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
William T. O’Brien, Sr., D.O.
San Antonio, TX

Design Editor
Jessica Roberts
Communications Director, AOCR

Managing Editor
Tammam Beydoun, D.O.
Farmington Hills, MI

Editorial Board
Susann Schetter, D.O.
Daniel J. Abbis, D.O.
Les R. Folio, D.O.
Michael W. Keleher, D.O.
Rocky Saenz, D.O.
Kipp A. Van Camp, D.O.
John Wherthey, D.O.
### Table of Contents

**Neuroimaging**

Editor-in-Chief: William T. O’Brien, Sr., D.O.

<table>
<thead>
<tr>
<th>Title/Author(s)</th>
<th>Page No.</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>From the Editor</strong></td>
<td>1</td>
</tr>
<tr>
<td><strong>Review Articles</strong></td>
<td></td>
</tr>
<tr>
<td>Imaging of Primary Posterior Fossa Brain Tumors in Children</td>
<td>2</td>
</tr>
<tr>
<td>William T. O’Brien, Sr., D.O.</td>
<td></td>
</tr>
<tr>
<td>Intradural Spinal Neoplasms: A Case Based Review</td>
<td>13</td>
</tr>
<tr>
<td>Bradley J. Carra, M.D., Paul M. Sherman, M.D.</td>
<td></td>
</tr>
<tr>
<td><strong>Case Reports</strong></td>
<td></td>
</tr>
<tr>
<td>Expansile Lesion of the Petrous Apex</td>
<td>22</td>
</tr>
<tr>
<td>Jason Romesburg D.O., Michael E. Zapadka D.O.</td>
<td></td>
</tr>
<tr>
<td>Enhancing Sellar/Suprasellar Mass in an Adolescent</td>
<td>27</td>
</tr>
<tr>
<td>Jessika Chumak, M.D., Anthony I. Zarka, D.O.</td>
<td></td>
</tr>
<tr>
<td><strong>JAOCR at the Viewbox</strong></td>
<td></td>
</tr>
<tr>
<td>Balo Concentric Sclerosis</td>
<td>30</td>
</tr>
<tr>
<td>Shannon Gaffney, D.O., William T., O’Brien, Sr., D.O.</td>
<td></td>
</tr>
<tr>
<td>Glomus Vagale</td>
<td>31</td>
</tr>
<tr>
<td>Anthony I. Zarka, D.O.</td>
<td></td>
</tr>
<tr>
<td>Caudal Regression Syndrome</td>
<td>32</td>
</tr>
<tr>
<td>Lauren May, M.D., William T., O’Brien, Sr., D.O.</td>
<td></td>
</tr>
</tbody>
</table>
The JAOCR is now in its second year of publication and has made tremendous strides since its inception. Over the past year, the Journal has been approved for Category 1-B CME credits by the AOA and is now indexed through Google Scholar™; an application for indexing with Index Copernicus® is also currently under review. Our next goal is to submit for indexing in PubMed® once we are eligible in terms of the number of articles required. This would be an important milestone for the Journal, since only a handful of submissions are initially approved, and a Journal may only apply once every two to three years. I have no doubt that the scientific content of the Journal merits this prestigious indexing; hopefully, the reviewers will agree.

None of the accomplishments of the Journal would be possible without the behind-the-scenes work of the AOCR staff, particularly Ms. Jessica Roberts and Tammam Beydoun, D.O. These individuals spend countless hours copyediting and formatting each article in preparation for final publication. Ms. Pam Smith, AOCR Executive Director, has been crucial in identifying and coordinating with guest editors for the first round of issues prior to her well-deserved retirement. This Journal would not be possible without them.

The continued success of the JAOCR ultimately relies on the contributions from the members of the AOCR and their colleagues, since without contributors the Journal would cease to exist. If any individuals are interested in serving as guest editors for a subspecialty issue, please contact the AOCR at info@aocr.org. Guest editors recruit high-quality review articles, differential-based case reports, and Viewbox articles from experts in their subspecialty field. Guest editors then provide initial review and editing of each article prior to submission to the JAOCR. Although invited, all articles are subject to peer review and final acceptance by the JAOCR editorial staff.

It is a privilege to present the second Neuroimaging subspecialty issue of the JAOCR. In this issue, we have comprehensive review articles covering primary posterior fossa brain tumors in children and intradural spinal neoplasms. The differential-based case reports include high-yield reviews of suprasellar masses in adolescents and lesions of the petrous apex. The final Viewbox section includes images from interesting cases with short captions. I would like to thank the residents, fellows, and staff at Wilford Hall USAF Ambulatory Surgical Center in San Antonio, TX, and Wake Forest Baptist Health in Winston-Salem, NC, for contributing their time and expertise to this issue.

I hope that you find this issue of the JAOCR informative, practical, and educational. The contributors thoroughly enjoyed pooling their academic resources to put it together.
Introduction

Brain tumors represent the most common solid neoplasm in children and second most common pediatric malignancy overall, following leukemia. Outside of infancy, the majority of primary childhood brain tumors occur in the infratentorial compartment and include medulloblastoma, juvenile pilocytic astrocytoma (JPA), ependymoma, and brainstem/pontine glioma; atypical teratoid rhabdoid tumor (ATRT) is an additional rare but important primary brain tumor of early childhood.

Children with posterior fossa brain tumors typically present with signs and symptoms related to increased intracranial pressure, gait disturbances, and/or cranial nerve deficits, depending upon the type, size, and location of the tumor. CT and MR evaluation is critical in the work-up, management, and follow-up of these patients. This article aims to provide an overview of the imaging manifestations and appearances of the most common primary posterior fossa brain tumors in children.

Medulloblastoma

Background.

Medulloblastoma is a highly malignant (grade IV) embryonal cerebellar tumor which occurs most frequently in children but may also affect adults. It is the second most common pediatric brain tumor overall, following astrocytoma, but is the most common pediatric posterior fossa tumor, accounting for up to 40% of cases. In the pediatric population, there are two age peaks – one at 3 and one at 7 years of age; in adults, the peak age of presentation is between 20 and 40 years of age. Boys are affected more often than girls by a ratio of greater than 2:1. There is an association and increased incidence in the setting of basal cell nevus, or Gorlin, syndrome.

Medulloblastomas are categorized into various pathologic subtypes, including classic, desmoplastic, extensively nodular, large cell, and anaplastic. The classic subtype is by far the most common. An additional rare subtype is the melanotic medulloblastoma, which appears similar to a classic medulloblastoma on imaging but has melanotic tint on gross inspection. The vast majority of medulloblastomas (85-90%) arise from the midline cerebellar vermis dorsal to the fourth ventricle. Approximately 10-15% occur within the cerebellar hemispheres, which is more common in older patients with the desmoplastic variant. More aggressive variants may be infiltrative and invade the fourth ventricle and adjacent brainstem or cerebellar parenchyma.

Children with medulloblastomas most often present clinically with a relatively rapid onset of symptoms - over the course of weeks or a few months - due to the rapid growth and malignant features of the neoplasm. The most common symptoms include headaches and nausea/vomiting due to obstructive hydrocephalus; truncal ataxia and papilledema are common clinical findings. Tumoral seeding of the cerebral spinal fluid (CSF) is present in approximately one-third of cases at the time of initial diagnosis.

Figure 1. Medulloblastoma. Unenhanced axial CT image reveals a well-circumscribed midline hyperdense posterior fossa mass with small hypodense cystic regions. There is compression of the 4th ventricle with obstructive hydrocephalus, as evidenced by enlargement of the third (arrow) and visualized portions of the lateral ventricles.
Posterior Fossa Tumors, O’Brien

Imaging Findings.

On CT, medulloblastomas typically present as well-defined, midline posterior fossa masses. The high cellularity with increased nuclear-to-cytoplasmic ratio leads to increased attenuation on unenhanced CT in approximately 90% of cases (Fig. 1); the remainder are isodense to brain parenchyma. Surrounding parenchymal vasogenic edema is noted in >90% of cases. After contrast administration, there is often avid enhancement which may be homogeneous or heterogeneous. Calcifications are seen in approximately 20% of cases; cysts are more common and occur in 50-60% of cases. As the lesions grow, there is anterior displacement and compression of the fourth ventricle, which often leads to obstructive hydrocephalus in approximately 90% of cases. If uncompensated, transependymal flow of CSF may be seen along the margins of the lateral ventricles.

On MRI, the majority of medulloblastomas are iso- to hypointense compared to white matter on T1 sequences and variable in signal on T2 sequences. The T2 signal variability has to do with the cellularity of the tumor. More cellular components are hypointense, while less cellular components are iso- to mildly hyperintense compared to white matter. Increased cellularity also leads to increased signal intensity on diffusion weighted sequences; although helpful, this finding has some overlap with other posterior fossa tumors. Medulloblastomas commonly demonstrate avid but heterogeneous enhancement on MRI (Fig. 2). Regions of CSF dissemination present with focal (more common) or diffuse leptomeningeal enhancement, which is typically nodular (Fig. 3). MR spectroscopy demonstrates a tumoral spectra with increased choline and decreased N-acetyl aspartate (NAA), along with decreased creatine. Lipid-lactate doublets may also be seen. An elevated taurine peak has recently been shown to be a specific MRS finding for medulloblastoma (Fig. 4).

Medulloblastomas grow in a circumferential pattern and maintain rounded borders. Unlike ependymomas, which are soft and pliable, medulloblastomas infrequently extend through CSF outlet foramina. When aggressive, however, medulloblastomas may be more infiltrative and invade the fourth ventricle, brainstem, and/or adjacent cerebellar parenchyma.

Preoperatively, it is critical to evaluate the entire neuroaxis on MRI to look for disseminated disease (Fig. 5). Failure to do so could complicate management decisions. Postoperatively, surveillance imaging of the brain and spine is typically performed at 3-6 months intervals for the first 5 years following initial therapy to evaluate for early recurrence or new CSF dissemination. Although the long-term impact of surveillance imaging on overall survival is debated, early detection of new or recurrent disease may alter clinical management.

Treatment.

Treatment options depend upon the age of the patient and extent of disease and include surgical resection with radiation and/or chemotherapy. Although medulloblastomas are relatively radiation-
sensitive, radiation therapy is typically avoided prior to 3 years of age due to the potentially devastating effects of craniospinal radiation to the developing CNS. The primary exceptions are in the presence of known CSF dissemination or with tumor recurrences. Neoadjuvant chemotherapy has produced promising results; its use prior to or after surgical resection has allowed for avoidance of craniospinal radiation in many cases of patients under 3 years of age and decreased the overall radiation dose needed to minimize the likelihood of tumor dissemination or recurrence. For children over 3 years of age, craniospinal radiation is typically performed after surgical resection – with or without adjuvant chemotherapy.

Prognosis.

The prognosis for medulloblastoma is based upon age at the time of presentation, presence or absence of CSF dissemination, and residual tumor following initial surgical resection. The best prognosis (approximately 80% 5-year survival) is for children greater than 3 years of age and adults at the time of presentation with no evidence of CSF dissemination and no or minimal residual post-operative disease. The presence of CSF dissemination at the time of diagnosis drops the 5-year survival rate to between approximately 30 and 50%.
Cerebellar Juvenile Pilocytic Astrocytoma (JPA)

Background.

JPAs are low grade neoplasms (WHO grade I) which occur most often in children and young adults. Common locations include the cerebellum, optic pathways, and hypothalamus. Supratentorial hemispheric involvement is more common in adults than children. There is an association and increased incidence of JPAs in the setting of NF-1, particularly involving the optic pathways. JPAs are the most common subtype of pediatric gliomas and account for 85% of all cerebellar astrocytomas;19 diffuse cerebellar astrocytomas are less common variants. Cerebellar JPAs represent approximately one-third of posterior fossa tumors in children, second in incidence only to medulloblastoma. The peak age of presentation is between 5 and 15 years of age, and boys and girls are affected equally.

Patients with cerebellar JPAs typically present with a gradual onset of symptoms due to slow growth of the tumor. Common presenting symptoms include headache, nausea and vomiting, gait imbalance, and visual disturbances. Common clinical findings include truncal ataxia and papilledema (due to increased intracranial pressure).

Imaging Findings.

Posterior fossa JPAs demonstrate imaging features, such as enhancement patterns and MR spectra, which are incongruent with their biological nature, meaning that some of their imaging features are more often associated with higher grade tumors, rather than a low grade astrocytoma. JPAs may present as midline or off-midline (more common) masses due to vermician or hemispheric involvement, respectively. They are typically well-circumscribed, round or ovoid, and have a large cystic component with a mural nodule. A less common appearance is a solid component peripherally with central necrosis.

On CT, the cystic component is hypodense and the solid nodular component is isodense compared to surrounding brain parenchyma. Adjacent parenchymal edema may occur but is less common due to the indolent nature of JPAs. With contrast administration, there is avid enhancement of the solid nodule and occasionally the walls of the cyst; enhancement of the cyst wall suggests but is not diagnostic of the presence of tumor cells within the cyst wall lining. A less common appearance is solid peripheral enhancement with central necrosis. Larger masses result in compression and obstruction of the fourth ventricle with associated hydrocephalus. If uncompensated, transependymal flow of CSF may be seen along the margins of the lateral ventricles.

On MRI, the cystic component is hypointense on T1 and hyperintense on T2 sequences, similar to CSF signal intensity. The solid components are T1 hypointense and T2 hyperintense compared to surrounding brain parenchyma. Vasogenic edema, when present, is relatively mild compared to the size of the lesion and is T2 hyperintense. After contrast administration, there is avid enhancement of the solid

Figure 6. Juvenile Pilocytic Astrocytoma. Axial T2 MR image (A) demonstrates a large cystic mass with solid mural nodule centered within the right cerebellar hemisphere. There is surrounding vasogenic edema and compression of the 4th ventricle. Post-contrast T1 image (B) reveals avid enhancement of the solid nodule.
nodular component (Fig. 6). As discussed above, enhancement of the cyst wall may occasionally be seen and is suggestive but not diagnostic of tumor cells lining the cyst wall (Fig. 7). Rarely, the nodule of a JPA may show increased signal on DWI; however, the high ADC will typically allow differentiation from high grade differentials, such as medulloblastoma. On MR spectroscopy, JPAs have a spectra which may be confused with higher grade tumors – increased choline/creatine due to very low creatine levels, decreased N-acetylaspartate (NAA), decreased myo-inositol, and increased lactate. Obstructive hydrocephalus will lead to ventriculomegaly proximal to the obstruction with transependymal flow of CSF, if uncompensated; the transependymal flow presents as a rind of increased T2 signal intensity along the margins of the lateral ventricles.

**Treatment.**

Surgical resection of cerebellar astrocytomas is the primary treatment and is considered curative in the setting of gross total resection. Repeat surgical resection is the treatment of choice for recurrences or regrowth after subtotal resection. Radiation and/or chemotherapy is reserved for rare instances of disseminated disease or in cases which are not amenable to surgical resection.

**Prognosis.**

The overall prognosis for cerebellar astrocytomas is excellent and is primarily based upon the lesion location and presence or absence of neurological deficits at the time of presentation. The 10-year survival rate is greater than 90%; complete surgical resection is generally considered curative.

**Ependymoma**

**Background.**

Ependymomas arise from ependymal cells which line the ventricles and the central canal of the cord. They may occur at any age but are most common in children and young adults, especially when located within the posterior fossa/fourth ventricle. Approximately 70% of ependymomas are infratentorial, and they are the third most common pediatric posterior fossa tumor, following medulloblastoma and cerebellar juvenile pilocytic astrocytoma. They occur along the floor (more common) or roof of the fourth ventricle. The mean age of presentation for infratentorial ependymomas is 6 years of age, and there is a slight male predominance. Supratentorial intraventricular and intraparenchymal (extraventricular) ependymomas are more common in adults.

The majority of ependymomas are WHO grade II tumors; a more aggressive anaplastic variant is less common. Ependymomas are soft, pliable tumors which have a propensity for spread through ventricular outlet foramina, which is fairly characteristic. CSF dissemination at initial presentation is estimated in approximately 12% of cases and is more common with...
more aggressive histology.\textsuperscript{23,25}

Children with ependymomas most often present clinically with symptoms related to increased intracranial pressure due to hydrocephalus; the most common symptoms include headaches, nausea/vomiting, and gait imbalance; truncal ataxia and papilledema are the most common clinical findings.

Imaging Findings.

On CT, posterior fossa ependymomas appear as fourth ventricular masses with solid components which are typically isodense to mildly hyperdense compared to surrounding brain parenchyma.\textsuperscript{23} Lesions result in obstructive hydrocephalus with nausea and vomiting in roughly 90\% of patients.\textsuperscript{23,26} If uncompensated, transependymal flow of CSF may be seen along the margins of the lateral ventricles. Compared to medulloblastoma, ependymomas are more heterogeneous; calcifications are noted in roughly 50\% of cases, cysts in approximately 20\% of cases, and hemorrhage in 10\% of cases.\textsuperscript{23} Solid components avidly but heterogeneously enhance. Ependymomas are soft and pliable, insinuating along tissue margins and through foramina; extension through the foramina of Luschka and/or Magendie is highly characteristic, although not pathognomonic. They may also encase surrounding nerves and vessels.

On MRI, posterior fossa ependymomas are iso- to hypointense on T1 and hyperintense on T2 sequences compared to surrounding brain parenchyma and demonstrate heterogeneous enhancement of solid components (\textbf{Fig. 8}). Signal characteristics of solid components are more heterogeneous compared to medulloblastoma, especially with foci of calcification, hemorrhage, or cystic change. More cellular components of ependymomas may show increased signal on DWI sequences; ADC maps are typically intermediate in signal with some overlap compared to medulloblastomas.\textsuperscript{20} Ependymomas fill and obstruct the fourth ventricle as they growth, often resulting in hydrocephalus. Extension through fourth ventricular outlet foramina and encasement of surrounding structures is better depicted on MRI compared to CT (\textbf{Figs. 9 and 10}). MR spectroscopy often reveals a nonspecific tumor spectra with elevated choline and decreased N-acetylaspartate (NAA).\textsuperscript{23}
Posterior Fossa Tumors, O'Brien

Brainstem/Pontine Glioma

Background.

Brainstem gliomas vary in histology and prognosis based upon location and are fourth in incidence in terms of pediatric posterior fossa tumors, accounting for approximately 15% of cases. There is an equal incidence between boys and girls.27

Diffuse intrinsic pontine glioma (DIPG) is the most common and most aggressive subtype of brainstem glioma. The majority are fibrillary astrocytomas (WHO grade II) with focal progression to anaplastic or even glioblastoma multiforme variants. Low-grade variants occasionally occur in the pons but are more common within the medulla and tectal plate; the discussion for this review will focus on the more common DIPG variant.

Children with brainstem gliomas most often present with relatively rapid onset (weeks) of cranial nerve deficits, long tract signs, and occasionally ataxia.28

Imaging Findings.

On CT, DIPG presents as diffuse, infiltrative hypodensity with expansion of the pons, often affecting more than 50-75% of the cross-sectional area.28 The MR appearance is similar with diffuse decreased T1 and increased T2 signal abnormality, which is ill-defined. There is heterogeneous enhancement of the solid components following contrast administration; enhancement characteristics vary during treatment. Cystic or necrotic components

Prognosis.

The prognosis of posterior fossa ependymoma is primarily based upon the degree of initial surgical resection and presence or absence of disseminated or recurrent disease. The overall 5-year survival rate is between roughly 50% and 75%; a younger age at presentation, CSF dissemination, and recurrence correlate with a worse prognosis.24

Foci of CSF dissemination are best seen on post-contrast sequences as regions of leptomeningeal enhancement, which is most often nodular and located along the cerebellar sulci and spinal canal (Fig. 11); pre-operative evaluation of the spine to evaluate for drop metastases is important for both management and post-operative follow-up. Postoperatively, surveillance imaging of the brain and spine is typically performed to evaluate for recurrence or new CSF dissemination.

Brainstem/Pontine Glioma

Background.

Brainstem gliomas vary in histology and prognosis based upon location and are fourth in incidence in terms of pediatric posterior fossa tumors, accounting for approximately 15% of cases. There is an equal incidence between boys and girls.27

Diffuse intrinsic pontine glioma (DIPG) is the most common and most aggressive subtype of brainstem glioma. The majority are fibrillary astrocytomas (WHO grade II) with focal progression to anaplastic or even glioblastoma multiforme variants. Low-grade variants occasionally occur in the pons but are more common within the medulla and tectal plate; the discussion for this review will focus on the more common DIPG variant.

Children with brainstem gliomas most often present with relatively rapid onset (weeks) of cranial nerve deficits, long tract signs, and occasionally ataxia.28

Imaging Findings.

On CT, DIPG presents as diffuse, infiltrative hypodensity with expansion of the pons, often affecting more than 50-75% of the cross-sectional area.28 The MR appearance is similar with diffuse decreased T1 and increased T2 signal abnormality, which is ill-defined. There is heterogeneous enhancement of the solid components following contrast administration; enhancement characteristics vary during treatment. Cystic or necrotic components

Prognosis.

The prognosis of posterior fossa ependymoma is primarily based upon the degree of initial surgical resection and presence or absence of disseminated or recurrent disease. The overall 5-year survival rate is between roughly 50% and 75%; a younger age at presentation, CSF dissemination, and recurrence correlate with a worse prognosis.24

Foci of CSF dissemination are best seen on post-contrast sequences as regions of leptomeningeal enhancement, which is most often nodular and located along the cerebellar sulci and spinal canal (Fig. 11); pre-operative evaluation of the spine to evaluate for drop metastases is important for both management and post-operative follow-up. Postoperatively, surveillance imaging of the brain and spine is typically performed to evaluate for recurrence or new CSF dissemination.
Posterior Fossa Tumors, O’Brien

Figure 12. DIPG. Axial FLAIR image (A) reveals a hyperintense pontine mass with expansion of the brainstem. Anteriorly, the mass engulfs the basilar artery (arrow), and posteriorly, the mass extends into and compresses the 4th ventricle. Post-contrast T1 image with fat suppression (B) shows prominent enhancement with a region of central necrosis posteriorly on the right.

Figure 13. DIPG. Axial T2 MR image (A) demonstrates a diffuse, infiltrative T2 hyperintense mass with expansion of the pons. Anteriorly, the mass engulfs the basilar artery (arrow). Posteriorly, the mass projects into the 4th ventricle on the left. Pre (B) and post (C) contrast T1 images reveal patchy, heterogeneous enhancement.

are not uncommon with higher grade lesions (Fig. 12). Perfusion and diffusion-weighted imaging are useful for evaluation of focal progression to a higher grade or to guide biopsy on the rare occasion where the diagnosis is unclear. Regions of increased cellularity will show increased signal on DWI, decreased signal on ADC maps, and increased perfusion. MR spectroscopy shows a tumor spectrum with increased choline and decreased N-acetylaspartate (NAA); the degree of increase (choline) or decrease (NAA) is proportional to the grade of the tumor. Higher grade lesions or regions will also commonly show a lactate peak.

When large, exophytic components of DIPG may be seen anteriorly with engulfment of the basilar artery (Figs. 12, 13, and 14) or posteriorly projecting into the fourth ventricle (Figs. 12, 13, and 14). Obstruction of the fourth ventricle is typically a late finding. As the tumor progresses, it may extend posteriorly, cephalad, and caudad along white matter tracts (Fig. 14).

Treatment.

Due to the critical brainstem location, surgery is not a viable option for the treatment of brainstem gliomas, aside from debulking of exophytic components. Radiation and adjuvant chemotherapy
are the mainstays of treatment in children older than 3 years of age; in children under 3 years of age, attempts are made to minimize or avoid radiation therapy due to its potentially devastating effects to the developing CNS.

**Prognosis.**

Prognosis for DIPG is dismal with a nearly 100% mortality rate.\(^{29}\) Median survival is approximately 9-12 months with aggressive treatment.\(^{28}\)

### Atypical Teratoid Rhabdoid Tumor (ATRT)

**Background.**

ATRT is an uncommon, highly malignant tumor which is composed of rhabdoid cells plus primitive neuroectodermal tumor components. It occurs most often in very young children (<3 years of age), although it may occasionally occur in older children and adults. More than half are infratentorial, while the remainder are supratentorial in location.\(^{30,31}\)

The precise incidence of ATRT is difficult to determine, since it is believed that ATRT is significantly under-diagnosed and misdiagnosed as medulloblastoma, especially in infants and young children.\(^{32,33}\) If not suspected prospectively, routine histological results will favor medulloblastoma. Therefore, it is important to consider ATRT in the appropriate setting in order to guide the histological evaluation;\(^33\) the presence of rhabdoid cells is a key distinguishing feature from medulloblastoma.

Children with posterior fossa ATRTs present with a rapid onset of symptoms – over the course of days to weeks; headaches and nausea and vomiting are most common. Older children also present with truncal ataxia, while infants present with increase in head size. CSF seeding is common at presentation and may result in multifocal disease.

**Imaging Findings.**

There is significant overlap in the imaging appearance of posterior fossa ATRT and medulloblastoma; however, some distinguishing features may help suggest ATRT, especially in children under 3 years of age.\(^{34}\)

On CT, ATRT most often presents as a large, iso- to hyperdense mass which may be midline or off-midline (more common) (Fig. 15). A cerebellopontine angle (CPA) location is highly characteristic for ATRT.\(^{33,34}\) ATRT is more likely to appear heterogeneous compared to medulloblastoma with large eccentric cystic components, visible calcifications, and intratumoral hemorrhage.\(^{33,34}\) Despite the aggressive features, there is often little or no vasogenic edema within the surrounding parenchyma. Solid components demonstrate avid but heterogeneous enhancement. CSF dissemination may present as regions of leptomeningeal enhancement or multifocal masses with similar imaging characteristics to the primary tumor. Hydrocephalus due to compression and obstruction of the fourth ventricle occurs in approximately 60-65% of cases, which is less frequent than with medulloblastoma.\(^{34}\) If uncompensated, transependymal flow of CSF may be seen along the margins of the lateral ventricles.

On MRI, ATRTs are typically iso- to hypointense compared to white matter on T1 sequences; however, intratumoral hemorrhage will demonstrate regions of T1 hyperintensity, which are more characteristic of ATRT compared to medulloblastoma. As with medulloblastomas, T2 signal intensity is variable; more cellular components are T2 hypointense, while less cellular components are iso- to mildly hyperintense compared to white matter. Eccentric cysts are commonly seen and are T2 hyperintense. Foci of calcification are hypointense on all sequences and may demonstrate blooming on gradient echo sequences. High cellularity of solid components leads to hyperintensity on DWI, similar to medulloblastoma.
ATRT demonstrates avid but heterogeneous enhancement of solid components (Fig. 15), including the walls of the eccentric tumoral cysts. CSF dissemination may present with foci of leptomeningeal enhancement, which is often nodular, or multifocal masses.

Treatment.
ATRT is aggressive and less responsive to therapeutic options compared to medulloblastoma. Treatment options are dependent upon patient age and the extent of disease and include surgical resection with radiation and/or adjuvant chemotherapy. Radiation therapy is typically decreased in dosage or avoided prior to 3 years of age due to the potentially devastating effects of craniospinal radiation to the developing CNS; however, each case is evaluated independently with considerations for CSF dissemination or multifocal disease.

Prognosis.
The overall prognosis for ATRT is dismal, and a younger age at presentation corresponds to a worse prognosis. The mean survival for patients under 3 years of age is roughly 3-6 months; mean survival in older patients is roughly 12 months from the time of initial presentation. There have been reports of curative treatment and survival measured in years, however, that is relatively uncommon and occurs more frequently in older patients.

Conclusion
CT and MRI are instrumental in the work-up, management, and follow-up of patients with primary posterior fossa brain tumors. Although imaging alone is insufficient to reliably predict histology prospectively, characteristic imaging features may help provide the most likely diagnosis, along with appropriate differential considerations. Therefore, it is essential that radiologists involved with interpretation of pediatric neuroimaging have a working knowledge of the imaging appearances associated with the most common pediatric posterior fossa brain tumors.

The views expressed in this material are those of the author, and do not reflect the official policy or position of the U.S. Government, the Department of Defense, or the Department of the Air Force.
References

Intradural Neoplasms, Carra et al.

Introduction

Spine neoplasms are classified based on their location as either extradural, intradural extramedullary (IDEM), or intramedullary. This review will focus on intradural neoplasms including both extra- and intramedullary. While neoplasms are relatively uncommon compared with other types of spine pathology, recognition of their imaging appearance and understanding the principles of spine tumor imaging is necessary as MRI is the primary tool for establishing a differential diagnosis, guiding the diagnostic workup, and formulating a treatment plan.

Spine Tumor Imaging

MRI is the imaging modality of choice for assessing the spine because of its high soft tissue contrast; therefore, its use will be emphasized in this review. The first step in forming a pertinent differential diagnosis is assessing the location of the abnormality. Intradural extramedullary neoplasms are characterized by abnormal cord signal and expansion. IDEM tumors are contained within the thecal sac but displace rather than expand the spinal cord. A CSF cleft may be seen separating the tumor from the cord. Use of gadolinium based contrast agents should be standard in suspected spinal tumors. Enhancement is the rule, and unlike most intracranial neoplasms, even low-grade spinal tumors characteristically enhance. Syringohydromyelia and cyst formation is common. Two types of cysts exist: tumoral cysts and rostral/caudal cysts. The former are lined with neoplastic cells and typically demonstrate rim-enhancement; the latter are reactive and do not contain neoplastic cells or enhance. Distinction is important as the former need to be surgically resected while the latter can simply be aspirated.

Intramedullary Neoplasms

Intramedullary tumors represent 5-10% of all spine tumors and are more common in children. The most common primary intramedullary neoplasms are ependymoma, astrocytoma, and hemangioblastoma. Ganglioglioma, metastasis, and primary lymphoma are less common. Intramedullary neoplasms share many common imaging features. Abnormal cord signal intensity and expansion, whether from tumor itself or from cord edema, commonly spans several spinal levels, and may involve the entire spinal cord. Contrast enhancement is the rule, and unlike most intracranial neoplasms, even low-grade spinal tumors characteristically enhance. Syringohydromyelia and cyst formation is common. Two types of cysts exist: tumoral cysts and rostral/caudal cysts. The former are lined with neoplastic cells and typically demonstrate rim-enhancement; the latter are reactive and do not contain neoplastic cells or enhance. Distinction is important as the former need to be surgically resected while the latter can simply be aspirated.

The majority of intramedullary neoplasms are low grade and slow-growing. Therefore, patients typically have a prolonged clinical course extending over many years prior to diagnosis. Common symptoms include back pain and sensory disturbances. In general, treatment consists of surgical resection, with or without adjuvant radiation and chemotherapy.
Ependymoma.

Ependymomas are the most common intramedullary neoplasm in adults and the second most common in children.\(^2\) Mean age of presentation is in the 4\(^{th}\) to 5\(^{th}\) decades of life, and there is no gender predilection.\(^3\) They arise from ependymal cells lining the central canal, and are therefore typically centrally located. The cervical cord is the most common location.\(^4\) While they share many of the histologic characteristics of intracranial ependymomas, they are considered genetically different and carry a better prognosis. Gross total resection results in extremely low recurrence rates.\(^1\)

Five histologic subtypes of ependymoma exist, including cellular, clear cell, papillary, tanycytic, and myxopapillary (discussed in the IDEM section). The cellular subtype is most common and is typically WHO grade II. Sensory symptoms predominate in the clinical presentation, possibly from involvement of the central crossing spinothalamic tracts. Ependymomas can be seen as part of neurofibromatosis (NF) type 2 ("MISME" syndrome — multiple inherited schwannomas, meningiomas, and ependymomas), though the majority are sporadic.

Ependymomas are characteristically well-circumscribed masses in the central portion of the cord. They generally demonstrate T1 iso- to hypointensity and T2 hyperintensity relative to the spinal cord. Cyst formation is common. Due to their highly vascular nature, profound low T2 signal at the cranial or caudal margins, known as the "hemosiderin cap", is a distinguishing feature (Fig. 1). While the classic enhancement pattern is homogeneous and intense, this was seen in only 38% of cases according to one series, with heterogeneous, rim, and minimal enhancement patterns collectively making up the remaining majority.\(^5\)

Astrocytoma.

Intramedullary astrocytomas arise from astrocytic cells within the cord and represent the most common cord neoplasm in children and the second most common in adults. There is a slight male predominance.\(^3,6\) The majority are low-grade WHO I and II, pilocytic and fibrillary variants, respectively. WHO grade III and IV tumors, anaplastic and glioblastoma multiforme (GBM) subtypes, together

Figure 1. **Ependymoma**. 34 year-old patient with a centrally located mass enlarging the cervical cord. Sagittal T1W (A) and T2W (C) images demonstrate a heterogeneous, solid intramedullary mass with rostral and caudal cyst formation and cord edema. Low T2 signal along the caudal margin is the "hemosiderin cap" (arrow). Diffuse heterogeneous enhancement is seen on post-gadolinium (B) sagittal T1-weighted sequences.

Figure 2. **Astrocytoma**. Cervical spinal cord intramedullary mass in this child causes fusiform enlargement of the cord spanning several vertebral levels. Pre- (A) and post-gadolinium (B) sagittal and axial (C and D) T1W images demonstrate heterogeneous, irregular ring-enhancement with eccentric involvement of the cord and poorly defined rostral margins. Pathology revealed a WHO grade III anaplastic astrocytoma.
form only 10-15%.\textsuperscript{7} GBM’s are rare, representing a minority of the higher grade cord astrocytomas. Expectedly, high-grade tumors have a higher rate of recurrence and metastasis and carry a worse prognosis with average survival on the order of months.\textsuperscript{8} All subtypes demonstrate infiltrative growth along non-neoplastic scaffolding of the cord. Cyst formation is frequently seen. Cervical and thoracic cord involvement predominates.\textsuperscript{1}

Many imaging features suggest a diagnosis of astrocytoma over ependymoma. As opposed to ependymomas, astrocytomas are more characteristically eccentrically located and are less well-defined secondary to their infiltrative growth pattern (Fig. 2). MRI signal characteristics are similar, with T1 iso- to hypointensity and T2 hyperintensity compared to cord, though hemorrhage is less common. Partial mild-moderate intensity heterogeneous enhancement is characteristic, though no enhancement was seen in 20-30\% in one series.\textsuperscript{9} Despite the lack of enhancement, neoplasm should still be suggested based on characteristic cord expansion and signal abnormality.

**Hemangioblastoma.**

Hemangioblastomas are far less common than ependymomas and astrocytomas, accounting for 3.3\% of spinal cord tumors.\textsuperscript{10} They represent a low-grade (WHO grade I) vascular neoplasm, though their cell of origin is yet to be delineated. Association with von Hippel-Lindau (VHL) syndrome ranges from 17-38\% in the literature,\textsuperscript{11,12} in conjunction with cerebellar hemangioblastomas, retinal angiomas, and endolymphatic sac tumors in the CNS and head and neck. There is a slight male predominance, and the mean age of diagnosis is in the 4\textsuperscript{th} decade of life, though they tend to present about a decade earlier when associated with VHL.\textsuperscript{13}

MR imaging characteristics include a well-delineated, uniformly T2 hyperintense mass in smaller lesions. Location is subpial, typically along the posterior aspect of the cord. Cyst and syrinx formation is characteristic, and the cyst-mural-nodule appearance classically seen in cerebellar hemangioblastomas may be seen with larger lesions (Fig. 3). Due to the highly vascular nature of this entity, enlarged feeding vessels and flow voids in larger masses may be seen. Uniform and intense enhancement is characteristic of smaller masses.

**Metastases.**

Spinal cord metastases are uncommon, seen in only 0.9-2.1\% of autopsied cancer patients in one series.\textsuperscript{14} They represent approximately 3\% of metastases to the spine.\textsuperscript{15} The most common primary tumors are lung\textsuperscript{15} and breast and arise either through hematogenous dissemination or CSF spread through the central canal. The cervical cord is the most common site of involvement.\textsuperscript{16} Intratumoral hemorrhage may be seen with certain types of primary neoplasm. While they may be solitary, multiple lesions or multifocal involvement, including the osseous spine, may be key
Intradural Neoplasms, Carra et al.

to distinguishing the diagnosis. Unlike primary spinal cord neoplasms, metastases tend to have a more rapid onset and disease progression, on the order of weeks to months rather than years. Prognosis is uniformly poor with life expectancy on the order of months, as this represents an advanced stage of disease.

Imaging features vary based upon the primary tumor. Common features include a relatively small mass with extensive edema that is out of proportion for that expected based upon the size of the mass. Cord enlargement, if present, is not pronounced, and cysts are uncommon. Enhancement is common (Fig. 4). T1 hyperintensity can be seen with melanoma metastases and with hemorrhage.

Lymphoma.

Lymphoma has a variety of disease patterns involving the spine, including osseous, epidural, leptomeningeal, and spinal cord involvement. Intramedullary spinal involvement is the least common, accounting for only 3.3% of CNS lymphoma cases. As in other areas of the CNS, non-Hodgkin lymphoma predominates over Hodgkin disease, though some of the reported cases of spinal lymphoma are of T-cell lineage, an atypical feature of CNS lymphoma. Cord involvement favors the cervical spine.

![Figure 4. Intramedullary and Osseous Metastases.](image)

Abnormal high T2 signal (arrows) on sagittal (A) and axial (C) images with avid irregular contrast enhancement on T1W images (B, D, and E) represent intramedullary metastases from a known cerebral GBM. Multiple heterogeneously enhancing masses in the lumbar and sacral vertebrae (*) are consistent with metastatic disease.

The MRI appearance is nonspecific with respect to other intramedullary neoplasms, typically demonstrating T1 isointensity and T2 hyperintensity to the spinal cord. The T2 hyperintensity reported in cases within the literature is in distinction to the typical T2 hypointensity seen in primary lymphoma of the brain because of the high cellular content. Contrast enhancement is heterogeneous.

Ganglioglioma.

Ganglioglioma is uncommon, accounting for only 1.1% of spinal neoplasms. It is a low grade slow growing tumor composed of a mixture of neoplastic neuronal and glial elements. Children are more commonly affected with a classic clinical presentation of painful scoliosis. Cervicothoracic cord involvement predominates but holocord involvement is not uncommon.

Several MRI findings are suggestive of ganglioglioma. Length of involvement tends to be larger with an average of 8 vertebral segments, and they are eccentrically located within the cord. Enhancement is patchy with more than half demonstrating extension to the pial surface. Tumoral cysts are common. Signal intensity on T1-weighted images is heterogeneous, and the amount of cord edema is relatively unimpressive based upon the size of the tumor.

Intradural Extramedullary Neoplasms

IDEM tumors are more common in adults. Schwannomas and meningiomas are the most common, together accounting for 45% of all primary spinal cord neoplasms. The differential diagnosis also includes neurofibroma, malignant nerve sheath tumor, lymphoma, myxopapillary ependymoma, paraganglioma, and metastatic disease.

Schwannoma.

Schwannomas arise from support cells of the nerve sheath and are the most common IDEM spinal neoplasm. Nerve sheath tumors account for 25% of all intradural spinal tumors in adults, with the majority being schwannomas. While they may occur in any spinal compartment, they are most commonly
Intradural Neoplasms, Carra et al.

Intradural and extramedullary in location within the thoracic or lumbar spine. The classic “dumbbell” lesion extending through the neural foramen is considered both intradural and extramedural, occurring in 10-15% of cases (Fig. 5). Most are solitary and sporadic. Syndromic associations include NF2, Carney complex, and schwannomatosis. Schwannomas are typically low grade (WHO grade I) tumors. Total resection of an intradural schwannoma is considered curative.

Spinal schwannomas can be difficult to distinguish from meningiomas on imaging. They are typically well-circumscribed T2 hyperintense masses with signal intensity approaching that of fluid. Cyst formation is more common than with meningiomas. Their enhancement pattern is typically uniform (Fig. 6), though heterogeneous and ring-enhancement patterns do occur. Imaging features useful in distinguishing schwannoma from meningioma include foraminal enlargement and osseous remodeling, involvement of the cauda equina, fluid intensity T2 signal, and ring-enhancement.

Meningioma.

Meningiomas are the second most common IDEM spinal neoplasm. They arise from arachnoid cap cells within the dura and are most commonly seen in females in the 5th-7th decades of life. The majority are solitary and sporadic; however, there are multiple syndromic associations, the most common of which is NF2 (see “MISME” in ependymoma section above). Thoracic cord involvement is typical and unlike schwannomas, cystic degeneration is uncommon. WHO grade I tumors are by far the most common, and total resection is generally curative.

Meningiomas are typically seen as well-circumscribed masses along the ventral or ventrolateral aspect of the cord (Fig. 7). MR signal
characteristics include T1 iso- and T2 iso- to hyperintensity relative to the spinal cord. A broad dural attachment, or “dural tail”, was seen in 58% of meningiomas in one series, best demonstrated on post-contrast imaging. In this same series, calcification, which is easily identified on CT, was also identified in 58%. Post-contrast imaging typically reveals diffuse and prominent enhancement. A solitary IDEM thoracic spine mass in a middle-age female should suggest the diagnosis.

**Neurofibroma.**

Neurofibromas are much less common than schwannomas within the spine. They are most commonly seen in the 3rd-4th decades of life and have no gender predilection. The classic association of NF1 with plexiform neurofibroma is virtually pathognomonic. However, the majority of lesions are sporadic. They can be intradural and/or extradural and can be seen at any spinal level. Neurofibromas are WHO grade I neoplasms. While total resection is considered curative, tumors with extensive paraspinal involvement and subtotal resection have a propensity to recur and require follow up imaging.

It is often difficult if not impossible to distinguish solitary spinal neurofibromas from schwannomas based on imaging alone. Peripheral T2 hyperintensity, the “target sign,” is more suggestive of neurofibroma but not pathognomonic (Fig. 8). Cystic degeneration and hemorrhage are also less common than schwannoma. Involvement of a ventral nerve root is more suggestive of neurofibroma. Extrudal extension can be prominent as a plexiform or infiltrative soft tissue mass.

**Malignant Peripheral Nerve Sheath Tumor.**

Malignant peripheral nerve sheath tumor (MPNST) represents a soft tissue sarcoma with histologic features of peripheral nerves either occurring de novo or arising from malignant degeneration of a neurofibroma. They are WHO grade II-IV tumors. Roughly half are associated with NF1 while the remainder are sporadic. They most commonly involve the paravertebral soft tissues with rare intraspinal involvement. Masses are typically markedly heterogeneous and infiltrating (Fig. 9). Prognosis is poor.

![Figure 8. Neurofibromas in NF1.](image)
Several IDEM masses of the cauda equina with peripheral T2 hyperintensity (“target sign”), suggestive of neurofibromas. Multiplicity suggests NF1.

![Figure 9. Malignant Peripheral Nerve Sheath Tumor.](image)
A large heterogeneous T2 hyperintense dumbbell mass (A and C) extends through and widens the neural foramen in this patient with NF1. Contrast enhancement is avid (B and D). Findings are consistent with a peripheral nerve sheath tumor. However, the large size and infiltration of the paraspinal musculature (*) (C and D) raises suspicion for malignancy. The serpentine enhancing structure (arrow) posterior to the cord in (B) is an enlarged vein from tumor compression. (Case courtesy of Dr. William T. O’Brien, Sr.)

**Myxopapillary Ependymoma.**

The myxopapillary variant represents roughly 30% of all intraspinalependymomas. It can be distinguished based on its characteristic location within the conus medullaris, filum terminale, or cauda equina where it is the most common tumor. Peak incidence is in the 3rd-5th decades of life, and there is a slight male predominance. Despite its WHO I grading, it may cause subarachnoid seeding. Hemorrhage, calcification, and cyst formation are common. The tumor can become intimately involved with the nerve roots of the cauda equina making total resection difficult. When accomplished, recurrence rates are low.
Intradural Neoplasms, Carra et al.

Figure 10. **Myxopapillary Ependymoma.** A heterogeneous, mixed solid and cystic (axial T2W images B and C) sausage-shaped mass occupies much of the lower lumbar spinal canal and enhances avidly (post-gadolinium T1W images A, D, and E).

Figure 11. **CSF Dissemination of Mantle Cell Lymphoma.** 64 year-old woman with known lymphoma presented with cauda equina syndrome. Imaging shows abnormal low T2 (A) and intermediate T1 (B) signal that fills the lumbar spinal canal and encases the conus medullaris and cauda equina. Post-gadolinium T1W images (C and D) best depict the mass which is avidly enhancing.

Figure 12. **Paraganglioma.** Markedly T2 hyperintense (A), circumscribed mass along the dorsal aspect of the distal spinal cord shows avid enhancement on post-gadolinium T1W images (B and C).

Myxopapillary ependymomas are sausage-shaped masses that can occupy the entire lumbosacral canal (Fig. 10). Unlike other histologic variants of spinal ependymoma, the myxopapillary type may have intrinsic T1 hyperintensity secondary to its high mucin content, seen in 67% of cases in one series. Other imaging features, such as the hemosiderin cap and intralesional flow voids, are similar to other ependymoma variants.

**Lymphoma.**

As discussed above, spine lymphoma has a variety of different disease patterns, which include leptomeningeal involvement. The majority occur as spread from intracranial lymphoma. Patient demographics are similar to that of other types of lymphoma, most commonly occurring in middle age adults. Imaging hallmarks include thickened nerve roots and smooth or nodular leptomeningeal enhancement; IDEM mass formation can occur in extreme cases (Fig. 11).

**Paraganglioma.**

Paragangliomas are neural crest tumors of which the intra-adrenal form, pheochromocytoma, is the most common. Extra-adrenal paragangliomas most commonly occur in the head and neck near the carotid bulb, jugular foramen, and within the temporal bone. When they occur in the spine, they are
characteristically located within the lumbar spine, involving the cauda equina or filum terminale where they are almost always non-secreting.

Paragangliomas typically appear as a well-circumscribed mass within the cauda equina. They are T2 hyperintense and highly vascular (Fig. 12). Hemosiderin cap and flow voids may be seen in larger lesions. MR imaging features are nonspecific and may mimic myxopapillary ependymoma, schwannoma, and meningioma, which are much more common. The diagnosis can be suggested with avid uptake on nuclear medicine metaiodobenzylguanidine (MIBG) scan.

Metastasis.

CSF seeding with malignant cells may occur through a hematogenous route or may represent spread from a CNS primary. The most common primary malignancies outside of the CNS include lung, breast, and melanoma. CNS primary tumors with a tendency for CSF dissemination include high-grade astrocytomas, ependymomas, germ cell tumors, medulloblastomas, and choroid plexus neoplasms.

Imaging findings mirror patterns of disease involvement, which include solitary masses (Fig. 13), diffuse smooth or nodular coating of the cord and nerve roots (Fig. 14), and thickening of the cauda equina. Use of intravenous contrast increases the sensitivity for detection of metastatic lesions. MRI is an important adjunct to lumbar puncture with CSF analysis in diagnosing CSF metastasis; cytology alone has a significant false-negative rate and frequently must be repeated. It is important to remember, however, that chemical meningitis can cause enhancement and thickening of nerve roots, mimicking metastatic disease. Imaging should therefore precede lumbar puncture and craniotomy.

Summary

While neoplasms are a relatively uncommon form of spine pathology, recognition of their imaging features plays an essential role in providing a meaningful differential diagnosis and guiding further evaluation and treatment. While many of the imaging characteristics are nonspecific with significant overlap, when tumors are suspected, certain diagnoses can be suggested based upon location, clinical and patient demographics, and certain specific imaging features.

The views expressed in this material are those of the author, and do not reflect the official policy or position of the U.S. Government, the Department of Defense, or the Department of the Air Force.
Intradural Neoplasms, Carra et al.

References


Expansile Lesion of the Petrous Apex

Jason Romesburg D.O., and Michael E. Zapadka, D.O.
Department of Neuroradiology, Wake Forest Baptist Health, Winston-Salem, NC

Case Presentation

A 58-year-old woman underwent a noncontrast head CT after trauma. Past medical history was non-contributory. An incidental finding is discovered (Fig. 1) and further evaluated by MRI (Fig. 2).

Figure 1. Axial CT in bone algorithm shows a lucent, expansile lesion in the right petrous apex (arrows) with smooth, scalloped margins.

Figure 2. Axial T2 weighted (A), axial high-resolution heavily T2 weighted FIESTA sequence (B), axial T1 weighted (C), and axial T1 post-contrast (D) MR images demonstrate a homogenous T2 hyperintense, T1 hypointense non-enhancing lesion that follows CSF signal and is continuous with right Meckel cave. Although not shown, the lesion completely suppressed on FLAIR and did not restrict diffusion.
Key imaging finding

Expansile lesion of the petrous apex

Differential diagnoses

- Cholesterol granuloma
- Meningocele
- Epidermoid cyst
- Mucocele
- Chondrosarcoma (and other neoplasms)
- Petrous apicitis
- Petrous carotid aneurysm

Discussion

The differential diagnosis for petrous apex lesions is broad, but careful attention to certain imaging features can often help distinguish between the various entities. The radiologist plays a key role in the evaluation of lesions of the petrous apex, since this area is beyond direct inspection by the clinician and surgical access is difficult. Therefore, the radiologist’s role should go beyond detection, localization, and characterization and make every effort to arrive at the most specific diagnosis – when possible – to appropriately direct patient care.

When evaluating petrous apex lesions, imaging findings are often nonspecific by CT or MRI alone. However, CT and MRI are complementary and both studies are typically required for more definitive characterization. On CT, the osteolytic component and margins are better evaluated. If the margins are smooth and scalloped, this suggests a slow-growing benign process with pressure erosion or remodeling of adjacent bone. This is more typical of cholesterol granuloma, meningocele, mucocele, schwannoma, and other benign lesions, while ill-defined margins or a permeative moth-eaten pattern are worrisome for aggressive neoplasm or infection. The presence of internal matrix/calcification is also an important feature to assess, and – if present – suggests a chondroid lesion such as chondrosarcoma. MR imaging, to include post-contrast sequences, is essential to narrow the differential diagnosis; it is superior to CT in assessing the lesion’s relationship to adjacent soft tissue structures.

While solid lesions often require biopsy to determine appropriate treatment, if resection is necessary, and the extent of surgery, cysts may be treated with simple drainage and fistulization if proper preoperative diagnosis is made and treatment is necessary. Potential pitfalls to be aware of, primarily on MR imaging, are asymmetric pneumatization of the petrous apex and unilateral retention of secretions in apical air cells. With the former, there is unilateral T1 hyperintensity from marrow of non-pneumatized petrous apex (Fig. 3). Fat suppression techniques can usually resolve this issue, as this area will follow suppression of marrow signal elsewhere. In the latter instance, unilateral T2 hyperintensity is seen in the petrous apex from retained secretions, which is usually T1 hypointense, similar to sinus secretions. CT imaging in both of these circumstances will show absence of an expansile lesion or bony erosion, and therefore usually resolves the question.

Petrous apex lesions are often incidentally seen and usually attain considerable size before causing cranial nerve symptoms that lead to their clinical presentation. Most of the lesions that may be encountered in clinical practice are specifically discussed below.

Figure 3. Normal Marrow as Possible Pitfall on MRI.
Axial T1 weighted MR image (A) shows asymmetric T1 hyperintensity in the right petrous apex due to normal marrow in a non-aerated apex, confirmed to have no lesion by CT (B).
Cholesterol Granuloma.

Cholesterol granuloma (also known as cholesterol cyst or giant cholesterol cyst) is the most common primary petrous apex lesion. A theorized mechanism for its development is obstruction of ventilation outlet by thickened mucosa, leading to repeated cycles of hemorrhage and granulomatous reaction. When gas is trapped in the pneumatized space and absorbed over time, negative pressure in the enclosed space leads to tissue edema, breakdown of blood vessels in the region, and subsequent accumulation of blood degradation products, including cholesterol. These lesions typically occur in young and middle-aged adults of both sexes. If symptomatic, hearing loss, tinnitus, and hemifacial spasm are the most common associated clinical findings. On CT, they are homogenous lucent expansile lesions with smooth, sharply defined margins, and no internal calcification (Fig. 4A). Overlying bone may be thinned or even absent and one may see dehiscence of the carotid canal. On MRI, the characteristic feature of this lesion is strong T1 hyperintensity (Fig. 4B). They are also typically bright on T2 weighted images and frequently contain non-homogenous hypointense internal areas that probably represent hemosiderin from previous hemorrhage. Note that normal unilateral T1 hyperintensity from marrow of non-pneumatized petrous apex can be a pitfall, and CT can usually sort this out if there is a question based on the MRI (Fig. 3).

Meningocele.

A petrous apex meningocele is an uncommon lesion that represents a protrusion of meninges and CSF from the posterolateral aspect of Meckel cave. On CT, these lesions mimic cholesterol granulomas with an expansile homogenous lesion of the petrous apex. MRI is key to demonstrating that the lesion follows CSF signal on all sequences and directly communicates with Meckel cave. These are usually asymptomatic and incidental but rarely have been associated with symptoms such as cranial nerve palsy.

Epidermoid Cyst.

Epidermoid cysts contain desquamated keratin, which appears grossly as whitish friable material. For this reason, they are also referred to as primary cholesteatomas. As the keratin accumulates, the mass slowly enlarges. On CT, epidermoid cysts appear similar to cholesterol granulomas with a homogenous, nonenhancing, sharply defined, expansile lesion. On MR imaging, they are strongly T2 hyperintense; the presence of restricted diffusion will help differentiate these from mucoceles (see below). Although these are often referred to as “cysts,” they often contain solid or semi-solid material and are usually treated with complete surgical resection to prevent recurrence.

Mucocele.

Mucoceles have very similar imaging features compared to epidermoid cysts on routine MR sequences but are less common. They are hyperintense on T2, hypointense on T1, and there may be a thin enhancing rim. In contrast to epidermoid
cysts, they do not restrict diffusion, which is an important distinguishing feature.¹

**Petros Apicitis.**

Petrosum apicitis refers to overt infection of the petrous apex, usually as a complication of otomastoiditis.³ This is thought to develop only in individuals that have a pneumatized apex, because if aerated (as is the case in approximately 30% of individuals), the petrous apex communicates with mastoid air cells and middle ear.⁴ On CT, there is debris and fluid in the petrous apex with destruction of bony septa (analogous to coalescent mastoiditis).⁴ Destruction of bony cortex can result in intracranial complications, which can be better evaluated by MRI. This can cause the classic Gradinego triad of petromastoiditis, sixth nerve palsy, and pain in the distribution of the fifth nerve, although patients rarely have all of these symptoms.⁴ Trapped fluid, or "petrous apex effusion," occurs commonly as an incidental finding on both CT and MRI and should not be confused with clinically significant disease. There should not be bony destruction or expansion with an effusion. Also, MRI typically shows enhancement with petrous apicitis, but that would not be the case with retained secretions.

**Chondrosarcoma.**

Petroclival chondrosarcoma is a rare tumor, yet probably the most common primary malignant neoplasm in the region of the petrous apex.¹ These tend to occur off midline along synchondroses, centered along the petrosphenoidal and petrooccipital fissures within the petrous apex. On CT, they cause bone destruction, enhance, and may contain chondroid-like matrix (Fig. 5). On MR imaging, they are characteristically hyperintense on T2, iso to hypointense on T1, and usually enhance avidly. Occasionally, their location may overlap with chordomas which arise from notochord remnants, typically occur in midline, and may contain calcifications.¹

**Other Neoplasms.**

A trigeminal schwannoma may occasionally attain such size that it straddles the petrous apex, extending into the anterior and middle cranial fossae. The petrous apex may also be invaded by direct extension from nasopharyngeal squamous cell carcinoma or glomus jugulare paragangliomas. On CT, these have permeative margins and associated soft tissue components seen as enhancing masses on postcontrast imaging. Squamous cell carcinoma will be associated with an infiltrative nasopharyngeal mass. Paraganglioma will characteristically have osseous destruction with a “moth-eaten” appearance on CT; on MRI, the soft tissue mass may have a “salt and pepper” appearance. Myeloma, lymphoma, metastases, and giant cell tumors are additional tumors that may occasionally involve the petrous temporal bone and typically have aggressive imaging features. Endolymphatic sac tumors are rare papillary cystadenomatous tumors that can cause facial palsy and sensorineural hearing loss.¹ These are associated with Von-Hippel-Lindau disease but may also occur sporadically; involvement tends to occur within the retrolabyrinthine petrous bone, although extension towards the apex may be seen.

**Petros Carotid Aneurysms.**

Aneurysms of the petrous portion of the internal carotid artery are extremely rare,¹ but are an important part of the differential diagnosis to consider given the serious implications of misdiagnosing this
entity. On CT, you would expect to see a lobulated soft tissue mass expanding the carotid canal with smooth, scalloped bony margins. On MR imaging, laminated mural thrombus may be apparent, showing varying signal intensity; the patent lumen appears as a flow void.

Diagnosis

Petrous apex meningocele

Summary

Petrous apex meningocele is typically an incidental finding, but is important to recognize as a potential mimicker of other lesions on CT. MRI confirms the diagnosis by demonstrating a CSF signal lesion that communicates with Meckel cave. As in this scenario, it is often possible to make a specific imaging diagnosis of a lesion involving the petrous apex. Every attempt should be made to do so, particularly because this area is not amenable to direct inspection and surgical access is difficult. Therefore, radiologists play a key role in diagnosis and determining if further management is necessary.

References

Enhancing Sellar/Suprasellar Mass in an Adolescent

Jessika Chumak, M.D., and Anthony I. Zarka, D.O.
Department of Radiology, Wilford Hall Ambulatory Surgical Center, Joint Base San Antonio-Lackland, TX

Case Presentation

A 16-year-old boy with a history of acute lymphoblastic leukemia (ALL) in remission presented with increasing headache, ptosis, myalgias, fatigue, and numbness and pain in his lower back and sacrum. Initial workup, including CT and MRI of the brain is shown (Fig.).

Figure. Axial noncontrast CT image of the brain (A) shows a round, hyperdense non-calcified mass in the suprasellar region. Post-contrast sagittal (B) and coronal (C) T1 MRI images reveal a circumscribed sellar and suprasellar mass with diffuse homogenous enhancement.
Key imaging finding

- Enhancing sellar/suprasellar mass in an adolescent

Differential diagnoses

- Craniopharyngioma
- Chiasmatic-hypothalamic glioma
- Germ cell tumor
- Recurrent leukemia
- Langerhans cell histiocytosis

Discussion

Approximately 2.5-4 per 100,000 children will be affected by a tumor of the central nervous system, which is second in incidence for childhood malignancies behind leukemia. The differential diagnosis for CNS tumors varies based upon location, age, sex, and clinical symptoms and history of any known malignancy. The differential diagnosis for an enhancing sellar/suprasellar mass in an adolescent includes more common entities, such as craniopharyngioma, chiasmatic-hypothalamic glioma, and germ cell tumor. Pituitary macroadenomas (in children), Langerhans cell histiocytosis, and hypothalamic hamartoma are less common. Leukemia should be a consideration in a patient with a history of prior or current disease. Magnetic resonance imaging is the modality of choice for further characterization of masses of the sellar/suprasellar region with computed tomography playing a complimentary role in evaluating for calcifications.

Craniopharyngioma.

Craniopharyngioma is a low-grade (WHO grade 1) neoplasm and represents the most common sellar/suprasellar tumor in children, accounting for approximately 6-9% of CNS tumors. Common presenting symptoms include headache, visual disturbances, hormonal imbalances, and behavioral changes. There are two pathologic variants which determine the age of presentation (bimodal) and overall prognosis. The adamantinomatous variant occurs in children with a peak age of presentation between 10 and 14 years. It presents as a cystic and solid sellar/suprasellar mass with avid enhancement of solid components and calcifications (>90% of cases). The calcifications tend to be stippled and are often peripheral in location. The papillary variant occurs in middle-aged and older adults and typically presents as a solid enhancing sellar/suprasellar mass; cystic changes and calcifications are less common.

Chiasmatic-Hypothalamic Glioma.

Gliomas are common CNS tumors in children, the majority of which are low grade. Common locations in pediatric patients include the posterior fossa and optic pathways. Gliomas may occur sporadically or in association with neurofibromatosis type 1 (NF1). Chiasmatic-hypothalamic gliomas have no sex predilection and are typically seen in children; they rarely occur in adults. Presenting symptoms include painless proptosis, visual impairment, diabetes insipidus, diencephalic syndrome, and precocious puberty. Radiographs demonstrate a classic J-shaped sella turcica, as well as enlarged optic canals. On MRI, gliomas are typically T1 isointense to mildly hypointense, T2 hyperintense, and show variable degrees of enhancement.

Germ Cell Tumors.

Intracranial germ cell tumors are fairly uncommon, accounting for approximately 0.4-3.4% of intracranial neoplasms. Boys have an incidence rate approximately twice that of girls. Germ cell tumors have a propensity for the suprasellar and pineal regions. Symptoms depend upon the aggressiveness of the tumor and its location. Common presenting symptoms include headache, hormonal imbalance, and visual disturbances. Prognosis is dependent upon the tumor type, the most common of which is a germinoma which has a peak incidence during puberty. Prognosis for germinomas is generally good with a 10-year survival rate over 90% after radiation therapy. Nongerminomatous germ cell tumors present in slightly older aged children (between 10-21 years of age) and have a worse prognosis with cure rates of approximately 40-60%. Tumor markers are important for the diagnosis and classification of germ cell tumors. On MRI, germinomas present as infiltrating, homogeneously enhancing masses which typically...
follow gray matter signal intensity on pre-contrast sequences. CSF seeding is common; therefore, imaging of the entire neuroaxis should be performed. A teratoma typically presents as a heterogeneous cystic and solid mass with macroscopic fat. Less common nongerminomatous tumors have nonspecific imaging and often heterogeneous characteristics. Biopsy is typically required for specific diagnosis.

Recurrence Leukemia

Leukemic infiltration can be difficult to differentiate from more common sellar/suprasellar tumors. The presentation of recurrent leukemia as a solid homogeneous mass in the sellar region is relatively rare and is asymptomatic in most cases. When symptomatic, common presentations include headache, diabetes insipidus, and visual disturbances. Bilateral hemianopsia is the most common visual field impairment due to compression of the optic chiasm. If tumor infiltrates into the cavernous sinus, patients may present with cranial neuropathy. On MRI, leukemic infiltration may appear circumscribed or irregular, enhances avidly, may demonstrate thickening of the infundibulum, and most importantly, shows rapid growth on interval exams. Some clues which are particularly suspicious for malignancy include rapid growth with sudden onset of symptoms, especially in the setting of cranial nerve VI palsy, since it is typically well sheltered within the cavernous sinus; its involvement indicates advanced tumor infiltration.

Langerhans Cell Histiocytosis (LCH)

LCH is a rare disease of uncertain etiology which typically presents in children under two years of age, is more common in boys, and may involve any organ system. There is an inverse relationship between the severity of the disease and the patient’s age at the time of presentation. The most common form of intracranial involvement includes infiltration of the hypothalamic-pituitary axis, resulting in hypopituitarism and diabetes insipidus, with or without lytic calvarial lesions. On MRI, the infundibular stalk is thickened, slightly T2 hyperintense, and avidly enhances. There is often absence of the posterior pituitary bright spot on pre-contrast T1 sequences.

Diagnosis

Recurrent Leukemia

Summary

In summary, the differential diagnosis for CNS tumors is based upon several factors, to include location, age, sex, imaging characteristics, and patient demographics. When presented with an enhancing sellar/suprasellar mass in an adolescent, the most common etiologies include craniopharyngioma, chiasmatic-hypothalamic glioma, and germ cell tumor. Leukemia should be a consideration in a patient with a history of prior or current disease, especially if the lesion demonstrates rapid growth on subsequent imaging exams. Magnetic resonance imaging is the primary modality of choice for characterization of masses in the sellar/suprasellar region; CT plays a complimentary role in evaluating for calcifications.

References

Balo Concentric Sclerosis.

When attempting to characterize white matter lesions, important considerations include morphology and distribution, as well as patient demographics. The majority of cases represent nonspecific gliosis secondary to some form of parenchymal insult, typically microvascular ischemic, post-infectious and/or inflammatory, or traumatic in nature. Demyelinating processes, on the other hand, often occur in young adults, especially women, and demonstrate characteristic morphology and distribution, including involvement of the periventricular white matter, corpus callosum, optic pathways, posterior fossa structures, and spinal cord.

The axial FLAIR image (above left) reveals multiple characteristic demyelinating lesions which are ovoid and oriented perpendicular to the ventricles along the perivenule spaces, referred to as Dawson fingers. The T1 image (above right) shows pathognomonic findings of a subset of demyelination, Balo concentric sclerosis (BCS), which is characterized by concentric rings of alternating signal intensity which represent rings of demyelination (hypointensity) adjacent to rings of spared white matter (isointensity).

BCS is a rare acute inflammatory demyelinating disease, classically considered to be a rapidly progressive and severe variant of multiple sclerosis. On MRI, BCS lesions are defined as two or more alternating bands of differing signal intensities. Lesions can be solitary or multiple and are typically without significant surrounding edema or mass effect. Enhancement can occur within the demyelinating rings, and – as with other demyelinating diseases – if present, is considered to represent active demyelination. Treatment of BCS is typically with steroids. If left untreated, many patients with BCS have aggressive disease. Recognizing the characteristic MRI pattern of BCS can aid in early diagnosis and treatment, ultimately improving prognosis.

The views expressed in this material are those of the author, and do not reflect the official policy or position of the U.S. Government, the Department of Defense, or the Department of the Air Force.
Glomus Vagale.

A 37-year-old woman presented with a several year history of a painless left neck mass. She became increasingly concerned, since her mother also had a neck mass which had been recently diagnosed as a carotid body paraganglioma. A contrast-enhanced CT shows intense enhancement of a left carotid space mass. Lateral projection from a digital subtraction angiogram obtained with a left common carotid artery injection demonstrates the hypervascular mass displacing the common carotid artery anteriorly. Indium-111 octreotide scan reveals avid uptake by the tumor.

Paragangliomas are highly vascular tumors of the paraganglia. The most common location in the head and neck is at the carotid bifurcation, termed carotid body tumor. Additional locations include the jugular fossa (glomus jugulare), tympanic cavity (glomus tympanicum), and along the course of the vagus nerve (glomus vagale). This neck mass has all the characteristic clinical and radiologic manifestations of a glomus vagale. The lesions typically present as a slow-growing, painless, lateral neck masses, most commonly located behind the angle of the mandible, with anterior displacement of the carotid artery. A familial occurrence is well documented. The hypervascularity of the tumor results in homogeneous, intense enhancement on contrast-enhanced CT and MRI (not shown), as well as catheter angiography. Additional MR findings include a classic “salt and pepper” appearance secondary to regions of calcification and flow voids (pepper) surrounded by hyperintense and hyperenhancing tumor (salt). The tumor also has early, intense octreotide avidity. This patient's imaging workup revealed no multicentric disease, and the tumor was successfully treated with preoperative embolization followed by surgical excision.

The views expressed in this material are those of the author, and do not reflect the official policy or position of the U.S. Government, the Department of Defense, or the Department of the Air Force.
Caudal Regression Syndrome.

How would you guess this patient presented and what associated anomalies would you expect to see?

This sagittal T2 image of a pediatric patient shows a blunted conus medullaris with sacral dysgenesis. There is also prominence of the central canal in the distal cord. A result of an insult before the fourth gestation week, caudal regression syndrome (CRS) encompasses a spectrum of lumbosacral, anorectal, genitourinary, and lower extremity anomalies. Although most cases are sporadic and the pathogenesis is not completely understood, there is a strong association and increased incidence in infants of diabetic mothers. Clinical manifestations vary from relatively asymptomatic to severe malformations such as sirenomelia with lumbosacral anomalies and fused lower extremities (mermaid syndrome). The most common presenting symptoms are related to a neurogenic bladder. As multiple systems may be affected, it is important to evaluate for genitourinary and anorectal anomalies when presented with spinal anomalies - and vice versa.

There are two types of CRS. Type 1 results in a high-riding, blunted conus and severe sacral dysgenesis, while type 2 is characterized by a low-lying, elongated cord and conus with less severe lumbosacral anomalies. Severe cases may be identified during antenatal ultrasound screening; however, more mild forms may go undiagnosed into childhood and even early adulthood. MRI allows for full characterization of the extent of anomalies.

The views expressed in this material are those of the author, and do not reflect the official policy or position of the U.S. Government, the Department of Defense, or the Department of the Air Force.