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

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. 1 a, 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. 2c). 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 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 hypoechogenic 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.