Proximal-Type Epithelioid Sarcoma—A Rare Soft Tissue Sarcoma of Thigh in a Child

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Introduction

Epithelioid sarcoma, first described by Enzinger in 1970 [1, 2], is a rare soft tissue sarcoma of adolescents and young adults usually involving the distal part of upper extremities [1, 2]. The epithelioid sarcomas are difficult to diagnose and can be confused with a variety of malignant and benign conditions; especially granulomatous process, synovial sarcoma and ulcerating squamous cell carcinoma [2]. This is usually a slow growing tumour which commonly involves the dermal or subcutaneous area of distal part of upper extremities. Sometimes it can be deeply seated attached to tendons, tendon sheaths or fascial structures [4]. The “proximal type or axial type” variant was later recognized in 1997 [2], with very few reports on such cases. It is usually seen in adults and are commonly deep seated and occurs in perineum, sex organs, pelvis and thighs. It is more aggressive with poorer prognosis with tendency for local recurrence and early metastases. The proximal type epithelioid sarcomas are rare undifferentiated soft tissue sarcomas usually seen in

adults with epithelioid features and a frequent rhabdoid phenotype on histopathology [1] thus posing diagnostic difficulty and has to be differentiated from extrarenal rhabdoid tumour, synovial sarcoma, angiosarcoma and melanoma [1, 5] (Figs. 1, 2, and 3).

Case Report

A 5 year old boy presented with painless swelling in the posterior aspect of the right thigh of 1 month duration. There was no history of trauma, fever or weight loss. On local examination a well defined swelling measuring approximately 8×6 cms was noted in the posterior aspect of the middle one-third of the thigh. The swelling was non-tender, firm, non-mobile and not associated with local signs of inflammation. Skin over the lesion was pinchable.

Radiograph of the right thigh was done which showed focal soft tissue swelling in the mid thigh posteriorly but no focus of calcification was identified. The femur was normal. Ultrasonogram of thigh was done in GE Voluson Pro using 11 Mhz probe which revealed a well circumscribed isoechoic mass in posterior aspect of thigh. Colour Doppler showed mild internal vascularity within the lesion. Contrast enhanced CT scan of thigh was performed in Asteion Toshiba CT scanner. Non contrast CT scan showed a well circumscribed homogenous oval mass measuring 30×38×45 mm which was slightly hypodense to adjacent skeletal muscles with HU of 50. No calcification was seen in this lesion. The lesion was in intermuscular plane between the posterior compartment and medial compartment of thigh. The lesion was closely abutting and indenting the adjacent muscles i.e. biceps femoris, semimembranosus and semitendinosus muscles laterally and adductor magnus muscle medially. In post contrast CT scan there was mild

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Fig. 1 (a & b): Ultrasound and colour Doppler images showing the mass (white arrow), femoral vessels (thin white arrow) in relation to the lesion

heterogeneous enhancement of the lesion. A few small areas of necrosis were seen in inferior part of the lesion. The lesion was closely abutting the femoral vessels but not encasing it. The lesion was in close proximity to the sciatic nerve. The femur was normal. The possibility of soft tissue sarcoma of thigh probably rhabdomyosarcoma was considered.

To rule out metastatic foci, radiograph of chest, contrast enhanced CT scan of chest, abdomen and pelvis were done, which were normal. No metastatic foci were identified. Bone marrow biopsy was also done which was also normal. Since no distant metastases was seen, the patient was an ideal candidate for surgery.

The patient underwent wide local excision of the tumour with resection of adjacent margins. The lesion was well

encapsulated and measured $6.5 \times 4 \times 3$ cms. There was no perineural invasion or muscular infiltration. Histopathological examination revealed densely cellular non descript sheets and nests of moderately pleomorphic predominantly polygonal epithelioid cells with moderate anisonucleosis, vesicular to hyperchromatic mitotically active nuclei and moderate amount of eosinophilic to clear cytoplasm. Based on histopathological features, diagnosis of undifferentiated sarcoma was considered with possibilities of extrarenal malignant rhabdoid tumour or alveolar soft tissue sarcoma or solid variant of alveolar rhabdomyosarcoma. The tumour was high grade and undifferentiated with TNM stage pT2bNxMx.

Immunohistochemistry revealed that the neoplastic cells are diffusely positive for cytokeratins, epithelial membrane antigen and vimentin and focally positive for synaptophysin and chromogranin. Negative for desmin and myo D. The final diagnosis after histopathology and immunohistochemistry was proximal- type epithelioid sarcoma of thigh.

Discussion

Epithelioid sarcomas are rare soft tissue sarcomas of young adults commonly occurring in distal part of upper extremities like hand [1–3, 5]. Apart from conventional epithelioid sarcomas, fibrous histiocytoma like and angiomatoid subtypes have also been described in literature. Lately, an aggressive subtype has been identified known as “proximal-type or axial-type” epithelioid sarcoma which is usually seen in young to middle aged adults and commonly occurs in perineum, genital tract, pelvis and thigh [1–3]. Peak incidence is between 20 and 40 years [1, 2, 5]. It is more common in males [2, 5]. On hispathology, it usually lacks the “granuloma-like appearance” of conventional epithelioid sarcomas. It is characterized by prominence of epithelioid cytomorphology, sheet like growth pattern of large cells with vesicular nuclei and prominent nucleoli, and frequent occurrence of rhabdoid features [1, 2, 5]. Immunohistochemistry is positive for cytokeratins, epithelial membrane antigen and vimentin [1, 2, 5, 6]. There is consistent staining for CA-125, and some have even suggested to use it as serum marker to monitor for metastases [2]. CD34 can be seen in about 50 to 60% of cases [1, 2, 5, 6].

Role of imaging is to determine the exact location of the lesion, their extent and involvement of adjacent structures and also to rule out metastases. On plain radiograph soft tissue mass or swelling can be seen in some cases. Rarely speckled pattern of calcification or ossification can be seen [2]. In CT scan epithelioid sarcomas are usually seen as well circumscribed masses isoattenuating to the skeletal muscles with variable degree of enhancement. In MRI these sarcomas are isointense to muscle on T1 and heterogeneously

Fig. 2 (a, b, c & d): CT scan images showing the mass as slightly hypodense to adjacent skeletal muscle (white arrow). Neurovascular bundle in close proximity to the lesion. The mass shows mild enhancement in post contrast scan (fig b). Coronal reformation shows a well circumscribed oval mass in inter-muscular plane (white arrows). Sagittal reformation shows the inter-muscular mass in close proximity to the sciatic nerve

hyperintense on T2 with variable degree of enhancement depending on their size and degree of necrosis [2]. Metastases to lymph nodes are commonly seen in epithelioid sarcomas unlike other sarcomas [2]. To rule out metastases CT scan of chest and abdomen and bone marrow biopsy should be done.

Fig. 3 PhotoMicrograph(H&Ex 400) showing densely cellular sheets and nests of moderately pleomorphic predominantly polygonal to epithelioid cells with vesicular to hyperchromatic mitotically active nuclei and moderate amount of eosinophilic to clear cytoplasm

Since imaging features of soft tissue sarcomas overlap the final diagnosis and management depend upon histopathology and immunohistochemistry. The differential diagnosis of proximal-type epithelioid sarcomas are conventional epithelioid sarcoma, extrarenal malignant rhabdoid tumour, epithelioid malignant peripheral nerve sheath tumor, rhabdomyosarcoma, melanoma and undifferentiated carcinomas [6]. The management protocol for patients with epithelioid sarcoma in absence of metastases is wide local excision with resection of tumour margins. Sometimes amputation can be done for recurrent disease or if the tumour is in digits [1, 2, 6, 7]. After surgery, chemotherapy and radiotherapy is also advocated due to high incidence of local recurrence and distant metastases [1, 6, 7]. Prognosis of proximal-type epithelioid sarcoma depends on tumour size and presence of rhabdoid features [2]. The presence of rhabdoid features is related to aggressive behavior, multimodality therapy resistance and rapidly fatal outcome [2].

To conclude we would like to emphasize that it is important to diagnose this rare variety of soft tissue sarcoma as early as possible due to better patient outcome in absence of metastases and smaller size of the tumour. Due to inconsistent pathologic features of epithelioid sarcoma, it is of utmost importance that the biopsy and interpretation of the microscopic specimen is done by an experienced musculo-skeletal oncologist and pathologist, respectively.

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