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Case Report

PROXIMAL EPITHELIOID SARCOMA IN A 4 YEAR OLD CHILD - A CASE REPORT

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ABSTRACT
Epithelioid Sarcoma (ES) is an uncommon malignant soft tissue tumor occurring in children. We report a case of proximal variety of ES occurring in a four year old boy. Since it is a rare tumor in this age group, the authors faced a diagnostic dilemma. The clinical presentation, diagnosis and management have been discussed and the importance of immunohistochemistry (IHC) in clinching the diagnosis has been highlighted in this case report.

Key Words: *Epithelioid Sarcoma (ES), Immunohistochemistry (IHC), Pediatric*

INTRODUCTION
Epithelioid Sarcomas (ES) are rare tumors and are even rarer in young children. Incidence being common between 20-40 years of age, although no age is exempt (Casanova *et al.*, 2006). It is an uncommon, malignant, soft tissue tumor that generally presents in fascial planes, aponeuroses and tendon sheaths of the extremities. It represents 5-8% Non-Rhabdomyosarcoma Soft Tissue Sarcomas (NRSTS).

CASES
A four year old boy presented with a swelling over the posterior aspect of his right thigh, which was incidentally noticed by his mother while giving bath. It rapidly and painlessly progressed over one month to attain the present size, which was significant enough to seek medical attention. There was no history of trauma, fever or restriction or alteration in the movements of his limb. On examination, a single, hard, non-warm, non-tender well defined swelling measuring 10x4 cm was found in the middle one-third of the posterior compartment of the right thigh. A provisional diagnosis of Alveolar Rhabdomyosarcoma (RMS) was provided by the referring doctor based on an FNAC report. Apart from the routine hematological investigations, a contrast enhanced computerized Tomography (CECT) scan of chest, abdomen, pelvis and limbs were done. This showed a minimally enhancing, well-circumscribed, soft tissue lesion in the posterior compartment of the right thigh, in the inter-muscular plane, measuring 38x50x41 mm with no calcification. There was no involvement of the bone. There was no evidence of regionallymphadenopathy (Figure1).

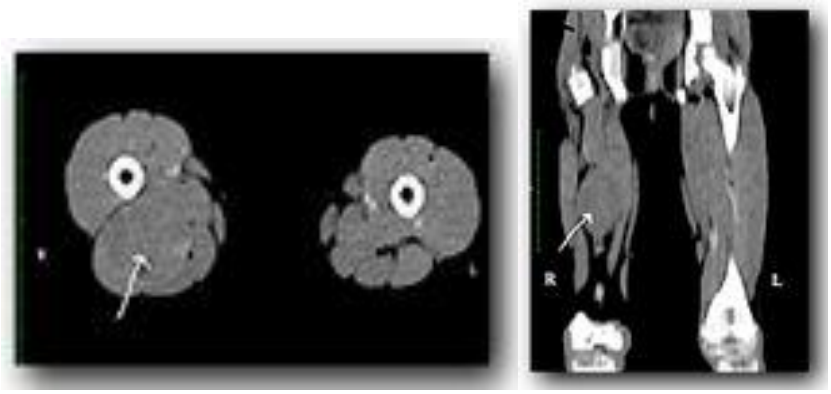

Figure 1:CECT showing well circumscribed Inter muscular ES

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At surgery the tumor was found to be arising from the muscle sheath of adductor magnus. Wide local excision was done, with a 2 cm margin of normal tissue, which was found to be negative for tumor cell as confirmed by frozen section and permanent section later on (Figure 2).

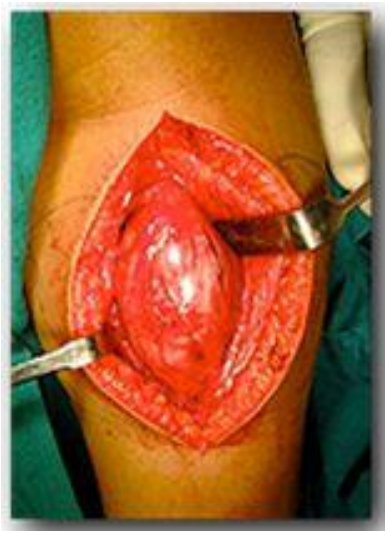

Figure 2: Intraoperative photo showing the lesion

Histopathology was reported, as an well encapsulated tumor comprised of densely cellular non-descript sheets and nests of predominantly polygonal to epithelioid cells with vesicular hyperchromatic mitotically active nuclei with prominent nucleoli with moderately eosinophillic to clear cytoplasm and rhabdoid phenotype. (Figure 3)

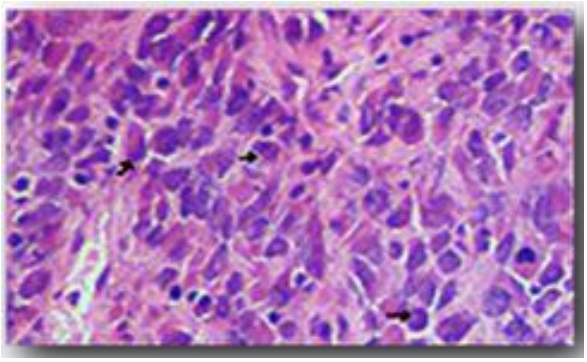

Figure 3: Microscopy slide of H & E staining, showing Rhabdoid cells

A provisional diagnosis of alveolar RMS or extra renal rhabdoid tumor was made. However, IHC showed tumor cells, which were diffusely positive for MIC2, EMA, CD34 and focally positive for CK, Chromogranin and Synaptophysin. While it was negative for Desmin, S-100 protein, MYOD1, CD31 and INI 1. A final diagnosis of proximal ES was made. Post-operative recovery was uneventful. Presently, the child is on chemotherapy with Vincristine, Ifosfamide and Etoposide (VIE), which is to be followed by Vincristine, cyclophosphamide and doxorubicin (VCD).

DISCUSSION

Epithelioid Sarcoma, sub-categorized as NRSTS (Casanova *et al.,* 2006) is rarely reported under the age of 20 years (Al-Salam and Al Ashari, 2010; Chowdhary *et al.*, 2000; Nagoshi *et al.*, 2006; Stang *et al.*,

2010). Histologically, it is classified into the classic type (distal) and the proximal type. The classic type shows distinctive nodular, granuloma like pattern with spindle and epithelioid cells circumscribing areas of central degeneration and necrosis. The proximal form is more aggressive, characterized by sheets of large, atypical, epithelioid cells with vesicular nuclei and prominent nucleoli (Casanova *et al.*, 2006). The later is also characterized by phenotypic features of presence of rhabdoid cells with intra cytoplasmic inclusions. Differential diagnosis for ES include extra renal rhabdoid tumor, atypical melanoma, epithelioid type of malignant peripheral nerve sheath tumor, RMS, angiosarcoma, synovial sarcoma and undifferentiated carcinoma **(**Casanova *et al.*, 2006; Hasegawa *et al.*, 2001; MD Julie *et al.*, 1998) IHC is positive for vimentin and cytokeratin in almost all the cases, moderately positive for CD34 and MIC2 and mildly positive for S100 protein, NSE, CD56 and smooth muscle actin (Hasegawa *et al*., 2001).

Classic ES has typical clinical features of location at superficial distal sights, indolent growth rate and tendency towards loco regional recurrence (Casanova *et al.*, 2006). Proximal type of ES arises in deep soft tissues, has male predominance; local recurrence, metastasis are very common; mortality is approximately 60-65% and has no association with tumor size and metastasis (Hasegawa *et al*., 2001). The mainstay of treatment is wide local excision with or without loco regional lymph node dissection with adjuvant chemotherapy and Radiotherapy.

In our case, the histopathological diagnosis of extra renal rhabdoid or alveolar RMS was proposed, which was ruled out by IHC being negative for desmin, MYOD1 and INI 1. Moreover, the definitive diagnosis of proximal ES was made on the basis of:

Typical appearance of sheets of epithelioid cells, vesicular nuclei, prominent nucleoli and rhabdoid cells; Clinically rapidly growing tumor in deep soft tissue of the thigh; IHC being positive for cytokeratin, EMA and CD34 predominantly

CONCLUSION

The diagnosis of ES should be included in the list of differential diagnosis of soft tissue tumors occurring in children. Moreover, the proximal form of ES, which is a highly malignant and aggressive tumor, requires urgent diagnosis and aggressive treatment. Timely intervention and multi modal approach of managing these patients involving the Pediatric Surgeons, Oncopathologists, Pediatric Oncologists and Radiotherapists may help to bring down the morbidity and mortality.

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