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Case Report

Alveolar soft part sarcoma masquerading as lipoma: a case report

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ABSTRACT

We report herein, a rare case of a 14-year-old child with alveolar soft part sarcoma. The patient initially noticed a progressively growing mass over medial side of right distal thigh. Although clinically the swelling appeared to be lipoma, the needle biopsy confirmed it to be alveolar soft part sarcoma. Patient was managed by wide excision, followed by adjuvant chemotherapy.

Keywords: Lipoma, Alveolar, Sarcoma

INTRODUCTION

Alveolar soft part sarcoma (ASPS) is a very rare sarcoma accounting for approximately 0.5-1% of all soft tissue sarcomas.[1] It is a slow-growing but nevertheless malignant soft tissue tumor arising in muscles, commonly seen in the age group between 15 and 35 years of age with slight increased predilection in young women by a ratio of 3:2.[2] Clinically it presents as a painless, slow, indolent soft growing lesion which rarely causes functional impairment. Primary tumors are often large and highly vascular, and often present as pulsatile mass. Head and neck, especially the tongue and orbit are common sites of involvement in children, whereas it occurs in muscles of upper and lower extremities in older adults.

Lesion most commonly metastasises to lungs followed by central nervous system. Taking into consideration the rarity of the disease, thorough knowledge of its clinical behaviour, pathology and optimal treatment is still obscure. According to previous literature data, the 5-year survival rate is reported to be 45–88%, 38% at 10 years and 15% at 20 years.[3] We hereby present a case of soft tissue swelling medial side of right thigh, which clinically appeared to be lipoma but on further work up came out as alveolar soft part sarcoma. The case highlights the importance of oncological workup even in cases that appear seemingly benign at presentation.

CASE REPORT

A 14-year-old male child presented to us with chief complaint of swelling over right thigh medially. The patient was apparently well 2 month ago when he noticed a swelling over medial side of distal thigh which was insidious in onset, and progressively increasing in size. There was no pain or any difficulty in movement. There was no history suggestive of antecedent trauma, fever, night sweats, weight loss, and decrease in appetite. On examination swelling 4×3 cm was present over anteromedial aspect of lower thigh. Swelling was nonadherent to surrounding structures, firm to hard in consistency and mobile. Swelling apparently looked benign with firm consistency; hence, provisional diagnosis of benign growth (Lipoma) was kept. Lab investigations were normal. Magnetic resonance imaging (MRI) reported well defined, slightly lobulated lesion of size 22×21×36 mm, observed on medial aspect of lower thigh in intramuscular plane in vastus medialis muscle. Possibility of highly vascular soft tissue tumor or sarcomatous mass was suggested (Figure 1).

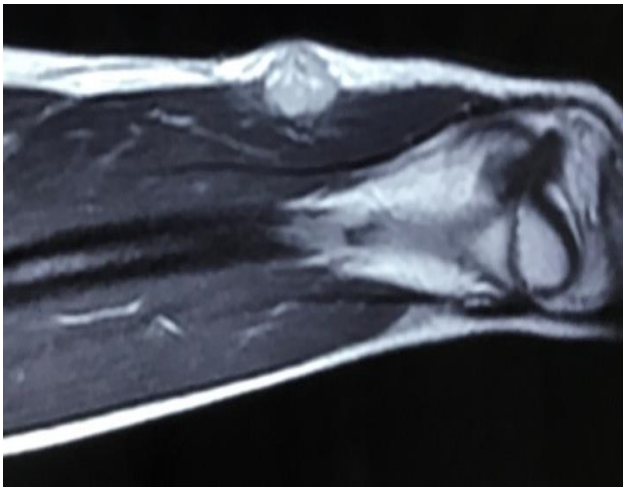

Figure 1: MRI scan suggestive of highly vascular soft tissue tumor or sarcomatous mass.

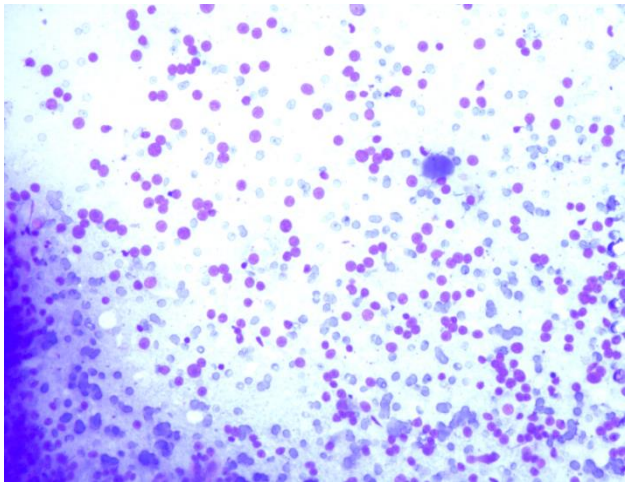

Figure 2: Needle biopsy showed clusters and scattered population of round to oval cells which shows minimal pleomorphism and prominent nucleoli suggestive of malignant round cell tumor.

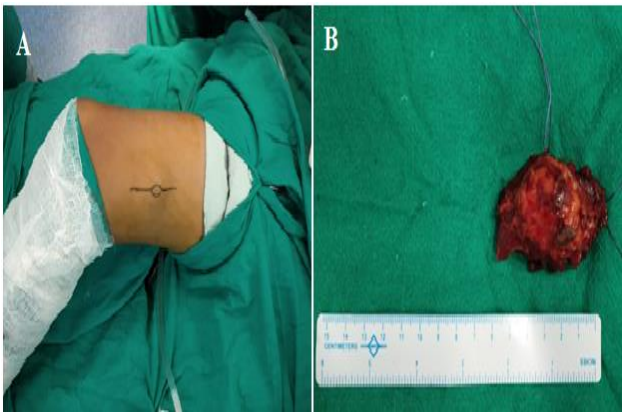

Figure 3: (A) Wide excision of soft tissue mass performed through a medial longitudinal incision centralized over the mass (B) excised mass.

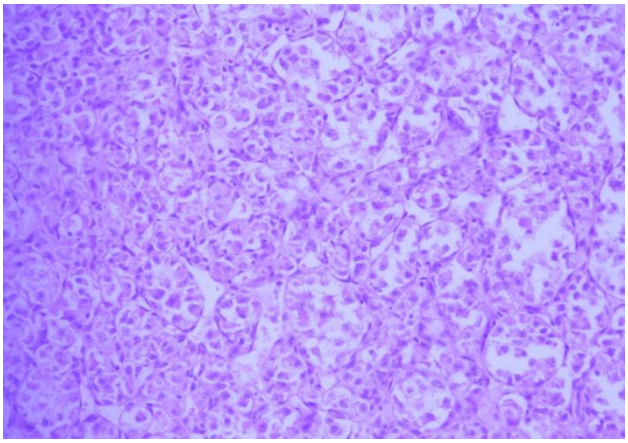

Figure 4: Postoperative histopathological analysis showing loss of cohesion among cells resulting in pseudo alveolar pattern. Individual cells are large rounded to polygonal with minimal nuclear pleomorphism with prominent nucleoli and abundant eosinophilic to vacuolated cytoplasm.

Needle biopsy was done to confirm the diagnosis. Clusters and scattered population of round to oval cells with minimal pleomorphism and prominent nucleoli were noted. Features were suggestive of malignant round cell tumor (Figure 2). Patient was operated by wide excision of the mass followed by skin closure. (Figure 3A, B). Post-surgery histopathology of specimen revealed loss of cohesion resulting in pseudo alveolar pattern. Individual cells were large rounded to polygonal with minimal nuclear pleomorphism with prominent nucleoli and abundant eosinophilic to vacuolated cytoplasm. Periodic acid–schiff (PAS) stain showed intracellular crystalline material. Margins were free from tumor. Postoperative histopathological findings confirmed it to be alveolar soft part sarcoma (Figure 4).

DISCUSSION

Alveolar soft part sarcoma (ASPS) is a rare tumor and accounts for 0.5–0.9% of all soft tissue sarcomas.[4] Christopherson first described its unique histological and cytological features in 1952.[5] ASPS comprises only 1% of all soft tissue sarcomas.[6] ASPS is seen mostly in children and adolescents. However, its usual age range of presentation is from 15 to 35 years, our patient falling into the same group. Extremity involvement, particularly lower limb, is commonly described in adults.[7] head and neck area; particularly the tongue and orbit are the favored sites in children. Most studies have found a female preponderance in adult patients although no such predilection has been noted in children.

ASPS arise in association with skeletal muscles or musculofascial planes, a fact that explains the strong predilection of this tumor for the thighs, buttocks and abdominal or chest walls. It tends to grow slowly and insidiously, often with a long clinical history and a large mass at presentation, similar to our patient, who had a

large swelling in the thigh. Despite the slow growth rate of the primary tumors, metastases are common. Metastases are found most often in the lungs, followed in frequency by bone and brain.[8] The prognosis for children with ASPS may be considerably better; lingual and orbital tumors also have very high survival rates, possibly reflecting a combination of small size at the time of diagnosis and younger patient age.[9]

ASPS tumor cells exhibit characteristic PAS-positive, diastase-resistant, intracytoplasmic rhomboid crystals and act as a diagnostic marker for ASPS. In addition, tumour cells exhibit characteristic round, regular, eccentrically placed nuclei with vesicular chromatin and prominent nucleolus.[10] These histopathological findings were present in our patient, confirming the diagnosis of ASPS.

CONCLUSION

The present study highlights the role of meticulous diagnostic workup in management of seemingly benign soft tissue lesions and emphasizes role of MRI and needle biopsy before embarking on final treatment plan in soft tissue lesions.

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REFERENCES

1. Portera Jr CA, Ho V, Patel SR, Hunt KK, Feig BW, Respondek PM, et al. Alveolar soft part sarcoma: clinical course and patterns of metastasis in 70 patients treated at a single institution. Cance. 2001;91(3):585-91.
2. Mitton B, Federman N. Alveolar soft part sarcomas: molecular pathogenesis and implications for novel targeted therapies. Sarco. 2012;2012:428789.
3. Sidi V, Fragandrea I, Hatzipantelis E, Kyriako poulos C, Papanikolaou A, Bandouraki M, et al. Alveolar soft–part sarcoma of the extremity: a case report. Hippokra. 2008;12(4):251.
4. Enzinger FM, Weis SW. Eds Malignant tumors of uncertain type. In: Soft tissue tumors. 4th edn St Louis Lon. 2001;1509-21.
5. Christopherson WM, Foote FW, Jr, Stewart FW. Alveolar soft part sarcoma: structurally chara-cteristic tumors of uncertain histogenesis. Canc. 1952;5:100-11.
6. Sarkar P, Mukherjee S, Saha ML, et al. Alveolar soft part sarcoma: a rare diagnosis. Ind. J Derm. 2013;58:244.
7. Fassenbender HG. Alveolar myoblastic sarcoma of the skeletal musculature. Onco. 1960;3:184-91.
8. Arqyris PP, Reed RC, Manivel JC, Lopez-Terrada D, Jakacky J, Cayci Z. Oral alveolar soft part sarcoma in childhood and adolescence: report of two cases and review of literature [J]. Hea Nec Pathol. 2013;7(1):40-9.
9. Folpe AL, Deyrup AT. Alveolar soft-part sarcoma: a review and update. J Clin Pathol 2006;59:1127-32.
10. Anbarasi K, Sathasivasubramanian S, Kuruvilla S. Alveolar soft-part sarcoma of tongue. Ind J Pathol Microbiol. 2011;54(3):581.

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