft-tissue lesions¹² and stage 1 or 2 disease.¹³ Other studies have shown lower overall and event free survival in tumors with higher SUV_{max}.¹⁴

Image-Defined Risk Factors

The IDRF is a central component to the INRG Staging System. As the name implies, IDRFs are imaging features associated with a high risk of surgical complication. To create uniformity and aid in reproducibility across clinical trials, a lexicon has been suggested to describe tumor IDRFs.¹⁵ As stated in the INRG,⁸ either CT or MR can be used to determine IDRFs, with usage based on local availability and clinician or radiologist preference.

Summary of IDRF Terms and Descriptions

Separation – “visible layer, usually fat, is present between the tumor and the neighboring structure.”¹⁵ An IDRF is *not* present if there is *separation*.

Contact – “no visible layer is present between the tumor and the neighboring structure.”¹⁵ For an artery, contact occurs when less than 50% of the vessel circumference is enveloped by tumor. For a vein, the term flattened is used to describe when a vein is decreased in diameter, but the lumen remains partly visible. An IDRF is *not* present if there is *contact*, except for renal vessels (see *Invasion*).

Encasement – “neighboring structure is surrounded by the tumor.”¹⁵ For an artery, encasement requires more than 50% of the vessel circumference to be enveloped by tumor. For a vein, encasement is present when the lumen is no longer visible. An IDRF is present if there is *encasement*.

Compression – “tumor is in contact with the airways and causes the short axis of the airway to be reduced.”¹⁵ This only applies to airways. An IDRF is present if there is *compression*.

Infiltration – “extension into a neighboring organ, causing the margins between the tumor and the infiltrated

advantageous due to the lack of radiation or need for sedation; however, it is insufficient for the evaluation of IDRFs according to the INRG staging system. This is due to poor interobserver reproducibility as well as limited assessment of highly calcified tumors due to posterior acoustic shadowing. As outlined in the INRG,⁸ CT or MR are mandatory imaging studies to assess for the extent of tumor and to document IDRFs. CT is widely available and can be performed rapidly often without the need for sedation. It also has higher resolution than MR and is the best modality to evaluate for airway compression or lung parenchymal involvement. MR

has better soft-tissue contrast and is the best modality to evaluate spinal canal extension; however, it is not as widely available as CT and often requires sedation. MIBG is a norepinephrine analog that demonstrates uptake in approximately 90% of neuroblastomas¹⁰ and is mandatory for the evaluation of metastatic disease. A variety of MIBG scoring systems have been published and are reviewed in the INRG Task Force publication by Matthay et al.¹¹ F-18 fluorodeoxyglucose (FDG) PET/CT is occasionally used when MIBG is not widely available or for MIBG negative tumors; however, studies have been inconclusive, and its use is not clearly



FIGURE 1. Partially calcified contiguous right thoracic and abdominal neuroblastoma (IDRF positive).

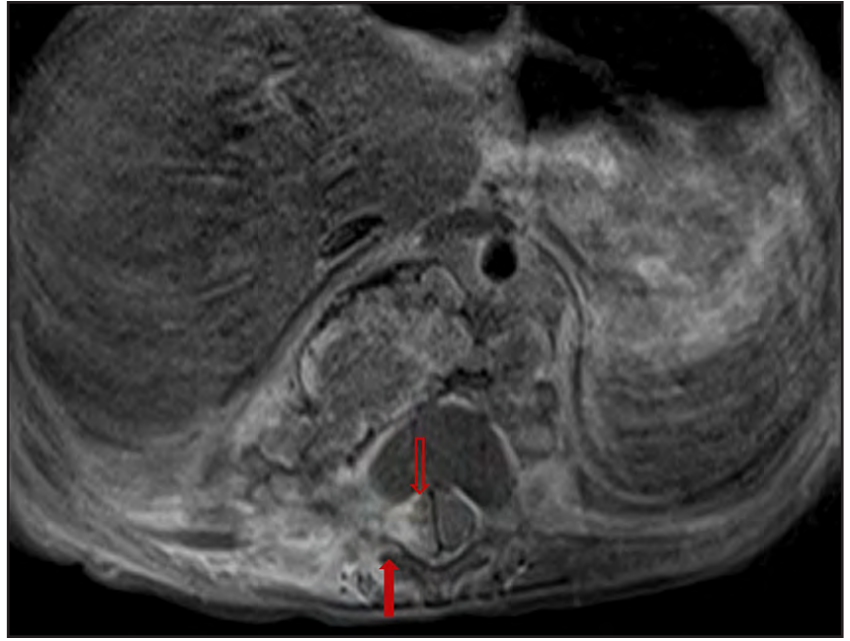


FIGURE 2. Axial postcontrast T1-weighted MRI with fat saturation demonstrates extension of the paraspinal neuroblastoma into the spinal canal, invading more than one-third of the spinal canal (arrow). IDRF positive. The spinal cord (open arrow) is displaced by the tumor.

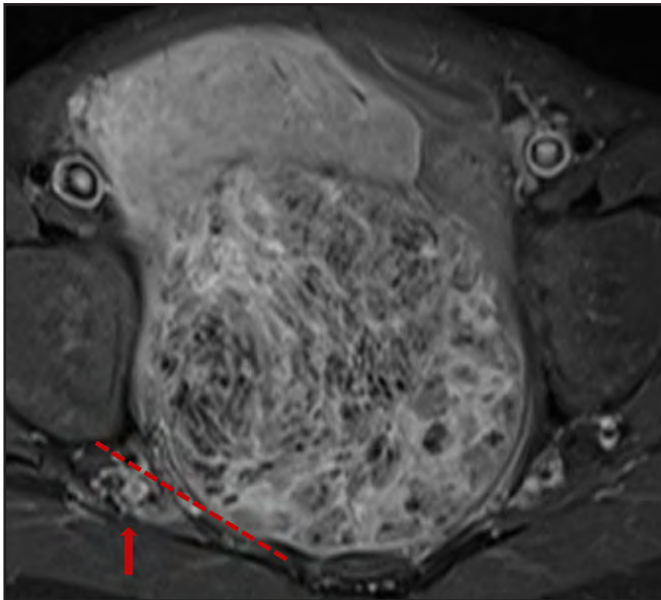


FIGURE 3. Axial postcontrast T1-weighted MRI with fat saturation demonstrates a large presacral and pelvic mass. The tumor (arrow) extends through the right greater sciatic foramen (dotted line). IDRF positive.

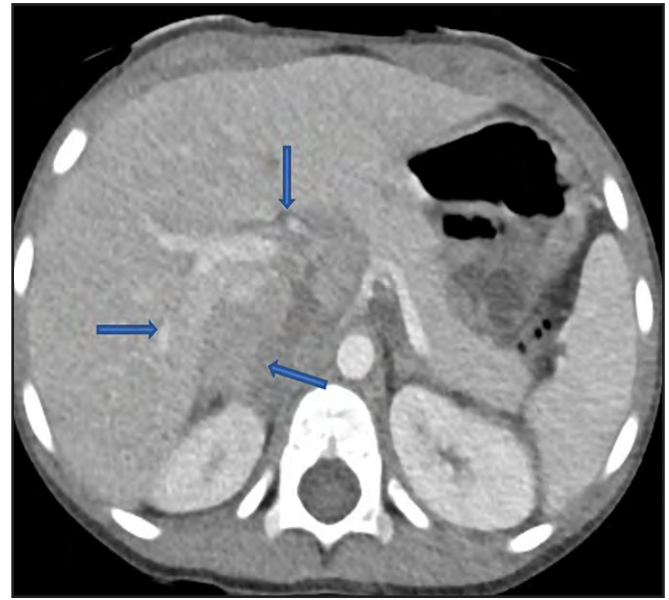


FIGURE 4. Tumor infiltrates (arrows) porta hepatis. IDRF positive.

structure to be lacking or ill defined.”¹⁵ This applies to vital structures other than vessels. An IDRF is present if there is *infiltration*.

Invasion – This term applies to tumor relationship with the renal pedicle, as studies have shown that sur-

gical resection of a tumor with renal invasion often requires nephrectomy or leads to segmental renal infarction.¹⁶ Invasion is present if the tumor is in contact (see above) with the renal artery, renal vein, or renal pelvis. An IDRF is present if there is *invasion*.

Summary of IDRFs Based on Anatomic Location as Defined by the INRG Staging System

Multiple body compartments – Tumors that involve multiple contiguous body compartments are considered IDRF positive (**Figure 1**). Multifocal

primary neuroblastomas are rare. They may be familial or seen in patients with syndrome-associated neuroblastic tumors. When found, multifocality should be documented and each neoplasm should be staged according to the greatest extent of disease. Multifocality alone is not considered IDRF.

Neck – Primary cervical neuroblastoma is uncommon, occurring less than 5% of the time.³ Cervical neuroblastomas arise from the superior cervical sympathetic chain, and tumors that encase the carotid or vertebral arteries, encase the jugular vein, compress the airway, and/or infiltrate the skull base are considered IDRF positive.

Cervicothoracic junction – Similar to cervical neuroblastoma, tumors that encase the major vessels (subclavian, carotid, and/or vertebral) or compress the trachea are considered IDRF positive. Additionally, tumors encasing the brachial plexus roots are IDRF positive.

Thorax – Thoracic neuroblastomas account for approximately 20% of neuroblastomas³ and arise from the paraspinal sympathetic chain in the posterior mediastinum. In addition to encasement of the aorta and trachea, tumors that involve the lower mediastinum, infiltrating the costo-vertebral junction between T9 and T12, are also considered IDRF positive. This is because injury to the anterior spinal artery may result in spinal cord infarction.

Thoraco-abdominal junction – Tumors that encase the aorta and/or vena cava are IDRF positive.

Abdomen and pelvis – The vast majority of neuroblastomas arise from the abdomen and pelvis,³ originating in the

adrenal glands or from the sympathetic ganglia or sympathetic fibers along the aorta and major branches. Vascular involvement is common, and encasement of the origin of the celiac artery and/or superior mesenteric artery, aorta, inferior vena cava, or iliac vessels are considered IDRF positive. As stated above, the risk of renal complications is high, so renal invasion is also considered IDRF positive. Additional IDRFs are listed in **Table 2**.

Spinal – The paraspinal course of the sympathetic ganglion chain commonly enables tumor extension into the spinal canal common. Spinal IDRF positive features include 1) invasion of more than one-third of the spinal canal (on axial plane) (**Figure 2**), 2) perimedullary leptomeningeal spaces that are effaced, and/or 3) abnormal spinal cord signal.

Summary

Neuroblastoma is a common pediatric malignancy with a wide range of biological behaviors, from spontaneous regression to progressive disease and death. Optimal treatment requires personally tailored treatment regimens based on individual patient risk. The INRG staging system is a robust and widely used staging system that incorporates image-defined risk factors to stratify patients in treatment categories according to risk.

REFERENCES

1. Maris JM, Hogarty MD, Bagatell R, Cohn SL. Neuroblastoma. *Lancet* 2007;369:2106-2120.
2. Park JR, Eggert A, Caron H. Neuroblastoma: biology, prognosis, and treatment. *Hematol Oncol Clin North Am* 2010;24(1):65-86.
3. Lonergan GJ, Schwab CM, Suarez ES, Carlson L. Neuroblastoma, ganglioneuroblastoma, and gangli-

oneuroma: radiologic-pathologic correlation. *RadioGraphics* 2002;22:911-934.

4. Cohn SL, Pearson AD, London B, Monclair T. The International Neuroblastoma Risk Group (INRG) classification System: an INRG Task Force report. *J Clin Oncol* 2009;27:289-297.

5. Morris JA, Shochat J, Smith EI, Look AT. Biological variables in thoracic neuroblastoma: a Pediatric Oncology Group study. *J Pediatr Surg* 1995;30:96-302.

6. Park JR, Kreissman SG, London WB, Naranjo A. Effect of tandem autologous stem cell transplant vs single transplant on event-free survival in patients with high-risk neuroblastoma – a randomized clinical trial. *JAMA* 2019;322:746-755.

7. Brodeur GM, Pritchard J, Berthold F, Carlsen NL. Revisions of the International Criteria for Neuroblastoma Diagnosis, Staging, and Response to Treatment. *J Clin Oncol* 1993;11:1466-1477.

8. Monclair T, Brodeur GM, Ambros PF, Brisse JJ. The International Neuroblastoma Risk Group (INRG) Staging System: an INRG Task Force report. *J Clin Oncol* 2009;27:298-303.

9. Chen A, Trout A, Towbin A. A review of neuroblastoma image-defined risk factors on magnetic resonance imaging. *Pediatr Radiol* 2018;1337-1347.

10. Vik TA, Pfluger T, Kadota R, Castel V. 123I-MIBG scintigraphy in patients with known or suspected neuroblastoma: results from a prospective multicenter trial. *Pediatr Blood Cancer* 2009;52:784-790.

11. Matthay KK, Shulkin B, Ladenstein R, Michon J. Criteria for evaluation of disease extent by 123I-metaiodobenzylguanidine scans in neuroblastoma: a report for the International Neuroblastoma Risk Group (INRG) Task Force. *Br J Cancer* 2010;102:1319-1326.

12. Papathanasiou ND, Gaze MN, Sullivan K, Aldridge M. 18F-FDG PET/CT and 123I-metaiodobenzylguanidine imaging in high-risk neuroblastoma: diagnostic comparison and survival analysis. *J Nucl Med* 2011;52:419-525.

13. Sharp SE, Trout AT, Weiss BD, Gelfand MJ. 123I-MIBG Scintigraphy and 18F-FDG PET in neuroblastoma. *J Nucl Med* 2009;50:1237-1243.

14. Lee JW, Cho A, Yun M, Lee JD. Prognostic value of pretreatment FDG PET in pediatric neuroblastoma. *Euro J Radiol* 2015;84:2633-2639.

15. Brisse HJ, McCarville MB, Granata C, Krug KB. Guidelines for imaging and staging of neuroblastic tumors: consensus report from the International Neuroblastoma Risk Group Project. *Radiology* 2011;261:243-257.

16. Shamberger RC, Smith EI, Joshi VV, Rao PV. The risk of nephrectomy during local control in abdominal neuroblastoma. *J Pediatr Surg* 1998;33:161-164.

Abdominal Mass in a Female Pediatric Patient

Tuan Dao, M.D., Hollie Lai, M.D.

Department of Radiology, Children's Hospital of Orange County, Orange, CA

Case Presentation

A previously healthy premenarchal 9-year-old girl presented to the emergency department after the patient's mother felt a nontender abdominal mass that had been noticeably larger and growing for the past week. The patient was initially evaluated by her pediatrician and subsequently had an outpatient abdominal ultrasound that revealed a large abdominal mass (**Figure 1**). Follow-up CT was also ordered by the primary team (**Figure 2**). She had a good appetite, good energy, and was able to participate normally in daily activities. Family history was negative for malignancy.

Key Imaging Findings

Solid mass with intrinsic areas of necrosis in the lower abdomen and pelvis, most likely arising from the adnexa, and ovarian in origin.

Differential Diagnosis

- Germ cell tumors
- Yolk sac tumor
- Dysgerminoma
- Embryonal carcinoma
- Teratoma
- Lymphoma

Discussion

Although ovarian tumors in children are uncommon, with an estimated incidence of 2.6 cases per 100,000 girls per year,¹ enlarging abdominal masses in the premenarchal female pediatric

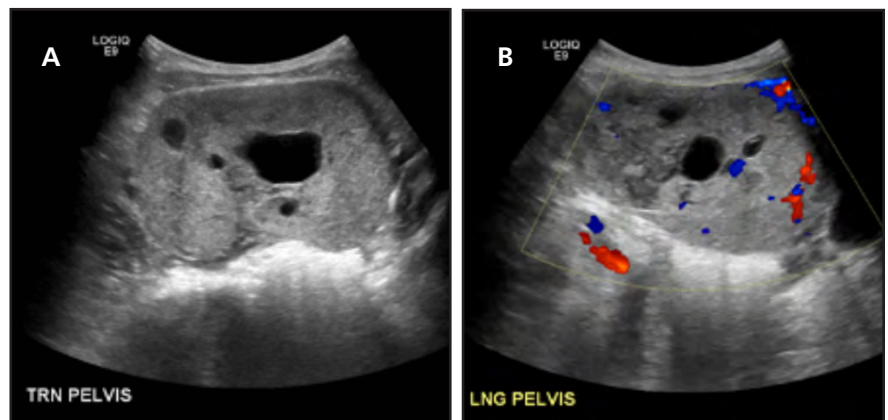


FIGURE 1. Grayscale transverse ultrasound image (A) and grayscale longitudinal ultrasound image with color Doppler (B) of the pelvis demonstrates an echogenic well-margined lobulated mass with internal vascularity and intrinsic anechoic cystic components.

population should raise suspicion for ovarian origin. Sonographic images revealed a large, well-margined, lobulated solid mass with internal cystic components. Further evaluation with contrast-enhanced CT of the abdomen and pelvis demonstrated a large lower abdominal/pelvic mass that appeared to arise from the adnexa. The heterogeneous mass was well-margined and had enhancing solid components and extensive areas of necrosis. Although germ cell tumors (GCTs) share imaging characteristics, the heterogeneous enhancing solid lesion with extensive necrosis, in addition to rapid growth per clinical history, suggested a GCT such as a yolk sac tumor (YST). Dysgerminoma was less favored since fibrovascular septa were not appreciated on the imaging. Teratoma was unlikely given

the lack of calcifications or fat. For ovarian masses, tumor markers such as alpha-fetoprotein (AFP), beta-human chorionic gonadotropin (b-hCG), cancer antigen 125 (CA-125), inhibin A and B, and carcinoembryonic antigen (CEA) can be obtained to aid in diagnosis and for postsurgical follow-up. For this patient, only AFP was significantly elevated at > 20,000 ng/mL. Given imaging findings and elevated AFP, YST was the primary differential diagnosis.

Yolk Sac Tumor

YST, also known as an endodermal sinus tumor, is a rare malignant GCT that usually occurs in the second and third decades of life, with an average age of 19 years.¹ YST is the second most common malignant GCT, and abdominal pain or mass is the most

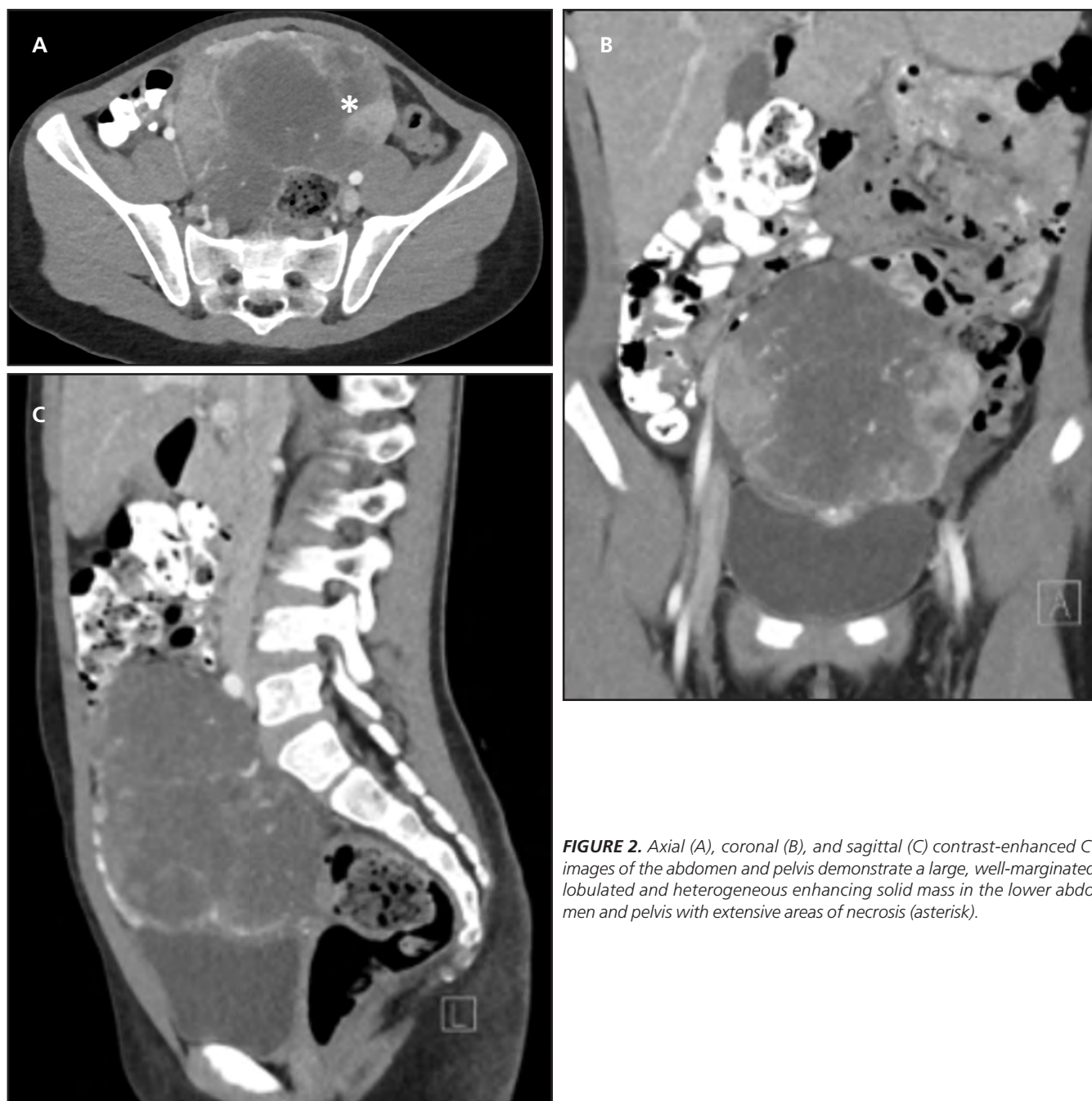


FIGURE 2. Axial (A), coronal (B), and sagittal (C) contrast-enhanced CT images of the abdomen and pelvis demonstrate a large, well-margined, lobulated and heterogeneous enhancing solid mass in the lower abdomen and pelvis with extensive areas of necrosis (asterisk).

common presenting complaint. In early stage disease, they are usually unilateral. Due to their rapid growth, duration of symptoms is usually 1 to 4 weeks.^{4,5} Because the tumor produces AFP, this tumor marker can be used at initial diagnosis and for postoperative follow-up.

Plain films can demonstrate a pelvic mass with mass effect on adjacent bowel loops. Sonography usually shows a large pelvic mass that extends

into the abdomen with both solid and cystic components. Sometimes, concurrent dermoid cyst of the ovary, ascites, or urinary obstruction can be seen as well.⁴ CT and MR imaging often demonstrate a large heterogeneous solid mass with intrinsic cystic components representing necrosis and hemorrhage. MR imaging can better detect intratumoral signal voids and intratumoral hemorrhage.

Pathologic features include extensive areas of hemorrhage and necrosis. Treatment can include conservative surgery with multiagent chemotherapy.

Dysgerminoma

Ovarian dysgerminoma is identical to testicular seminoma, originating from undifferentiated germ cells. It is the most common malignant GCT in children and usually occurs in the second and third

decades of life, although 10% can occur in the first decade. Lactate dehydrogenase (LDH) is a useful marker for diagnosis and follow-up. Rarely, β -HCG can be elevated.

Gross pathology often shows a well-encapsulated and lobulated solid tumor with a white to tan color. Irregular areas of necrosis, cystic changes, or calcifications can be seen. Intrinsic areas of hemorrhage can also exist, as in the case of YST.

Histologically, the tumor is composed of monotonous polygonal tumor cells aggregated into cords and clumps with abundant cytoplasm and flattened central nuclei.¹ Tumor cells are separated by fibrous or fibrovascular septa, which can be seen on imaging.

On imaging, dysgerminomas usually are multilobulated, well-marginated masses with prominent fibrovascular septa. Because of the fibrous component of the septa, these will appear hypointense on both T1- and T2-weighted images on MR imaging.³ In addition, the septa may show intense enhancement. Doppler imaging can show flow signal within the septa.¹ Areas of necrosis, hemorrhage, or speckled calcifications can be seen. As opposed to other malignant GCTs, dysgerminomas can often be bilateral and can spread to retroperitoneal lymph nodes.¹

Embryonal Carcinoma

Embryonal carcinoma occurs primarily in children, with a median age of 14 years. The tumor can produce AFP and β -HCG, which can aid diagnosis and post-treatment follow-up. CT and MR imaging characteristics are nonspecific and share similarities to other malignant GCTs, as detailed above. Again, internal areas of hemorrhage, necrosis, and cystic components are common.

Teratoma

Ovarian teratomas are GCTs composed of mature or immature tissues derived from more than one of the three

primitive embryonic layers (ectoderm, mesoderm, and endoderm). Benign teratomas comprise 67% of pediatric ovarian neoplasms and can be bilateral in up to 25% of cases.²

Mature cystic teratoma (MCT) comprises about 50% of all pediatric ovarian neoplasms. When ectodermal elements predominate, they are called dermoid cysts. MCT is the most common ovarian neoplasm in children. In 10% of pediatric cases, it can be bilateral.¹ Treatment for MCT is surgical excision with fertility preservation.

Imaging characteristics of MCT include a cystic adnexal mass that contains fat and/or calcifications. Ultrasound can reveal areas of echogenicity corresponding to fat, although this can be challenging to definitively determine. Helpful signs include the “tip-of-the-iceberg” sign represented by coarse shadowing calcifications; dermoid mesh seen as linear interfaces corresponding to hair; and a dermoid plug represented by an echogenic nodule with fat, hair, and teeth. However, sometimes CT or MR is needed for further characterization. Fat density can be definitively seen on CT. On MR imaging, hyperintense T1 signal with signal loss on fat-saturated sequences can confirm fat composition. Calcifications are often seen as well.

Immature teratoma affects a younger age group and frequently occurs between ages 10 and 20 with a median age of 17 years. It represents 10% to 20% of ovarian malignancies in the pediatric population.¹ Elevated AFP has been reported in cases of immature teratoma. Imaging usually reveals a unilateral, large, heterogeneous, predominantly solid or mixed solid and cystic mass with scattered calcifications and fat.

Lymphoma and Leukemia

Ovarian lymphoma is rare, accounting for less than 2% of all pediatric ovarian tumors.¹ On CT and MR imaging, ovarian lymphoma is often seen as unilateral or bilateral predominantly solid masses

with homogeneous enhancement and preserved ovarian follicles. Small cysts can be seen along the periphery or within the solid portions.¹ Although signal intensity on MR imaging is variable, leukemic involvement can often demonstrate hypointense T2 signal, likely due to high levels of myeloperoxidase.¹

Diagnosis

Ovarian YST. The patient underwent right salpingo-oophorectomy without complications. The patient was also started on multiagent chemotherapy with subsequent decreasing AFP levels.

Summary

Ovarian tumors are uncommon in the premenarchal female pediatric population. Patients in this age group presenting with abdominal masses should raise suspicion for ovarian neoplasm. Predominantly solid masses arising from the adnexa seen on cross-sectional imaging suggest malignant lesions such as GCTs. Tumor markers are helpful in favoring one GCT over another as imaging characteristics often overlap. Ovarian YSTs are large, heterogeneous, well-marginated solid masses with intrinsic necrosis and hemorrhage. Fibrovascular septa should point toward dysgerminoma. Fat or calcifications suggest an ovarian teratoma. Lymphoma and leukemia can be in the differential diagnosis as well. Treatment includes conservative surgery and multiagent chemotherapy.

REFERENCES

1. Heo SH, Kim JW, Shin SS, et al. Review of ovarian tumors in children and adolescents: radiologic-pathologic correlation. *RadioGraphics* 2014;34(7):2039-2055.
2. Anthony EY, Caserta MP, Singh J, Chen MYM. Adnexal masses in female pediatric patients. *Am J Roentgenol* 2012;198: W426-W431.
3. Jeong YY, Outwater EK, Kang HK. Imaging evaluation of ovarian masses. *RadioGraphics* 2000;20(5):1445-1470.
4. Levitin A, Haller KD, Cohen HL, et al. Endodermal sinus tumor of the ovary: imaging evaluation. *Am J Roentgenol* 1996;167(3):791-793.
5. Choi HJ, Moon MH, Kim SH, Cho JY, Jung DC, Hong SR. Yolk sac tumor of the ovary: CT findings. *Abdom Imaging* 2008;33(6):736-739.

Patellar Bone Lesion

Danny Nahl, D.O., Andrew Sung, M.D.

Department of Radiology, Children's Hospital of Orange County, Orange, CA

Case Presentation

A 14-year-old boy with no significant past medical history presented to the pediatric orthopedic clinic with left knee pain that began four months prior. The patient localized the pain to underneath/inside his kneecap and denied any history of trauma. He reported the pain to be worse with running and jumping activities. There was mild pain with patellar compression. All other focused physical examination findings of the left lower extremity, past medical history, and review of systems were unremarkable.

Radiographs of the left knee were initially obtained (**Figure 1**) followed by MRI of the left knee without and with intravenous contrast (**Figure 2**). CT-guided biopsy was performed for definitive diagnosis.

Differential Diagnosis

Chondroblastoma
Langerhans cell histiocytosis
Osteomyelitis
Chondromyxoid fibroma
Giant cell tumor
Malignant lesions (chondrosarcoma, osteosarcoma, metastases)

Discussion

The patella is a sesamoid bone, embedded within the quadriceps tendon superiorly and patellar tendon inferiorly. The patella demonstrates similar architecture to the epiphyses of long bones and is, therefore, termed an

“epiphyseal equivalent.” Examples of other epiphyseal equivalents include the greater and lesser trochanters of the femur, the carpal bones, calcaneus, as well as most apophyses. Knowledge of this information helps narrow the differential diagnostic considerations for lesions typically encountered in the epiphysis.

Chondroblastoma

Chondroblastoma is a benign cartilaginous neoplasm typically seen in the epiphyses of long bones or epiphyseal equivalents. Although benign, the tumor is locally aggressive and typically produces surrounding edema of the bone marrow. On radiographs, typical features are of a well-defined osteolytic lesion with thin peripheral sclerotic border. They typically arise eccentrically and may extend to the metaphysis, although breach of the cortex is rare. Internal calcifications may be present, and a joint effusion may be seen. Periosteal reaction distal to the lesion (along the diaphysis when the lesion is in the epiphysis) distinguishes chondroblastoma from other epiphyseal lesions.¹ CT shows similar findings to radiographs, with better delineation of extent, periosteal reaction, and internal matrix. MRI is optimal for evaluating transphyseal extent and degree of surrounding marrow edema. Chondroblastoma typically demonstrates isointense T1 signal, variable hyperintense T2 signal within the lesion, hyperintense surrounding STIR signal

due to edema, and moderate heterogeneous enhancement with enhancement of the surrounding bone and soft-tissue edema.² A thin hypointense rim with lobulated margins is characteristic. Fluid-fluid levels can be seen due to association with secondary aneurysmal bone cyst (ABC), which have been reported to occur with chondroblastoma.

Langerhans Cell Histiocytosis

The skeletal system is the most common site for Langerhans cell histiocytosis (LCH). LCH in the skeletal system is often referred to as eosinophilic granuloma. LCH primarily occurs in older children and young adults and may occur anywhere in the skeletal system with variable appearance. The imaging features on radiographs depend on the phase of disease. Features in long bones range from permeative appearance to focal lytic lesions with endosteal scalloping, cortical thinning, and periosteal reaction. An associated soft-tissue mass is often present. On MRI, LCH typically demonstrates hypointense to isointense T1 signal, hyperintense T2/STIR signal, and avid enhancement with or without enhancing soft-tissue component. Diaphyseal location is most common in long bones.⁴

Osteomyelitis

Pyogenic osteomyelitis with a Brodie abscess may mimic certain bone lesions. On radiographs, the typical appearance is of a round lucent lesion with



FIGURE 1. AP (A), lateral (B), and merchant (C) views of the left knee demonstrate a well-circumscribed osteolytic lesion centered in the patella. There is a thin rim of sclerosis. There is no periosteal reaction or joint effusion.



FIGURE 2. Sagittal T1- (A), axial T1- (B) weighted images of the left knee demonstrate the well-margined lesion centered in the patella. Lesion demonstrates isointense T1 signal with surrounding hypointense marrow signal. On the sagittal STIR (C) images the lesion demonstrates mildly hyperintense signal with surrounding hyperintense marrow signal consistent with edema. Sagittal (D) and axial (E) T1 post contrast with fat suppression demonstrate homogeneous enhancement of the lesion and enhancement of the surrounding marrow.

or without peripheral sclerosis depending on the time at which the bone is imaged in the course of infection. On MRI, the abscess demonstrates variable T1 signal and central increased T2 signal. On postcontrast images, enhancement is usually peripheral with enhancement of the surrounding marrow, and nonenhancement of the purulent core. Diffusion-weighted sequences, if performed, would demonstrate central diffusion restriction due to pus. Location is often an important distinguishing feature, as hematogenous osteomyelitis in older children tends to affect metaphysis/metaphyseal equivalents.⁶ In younger patients (infants and toddlers), epiphyseal location of infection is more common due to transphyseal vascular supply.

Chondromyxoid Fibroma

Chondromyxoid fibroma is an extremely rare, benign cartilaginous neoplasm that most commonly occurs in the second and third decades of life. It is typically located in the metaphysis of long bones and may extend to the epiphysis but is almost never exclusively epiphyseal. On radiographs, they are typically eccentrically located lobular lesions with a well-defined sclerotic margin. They are often expansile with geographic bone destruction. MRI features are nonspecific and generally include low T1 signal, intermediate to high T2 signal, and peripheral nodular enhancement on postcontrast images.⁵

Giant Cell Tumor

Giant cell tumor (GCT) is usually a benign tumor that typically arises in the metaphyses of skeletally mature patients. Epiphyseal location of the tumor is most often due to extension along the fused physis, helping distinguish this entity from other epiphyseal-based lesions. Four characteristic features of GCT in a long bone include occurrence in skeletally mature patients, abutment of the articular surface, being well-defined without sclerotic margins, and eccentric location. The overlying cor-

tex is often thinned, and periosteal reaction is rare. Association with ABC formation is documented.³ On MRI, GCT demonstrates low to intermediate T1 signal, heterogeneously hyperintense T2 signal, and enhancement of the solid components.³ Perilesional edema is typically much less than that seen in chondroblastoma. It is a rare tumor, comprising 1% to 2% of all primary bone tumors. However, it is the most common neoplasm of the epiphysis in skeletally mature patients. The typical presentation is localized pain, although due to its epiphyseal location, it may trigger synovial reaction.

Malignant Bone Lesions

Malignant bone lesions, both primary and metastatic, are rare in the epiphyses. Osteosarcoma usually occurs in the metaphyseal-diaphyseal region of long bones and may extend into the epiphysis. Osteoid matrix, permeative growth, and cortical destruction are typical features.⁷ Epiphyseal clear cell chondrosarcoma is a subtype of chondrosarcoma that may be found in the epiphysis. This tumor, however, usually affects an older population (third to fifth decades) and generally produces little to no surrounding marrow edema.⁸ While bony metastatic lesions can have a variable appearance depending on the primary tumor, epiphyseal hematogenous metastasis is rare and, therefore, would be considered unlikely when encountering a solitary epiphyseal lesion.

Diagnosis

Chondroblastoma

Discussion

When encountering lesions of the patella, the differential diagnosis is that of epiphyseal lesions. While tissue sampling is necessary for definitive diagnosis, it is important to account for the patient's age as well as specific imaging features to help narrow the differential. Chondroblastoma is the most common epiphyseal lesion in a

skeletally immature patient. Both chondroblastoma and GTC may involve the epiphysis and have an associated ABC component, but GCT classically enters the epiphysis by crossing a fused physis. Primary malignant tumors and metastases of the epiphysis/patella are rare. Langerhans cell histiocytosis may cause cortical breach and soft-tissue component, distinguishing it from chondroblastoma. Presence of surrounding marrow edema is a hallmark of chondroblastoma. Infection with Brodie's abscess may have a similar appearance, in which case clinical findings are helpful in differentiating the two. Additional considerations for patellar abnormalities in the pediatric population include osteochondral lesions and dorsal defect of the patella.

Treatment for chondroblastoma typically involves surgical curettage and packing. Radiofrequency ablation may also be used.⁹ Complete eradication of lesions may be difficult due to proximity to the growth plate; thus, lesions may recur and cause growth arrest with resultant limb-length discrepancy.

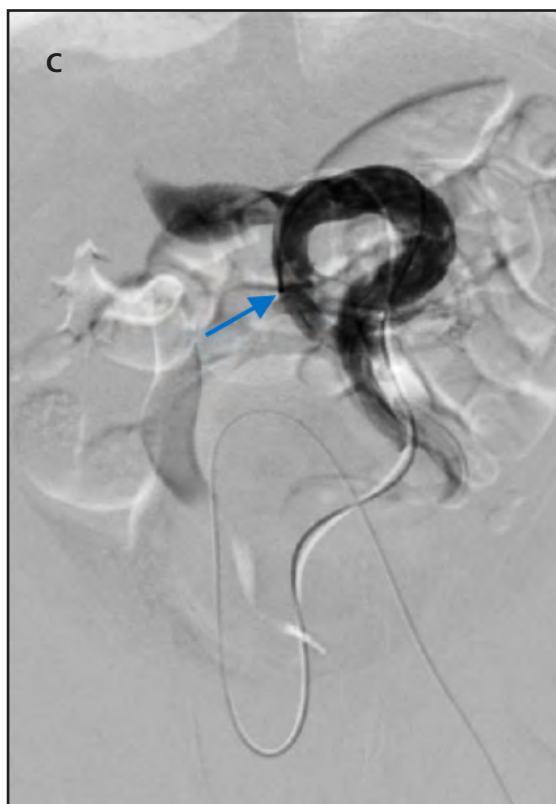
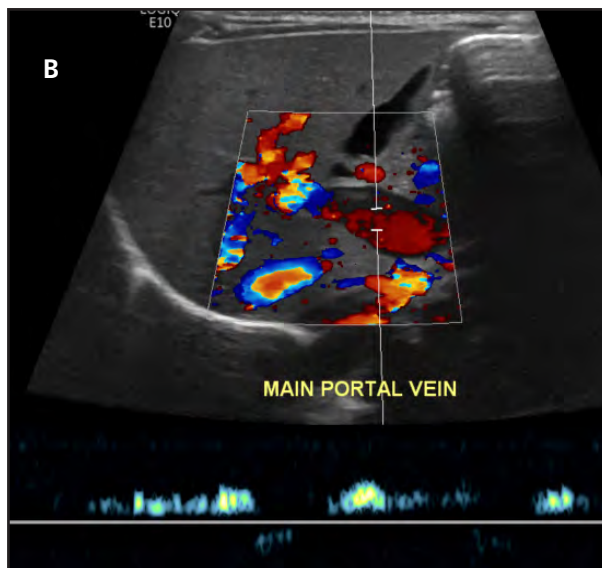
REFERENCES

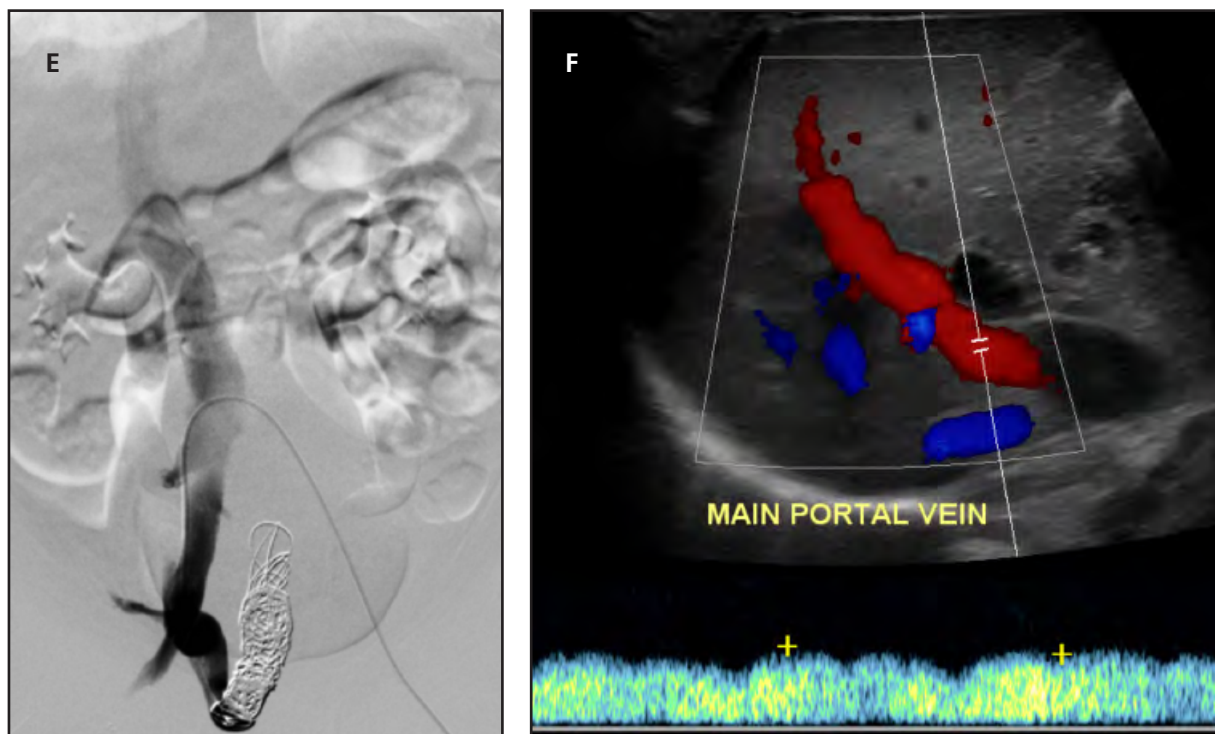
1. Brower AC, Moser RP, Kransdorf MJ. The frequency and diagnostic significance of periostitis in chondroblastoma. *Am J Roentgenol* 1990;154(2):309-314.
2. Oxtoby JW, Davies AM. MRI characteristics of chondroblastoma. *Clin Radiol* 1996;51(1):22-26.
3. Stacy GS, Peabody TD, Dixon LB. Mimics on radiography of giant cell tumor of bone. *Am J Roentgenol* 2003;181(6):1583-1589.
4. David R, Oria RA, Kumar R, et al. Radiologic features of eosinophilic granuloma of bone. *Am J Roentgenol* 1989;153(5):1021-1026.
5. Eisenberg RL. Bubbly lesions of bone. *Am J Roentgenol* 2009;193(2):W79-W94.
6. Gold RH, Hawkins RA, Katz RD. Bacterial osteomyelitis: findings on plain radiography, CT, MR, and scintigraphy. *Am J Roentgenol* 1991;157(2):365-370.
7. Murphey MD, Robbin MR, McRae GA, Flemming DJ, Temple HT, Kransdorf MJ. The many faces of osteosarcoma. *RadioGraphics* 1997;17(5):1205-1231.
8. Murphey MD, Walker EA, Wilson AJ, Kransdorf MJ, Temple HT, Gannon FH. From the archives of the AFIP: imaging of primary chondrosarcoma: radiologic-pathologic correlation. *RadioGraphics* 2003;23(5):1245-1278.
9. Christie-Large M, Evans N, Davies AM, James SLJ. Radiofrequency ablation of chondroblastoma: procedure technique, clinical and MR imaging follow up of four cases. *Skeletal Radiol* 2008;37(11):1011-1017.

JAOCR at the Viewbox

Tammam Beydoun, D.O., Daniel Nahl, D.O.

Department of Radiology, Children's Hospital of Orange County, Orange, CA





Congenital Extrahepatic Portosystemic Shunt (Abernathy Malformation)

A 3-day old girl, born at 36 weeks' gestation, presented with a cystic lesion of the pelvis seen on prenatal ultrasound and fetal MRI. On day one of life, an MRI/MRA of the abdomen/pelvis was obtained (**A**) and demonstrated a large abnormal vessel extending from the liver caudally with the tortuous path toward the right internal iliac vein (**arrows**). A large portosystemic shunt was suspected. A subsequent echocardiogram showed elevated right heart pressures. Abdominal ultrasound (**B**) was also performed demonstrating a “to and fro” flow within the main portal vein.

The patient was taken to the interventional radiology suite for a more detailed evaluation of the shunt. Left common femoral vein approach was chosen and accessed using a 21-gauge needle and microcatheter system without sheath. Through the right internal iliac vein, the shunt was accessed with retrograde navigation toward the liver. Contrast injection from the superior mesenteric vein (**C**, **blue arrow**) demonstrates a high-flow tortuous shunt directing flow away from the fusiform main portal vein to the right internal iliac vein (**D**, **green arrow**). Note the stasis of contrast within the dysgenic portal system (**D**, **yellow arrow**) demonstrating fusiform main portal vein and mildly hypoplastic portal veins.

The pressure gradient was measured without and with manual external pressure occluding the shunt to evaluate whether performing an embolization procedure would be safe. Occluding a shunt with a pressure gradient greater than 10 mmHg carries a risk of acute portal hypertensive complication.¹ The gradient measured 6 mmHg and the shunt was successfully occluded with coils (**E**). Liver ultrasound performed the following day demonstrated monophasic antegrade/hepatopedal flow (**F**).

Congenital portosystemic shunts (Abernathy malformations) were described by Dr. Abernathy in 1793 and result from persistence of embryonic vessels. With advances in imaging, earlier identification of these congenital anomalies has allowed for potential earlier intervention. When left untreated, they may go unnoticed for years presenting as heart failure or hepatic encephalopathy.² Smaller shunts may close spontaneously¹ and can be monitored in the absence of acute sequelae such as heart failure.

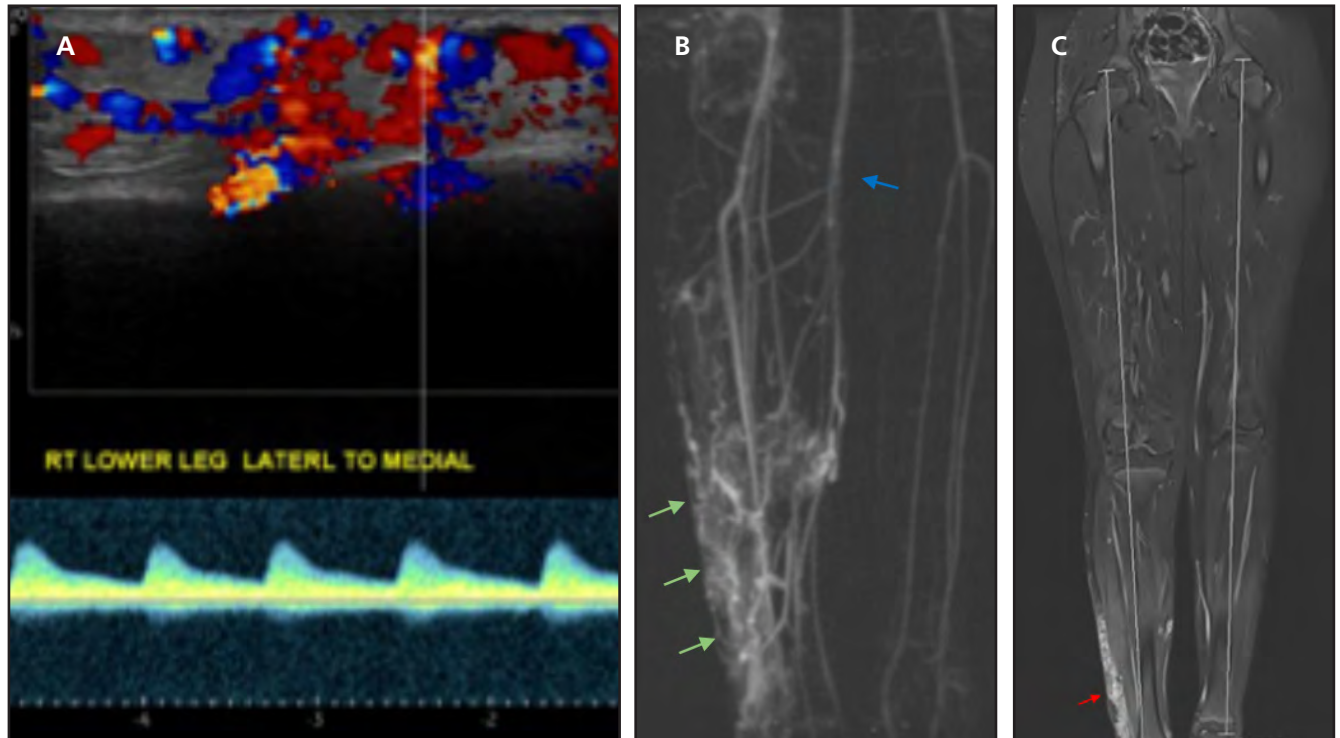
REFERENCES

1. Rajeswaran S, Johnston A, Green J, et al. Abernathy malformations: evaluation and management of congenital portosystemic shunts. *J Vasc Interv Radiol* 2020;31(5):788-794.
2. McLin VA, Franchi Abella S, Debray D, et al. Congenital portosystemic shunts: current diagnosis and management. *J Pediatr Gastroenterol Nutr* 2019;68(5):615.

JAOCR at the Viewbox

Tammam Beydoun, D.O., Tuan Dao, M.D.

Department of Radiology, Children's Hospital of Orange County, Orange, CA



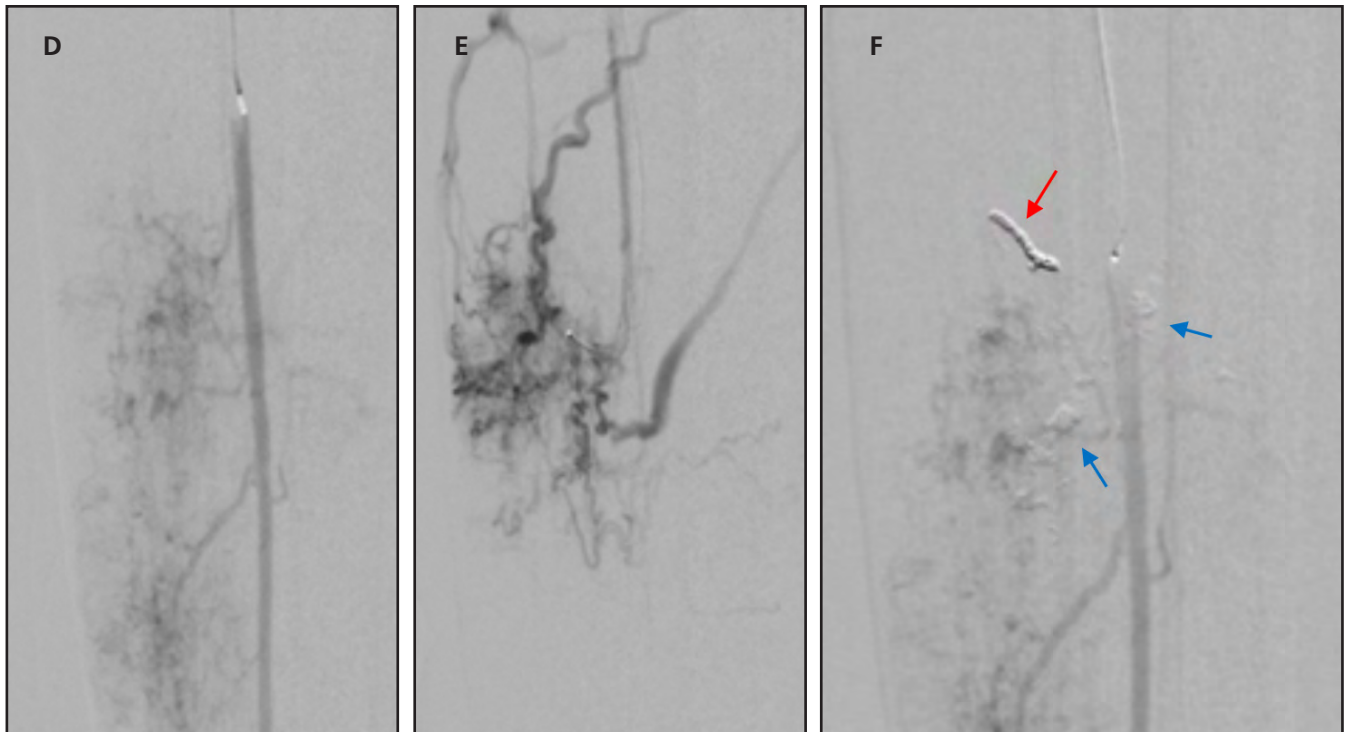
Parkes Weber Syndrome

A 10-year-old girl with no significant prior medical history was referred from her pediatrician to the interventional radiology clinic for persistent right lower extremity swelling and enlarging veins predominantly about her right shin. Ultrasound demonstrated multiple enlarged superficial veins (A) with arterialized waveforms, which was initially reported as a traumatic AV fistula.

A more thorough physical examination demonstrated two small 2- to 3-cm flat reddish-purple lesions on the patient's right posterior upper thigh and gluteal fold as well as a subtle 3-cm limb-length discrepancy. Mild right leg hypertrophy was initially thought to be secondary to swelling by the pediatrician, but likely represents limb overgrowth.

MRI of the right lower extremities was performed demonstrating the extent of disease, particularly with the time-resolved MRA sequence (TWIST) (B), which gives an overview of the numerous arteriovenous fistulous communications with capillary components (**green arrow**) and early draining embryonic sciatic vein (**blue arrow**). Coronal STIR sequence (C) showed mild leg-length discrepancy and part of the capillary malformation component in the lower leg (**red arrow**). The patient indicated persistent pain in her right shin and was taken to the interventional suite for further evaluation and possible embolization of the arteriovenous fistula.

Antegrade access was obtained through the right common femoral artery and angiography of the anterior tibial artery (D, E) was performed showing innumerable arteriovenous communications predominantly about the knee and shin, confirming suspicion for Parkes Weber syndrome (PWS).



Arteriovenous malformation at the symptomatic focus of pain was subsequently embolized (F) with coils from an arterial approach (**red arrow**) and n-Butyl cyanoacrylate glue (**blue arrow**) from a retrograde approach by direct puncture through the symptomatic superficial vein. The patient's local symptoms improved postoperatively.

PWS is a rare vascular anomaly that results in numerous abnormal vessels within the affected region and can clinically present with usually regional small capillary malformations on the face or limb in addition to overgrowth of the affected region/limb.¹ PWS is distinguished from Klippel-Trenaunay syndrome with presence of arteriovenous fistulas. Relatively recent association of PWS with RASA1 genetic mutation^{2,3} has opened a pathway for promising pharmacologic approaches to therapy. Nonetheless, at this time, overall progression of disease is expected and endovascular therapy of complex arteriovenous fistulas is predominantly focused on symptomatic relief of focal pain, bleeding or ulceration.

REFERENCES

1. Kumar V, Jorwal P, Biswas A, Deorari V. Parkes Weber syndrome. QJM 2019;112(12):936.
2. Monroe EJ. Brief description of ISSVA classification for radiologists. Tech Vasc Interv Radiol 2019;22(4):100628.
3. Boccara O, Eyries M, Pannier S, Ariche-Maman S, Hadj-Rabia S, Coulet F. Parkes-Weber syndrome related to RASA1 mosaic mutation. Clin Genet 2021;99(2):330-331.