

Case Report

Alveolar soft part sarcoma of extremity

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ABSTRACT

Alveolar soft-part sarcoma (ASPS) is an extremely rare connective tissue tumor, predominantly seen in adolescents and young adults, with a female preponderance. Alveolar soft-part sarcoma (ASPS) is a slow growing tumor, but with high likelihood of metastasis, leading to high mortality. A classical histopathological feature of an alveolar pattern from the biopsy of the lesion favors the diagnosis. We report a case of 14 years old male patient who presented with a history of single painless swelling over thigh for which surgical excision was done. Histopathology was suggestive of Alveolar soft-part sarcoma (ASPS). There was no evidence of distant metastases. He was treated with external beam radiotherapy in view of vascular invasion.

Keywords: Alveolar soft-part sarcoma, Pathology, Surgery, Radiotherapy

INTRODUCTION

Alveolar soft-part sarcoma (ASPS) is an extremely rare connective tissue tumour accounting for <1% of soft tissue sarcomas.¹ Alveolar soft-part sarcoma (ASPS) most commonly occurs in adolescents and young adults between 15-35 years of age with increased predilection for females.² Most common site of presentation is the lower extremities mainly involving the thigh.¹ ASPS often presents as a slow-growing painless mass, even with metastatic disease.¹ Complete surgical excision of the primary tumour in its early stages is the mainstay of treatment.³ It is resistant to standard chemotherapy agents and has a high likelihood to metastasize early, resulting in poor prognosis.¹

CASE REPORT

A 14 years old male presented with history of single painless swelling over right thigh for duration of 2 months. On examination, a firm swelling of size 4×3 cm was present over the medial aspect of right distal thigh, fixed to the underlying tissue, but free from the overlying skin. There were no neurovascular deficits. No regional

lymphadenopathy was noted. Rest of systemic examination was normal.

Routine investigations were normal including complete haemogram, liver and renal functions, chest X-Ray, and ultrasound of abdomen. Magnetic Resonance Imaging (MRI) revealed well defined slightly lobulated lesion of size 22×21×36 mm in medial aspect of right lower thigh in intramuscular plane in vastus medialis muscle. It was hyper intense on T1- weighted and T2- weighted images with evidence of multiple tortuous flow voids with post contrast enhancement. It had maintained fat planes with subcutaneous soft tissue. Possibility of highly vascular soft tissue tumour, most likely ASPS was kept.

Fine needle aspiration cytology suggested malignant round cell tumour. Complete surgical excision of the swelling was done. The resected specimen revealed a well circumscribed grey-white tumour of size 3×2×1.6 cm, with no areas of haemorrhage and necrosis.

Histopathology showed large, round to polygonal cells arranged in an organoid pattern, with abundant eosinophilic to vacuolated cytoplasm and minimal

nuclear pleomorphism, with prominent nucleoli (Figure1).

The cellular aggregates were separated by thin walled sinusoidal vascular channels. The cells showed loss of cohesion. Occasional mitotic figures were seen. Margin of the tumour showed vascular invasion (Figure 2).

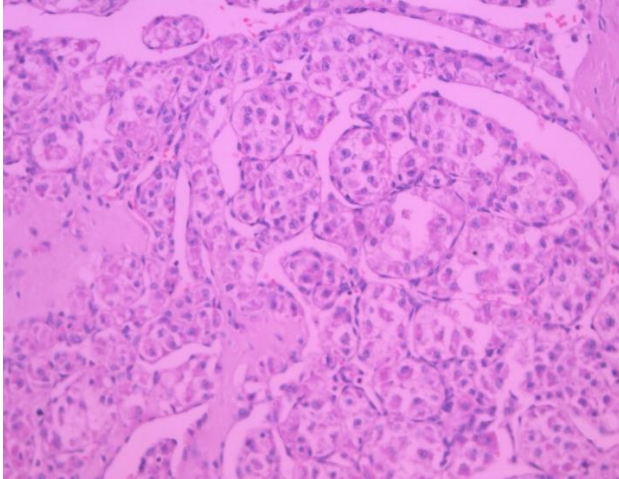


Figure 1: ASPS of thigh showing pseudoalveolar pattern.

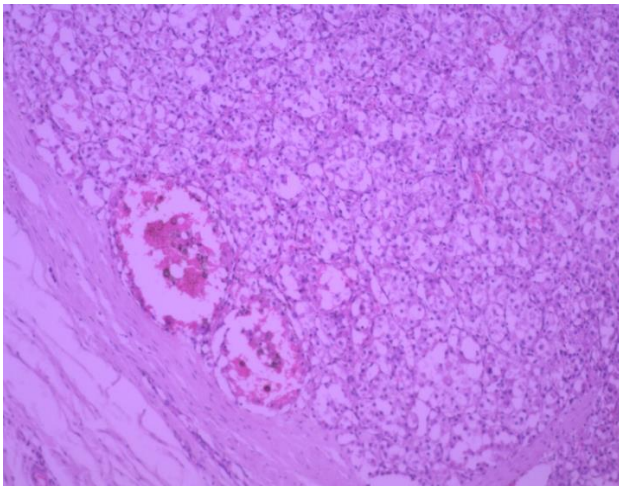


Figure 2: Vascular invasion of the tumor margin seen in ASPS.

Resection margins were free of tumour. Periodic acid-Schiff (PAS) stain revealed focal intracellular cry-stalline material. On immune histochemistry, the tumour cells expressed strong and diffuse nuclear positivity for TFE3 and were negative for Desmin. Thus, a diagnosis of ASPS was made.

In view of vascular invasion, post-operative external beam radiotherapy was given. There was no evidence of local recurrence or distant spread at 10 months of follow-up.

DISCUSSION

ASPS comprise <1% of all soft tissue sarcomas and commonly affects adolescents and young adults between 15- 35 years of age. It is rare before 5 and after 50 years of age. Most common site of presentation is thigh or buttock in adults. In children, it usually occurs in head and neck region, often in the orbit and tongue.² Our patient was 14- years old with history of swelling over thigh for 2 months. The patient often presents with slow growing painless mass. The patients may remain asymptomatic over years, even with metastatic disease.¹ Most common site of metastases are lungs, followed in frequency by bone and brain.^{4,5}

ASPS characteristically show the presence of a hyper intense lesion with central necrosis and flow voids within the lesion on T1-weighted and T2-weighted MRI.⁶

It is characteristically composed of large, uniform, epithelioid cells having abundant eosinophilic, granular cytoplasm arranged in solid nests and / or alveolar structures, separated by thin, sinusoidal vessels. In addition, tumour cells exhibit characteristic PAS-positive, diastase-resistant, intracytoplasmic rhomboid crystals. The distinguishing phenotype of ASPS is its strong nuclear staining with an antibody to the carboxy terminal portion of TFE3 retained in the fusion protein resulting from the ASPSCR1-TFE3 fusion gene.²

Complete surgical excision of the primary tumour in its early stages is the mainstay of treatment.³ Increased risk of metastases is seen with increasing age, large tumor size and initial presentation (localized versus metastatic disease).^{4, 7, 8} The prognosis is poor because ASPS has a high propensity to metastasize early and is resistant to standard chemotherapy agents.¹ Role of radiotherapy for treatment of ASPS is unclear.⁹ Benefit from immunotherapeutic agents is under evaluation.¹

CONCLUSION

ASPS is a rare tumor which runs an indolent course with high propensity for metastasis. Early diagnosis and complete excision of the localized disease are essential in the treatment of ASPS.

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Ethical approval: Not required

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