Aneurysmal Fibrous Histiocytoma: A Case Report and Review of the Literature

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Abstract

Dermatofibroma is one of the most common subcutaneous dermatologic tumors. In its classic variant, a dermatofibroma is easily recognized by dermatologists; however, studies have identified numerous variants of the dermatofibroma that do not present with a classic clinical picture. Aneurysmal fibrous histiocytoma, one of these variants, is not easily recognized given its bizarre growth and potentially malignant appearance. Microscopically, aneurysmal fibrous histiocytoma can be difficult to identify, as the lesion will display some similarities to a classic dermatofibroma along with distinguishing characteristics, like large blood-filled cavernous spaces. Aneurysmal fibrous histiocytoma is a benign lesion with a low risk for recurrence if adequately excised. In this paper, we present a case of aneurysmal fibrous histiocytoma and review the literature on this rare dermatofibroma variant and what to consider on the differential diagnosis.

Introduction

Dermatofibroma, also known as fibrous histiocytoma, is a common dermatologic subcutaneous tumor. It represents roughly 3% of tissue specimens received by dermatologic laboratories.[1] Among fibrohistiocytic tumors, the DF is second to acrochordons in prevalence.[2] The lesion typically develops on the extremities of middle-aged individuals. Clinically, a dermatofibroma is described as a typically dense, dome-shaped papule that ranges from a few millimeters to 5 cm in size.[2,3] When palpated, clinicians may describe the lesion as adhered to subcutaneous tissue, and pinching a dermatofibroma can produce a downward movement of the papule, called "dimple sign."[2] Clinicians feel that some dermatofibromas develop following an injury, such as blunt trauma or an insect bite to the skin.[3] When a classical fibrous histiocytoma is identified, the diagnosis is usually straightforward; however, numerous non-classical variants exist, namely aneurysmal, atypical, cellular, clear cell, myxoid and palisading, among others.[4] These varying presentations can make the diagnosis of dermatofibroma difficult. We present a case of an aneurysmal fibrous histiocytoma and review the literature on these unique dermatological neoplasms.

Case Report

A 28-year-old, healthy male presented to the clinic for evaluation of an enlarging nodule on his right scapula. He reported that it had been present for about one year and initially appeared as a 1 mm to 2 mm purple papule. It slowly grew for the first six months and then rapidly enlarged in size over the next six months. The patient regularly lifted weights and stated that the squat bar often rubs against the nodule, but it had never ruptured or bled. He reported no pain with the lesion unless firmly palpated. He had no family history of cutaneous malignancy.

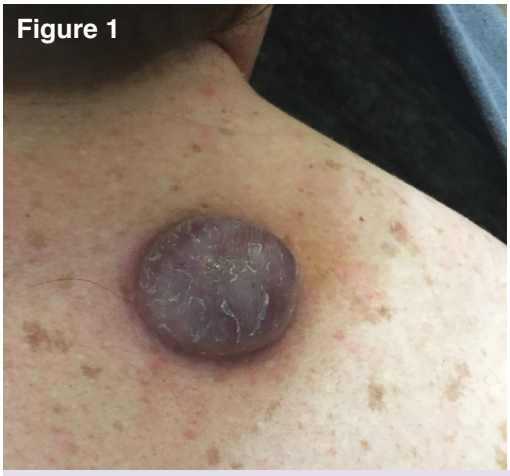
Physical examination revealed a 2.5 cm x 2.5 cm, purple, exophytic, slightly scaly, well-circumscribed, spherical nodule with minimal surrounding erythema that was slightly compressible, with some blanching upon palpation **(Figures 1, 2)**. The rest of his examination was unremarkable, including no lymphadenopathy. On the differential diagnosis, we considered malignant melanoma, nodular Kaposi's sarcoma, leiomyosarcoma, pyogenic granuloma, and dermatofibrosarcoma protuberans. An excisional biopsy was performed that day. Interestingly, upon removal, we noticed that the nodule was very well-circumscribed, encapsulated, and appeared completely removed.

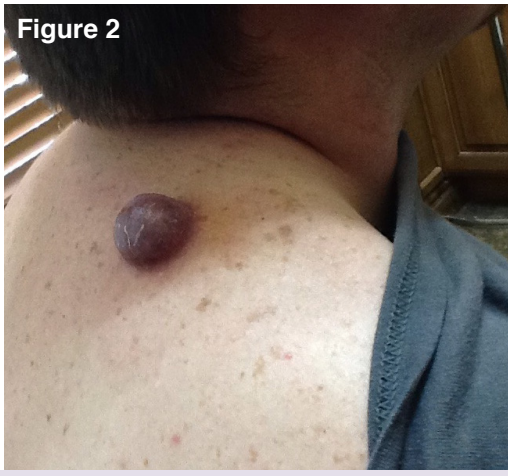
Histologically, hematoxylin and eosin (H&E) staining revealed a dense cellular dermal proliferation of spindled and plump fibroblasts, histiocytes with scattered giant cells, numerous extravasated erythrocytes, siderophages, and large blood-filled cavernous spaces that invaded into the superficial subcutis (**Figure 3**). Immunohistochemical stains FXIIIa, CD10, and CD68 confirmed that the lesion was a histiocytic tumor (**Figure 4**). CD34 highlighted the vascular component. Mart-1, S-100, and HMB-45 were all negative, which ruled out malignant melanoma.

As of this paper's submission, the patient has healed well, with no evidence of recurrence over the past three months. He will be reevaluated at six months.

Discussion

Aneurysmal fibrous histiocytoma was originally described by Santa Cruz and Kyriakos in 1981.[5] They described aneurysmal fibrous histiocytoma lesions with a range of color possibilities, from dark purple to red, brown, or blue, with a soft





Figures 1, 2. Right upper back: 2.5 cm x 2.5 cm, purple, exophytic, scaly nodule.

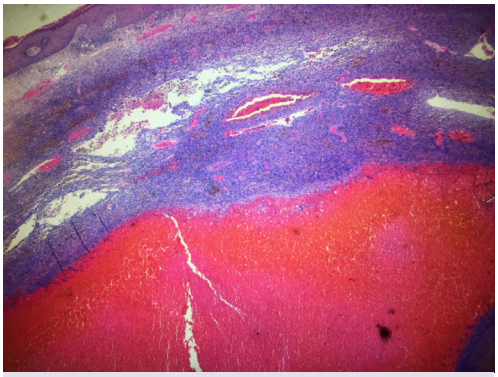


Figure 3. On H&E, large blood-filled spaces make up most the lesion. Note the surrounding hypercellularity.

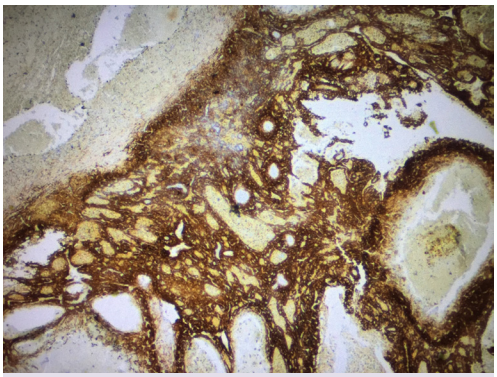


Figure 4. CD10 staining confirming the fibrohistiocytic nature of the lesion.

sensation when palpated.[5] When compared to a classical dermatofibroma, the aneurysmal fibrous histiocytoma is typically larger in diameter, more elevated, and has an accelerated growth phase.[6] The rapid growth is thought to be due to vast hemorrhage within the lesion.[7] Patients usually do not complain of pain or tenderness. While aneurysmal fibrous histiocytoma tumors can present in various locations, such as the head, neck, and trunk, they are more commonly seen on the extremities. Aneurysmal fibrous histiocytoma accounts for less than 2% of fibrous histiocytomas.[4]

The clinical appearance of aneurysmal fibrous histiocytoma potentially creates a scenario in which the clinician fears a more severe prognosis. With the rapid changes occurring in the aneurysmal fibrous histiocytoma tumor, one must include conditions such as malignant melanoma, nodular Kaposi's sarcoma, dermatofibrosarcoma protuberans, spindle cell hemangioendothelioma, and angiosarcoma in the differential diagnosis.[6,8,9] While nodular Kaposi's sarcoma can appear clinically similar to aneurysmal fibrous histiocytoma, nodular Kaposi's sarcoma is a multifactorial disease presenting simultaneously with patch and plaque stages.[9] Cutaneous angiosarcoma typically occurs on the scalp in the elderly population and displays dissection of collagen bundles caused by atypical endothelial cells with mitotic figures, which is not expected in aneurysmal fibrous histiocytoma.[10]

Histologically, aneurysmal fibrous histiocytoma varies from the common fibrous histiocytoma, displaying large blood-filled spaces owning up to one half of the tumor. These spaces have a range of possible appearances, from thin clefts to broad gaping cysts lacking an endothelial lining, and can be either focal or involve most of the lesion.[1,4,10] Aneurysmal fibrous histiocytoma usually contains some solid areas with the same hypercellularity found in common fibrous histiocytoma.[1,4] One can visualize numerous small capillaries in the stroma, which have the potential to hemorrhage and deposit hemosiderin.[4] Mitoses can be visualized, but atypical mitotic figures are not expected.[10]

A literature review revealed research on the expected invasion depth of aneurysmal fibrous histiocytoma. In 2011, Han et al. reviewed 122 cases of dermatofibromas to study their histological features, 7% of which were aneurysmal.[11] They analyzed nine cases of aneurysmal fibrous histiocytoma and found that two invaded into the dermis, six invaded into the superficial subcutis, and one invaded into the deep subcutis.[11] Interestingly, Han et al. found that 42.6% of all dermatofibromas in the study invaded into some level of the subcutis.[11] Aneurysmal fibrous histiocytoma, on the other hand, invaded into the subcutis 77.8% of the time.[11] Similarly, Alves et al. found that 81.8% of aneurysmal fibrous histiocytomas invaded into the subcutis.[4]

Because aneurysmal fibrous histiocytoma can present histologically in a variety of ways, one must recognize traits that distinguish it from other vascular or fibrous tumors. Dermatofibrosarcoma protuberans, for example, invades into the subcutaneous fat with a reticular pattern, whereas aneurysmal fibrous histiocytoma invades without a reticular pattern.[6] Nodular Kaposi's sarcoma often presents histologically with angiomatoid and lymphangiomatoid components in the periphery, which may represent early patch and plaque lesions.[9] Also, nodular Kaposi's sarcoma presents with numerous CD34-positive spindle cells that form small spaces with erythrocytes.[10] While both nodular Kaposi's sarcoma and aneurysmal fibrous histiocytoma can be CD34-positive, nodular Kaposi's sarcoma does not have fibrohistiocytic cells.[12] One would also expect to see nuclear positivity for human herpesvirus 8 with nodular Kaposi's sarcoma.[4] Aneurysmal fibrous histiocytoma appears similar to angiomatoid malignant fibrous histiocytoma, as they both present with large blood-filled vascular areas.[10] Angiomatoid malignant fibrous histiocytoma presents with an array of symptoms, such as fever and malaise, and laboratory findings like anemia and hypergammaglobulinemia.[10] On histological evaluation, angiomatoid malignant fibrous histiocytoma displays desmin-positive cells surrounded by a dense lymphohistiocytic and plasma-cell infiltrate.[10]

Aneurysmal fibrous histiocytoma has a good prognosis, but its recurrence rate is up to 19%.[10] This recurrence rate is significantly higher than with common fibrous histiocytoma, which recurs in less than 2% of cases. Most likely this is due to an incomplete removal of the tumor, given its large size, and not a biological component.[6,10] Regular reevaluations are thus recommended to ensure that the aneurysmal fibrous histiocytoma does not recur.

Conclusion

Aneurysmal fibrous histiocytoma is a rare variant of fibrous histiocytoma. While it is benign, the lesion can appear malignant, and one should consider an excisional biopsy to rule out malignant conditions like malignant melanoma, nodular Kaposi's sarcoma, and angiosarcoma. Histologically, aneurysmal fibrous histiocytoma presents with large cavernous blood-filled spaces along with the hypercellularity seen in classic fibrous histiocytoma. Our patient had regular repeated trauma to the lesion due to pressure from a squat bar. We believe this contributed to the rapid growth of the lesion secondary to increased hemorrhage. While not all cases of aneurysmal fibrous histiocytoma will increase in size from repeated trauma, we do consider trauma a potential cause for rapid growth. Given its propensity to recur if not adequately excised, we recommend regular, long-term reevaluations to check for recurrence.

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