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Angiosarcoma of the hand associated with pseudoaneurysm

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ABSTRACT

Angiosarcoma is a rare malignancy of vascular endothelial origin. We describe a case of angiosarcoma of the hand initially histopathologically diagnosed as a pseudoaneurysm, emphasizing the diagnostic importance of radiological pathologic concordance. Here we highlight the distinctive imaging and the histopathologic features of angiosarcoma, invaluable to its accurate and timely diagnosis.

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Introduction

Angiosarcoma is a rare, malignant tumor derived from the vascular or lymphatic endothelium. Angiosarcoma most frequently involves the head and neck, breast, and soft tissues, and comprises 2%-3% of all soft-tissue sarcomas [1]. Even though most cases are idiopathic, some are associated with chronic lymphedema or ionizing radiation [1]. Less common associations with foreign bodies and pseudoaneurysms have also been described. Soft-tissue angiosarcomas are aggressive tumors with a 1-year mortality rate of 53% [2]. Angiosarcomas are composed of malignant endothelial cells that demonstrate a range of cytologic atypia. Histologically, angiosarcomas are characterized by varying degrees of vasoformation [3]. Although imaging may help diagnose and delineate the extent of tumor and help treatment planning, definitive diagnosis requires biopsy. We discuss the imaging and pathologic findings of an angiosarcoma of the hand associated with a pseudoaneurysm.

Case report

A 72-year-old white man with no significant medical history presented with persistent bleeding from a left-hand wound.

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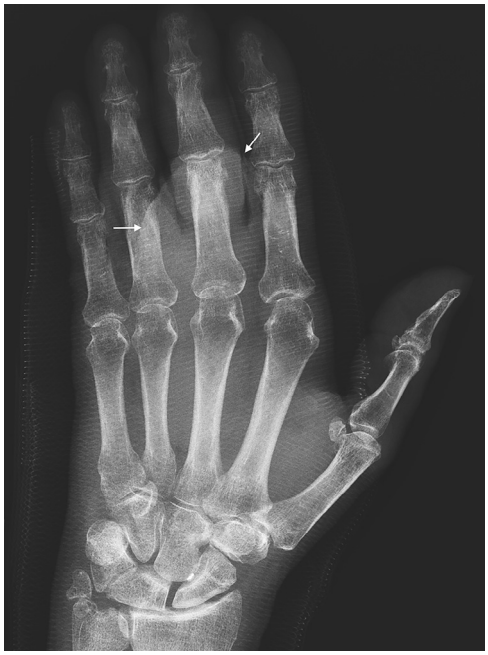

Fig. 1 – Frontal radiograph of the left hand shows soft-tissue swelling related to a mass (arrows) overlying the third and the fourth proximal phalanges. There is no bone erosion.

The patient reported a firm bump on his hand for the past year. Two months before, the patient felt a popping sensation in his left hand while lifting a heavy object and developed a persistent swelling in his left hand. Surgical exploration was performed in which multiple blood clots were removed. The patient presented to our institution due to persistent bleeding from the incision despite repeat surgical exploration.

Radiographs revealed soft-tissue swelling at the dorsum of the second through fourth proximal phalanges (Fig. 1). Magnetic resonance imaging (MRI) showed large hematomas of the hand with areas of nodular enhancement somewhat suspicious for sarcoma (Fig. 2). A thrombosed pseudoaneurysm is also seen at the palmar aspect of the hand on MRI (Fig. 3). Initial ultrasound-guided biopsy targeted toward enhancing components dorsally was composed histologically of vascular proliferation and inflammation suggestive of a benign reactive change and hematoma (Fig 4A). Subsequent surgical biopsy removed a palpable nodule, which was consistent with pseudoaneurysm pathologically. It was then thought that the recurrent bleeding may have been due to this pseudoaneurysm, with chronic hematoma mimicking tumor.

An excisional biopsy, performed a month later due to continued bleeding, demonstrated conclusive features of epithelioid angiosarcoma. Whole-body F18-fluorodeoxyglucose positron emission tomography-computed tomography was then performed and revealed an irregular hypermetabolic soft-tissue mass in the left hand consistent with malignancy, as well as left axillary hypermetabolic lymph nodes (Fig. 5). The patient underwent a left-hand and forearm amputation, exhibiting a high-grade malignant neoplasm in the deep muscular plane of the hand composed of variably vasoformative, well-differentiated, and poorly differentiated solid tumor

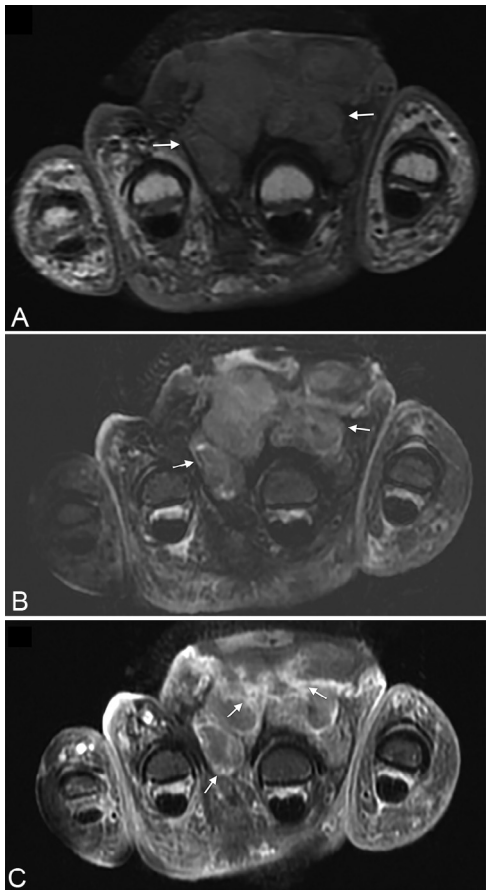

Fig. 2 – Axial magnetic resonance images of the hand using T1-W (TR: 700, TE: 10) (A), T2-W fat saturated (TR: 3405, TE: 55) (B), postcontrast T1-W (TR: 700, TE: 10) show a lobulated T1 and T2 hypointense mass with a predominantly peripheral and somewhat nodular enhancement (arrows) extending between the dorsal third and fourth proximal phalanges. T1-W, T1-weighted; T2-W, T2-weighted; TE, echo time; TR, repetition time.

histopathologies. The well-differentiated portions showed hemorrhagic irregular anastomosing vascular channels lined by hyperchromatic atypical endothelial cells, multilayering, intraluminal tufting, and infiltration through the subcutaneous

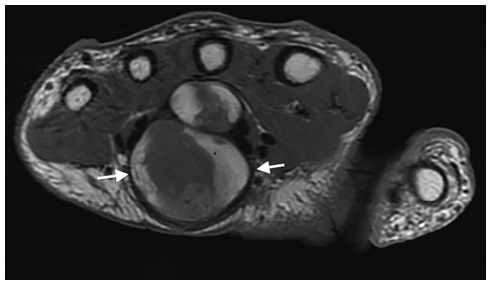

Fig. 3 – Axial T1-W (repetition time: 700, echo time: 10) image shows a thrombosed pseudoaneurysm with peripheral T1 hyperintensity in the volar aspect of the hand (arrows). No enhancement was present (not shown).

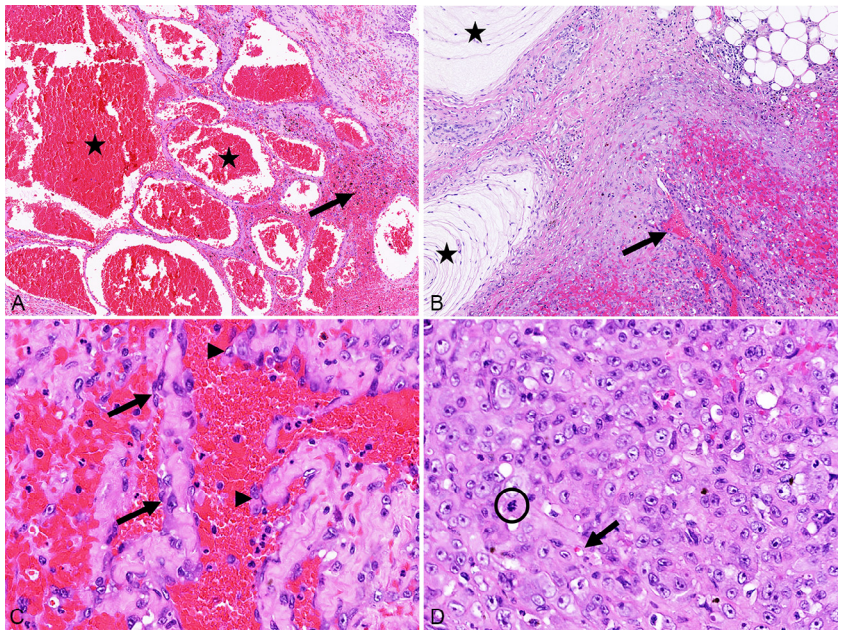

Fig. 4 – Epithelioid angiosarcoma. (A) Reactive appearing thin-walled vascular spaces (stars) with hematoma and hemosiderin-laden macrophages (arrow) in the surrounding soft tissue (hematoxylin and eosin, original magnification 50×). (B) Poorly demarcated hemorrhagic irregular anastomosing vascular channels (arrow) dissecting through the subcutaneous fibroadipose tissue adjacent to the Pacinian corpuscles (stars), characteristic of epithelioid angiosarcoma (hematoxylin and eosin, original magnification 100×). (C) A well-differentiated area of epithelioid angiosarcoma revealing irregular vascular channels lined by hyperchromatic atypical endothelial cells with multilayering (arrows) and intraluminal tufting (arrowheads) (hematoxylin and eosin, original magnification 400×). (D) Sheets of large atypical epithelioid endothelial cells with irregular vesicular nuclei, prominent basophilic nucleoli, and an abundant eosinophilic cytoplasm with focal intracytoplasmic lumina (arrow) demonstrating mitotic activity (circle) (hematoxylin and eosin, original magnification 400×).

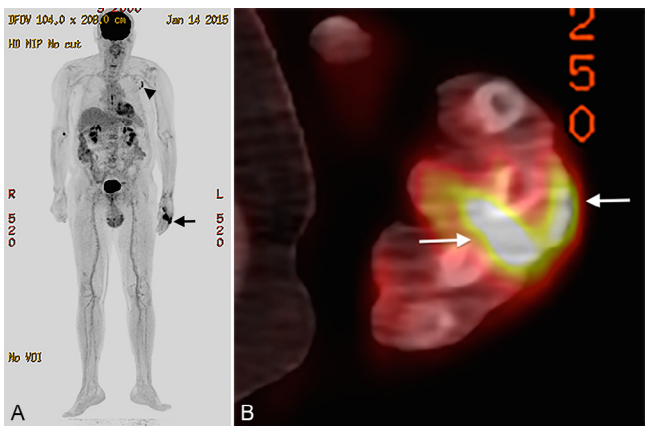

Fig. 5 – Whole-body positron emission tomography (A) and fused axial F18-fluorodeoxyglucose positron emission tomography-computed tomography (B) image shows the increased metabolic activity of the tumor of the hand extending between the third and the fourth proximal phalanges (arrows), as well as the mild increased uptake in the left axillary lymph nodes (arrowhead). Focal uptake in the right elbow represents the injection site.

fibroadipose tissue (Fig. 4B and C). The poorly differentiated areas showed solid sheets of large atypical epithelioid endothelial cells with irregular vesicular nuclei, prominent basophilic nucleoli, and an abundant eosinophilic-amphophilic cytoplasm with focal intracytoplasmic lumina and conspicuous mitosis (Fig. 4D) intermingled among more spindled endothelial cells associated with significant necrosis and inflammation. Immunohistochemical stains showed focal positivity for CD31 and diffuse nuclear positivity for ERG (V-ets erythroblastosis virus E26 oncogene homologue) within the malignant epithelioid cells. Although vascular invasion was identified, the final surgical resection margins and axillary lymph nodes were negative. However, 9 months following the amputation, the patient developed a recurrence at the dorsum of the left forearm. An above-elbow amputation was performed and chemotherapy is being considered.

Discussion

Angiosarcoma is a highly aggressive malignancy of endovascular origin. Soft-tissue angiosarcoma comprises 10% of all angiosarcomas, which also includes cutaneous angiosarcoma, angiosarcoma associated with lymphedema, breast angiosarcoma, bone angiosarcoma, and radiation-induced angiosarcoma [2]. Soft-tissue angiosarcoma can occur at any age with even

distribution throughout all ages [2]. The mortality rate of soft-tissue angiosarcoma is 53% within 1 year, with local recurrence in 20% and distant metastasis in lung, lymph nodes, bone, and soft tissue in 50% [2]. Risk factors include radiation, exposure to polyvinyl chloride and thorotrast contrast, and chronic lymphedema [4]. Angiosarcoma has been reported to develop near joint replacements, vascular grafts, or foreign bodies, such as a bullet, gauze, or beeswax. However, the majority of cases are idiopathic, and our patient had no known risk factors for angiosarcoma. Additionally, angiosarcoma associated with pseudoaneurysm has been reported in the popliteal, carotid, coronary, and pulmonary arteries [5–9].

A recent study identified mutations in 2 specific gene sequences. Twenty-six percent of investigated angiosarcomas contained mutations in the protein tyrosine phosphatase, receptor type B (*PTPRB*) gene, a tyrosine phosphatase of vascular endothelium that has an antiangiogenetic property, specifically in secondary angiosarcomas [10]. A gene sequence identified in 20% of angiosarcoma cases is phospholipase C, gamma 1 (*PLCG1*), which encodes a protein that transduces the tyrosine kinase signal in the phosphoinositide signaling pathway [10]. These findings may have therapeutic implications.

MRI is useful in diagnosis and local staging. T2-weighted signal intensity may be increased or decreased based on the amount of vascular spaces, tumor cells, and fibrosis [4]. Imaging findings of angiosarcoma may also differ depending on the etiology. In the Stewart-Treves syndrome, which is an angiosarcoma associated with chronic lymphedema, MRI shows a low signal intensity on T1-weighted images and high signal intensity on T2-weighted images, and heterogeneous enhancement following gadolinium contrast administration [4]. A heterogeneous, increased signal on the T1-weighted image indicates an ample blood supply with predisposition to internal hemorrhage [11]. F18-fluorodeoxyglucose positron emission tomography-computed tomography helps differentiate between malignant and benign lesions and also aids in staging and assessing response [12].

Ultimately, a tissue biopsy is needed for a definitive diagnosis of angiosarcoma. Nonrepresentative biopsy material, as in our case initially, may easily lead to misdiagnosis as chronic hematoma [2]. Although hematomas are usually gradually reabsorbed, they sometimes persist as chronic organized hematoma surrounded by a fibrous wall. On the other hand, angiosarcomas and other sarcomas can lead to hematoma formation after intratumoral hemorrhage [13]. On imaging, chronic hematoma also can be difficult to differentiate from malignancy. MRI features that favor tumor are a heterogenous T1-weighted signal intensity, necrosis, bone or neurovascular involvement, and a mean diameter of 66 mm [14]. However, chronic hematoma can also have a heterogeneous T1-weighted signal intensity. Nodular enhancement is suspicious for tumor; however, chronic hematomas may also have some enhancement due to fibrovascular tissue [15]. Contrast can also diffuse into hematomas on delayed images, mimicking central enhancement [15]. Because hematomas can occur in conjunction with underlying tumors, any hematoma with nodular areas of soft-tissue enhancement needs to be monitored to resolution or biopsied to exclude an underlying tumor [16]. Additionally, angiosarcoma has also been reported to be associated with chronic expanding hematomas [17].

In conclusion, angiosarcoma of the hand is rare and prompt diagnosis is important. Imaging is useful for diagnosis and staging, including preoperative surgical planning. In some cases, angiosarcoma may be associated with a pseudoaneurysm. Determining the concordance between pathologic and radiological results is important for timely diagnosis.

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