



Alveolar Soft Part Sarcoma, Lumbar Region - A Case Report

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Abstract

We present a case of Alveolar Soft Part Sarcoma in left lumbar region in a 14-year-old girl. Following its excision, histopathological examination was performed to arrive at the diagnosis.

Introduction

Alveolar Soft Part Sarcoma is a soft tissue tumour that typically affects adolescents, with a female preponderance¹. Although its histogenesis is still unknown, its genetic basis has been delineated.² Despite its slow growth, this tumour can be fatal, owing to its propensity to recur in distant sites and limited response to chemotherapy and radiotherapy³. The diagnosis can usually be made by histopathological examination.

Case Report

A 14-year-old female presented with a small swelling on her left loin. It had been present for 3 years. She had intermittent pain for 2 years. The swelling was first noticed as a strawberry-sized

lump, from which it had increased gradually to attain a size of 6x4x3 cm. She had no history of radiation exposure. Her menstrual cycles were regular. On examination, the swelling was firm in consistency with restricted vertical mobility. The overlying skin was pinchable.

Her routine laboratory investigations were normal. Her blood group was O negative.

Magnetic Resonance Imaging (Fig. 1) revealed a well-defined T1/T2 heterogeneous lesion with multiple flow voids in the left lumbar region, posteriorly in the deep subcutaneous plane, causing mass effect on the muscles with minimal inflammatory effect. The possibility of a low-flow vascular malformation or vascular soft tissue tumour was considered.

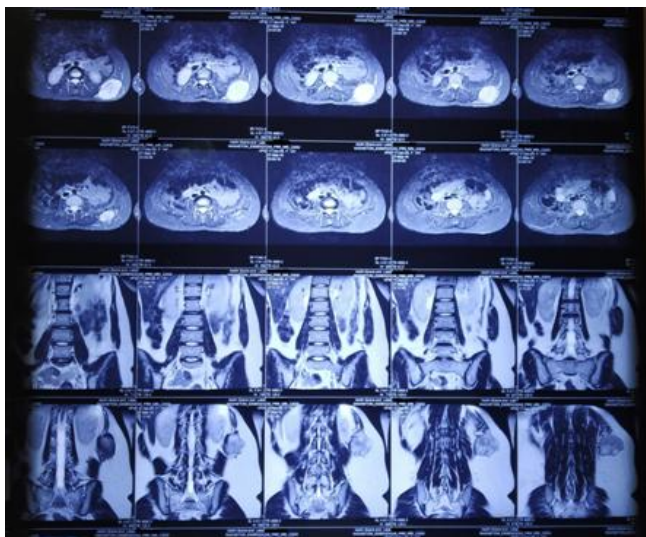


Fig. 1 Magnetic Resonance Imaging shows a mass in left lumbar region.

Fine Needle Aspiration yielded mainly blood and on cytology, only a few capillary fragments were visible in a background of hemorrhage.

Subsequently, an excision of the swelling was performed. During surgery, heavy bleeding was encountered. The excised specimen was sent for histopathological examination.



Fig. 2 Excision of mass from left lumbar region.

On gross examination (Fig. 3), the excised mass had a yellowish-white firm cut surface, extending up to the resected margins. Microscopic examination (Fig. 4-6) showed a tumour composed of polygonal cells arranged in nests separated by thin fibrovascular septa. The cells showed dyscohesion in the centres of the nests, thus imparting an 'alveolar' appearance. The pale granular cytoplasm of the cells contained PAS-positive, diastase-resistant crystalline rods. Hence

a diagnosis of Alveolar Soft Part Sarcoma was made



Fig. 3 Cut surface of the mass was yellowish-white and extending up to the resected margins.

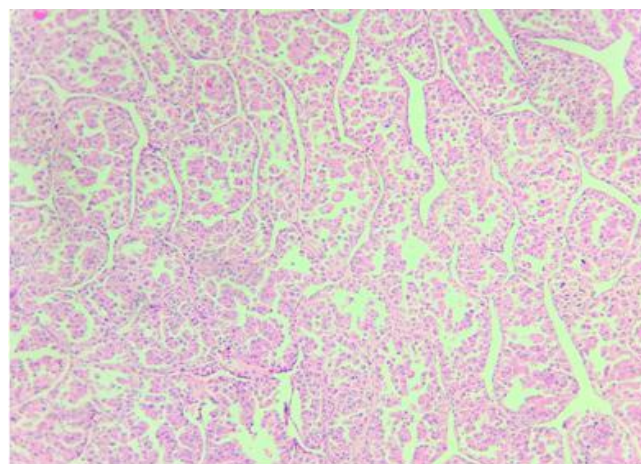


Fig. 4 Low-power microscopy shows nests of cells with central dyscohesion, giving a so-called 'Alveolar' appearance

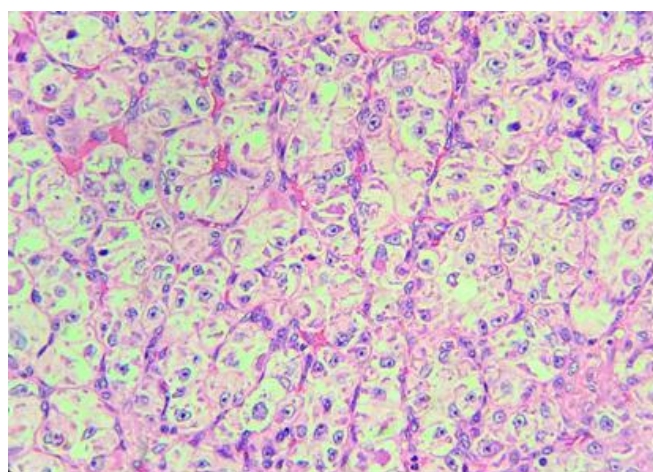


Fig. 5 High-power view shows nests of cells with pale cytoplasm and large nuclei with prominent nucleoli.

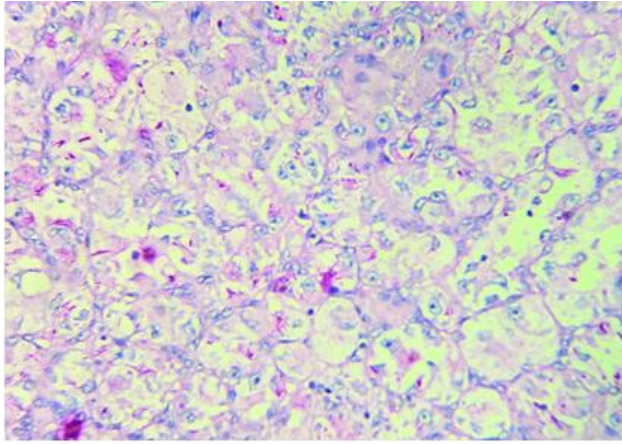


Fig. 6 PAS stain. Bright pink diastase-resistant crystalline rods in the cytoplasm of the cells

Discussion

Owing to recurrence at unusual sites and its fatality, this case mandates histopathological examination, at the earliest stage. Ancillary investigations may also be useful to confirm the diagnosis. TFE3 protein, is a reliable sensitive and specific marker for Alveolar Soft Part Sarcoma. Skeletal muscle origin nuclear markers Myogenin and MyoD1 are also known to be positive. Cytogenetic Analysis can identify the unbalanced translocation $t(x:17)(p11;q25)$, which causes the ASPL:TFE3 fusion gene.³

Acknowledgements

We are grateful to the Dean, Faculty and Staff of Sri Muthukumaran Medical College for enabling us to present this case.

References

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