Orthopaedic Case of the Month

Pulsatile Soft Tissue Mass

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History and Physical Examination

A 49-year-old healthy woman presented with a mass on the volar aspect of her left proximal forearm. She noticed the mass incidentally while rubbing her arm approximately 3 weeks before the visit. There was no pain associated with

the mass, although, she described a feeling of “fullness” in the area and occasional throbbing. She denied direct trauma to the area, but noted that she probably received contact to that area frequently with her avocation in martial arts.

On physical examination, a 6- by 3-cm mass was present just distal to the antecubital fossa crease. It was soft but not compressible. There was no overlying skin abnormality such as erythema, ecchymosis, or warmth. The mass was visibly and palpably pulsatile. Allen test revealed radial artery dominance with sluggish refill from the ulnar artery.

Laboratory studies, including a complete blood count, electrolytes, and coagulation studies, were normal. The patient was referred to our institution after an MR scan was suspicious for a possible soft tissue sarcoma (Fig. 1). An ultrasound of the mass was performed (Fig. 2).

Based on the history, physical examination, laboratory studies, and imaging studies, what is the differential diagnosis at this point?

Imaging Interpretation

The initial MR scan showed a soft tissue mass in the area of concern that was isointense to muscle on T1-weighted sequences (Fig. 1A) and intermediate to hyperintense in signal intensity compared with surrounding muscle on T2-weighted (Fig. 1B) and STIR sequences (Fig. 1C). A tubular low signal structure seen in the center of the mass on the T1- and T2-weighted images (Figs. 1A, B) was consistent with a central high flow vessel. The mass exhibited diffuse, avid enhancement after administration of gadolinium (Fig. 1D).

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Fig. 1A–D (A) An axial T1-weighted MR image shows a lobulated, relatively well-circumscribed 2×3.4 (axial) $\times$ 4.5-cm mass in the proximal forearm (arrows), that is isointense to muscle in signal intensity. A central low signal focus (arrowhead) represents a high-flow vessel in the mass. (B) This coronal T2-weighted MR image shows an elongated, relatively well-circumscribed mass in the proximal forearm (arrow) that is of intermediate signal intensity,

slightly hyperintense compared with surrounding muscle. A tubular low signal area seen centrally (arrowhead) represents a high-flow vessel. (C) On this STIR image, the mass (arrow) is of intermediate signal intensity, slightly hyperintense compared with surrounding muscle, and slightly heterogeneous, with trace surrounding edema. (D) A postcontrast fat saturated T1-weighted image shows diffuse, slightly heterogeneous enhancement throughout the mass (arrow).

Fig. 2A–B (A) The ultrasound images show a mass (arrows) with a large, high flow vessel (asterisks) and (B) only moderate peripheral vascularity (arrowhead) in the remainder of the mass.

The ultrasound showed a mass that appeared to surround the ulnar artery (Fig. 2A) with relatively moderate peripheral vascularity (Fig. 2B). An arteriogram (Fig. 3A) showed direct filling of the lesion from a tortuous and irregular ulnar artery with rapid shunting to the venous system through the mass. There was radial-dominant arterial supply to the hand, though the distal ulnar artery remained patent (Fig. 3B).

Differential Diagnosis

- Pseudoaneurysm
- Arteriovenous malformation
- Epithelioid hemangioma
- Angiosarcoma
- Epithelioid hemangioendothelioma

Fig. 3A–B (A) The digital subtraction arteriogram shows an irregular and tortuous proximal ulnar artery (arrowhead) with associated soft tissue tumor blush (arrow). There were multiple tortuous ulnar artery branches and rapid arteriovenous shunting into the cephalic vein. Findings were suggestive of a vascular tumor. (B) The remaining vessels in the forearm were normal. Note was made of the dominant radial arterial supply to the hand (thick arrow), but the distal ulnar artery remained patent (thin arrow).

An open biopsy of the mass was performed (Fig. 4). Based on the history, physical examination, laboratory studies, imaging studies, and histologic picture, what is the diagnosis and how should the patient be treated?

Histology Interpretation

The gross specimen had a homogenous, pale, cut surface. Microscopically, the lesion was poorly circumscribed and showed a proliferation of well-formed vascular spaces with an admixed, brisk inflammatory infiltrate (Fig. 4A). Examination at higher power showed these vascular spaces were lined by plump, cuboidal endothelial cells that were uniform in their cytology without evidence of malignancy. The inflammatory infiltrate was polymorphic with a mixture of lymphocytes and plasma cells; eosinophils were conspicuous (Fig. 4B).

Diagnosis

Epithelioid hemangioma

Discussion and Treatment

The finding of a tumor associated with a fairly large vessel and a history of repetitive trauma points to epithelioid hemangioma. Histologically, this was confirmed by the presence of vascular channels lined with benign epithelioid endothelial cells.

Fig. 4A–B (A) This photomicrograph shows the poorly circumscribed nature of the tumor microscopically. The well-formed vascular spaces have intervening inflammatory infiltrate (Stain, hematoxylin and eosin; original magnification, $\times 20$). (B) At higher

magnification, the cells lining the vascular spaces are benign-appearing endothelial cells. The inflammatory infiltrate is polymorphic with eosinophils predominant (Stain, hematoxylin and eosin; magnification, $\times 200$).

A pseudoaneurysm could result from repetitive trauma and also could present as a mass abutting or surrounding a vessel on imaging [9]. However, it would be more common to see a pseudoaneurysm after penetrating trauma as opposed to blunt trauma as in this case. Moreover, a pseudoaneurysm would show a characteristic swirling pattern of central blood flow on ultrasound (yin-yang pattern) and would have a discrete neck of blood flow connecting it to the artery [9]. In addition, a pseudoaneurysm would not show the tumor blush seen on this patient's angiogram (Fig. 3A). Furthermore, histologic analysis showed a solid tumor, not a nonneoplastic space filled with blood and organizing thrombus as one would expect for a pseudoaneurysm. An arteriovenous malformation could be a cause of a pulsatile soft tissue mass. However, the patient probably would have presented at a younger age with this condition since they are congenital and in this location would be noticeable. The imaging also would differ in that one would expect even more rapid filling of the ectatic vessels composing the lesion [9]. Histologically, the lesion would be composed of an admixture of medium-sized arteries and veins with abnormal perivascular smooth muscle as opposed to the proliferation of capillary-sized vessels with cuboidal endothelial cells with the admixed, eosinophil-predominant inflammation as in this case. Angiosarcoma and hemangioendothelioma are malignant vascular tumors. They can be excluded from the differential diagnoses mainly owing to the lack of cellular atypia histologically [3]. Angiosarcoma also tends to be multinodular, which was not the case in our patient. Hemangioendothelioma also may be intravascular rather than being located adjacent to a vessel as was the case here [3]. On imaging, angiosarcomas and hemangioendotheliomas show a network of prominent serpentine vessels, often at the periphery of the mass, and show arteriovenous shunting on Doppler ultrasound [9]. This pattern is distinct from the single, irregular ulnar artery surrounded by tumor blush seen in our patient.

Epithelioid hemangioma (EH) was first described by Wells and Whimster in 1969 [23] as "subcutaneous angiolymphoid hyperplasia with eosinophilia" (ALHE) and definitively differentiated from a similar lesion, Kimura's disease, by Rosai et al. in 1979 [16] and Urabe et al. in 1987 [19]. The term epithelioid hemangioma was suggested as a replacement for ALHE by Enzinger and Weiss in 1988 [4] "to reflect the belief that most were benign neoplasms and to clearly delimit these lesions from the more aggressive tumor, epithelioid hemangioendothelioma, a neoplastic process with metastatic potential" [6]. Errani et al. [5] recently reported that a chromosomal translocation, t(1;3)(p36;q25) identified in epithelioid hemangioendothelioma is not present in EH and thus can be used to differentiate these two entities. However, EH and epithelioid

hemangioendothelioma are histologically distinct, so cytogenetic analysis is rarely, if ever, indicated for accurate diagnosis.

It has been debated whether EH is a true neoplasm or reactive secondary to trauma to a vessel and repair therein. A history of trauma often can be elicited from the patient or can be inferred from changes in the vessel with which the tumor is associated. Fetsch and Weiss [6] reviewed 96 cases of EH and found that in the majority of cases, the mass was associated with a vessel and that the vessel showed clear signs of damage microscopically. They surmised that these findings pointed to a reactive, nonneoplastic origin for most cases of EH, perhaps in response to trauma. This was supported by Vadlamudi and Schinella [20] who noted traumatic pseudoaneurysms in four patients with a clear history of trauma. The masses in these patients were similar to classic EH lesions, but possibly earlier in a spectrum of development, perhaps indicating again a nonneoplastic reactive pathogenesis. Sun et al. [18] proposed that local hypoxia, often associated with trauma, may be important in the pathogenesis of EH.

Most cases of EH are superficial and arise in areas prone to trauma, such as the face and scalp. Only approximately 4% involve the deep soft tissues [4, 6, 16, 19, 23]. There are few reports of EH or ALHE involving a larger vessel in an extremity. A literature search resulted in only 13 in which this was clearly the case [1, 2, 7, 8, 10–15, 17, 21, 22]. Of these, nine were isolated and four arose in the setting of multiple associated superficial EH lesions in the regional subcutaneous and dermal tissues. Eleven were in an upper extremity and only two were in a lower extremity. All were treated with marginal excision including the vessel from which they arose. This required reconstruction of the vessel in all cases except one in which a ray amputation was necessary because of extensive involvement of the distal ulnar artery with limited ability to reconstruct such a small vessel. None of these masses recurred in the deep tissues with followup of 2 to 18 months. In the cases in which they were associated with several subcutaneous lesions, the superficial lesions often recurred or new lesions developed, without recurrences in the deeper tissues. Superficial EH have a reported rate of local recurrence of between 12% and 44% at 1 to 2 years [3, 4, 6, 16, 19, 23]. The collected reports of these deeper lesions associated with larger vessels, however, do not show the same tendency and so they may have a different natural history. Further followup and a truly systematic review would be necessary to determine this definitively.

With our patient, the tumor was excised with a marginal margin along with the involved portion of the ulnar artery. It was decided not to reconstruct the ulnar artery based on normal hand vascularity after excision and based on how small and atrophic the ulnar artery had become

preoperatively. Three lymph nodes located in the adjacent area were removed at the same time and showed no evidence of EH. The patient's wound healed without complication and the patient has returned to all preoperative normal functions. At last followup 4 months postoperatively she had no symptoms of dysvascularity, such as claudication, related to the removal of the atrophic ulnar artery. Her motor and sensory function were normal and aside from the absence of the ulnar pulse, a normal vascular examination. She had no evidence of disease recurrence on MR scans. She will be followed with MRI and physical examinations for local recurrence periodically for the first 2 years after surgery as most local recurrences, at least in superficial EH, occur within that time [4, 19, 23].

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