

Painless Angioleiomyoma of the First Web Space of the Hand

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Abstract. *Angioleiomyoma is a benign dermal or subcutaneous tumor originating from the tunica media of small veins and arteries, and rarely occurs in the hand. Because of its non-specific imaging features, a definite preoperative diagnosis is quite difficult. We present an unusual case of angioleiomyoma arising in the first web space of the right hand of a 56-year-old male. Physical examination showed a 3-cm, elastic-soft, mobile, non-tender mass. Magnetic resonance imaging (MRI) revealed a well-demarcated, subcutaneous, soft tissue mass with iso- to slightly high signal intensity relative to skeletal muscle on T1-weighted sequences and heterogenous high signal intensity with scattered foci of low signal intensity on T2-weighted sequences. Contrast-enhanced fat-suppressed T1-weighted sequences demonstrated heterogenous, strong enhancement throughout the mass. There was no vascular structure closely abutting the mass. Simple excision of the mass was performed, and the histology was characteristic of an angioleiomyoma. The patient had no evidence of local recurrence within four months of follow-up. Angioleiomyoma should be considered in the differential diagnosis of a well-defined, oval soft tissue mass in the first web space of the hand even when an adjacent tortuous vascular structure is not seen on MRI.*

Angioleiomyoma, also known as vascular leiomyoma, is a benign soft tissue tumor composed of mature smooth muscle cells arranged around many vascular channels. It belongs to the pericytic (perivascular) tumor group according to the 2013 World Health Organization Classification of Soft Tissue Tumors (1). Angioleiomyoma usually occurs in the fourth to six decades of life, with a predominance in females. The most common presentation is of a small, slow-growing, firm,

painful nodule arising in the extremities, particularly the lower leg (2). There are no specific imaging techniques for angioleiomyoma. Simple excision is curative and local recurrence is rare. The etiology of this condition remains unknown.

The preoperative diagnosis of angioleiomyoma in the hand is quite difficult (3). Herein we report an unusual example of angioleiomyoma without definite vascular involvement in the first web space of the hand.

Case Report

A 56-year-old, right-hand dominant male was referred to our hospital with a 3-year history of a slow-growing, painless mass in the first web space of the right hand. There was no history of antecedent trauma. Physical examination showed an elastic-soft, mobile, non-tender mass, measuring approximately 3.0×3.0 cm. No local heat or redness of the mass was present. Neurological and vascular examinations were unremarkable. The patient's medical history was noncontributory.

Plain radiographs of the right hand revealed an increased soft tissue shadow in the first web space (Figure 1). Magnetic resonance imaging (MRI) showed a well-demarcated ovoid soft tissue mass in the subcutis. The mass exhibited iso- to slightly high signal intensity relative to skeletal muscle on T1-weighted sequences (Figure 2A) and heterogenous high signal intensity with scattered foci of low signal intensity on T2-weighted sequences (Figure 2B). Contrast-enhanced fat-suppressed T1-weighted sequences demonstrated heterogenous, strong enhancement of the mass (Figure 2C). There was no vascular structure closely abutting the mass. Our preoperative differential diagnosis included schwannoma, hemangioma, and tenosynovial giant cell tumor.

The operative procedure was performed under general anesthesia with tourniquet control and loupe magnification. A gently curved longitudinal incision was made on the volar aspect of the first web space of the right hand. The mass was encapsulated and did not involve the flexor tendon sheath, local vasculature, or digital nerves. It was easily dissected from surrounding tissues. Microscopically, the tumor was composed of a proliferation of smooth muscle cells, which

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Figure 1. Plain radiograph of the right hand reveals an increased soft tissue shadow in the first web space.

were interspersed with many blood vessels (Figure 3A). Neither cellular atypia nor mitotic figures were observed. Immunohistochemically, the tumor cells were diffusely positive for smooth muscle actin (SMA) (Figure 3B), muscle-specific actin (MSA), calponin, and h-caldesmon (Figure 3C), and focally for desmin (Figure 3D). Based on these features, the tumor was diagnosed as an angioleiomyoma.

The postoperative course was uneventful. At the four-month follow-up, the patient was doing well without evidence of local recurrence.

Discussion

Angioleiomyoma is uncommon in the hand. Unlike angioleiomyomas located elsewhere, those arising in the hand are less commonly painful, have an equal gender distribution, and are not predominantly of the solid type as seen in the lower extremity (3). Most hand angioleiomyomas show a direct relationship to veins or arteries (4-7). In the current case, however, we were unable to demonstrate any vascular structure closely abutting the mass.

The MRI features of angioleiomyoma are non-specific. The lesions are usually well-defined, round, or oval masses in a subcutaneous or dermal location. On MRI, the tumors are usually isointense or slightly hyperintense relative to skeletal muscle on T1-weighted images and have a heterogeneous appearance on T2-weighted images (8-10).

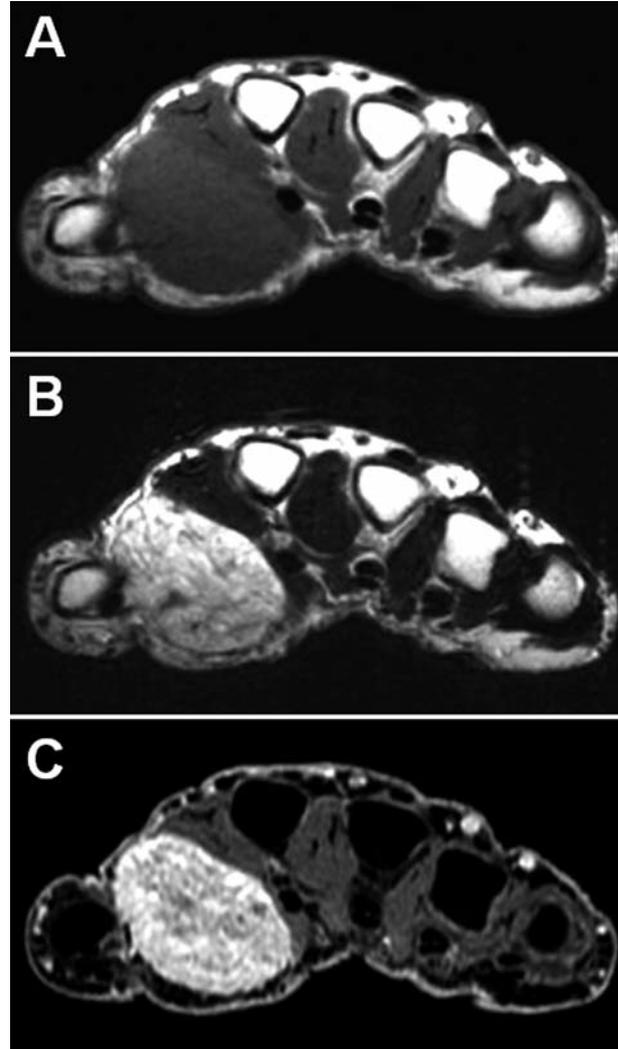


Figure 2. Axial magnetic resonance images of angioleiomyoma in the right hand. A: T1-Weighted sequence shows that the mass has iso- to slightly high signal intensity relative to skeletal muscle. B: T2-Weighted sequence shows that the mass has heterogeneous high signal intensity with scattered foci of low signal intensity. C: Contrast-enhanced fat-suppressed T1-weighted sequence shows heterogeneous, strong enhancement of the mass.

The high signal intensity areas on T2-weighted images have a linear or branching pattern, corresponding to numerous patent vessels (9). It is likely that a peripheral low signal intensity area on T1- and T2-weighted images corresponds to a fibrous capsule. With administration of intravenous contrast material, a marked enhancement with a heterogeneous or homogenous pattern is observed (9, 10). In the current case, the preoperative radiological differential diagnosis included benign neurogenic or vascular tumors. It is difficult to distinguish angioleiomyoma from benign neurogenic tumors in patients with atypical clinical features.

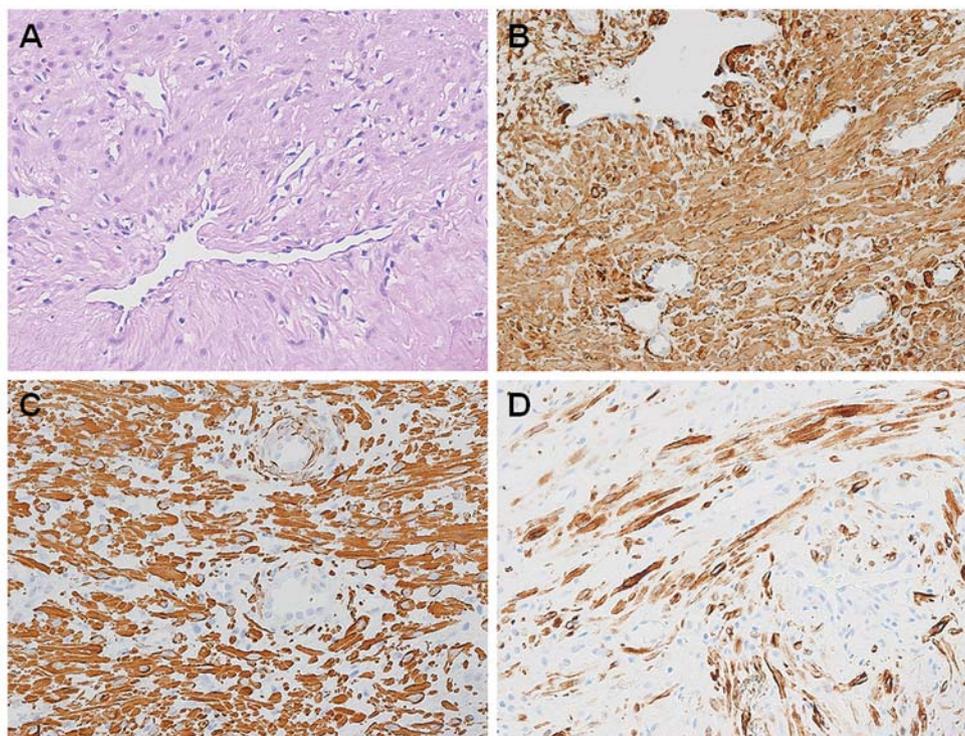


Figure 3. *Histological and immunohistochemical findings of angioleiomyoma. A: The tumor is composed of well-differentiated smooth muscle cells with intervening vascular channels (Hematoxylin and eosin staining, original magnification $\times 120$). The tumor cells are diffusely positive for smooth muscle actin (B) and h-caldesmon (C), and focally for desmin (D) (B, C, and D, original magnification $\times 100$).*

Histologically, the lesion is composed of well-differentiated smooth muscle cells with intervening vascular channels. Areas of myxoid change, hyalinization, calcification, and mature fat are occasionally present. Nerve fibers may be seen adjacent to or within the capsule (3). Based on a predominant histological pattern, angioleiomyomas are classified into solid, venous, and cavernous types (1). The solid form is the most common subtype (2, 11), and each histological subtype has different clinical features. In the current case, the histological features of both solid and cavernous types were seen. Immunohistochemically, the tumor cells are consistently and diffusely positive for SMA, MSA, and calponin and variably for h-caldesmon and desmin (11), as shown in the current case.

Cytogenetic abnormalities have been described in only six cases of angioleiomyoma (12-16). Angioleiomyomas are usually diploid with no consistent chromosomal aberrations. Previously, we applied comparative genomic hybridization to angioleiomyomas to screen for gains and losses of DNA sequences (17). Eight (35%) out of 23 cases exhibited DNA copy number changes involving one or two chromosomes. The most common recurrent change was a loss in 22q11.2 that may play a role in the development of angioleiomyoma.

In summary, the current case illustrates that angioleiomyoma can occasionally grow to a relatively large

size without pain. Angioleiomyoma should be considered in the differential diagnosis of a well-defined, oval soft tissue mass in the first web space of the hand, even when an adjacent tortuous vascular structure is not seen on MRI.

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