Ossified Intramuscular Hemangioma

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ABSTRACT

Although hemangioma is a common benign soft-tissue tumor, the formation of intratumoral ossification is rare. Ossified intramuscular hemangioma is an infrequent benign tumor, characterized by a progressively enlarging painless mass in the lower extremity of young female. Herein, we report a case of 27-year-old female suffered from a progressively enlarging painless mass in her left thigh. The mass showed a “Swiss cheese”-like calcification on plain radiographs. Magnetic resonance images revealed multiple cystic spaces with marked contrast enhancement suggesting vascular nature and intratumoral non-enhanced foci of very low signal intensity. The image finding may prompt the diagnosis of ossified intramuscular hemangioma, which was confirmed by the histologic examination. Our report emphasized that the characteristic imaging features of ossified intramuscular hemangioma might be helpful for the correct diagnosis preoperatively.

CASE REPORT

A 27-year-old female presented with a 10-year history of a painless progressively enlarging mass at her left upper thigh. Past medical or surgical history was unremarkable. She also denied any prior trauma. Physical examination was only notable for a palpable well-demarcated mass around 10 × 5 cm in the posteromedial aspect of left thigh. The overlying skin was normal. The mass was bone-hard and rough-surfaced without tenderness or local reddish swelling. There was no lymphadenopathy at the inguinal or popliteal area. Plain radiographs revealed a huge soft tissue mass with coarse trabecular rings and arcs-like ossification, the so-called “Swiss cheese” appearance, at the medial aspect of left thigh (Fig. 1), implying ossified intramuscular hemangioma. No periosteal reaction could be discerned. MR imaging disclosed a large well-circumscribed intramuscular mass involving the medial aspect of thigh muscle and abutting left femur. T1-weighted images showed an isointense mass as compared with normal muscle, surrounded by inhomogeneous hypointense area, and intermingled with spotted foci of very low signal intensity (Fig. 2a). Gadolinium-enhanced T1-weighted fat-suppressed images revealed marked enhancement of the mass, suggesting its...
vascular nature. Nonetheless, the foci of very low signal intensity remained unenhanced, consistent with the ossification (Fig. 2b, 2f). T2-weighted fat-suppressed images and proton density images revealed hyperintensity of the mass with intratumoral lacelike and linear fat signal (Fig. 2c, 2d, 2e). She received partial surgical excision of the mass and recovered uneventfully. Histopathologic sections of the mass discovered thick, mature lamellar bones interposed between numerous dilated vascular channels which were lined by benign endothelial cells, confirming the diagnosis of intramuscular hemangioma consisting of abundant ossification (Fig. 3). The patient had been followed up six years later and the tumor mass still presented without significant enlargement. Besides, progressive atrophy of left thigh muscle was noted.

**DISCUSSION**

Soft-tissue hemangioma is a common benign vascular tumor which is considered to be congenital and usually discernible during childhood or early adulthood [3]. Soft-tissue hemangioma may originate from the skin, subcutaneous tissue, synovium, and muscle [3]. Intramuscular hemangioma, first reported by Liston in 1843 [4], only accounts for 0.8% of all benign vascular tumors and usually locates in the lower extremities [5]. Intramuscular hemangioma is prevalent among people in their thirties with female preponderance [6]. The size of the tumor ranges from less than 4 cm to over 20 cm, but most is less than 9 cm in diameter [6]. Unlike superficial soft tissue hemangioma which classically displays a palpable soft tissue mass with overlying blue discoloration of the skin, intramuscular hemangioma seated in the trunk or extremities usually manifests as a slowly enlarging painless soft tissue mass, only occasionally accompanies with painful symptoms, with no overlying skin discoloration [3].

Calcification within hemangioma is a common radiographic finding, in the form of amorphous, curvilinear calcification, or multiple rounded, variable-sized phleboliths [1]. Nonetheless, ossified intramuscular hemangioma, namely hemangioma with the formation of massive mature bone

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**Figure 1**

1a

1b

**Figure 1.** **a.** Anteroposterior and **b.** lateral views of plain radiograph of the left thigh. A large soft tissue mass with massive irregular, rings-and-arcs ossification, namely “Swiss cheese” appearance (arrowheads) at the medial aspect of left thigh.
in the skeletal muscle of the extremities, is quite rare [1]. Among all types of hemangiomas, ossified intramuscular hemangioma often occurs in the cavernous hemangioma [1, 2, 6, 7].

Radiographically, ossified intramuscular hemangioma displays a soft-tissue mass associated with multiple rings and arcs-like ossification with a coarse trabecular pattern, the so-called “Swiss cheese” appearance as illustrated in our case (Fig. 1). Engelstad et al. first described this characteristic feature in ossified intramuscular hemangioma, which represented the architecture of mature bone interspersed with large cavernous vascular channels [2]. By contrast, phleboliths, which only indicate the calcification in hemangiomas, usually exhibit well-defined round or oval hyperdensity with central lucency. “Swiss cheese” appearance is unique in the ossified intramuscular hemangioma.

Figure 2. MR imaging of the mass in left thigh. a. An axial T1-weighted image (fast spin-echo sequence, TR/TE, 650/12.58 msec.) shows an irregular well-delineated mass of intermediate to high signal intensity (solid arrows) involving the medial aspect of left thigh muscles. There are multiple foci of low signal intensity within the tumor (open arrows), representing intratumoral ossification. b. Gadolinium-enhanced T1-weighted fat-suppressed image reveals heterogeneous enhancement of the tumor (solid arrows). Intratumoral foci of very low signal intensity remain unenhanced, compatible with ossification (open arrows). c. An axial T2-weighted fat-suppressed image (fast spin-echo sequence, TR/TE, 4000/86 msec.) shows hyperintensity of the tumor (solid arrows) with the intratumoral hypointense foci (open arrows). d. An axial proton density image (fast spin-echo sequence, TR/TE, 2000/12.58 msec.) reveals the hyperintensity of the tumor (arrows) with intratumoral linear fat signal (arrowheads). e. A coronal T2-weighted image (fast spin-echo sequence, TR/TE, 300/12 msec.) clearly demonstrates a large intramuscular mass involving the medial aspect of left thigh, associated with spotted hypointense areas (arrowhead). f. A coronal Gadolinium-enhanced T1-weighted fat-suppressed image (fast spin-echo sequence, TR/TE, 366/13 msec.) illustrates the heterogeneous enhancement pattern of the mass. The hypointense area is not enhanced and corresponds to the intratumoral ossification (arrowhead).
and therefore may allow that to be distinguished from other types of calcifications or soft tissue tumors [1]. On sonography, an ossified intramuscular hemangioma appears a complex poorly-demarcated mass with a mixture of hypoechoic and hyperechoic components [8]. Prominent vascular channels manifest as hypoechoic lesions on grayscale imaging and as hypervasculare lesions with abnormal low-resistance arterial signal with forward flow during both systole and diastole on color Doppler imaging, suggestive of low vascular resistance. Intratumoral hypoechoic lesions without hypervasculareity are in agreement with the smooth muscle and fibrous tissue. Moreover, hyperechogenicity in the mass with posterior acoustic shadowing corresponds to multiple irregularly-shaped islands of bone in the dilated vascular channels [8].

Computed tomography (CT) scan of ossified intramuscular hemangioma shows irregular increased density throughout the lesion, compatible with the ossification within the lesion. Decreased density may also be found around the lesion, indicative of the peritumoral fat attenuation. Post-contrast CT may fail to further enhance the lesion [1].

MR imaging of ossified intramuscular hemangioma shows a well-delineated mass of isointensity relative to skeletal muscle on T1-weighted images. The mass will become bright up after contrast enhancement. Serpentine or lace-like strands may be present in the lesion with the intensity approximating to the subcutaneous fat, representing the fibrofatty septa between the vessels [1, 9]. On T2-weighted images, ossified intramuscular hemangioma displays heterogeneous bright signal as compared with subcutaneous fat. The surrounding muscular tissue has inhomogeneous high signal intensity on T1-weighted and T2-weighted images, probably due to fatty degeneration of the muscle. Additionally, within the hemangioma are non-enhanced, spotted areas of very low signal intensity both on T1-weighted and T2-weighted images, consistent with the ossification [1, 9]. In the present case, contrast enhancement of the tumor indicated its vascular nature and the non-enhanced intratumoral lesions of very low signal intensity corresponded to the ossification.

Microscopically, ossified intramuscular hemangioma shows multiple dilated vascular channels lined by benign endothelial cells with interpositions of mature bony trabeculae and fibrous stroma surrounded by skeletal muscle [1, 8]. The vessels may contain thrombosed materials and the surrounding muscles may exhibit extensive fatty degeneration [1, 8]. Radiologic features in our case reflected these histologic findings.

The radiologic differential diagnosis of ossified intramuscular hemangioma includes myositis ossificans, ossifying fibromyxoid tumor of soft parts, fibrodyplasia ossificans, synovial sarcoma, and extraskeletal osteosarcoma [1, 8]. For myositis ossificans, the formation of calcification or ossification depends on the stage of the disease and will be shown in the periphery of the mass eventually, namely “zonal phenomenon”. Metaplastic bone formation in an ossifying fibromyxoid tumor of soft parts is usually faint.
and presented at the periphery of the lesion. A synovial sarcoma differs from ossified intramuscular hemangioma in the lack of peripheral fat component. Fibrodysplasia ossificans is a rare disease and mainly involves the soft tissue in the back, shoulder and upper arms in a diffuse manner, associated with symmetric malformed digits. In extraskeletal osteosarcoma, ossification is predominantly in the central portion of the lesion and solid tumor portion is often in the periphery [1, 8].

Wide excision of the ossified intramuscular hemangioma is usually adequate although malignant transformation rarely occurs. The local recurrence rate, however, could be up to 50% and therefore the role of imaging to determine how well the tumor is defined is of the most importance [8].

In conclusion, ossified intramuscular hemangioma, although rare, should be included in the differential diagnosis in young females with a progressively enlarging mass in the extremity. The “Swiss cheese” appearance on plain radiographs could prompt the diagnosis. Familiarity with the characteristic imaging findings would facilitate the correct diagnosis of ossified intramuscular hemangioma preoperatively.

REFERENCES

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