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ScienceDirect
Computerized Medical Imaging and Graphics 30 (2006) 479–482
Computerized Medical Imaging and Graphics
www.elsevier.com/locate/compmedimag

Case report

MRI of alveolar soft-part sarcoma

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Received 16 November 2005; accepted 19 July 2006

Abstract

Alveolar soft-part sarcoma (ASPS) is rare. We present a case of a 30-year-old woman with a 6-month history of a palpable mass in her left thigh. MRI showed an 8-cm mass and a satellite nodule at the left gluteus muscle. The main tumor exhibited an isointense signal on T1-weighted images and high-signal-intensity areas with low-signal-intensity scanty solid components on T2-weighted images. MR angiography showed dilated and tortuous veins around the tumor. Histologic findings were compatible with ASPS.

Keywords: Alveolar soft-part sarcoma; MR imaging (MRI); Sarcoma; Soft-tissue tumor; Thigh

1. Introduction

Alveolar soft-part sarcoma (ASPS) is rare, accounting for less than 1% of all soft-tissue sarcomas [1]. The term "alveolar soft-part sarcoma" is referred to the large granular cells dispersed in alveolar-like arrangements separated by vascular channels [2,3]. Previous authors have reported that ASPS typically shows high-signal-intensity on both T1-weighted and T2-weighted MRI images, with multiple intra- and extra-tumoral signal voids [1,4,5]. We report MRI and magnetic resonance angiography (MRA) findings of a 30-year-old woman with histologically proven ASPS in her left upper thigh.

2. Case Report

A 30-year-old woman complained of swelling of her left upper thigh for 6 months. Exaggeration of swelling with painful sensation was noted 2 months before this admission. She had no history of trauma or any systemic diseases. Physical examination revealed a tender mass without skin color changes over the left upper thigh. Chest radiographs showed multiple tiny nodules in the right lower lung zone. Chest CT scan revealed multiple nodules of uneven size in both lungs consistent with metastasis. MRI of the brain showed no evidence of metastasis. Whole body bone scan was negative.

MRI of bilateral thighs showed an 8 cm × 5 cm soft-tissue mass involving the left gluteus muscle and one satellite nodule (Fig. 1a). The main tumor exhibited an isointense signal relative to muscle on T1-weighted images. T2-weighted images showed tortuous high-signal-intensity areas and relatively low-signal-intensity scanty solid components within the tumor (Fig. 1b). Obvious enhancement of the tortuous areas within the main tumor was noted on gadolinium-enhanced T1-weighted images (Fig. 1c). The core and the margin of the tumor had some serpentine signal voids on all pulse sequence images. MRA showed dilated and tortuous vessels around the tumor (Fig. 1d).

At surgery, a grayish tumor and satellite nodule at the gluteus medius and between the gluteus medius and maximus muscles of the left thigh was noted. A wide excision of the tumor was performed. Histological examination revealed an alveolar soft-part sarcoma arranged in organoid structures separated by thin-walled, sinusoidal vessels. A prominently pseudoalveolar pattern in the organoid structures was noted. A focal sheet-like distribution was seen. The cut surface was homogeneously brownish with no hemorrhage or necrosis. The sarcoma cells were large, round or polygonal in shape with vesicular nuclei and abundant granularly eosinophilic to vacuolated cytoplasm (Fig. 1e). Mild to moderate nuclear pleomorphism

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doi:10.1016/j.compmedimag.2006.07.005

was noted. Mitotic figures were difficult to see. Dilated veins with tumor invasions were frequently identified in the periphery (Fig. 1f). There were focal cystic changes and hyalinization in the sarcoma. The sarcoma was focally surrounded by skeletal muscle. Characteristic intra-cytoplasmic rhomboid-shaped crystalline materials were noted via periodic acid-Schiff (PAS) and diastase-resistant PAS (DPAS) stains (Fig. 1g). Immunohistochemical study revealed the sarcoma was focally positive for S-100, focally equivocally positive for desmin, and negative for actin, CK, and EMA.

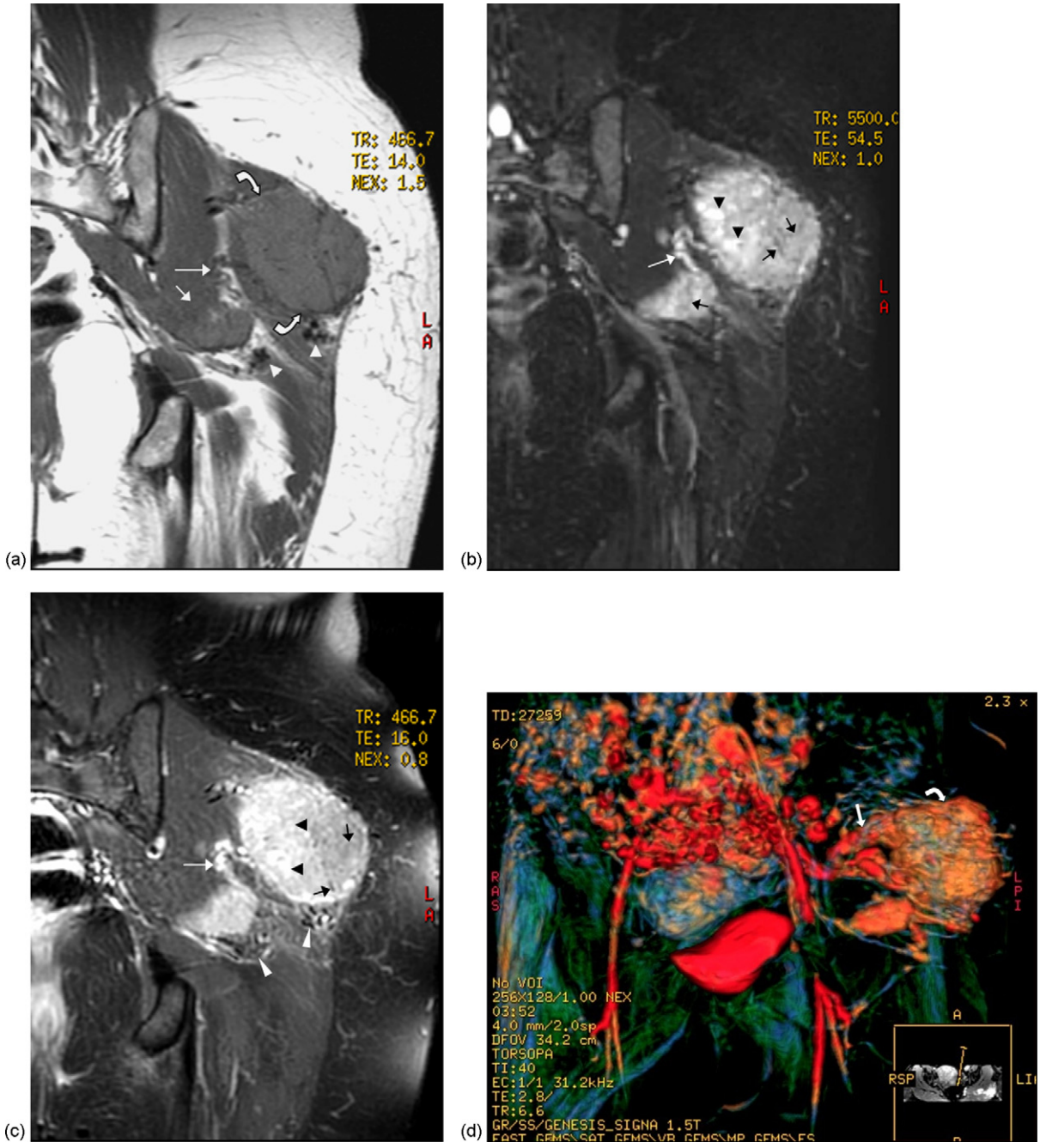

Fig. 1. (a) On T1-weighted MRI images (TR = 466.7 ms, TE = 14 ms) shows an isointense signal tumor (curved arrow) and one satellite nodule (short arrow) in the left upper thigh muscle. Note a tortuous vessel (long arrow) connecting the masses and extra-tumoral signal voids (arrowheads). (b) On T2-weighted MRI images (TR = 5500 ms, TE = 54.5 ms), there are tortuous high-signal-intensity areas (arrowheads) within the relative low-signal-intensity scanty areas of the tumor. Note serpentine signal voids (black arrows) within the tumor and the satellite nodule. The tortuous vessel (white arrow) becomes high-signal-intensity, suggesting slow-flowing blood. (c) Postgadolinium image (TR = 466.7 ms, TE = 16 ms) shows obvious enhancement of the tortuous vessels (black arrowheads) within the tumor and the one (white arrow) connecting the main mass and the nodule, suggesting slow blood flow. There are some areas of intra- (black arrows) and extra-tumoral signal voids (arrowheads), suggesting high blood flow. (d) MRA obtained from SPGR (TR = 6.6 ms, TE = 2.8 ms) dynamic gadolinium-enhanced late phase imaging study shows tortuous, dilated draining veins (arrow) in and around the tumor (curved arrow). (e) Photomicrograph shows large, round or polygonal tumor cells with vesicular nuclei and abundant granularly eosinophilic to vacuolated cytoplasms (H&E, ×200). (f) Photomicrograph shows dilated veins (arrows) with tumor invasion are frequently seen in the periphery (H&E, ×40). (g) Photomicrograph shows intra-cytoplasmic rhomboid-shaped crystalline materials (arrow) (DPAS stain, ×400).

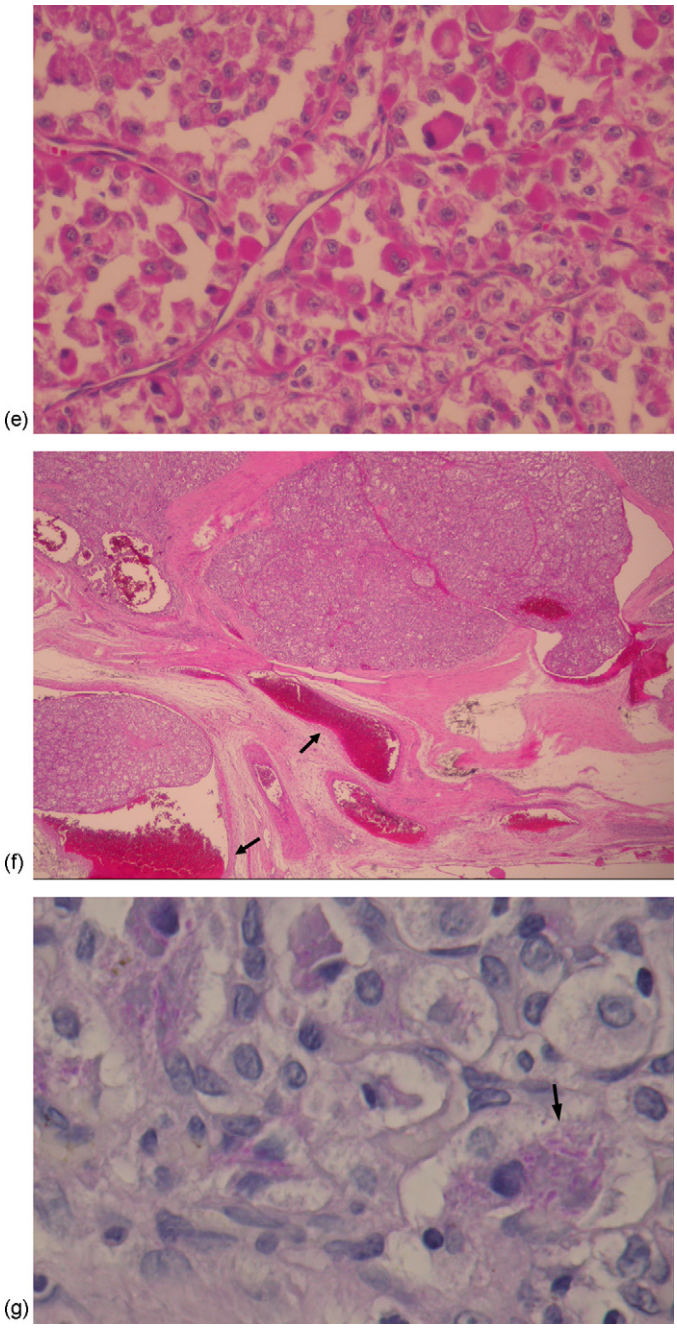

Fig. 1. (*Continued*).

3. Discussion

Alveolar soft-part sarcoma is a rare soft-tissue malignancy [6], which commonly occurs in women and in the first two decades of life [7]. The tumor most commonly involves the lower limbs, followed by the trunk and upper limbs. Although alveolar soft-part sarcoma grows slowly, metastases have been reported in 20–25% of patients at diagnosis [7]. Metastasis most frequently attacks the lungs, followed by bone and brain [6]. However, lymph node metastases are not common.

An ASPS typically is a well-circumscribed mass with a homogeneous appearance on the cut surface. The microscopic picture is uniform and characterized by a pseudoalveolar pattern with nests of tumor cells separated by sinusoidal vascular channels [3,8]. Individual cells have vesicular nucleoli and abundant eosinophilic cytoplasm and show little variation. PAS preparation frequently reveals rod- or sheaf-like crystals in the tumor cells, which is diagnostic for ASPS [3].

ASPS commonly show high-signal-intensity on T1-weighted and T2-weighted images and multiple intra- and extra-tumoral signal voids [1,4,5]. The high-signal-intensity areas of the tumors on T1-weighted sequence can be attributed to slow-flowing blood in or around the tumor [1,4,9]. However, the soft-tissue tumor of our case exhibited an isointense signal on

T1-weighted images. Some tortuous high-signal-intensity areas on T2-weighted images and areas with obvious enhancement on postgadolinium images can be attributed to slow blood flow, as documented by dilated veins within and around the tumor in our histological findings. Multiple serpentine signal voids within the tumor can be attributed to high blood flow, which has been frequently observed in ASPS [4]. MRA showed dilated feeding and draining vessels in and around the tumors. These dilated vessels can easily mimic arteriovenous malformation on MRA and conventional angiography [4,6].

Many other soft-tissue sarcomas may show an isointense signal or slightly increased signal on T1-weighted images [4]. These tumors include clear cell sarcoma, metastatic melanoma, hemangioma, liposarcoma and soft-tissue tumor with hemorrhage [4]. Clear cell sarcoma has melanocytic differentiation on histology, which is related to T1 shortening [4,10]. Multiple signal-void structures are infrequently observed in ASPS, which may be related to fast-flowing blood. Clear cell sarcoma does not have these signal-void structures.

Hemangioma also has slightly high-signal-intensity on T1-weighted images due to fibrofatty components, and has signal voids from fast-flowing blood vessels [11].

Arteriovenous malformations usually show a mixed fast and slow flow pattern relating to a continuous spectrum of vascular malformations [8,11]. On MRI, ASAP has abundant solid tissue components in addition to signal voids, whereas arteriovenous malformation has exclusively vascular components with scanty solid-tissue components in the tumor and frequently has fibrofatty tissue within the mass [4].

In summary, we report a case of ASAP in the left thigh with MRI findings of isointense signal on T1-weighted images and high-signal-intensity on T2-weighted images. MRA showed dilated feeding and draining vessels in and around the tumors. Findings of MRI and MRA correlated well with the histological findings of ASPS.

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