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## Alveolar soft-part sarcoma: a rare soft-tissue malignancy with distinctive clinical and radiological features

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**Abstract** Alveolar soft-part sarcoma (ASPS) is a rare tumour. Certain distinctive clinical and radiological features suggest the correct diagnosis. There is moderate predilection for young women. ASPS almost always arises in skeletal muscle and occurs most frequently in the lower limbs. There is often a long clinical history and a large mass at presentation. Two young females with ASPS presented with very vascular tumours in the thigh, with prominent intra- and extra-tumoural blood vessels. The imaging findings and the existing literature are reviewed.

### Introduction

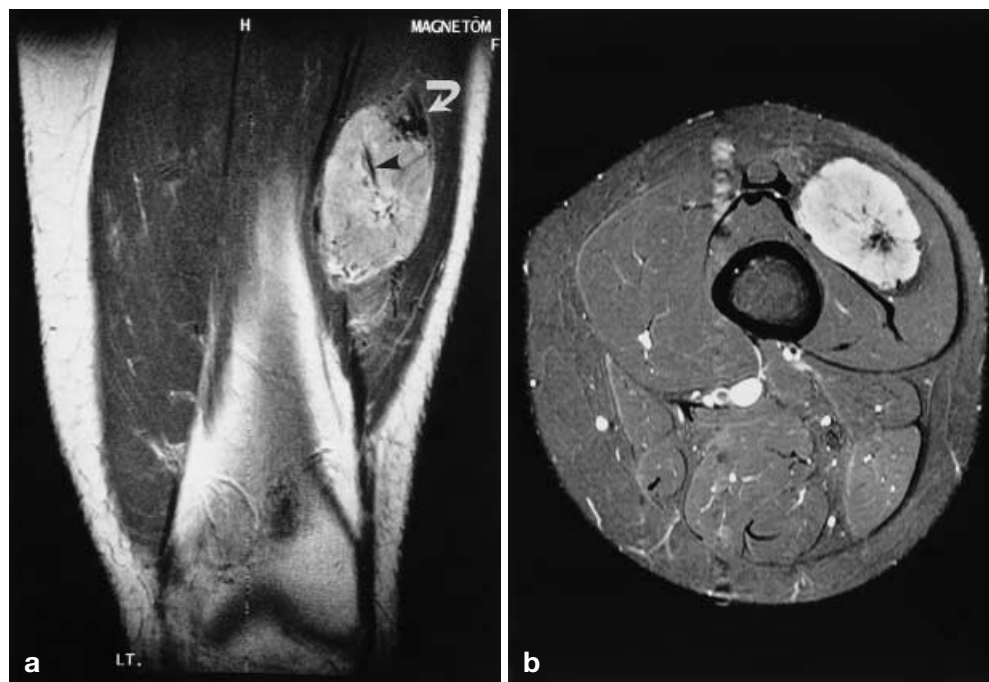
Alveolar soft-part sarcoma (ASPS) is a rare tumour, accounting for 0.5–1% of all soft-tissue sarcomas [1]. It tends to grow slowly and insidiously, often with a long clinical history and a large mass at presentation. The prognosis is poor due to the high incidence of metastatic disease and poor response to chemotherapy [2–4]. ASPS is typically very vascular, a feature reflected on imaging studies, which may show large intra- and extra-tumoural blood vessels. This paper describes the sonographic and MR imaging findings in two young females with ASPS.

### Case reports

#### Patient 1

A 14-year-old female presented with a left thigh mass, which had been present for 1 year. MRI revealed a small mass centred in the vastus lateralis muscle (Fig. 1). It was mildly hyperintense to skeletal muscle on T1-weighted (T1-W) images and showed prominent intratumoural blood vessels and contrast enhancement. There was no evidence of inguinal or pelvic lymphadenopathy. Bone scintigraphy was normal. Thoracic CT revealed pulmonary nodules, consistent with metastatic disease. Tissue obtained by needle biopsy was originally interpreted as being from a granular cell myoblastoma. Excisional biopsy showed histological features compatible with ASPS. The patient is currently being considered for further therapy.

**Fig. 1a, b** Case 1. **a** Coronal contrast-enhanced T1-W MRI shows a tumour in the left vastus lateralis. There is intense contrast enhancement. Several small intratumoural vessels are represented by signal voids (*arrows*). **b** Transverse contrast-enhanced T1-W MRI with fat suppression shows the mass in vastus lateralis. There is uniform enhancement, except for intratumoural vessels of various sizes



#### Patient 2

A 26-year-old female presented with a 1-year history of a right thigh mass. Dilated subcutaneous veins were visible superficial to the mass. An enlarged inguinal lymph node was palpable. Radiographs revealed nonspecific soft-tissue swelling in the left thigh.

Sonography showed a large, heterogeneous vascular tumour with prominent feeding arteries and draining veins (Fig. 2a). The enlarged inguinal lymph node was hypervascular and showed evidence of central necrosis (Fig. 2b).

MRI showed a large tumour in the adductor and hamstring compartments of the thigh (Figs. 2c, d). The tumour was minimally T1-hyperintense and markedly T2-hyperintense to skeletal muscle. The sciatic nerve, profunda femoris and superficial neurovascular bundles were encased. The tumour showed marked contrast enhancement except for a central area of presumed necrosis. MRA showed enlarged tumour vessels (Fig. 2e). Thoracic CT revealed numerous pulmonary nodules up to 30 mm in diameter, consistent with metastases. US-guided 18-gauge core biopsy of the primary tumour and lymph node metastasis showed histological features of ASPS. The patient is currently being treated with neoadjuvant cisplatin and doxorubicin.

#### Discussion

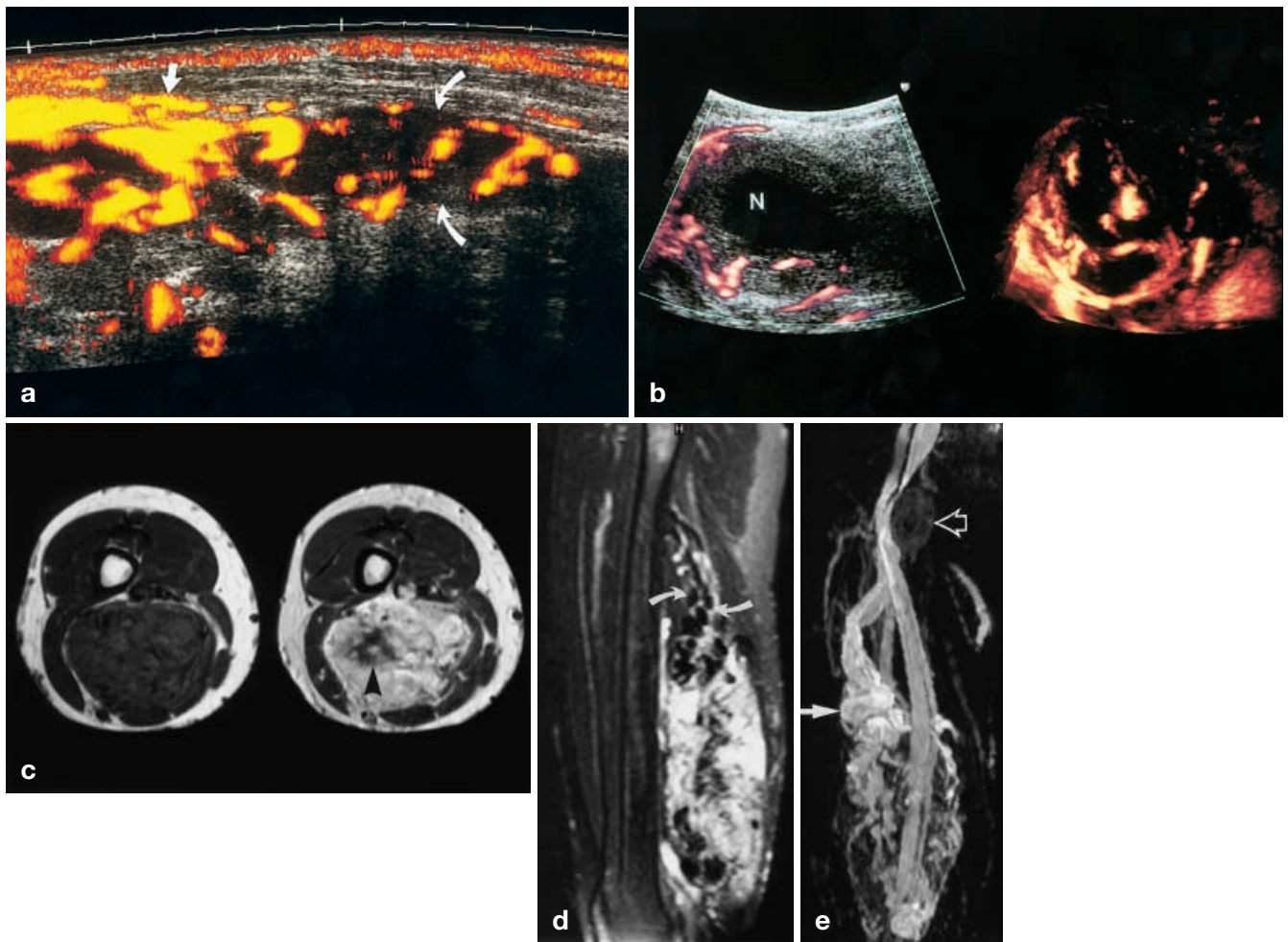
ASPS [5] is a rare soft-tissue malignancy. The mean age at diagnosis is about 22 years in females and 27 years in males [2], but the tumour can occur in children as young as 2 years [2, 3]. ASPS is more common in females, especially in the first two decades of life [2]. The most common site of origin is the lower limb, followed by the trunk and upper limb [2]. Almost all cases appear to arise in skeletal muscle [5]. Despite the slow growth

rate of the primary tumour, metastases are common, being detected in about 20–25% of patients at diagnosis [2]. Metastasis is most frequently to the lungs [4, 5], followed by bone [4, 6] and brain [4, 7, 8]. Lymph node metastases are uncommon [4]. In one series [2], 38% of metastases were detected more than 10 years after the diagnosis of the primary tumour. Response to systemic chemotherapy is usually poor [2, 3, 9], but the prognosis may be better in children than in adults [3].

The two cases illustrated show the characteristic features of ASPS. US typically shows a heterogeneously hypoechoic mass [6, 8], with large tumour vessels (Fig. 2a) [8] and poorly defined margins. A large, markedly hypervascular lymph node metastasis was present in one patient (Fig. 2b). This finding does not appear to have been previously reported. Metastases in the brain [7] and lung [10] have also been noted to be extremely vascular.

ASPS tends to be of equal or slightly higher signal intensity than skeletal muscle on T1-W images (Fig. 2c) [11, 12] and to show high and heterogeneous signal intensity on T2-W images (Fig. 2d) [11–13]. The presence of a large soft-tissue mass associated with large peritumoural vessels (Figs. 1a, 2d) is strongly suggestive of ASPS [13]. There is strong and almost uniform enhancement with IV contrast medium (Figs. 1, 2c) [13]. Small areas that fail to enhance (Fig. 2c) may represent tumour necrosis.

Conventional angiography typically shows enlarged feeding arteries and early filling of large draining veins. There is often delayed washout of contrast medium



**Fig. 2a-e** Case 2. **a** Extended field-of-view sagittal power Doppler image shows the large, heterogeneously hypoechoic mass (*curved arrows*) with enlarged blood vessels, especially proximally (*straight arrow*). **b** Biopsy-proven right inguinal lymph node metastasis. Power Doppler US (*left*) shows an enlarged vascular lymph node with probable central necrosis (*N*). Three-dimensional reconstruction (*right*) shows the vascular nature of the node better. **c** Transverse T1-W MRI before (*left*) and after contrast enhancement shows a poorly defined mildly T1 hyperintense tumour in the hamstring and adductor compartments. The tumour shows uniform enhancement except for a small central area (*arrowhead*), which may reflect necrosis. **d** Sagittal T2-W fast-spin-echo image shows a large hyperintense tumour in the hamstring compartment of the thigh. There are large intra- and extra-tumoural flow voids, especially proximally (*arrows*). **e** Oblique coronal first-pass MRA shows large feeding and draining vessels within and near the tumour (*solid arrow*). In isolation, this appearance simulates an arteriovenous malformation. The femoral vessels are displaced by a large inguinal lymph node metastasis (*open arrow*)

from the tumour [6, 10, 11, 13]. The angiographic findings have led to the misdiagnosis of ASPS as an arteriovenous malformation (AVM) [14]. It is important to be aware that MRA may show a similarly misleading appearance (Fig. 2e). However, the presence of an accompanying large soft-tissue mass should allow the diagnosis of soft-tissue neoplasm to be made. The combination of a mass and extensive flow voids is not consistent with either an AVM ('high-flow' lesion) or a capillary or capillary-venous malformation ('low-flow' lesion) [15].

In conclusion, ASPS occurs most frequently in females in the second and third decades and has distinctive and potentially misleading clinical and radiological features. It presents insidiously, but has a poor prognosis. Imaging studies that show a large intramuscular mass with very prominent vascularity, or the presence of highly vascular metastases, should suggest the correct diagnosis. Appearances at angiography or MRA may be misleading.

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