

Primary Alveolar Soft Part Sarcoma of the Scapula

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Key Words

Alveolar soft part sarcoma · Scapula · Lung metastasis

Abstract

Alveolar soft part sarcoma (ASPS) is an unusual soft tissue malignancy, occurring in less than 1% of sarcomas and typically found in the head and neck tissues in children or, in adults, in the deep soft tissues of the lower extremities. In this report, we present a 33-year-old male with primary ASPS in the right scapular bone and discuss the radiologic features of this tumor in the context of the current literature.

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Introduction

Alveolar soft part sarcoma (ASPS) is a rare soft tissue neoplasm of unknown cellular origin that occurs primarily among adolescents and young children. Pediatric ASPS occurs most commonly in the head and neck tissues; however, lower extremity involvement may occur in adults. The initial manifestation is commonly lung or brain metastasis, which is followed by an investigation locating the primary site of the tumor. Only 2 cases with primary involvement of the scapular bone have been reported previously in the literature.

Case Report

A 33-year-old male was referred to our institute complaining of right shoulder pain for the previous year due to a progressive tumor in the right scapular region. A swelling of the right shoulder was apparent during the physical examination. A lytic expansile scapular bone lesion with poorly distinguished margins and moth-eaten pattern was seen using standard X-ray imaging ([fig. 1a](#)). Multiple metastatic pulmonary nodular opacities were

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apparent on the chest radiogram (fig. 1b). The presence of a heterogeneous soft tissue mass within the right scapula was demonstrated by contrast-enhanced computed tomography (CT), resulting in expansion and destruction of the bone cortex (fig. 1c). An expanded and disrupted bone cortex was revealed by bone window level axial CT imaging (fig. 1d). An osseous soft tissue mass originating in the right scapular region and involving the glenohumeral joint and supraspinatus muscle was revealed by magnetic resonance imaging (MRI) with a tubular signal void of both the T1-weighted coronal (fig. 2a) and T2-weighted fat-suppressed axial (fig. 2b) images due to characteristic enlarged blood vessels; T1-weighted fat-suppressed coronal images were modified by heterogeneous contrast enhancement (fig. 2c). Ultrasound-guided Tru-Cut biopsy of the scapular lesion was performed, and the histopathology was consistent with ASPS and included positive periodic acid-Schiff (PAS) reaction of the cytoplasm in uniform cells with minimal pleomorphic nuclei (fig. 3). Wide resection of the tumor (subtotal scapulectomy) was performed and the patient's shoulder pain resolved completely after surgery.

Discussion

ASPS is an unusual soft tissue malignancy occurring in less than 1% of sarcomas and typically found in the head and neck tissues in children or, in adults, in the deep soft tissues of the lower extremities. Only a few cases of ASPS have been reported in the literature since the cancer was first described in 1951 by Smetana and Scott [1] as a malignant tumor of nonchromaffin paraganglia. Christopherson et al. [2] gave it the descriptive name 'alveolar soft part sarcoma' in 1952. The soft tissue of the extremities is the primary site of involvement, frequently the thigh or buttock, with a few cases involving other soft tissue locations including the arm, chest or retroperitoneum [3, 4]. ASPS occurring in the cervix, uterus, orbit, pulmonary vein, stomach and bone with absent concomitant skeletal muscle involvement has been reported. Metastases commonly occur in the brain regardless of the primary site [5]. Characteristic histopathology includes marked vascular invasion by polygonal cells with granular cytoplasm and prominent nucleoli and alveolar structure. The diagnosis is often established definitely by histochemical staining. Intracellular glycogen varies greatly according to PAS preparation, and diastase-resistant rhomboid or rod-shaped crystals that are characteristically PAS positive may occur. The standard treatment for both primary tumors and metastatic tumors in the brain and lung is surgical excision aiming at obtaining tumor-free margins. The 5-year survival rate of ASPS was 56% in a Japanese cohort reported by Ogose et al. [6]: in that study group, the tumor size, the presence of bone involvement, and the presence of metastasis were the primary factors influencing prognosis.

Radiological features of primary bone manifestations of ASPS have been described by Park et al. [7]: a common feature of all cases was bone destruction with poorly defined tumor margins. In many cases, ASPS is distinguishable at equal or slightly increased signal intensity relative to skeletal muscle on T1-weighted MRI with high and heterogeneous signal intensity apparent by T2-weighted MRI. Intravenous contrast medium results in strong, uniform enhancement of tumor imaging [8]. Serpentine flow voids due to peripheral feeding vessels and intratumoral vessels are characteristic findings to manifest tumor hypervascularity. In the case presented, intensive bone destruction activity and heterogeneous contrast enhancement of the right scapular soft tissue mass was consistent with the radiologic characteristics reported in the literature.

Only 2 previous reports of ASPS in the scapular region have been previously reported in the medical literature [9, 10]. Radiologic findings reported in these cases were similar to

ours: ill-defined osteolytic lesions were observed on plain radiographs, and an expansile bone tumor with large intra- and extraosseous soft tissue masses was observed on CT as well as moderately high signal intensity with foci of signal void corresponding to dilated blood vessels on T1- and T2-weighted MR images. The case presented is the third such case and includes extensive radiologic and scintigraphic findings. In conclusion, ASPS should be taken into consideration in the differential diagnosis of bone-originating highly vascularized soft tissue masses; characteristic features of the lesion (high-flow vascular structures and aggressive solid component) contribute to differentiating this entity from other hypervascular bone tumors such as rhabdomyosarcoma, hemangioendothelioma and hemangiopericytoma. The primary therapeutic approach consists of surgical exploration in combination with radiotherapy treatment.

Disclosure Statement

All authors declare that they have no conflicts of interest.

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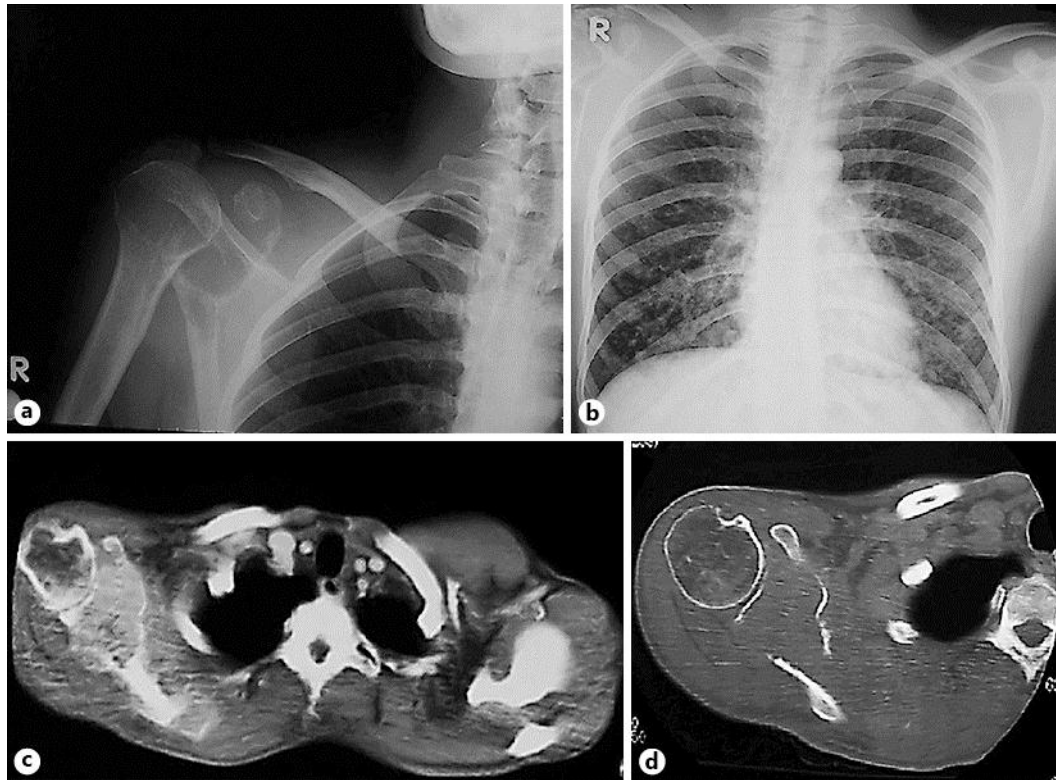


Fig. 1. Plain X-ray and CT findings. **a** X-ray examination because of right shoulder pain revealed a lytic scapular bone lesion with moth-eaten pattern. **b** Chest radiograph showed multiple nodular opacities due to bilateral lung metastasis. **c** Contrast-enhanced CT revealed a heterogeneous mass with intensive invasion to the scapula. **d** Bone window level CT revealed massive cortical destruction of the scapular bone.

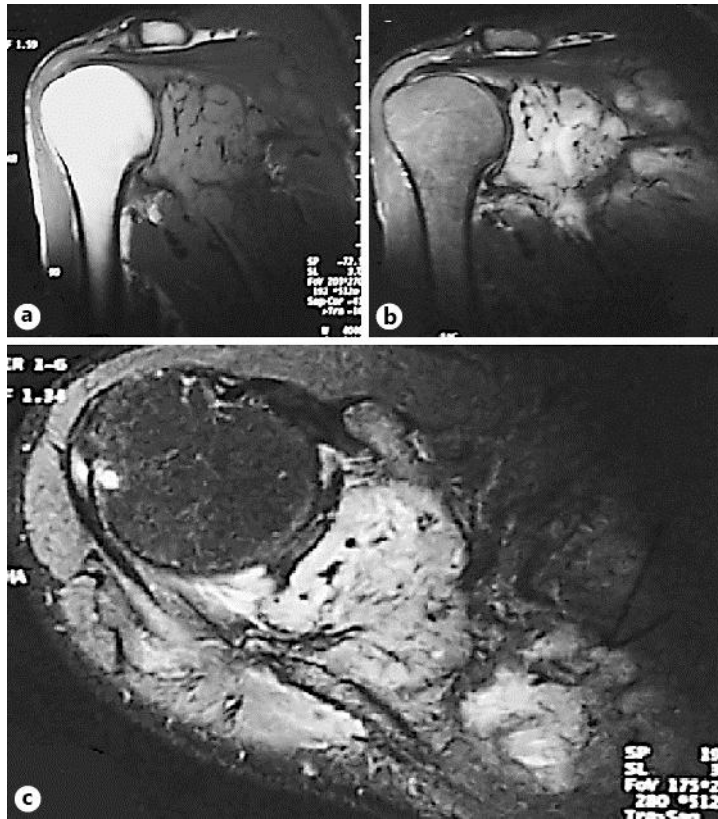


Fig. 2. MRI of the right shoulder. **a** T1-weighted coronal image showed a large mass of moderately high signal with signal-void areas in the central portion. **b** Contrast-enhanced T1-weighted fat-suppressed coronal image revealed heterogeneous contrast enhancement of the lesion. **c** T2-weighted fat-suppressed axial image showed a hyperintense soft tissue mass with lobulated contour.

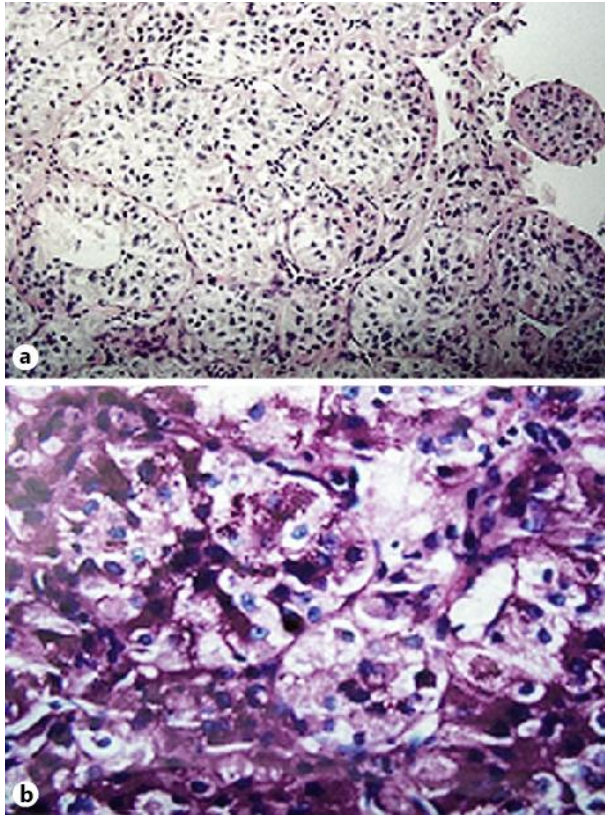


Fig. 3. Histological observation. **a** Large uniform tumor cells proliferated in an alveolar pattern (HE; 50×). **b** Cells with positive PAS reaction of the cytoplasm (PAS stain; 200×).