CASE REPORT

Giant Angioleiomyoma in Extremity: Report of Two Cases

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Angioleiomyomas are smooth muscle tumors that occur most often in the extremities and are usually less than 2 cm in diameter. We present 2 case reports of giant angioleiomyoma. Case 1 was a 51-year-old woman with a 5-year history of a slowly growing mass 12 × 10 cm in diameter in the right posterior thigh. The tumor was larger than any previously reported. Case 2 was a 72-year-old man with an 8-year history of a slowly growing mass 6 × 6 cm in diameter in the left palm. The tumor was unique in that it grew to a giant size in the hand of the elderly patient. Magnetic resonance imaging of both cases confirmed the tumors' presence in the subcutis. T1-weighted imaging revealed homogenous signal isointense to muscle. On T2-weighted images, areas of hyperintensity and hypointensity intermingled. Three-dimensional gadolinium-enhanced magnetic resonance angiography showed rapid initial enhancement followed by a gradual increase in enhancement, suggesting that the tumors were rich in vessels. Microscopic examination revealed numerous vascular channels embedded within a huge smooth muscle component.

Keywords: angioleiomyoma, extremity, MRI, 3D Gd-enhanced dynamic MRA

Introduction

Angioleiomyoma is a distinctive, relatively common, benign subcutaneous neoplasm composed of smooth muscle and thick-walled vessels. The tumors are most frequently found in persons between 30 and 60 years of age and most often in women. Usually occurring in an extremity, the tumor most often presents as a small (<2 cm), slowly growing mass of several years' duration.1 The tumor is usually round, forming a nodule that generally elevates the skin. Hachisuga’s group reported 562 cases of angiomyoma,2 with tumor ranging from 0.2 to 4.3 cm at the widest diameter, most (78%) less than 2 cm in diameter. To our knowledge, there are few reports of giant angioleiomyoma, and there is no report of examination with 3-dimensional gadolinium-enhanced dynamic magnetic resonance angiography (3D-MRA).

Case Report

Case 1

A healthy 51-year-old woman with no antecedent trauma presented with a 5-year history of a slowly growing mass in the right posterior thigh. The mass was hard, elastic, painless, tender, freely movable, and approximately 12 × 10 cm in diameter. The overlying skin was raised, thinned, and tan colored (Fig. 1). Magnetic resonance imaging (MRI) confirmed the tumor’s presence in the subcutis. T1-weighted imaging revealed a homogenous, well demarcated tumor with a signal isointense to muscle. On T2-weighted imaging, there were intermingled areas of hyperintensity and hypointensity, a well defined peripheral area of hypointensity, and multiple tubular hypointense structures (arrow) peripheral to the mass (Fig. 2A). On T2-weighted imaging, there were intermingled areas of hyperintensity and hypointensity, a well defined peripheral area of hypointensity, and multiple tubular hypointense structures (arrow) peripheral to the mass (Fig. 2B). A contrast-enhanced T1-weighted image revealed strong, heterogeneous enhancement (Fig. 2C). 3D-MRA showed a hypervascular ovoid tumor fed by a branch of the superficial femoral artery (arrow). The dynamic pattern was rapid initial enhancement followed by a gradual increase in enhancement,
suggesting that the tumor was rich in vessels (Fig. 3A-C).

Wide resection was performed because angioleiomyoma was diagnosed by open biopsy. The fascia that adhered to the tumor was exfoliated from the biceps femoris muscle. The resected mass was grossly whitish. Histopathologic examination revealed the characteristic appearance of angioleiomyoma: smooth-muscle thickening of the walls of some of the small vessels and edematous stroma with degeneration (Fig. 4).

**Case 2**

A healthy 72-year-old man with no remarkable medical history presented at our hospital with a tumor in the left palm that had been slowly growing for about 8 years. On physical examination, a hard, elastic, painless, and fixed tumor about 6 × 6 cm in diameter was detected in the left palm. The overlying skin was raised and tan colored. Flexion was moderately restricted at the metacarpophalangeal and proximal interphalangeal joints of the middle finger (Fig. 5).

MRI confirmed the presence of the tumor in the subcutis. The deep part of the tumor was between the third and fourth fingers, with no invasion of the bone. The superficial border of the tumor was comparatively clear; however, the deep border was poorly defined and adjacent to the third and fourth flexor digitorum tendon. A T1-weighted image

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**Fig. 1.** A hard, elastic, painless, tender, freely movable tumor about 12 × 10 cm in diameter is detected. The overlying skin is raised, thinned, and tan colored.

**Fig. 2.** Magnetic resonance imaging confirms the presence of the tumor in the subcutaneous tissue. **A:** A T1-weighted image reveals a homogenous, poorly demarcated soft tissue tumor with a signal isointense to the muscle. **B:** Areas of hyperintense and hypointense signals are intermingled, and a well-defined peripheral hypointense area and multiple tubular hyperintense structures (arrow) peripheral to the mass are observed on a T2-weighted image. They are flow void peripheral to the interior of the mass. **C:** The contrast-enhanced T1-weighted image is strong and heterogeneous.
Fig. 3. Three-dimensional magnetic resonance angiography is acquired 25 (A), 60 (B), and 150 (C) s after the administration of gadolinium-diethylenetriamine pentaacetic acid. The angiography shows a hypervascular ovoid tumor fed by a branch of the superficial femoral artery (arrow). The enhanced pattern is rapid initial enhancement followed by a gradual increase in enhancement, suggesting that the tumor is rich in vessels.

Fig. 4. The smooth-muscle thickening of the walls of some of the small vessels and edematous stroma with degeneration is observed. (hematoxylin and eosin, ×40)

Fig. 5. A hard, elastic, painless, fixed tumor about 6×6 cm in diameter is detected in the left palm. The overlying skin is raised and tan colored.

showed a homogenous, poorly demarcated tumor with a signal isointense to muscle (Fig. 6A). A T2-weighted image revealed intermingled areas of hyperintensity and hypointensity and multiple dot-like or tubular hypointense structures (arrow) inside the mass (Fig. 6B). A contrast-enhanced T1-weighted image revealed strong, heterogeneous enhancement (Fig. 6C). 3D-MRA revealed a hypervascular ovoid tumor fed by a branch of the ulnar artery (arrow). The dynamic pattern was rapid initial enhancement followed by a gradual increase in enhancement, suggesting that the tumor was rich in vessels (Fig. 7A-D).

Marginal resection was performed because angi-
MR imaging confirmed the presence of the tumor in the subcutaneous tissue. The deep part of the tumor is between the third and fourth fingers, but there is no invasion of the bone. A: A T1-weighted image reveals a homogenous, well demarcated soft tissue mass with a signal isointense to muscle. B: On a T2-weighted image, there are areas of intermingled hyperintensity and hypointensity and multiple dot-like or tubular hypointense structures (arrow) inside the mass. They are flow void peripheral to the interior of the mass. C: The contrast-enhanced T1-weighted image reveals strong, heterogeneous enhancement.

Angioleiomyoma was diagnosed by biopsy. After the subcutaneous tissue was divided, the capsule that adhered to the third and fourth flexor digitorum tendon was exposed. The tendons were exfoliated from the capsule. The resected tumor was grayish in color. Histopathologic examination revealed the characteristic appearance of angioleiomyoma—numerous vascular channels embedded within a huge smooth muscle component (Fig. 8).

Discussion

Hachisuga’s group reported 562 cases of angioleiomyoma, in which tumor size at excision ranged from 0.2 to 4.3 cm at the widest diameter, most (78%) less than 2 cm in diameter. The angioleiomyoma of Case 1 was 12×10 cm in diameter in the right posterior thigh. A review of the literature indicated it to be larger than any previously reported. The angioleiomyoma in the hand of Case 2 was 6×6 cm in diameter. Calle and associates reviewed 105 cases of vascular leiomyoma (so-called angioleiomyoma) in the hand, in which the mean size of the tumors was 1.5 cm in diameter. We found only 2 case reports in which a tumor in the hand was more than 4 cm in diameter. We assumed that the reason that our patients' tumors were so large was because they caused no pain and were slow growing. Freedman and colleagues reviewed the relationship between size and pain, however, in 39 angioleiomyomas in the extremities. They reported that the mean size of the lesions in the symptomatic group (0.94 cm) did not differ from that in the asymptomatic group (0.95 cm). Hachisuga's group also reported that the ages of patients ranged from 12 to 84 years (average, 47 years), and 67% of the patients were in their fourth, fifth, and sixth decades. Angioleiomyomas are found throughout the body, but are observed most often in the lower extremities. Angioleiomyomas of the hand are rare, accounting for approximately 10% to 17% of all such tumors. Thus, Case 2 was relatively rare regarding location and age.

MRI in both cases revealed an almost well defined round mass in the subcutis of homogeneously low intensity on T1-weighted images, and areas of hyperintensity and hypointensity areas intermin-
Fig. 7. Three-dimensional magnetic resonance angiography (3D-MRA) is acquired at 25 (A), 60 (B), 150 (C), and 270 (D) s after gadolinium-diethylenetriamine pentaacetic acid. A-D, 3D-MRA shows a hypervascular ovoid tumor fed by a branch of the ulnar artery (arrow). The enhanced pattern is rapid initial enhancement followed by a gradual increase in enhancement.

Fig. 8. Microscopic findings show numerous vascular channels embedded within a huge smooth muscle component. (Hematoxylin and eosin, ×20)

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the early, enhanced area of the mass on contrast-enhanced computed tomography corresponded to large vessels filled with blood, and the other area was composed mainly of bundles of smooth muscle, fibrous tissue, and minute blood vessels. 3D-MRA is a reliable method for noninvasive investigation of angioleiomyoma and provides the vascular information required by the surgeon.

Preoperative diagnosis is sometimes difficult. Hwang’s group reported that differential diagnosis of schwannoma is not easy because of the well defined capsules and mixed signal intensities.7 Stoller and associates also reported that hypervascular tumors had quite similar characteristics, even though hemangiopericytomas and some hemangiomas are often characterized by the presence of serpiginous vessels and a slight hyperintensity on T1-weighted images.10 Because the tumors in both of our cases were hypervascular and larger than any previously reported, they had to be distinguished from malignant tumors, for example, from malignant fibrous histiocytoma or hemangiopericytoma. Angioleiomyomas usually grow more slowly than malignant tumors, which we think is one of the points of clinical discrimination.

In conclusion, we present 2 cases of giant angioleiomyoma. To the best of our knowledge, the angioleiomyoma of Case 1 was larger than any previously reported. Case 2 was unique in that the tumor grew to a giant size in the hand of an elderly patient. Thus, angioleiomyoma should be considered in the differential diagnosis of any well circumscribed, hypervascular, subcutaneous soft tissue tumor that elevates the skin above the site of the tumor, even if it is a painless giant soft tissue tumor. 3D-MRA is a useful method for detecting the feeder artery and provides the vascular information required by the surgeon.

References