Case Report

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Rare Case of Metastatic Pleomorphic Liposarcoma of Thigh Presenting as Ewing's Sarcoma

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ABSTRACT

Liposarcoma is an uncommon group of tumors in the pediatric population. Of the various known subtypes,pleomorphic liposarcoma is the most infrequent in children.We present the case of a 7-year old child coming to the radiology department for evaluation of a right knee swelling. Plain radiograph and corresponding MRI images showed features of primary bone tumor possibly Ewing's sarcoma in right distal femur.The child developed right shoulder pain and CT thorax abdomen was done which revealed retroperitoneal mass with hepatic and pulmonary metastases.Subsequent core needle biopsy from the femur confirmed metastatic liposarcoma with pleomorphic features, extremely rare in childhood.The very rare incidence of metastasis that too presenting as a common primary tumor the misdiagnosis of which could result in catastrophic fate. So it's of prime importance that clinicians should be aware of this entity.

Keywords: FISH; liposarcoma; myxiod dedifferentiated liposarcoma; pleomorphic liposarcoma; softsarcoma

INTRODUCTION

Soft tissue sarcomas accounts for less than 1% of all malignant tumors,[1] liposarcoma representing 18% of them. The severity of liposarcoma varies with the different subtypes, distinguished by histopathologically and genetic testing.[2]

According to the World Health Organisation classification, there are four subtypes of liposarcoma: well-differentiated, dedifferentiated, myxoid, and pleomorphic.[3] Irrespective of the histopathological subtype, primary liposarcoma of the bone is rare presenting with differentiation towards adipocytes in histopathological studies. We present an extremely rare case of metastatic pleomorphic liposarcoma in femur resembling Ewing's sarcoma, with hepatic and pulmonary metastases and a synchronous tumor in the retroperitoneum.

Our case is one of its kind and first to be reported which many clinicians are unaware of. We aim to make our clinicians familiarize about this atypical presentation of the entity in the pediatric population for proper management.

CASE REPORT

A seven-year-old child presented to the pediatric oncology department with a two months duration of progressively increasing swelling over the right knee following trivial trauma and weight loss of four kilograms in the same duration. The child also complained of mild occasional abdominal pain, which was never evaluated. There was no history of fever. On clinical examination there was a firm swelling in the distal right thigh and knee and tenderness with local rise of temperature.

Radiograph of the right knee was obtained which showed a lytic lesion in the metadiaphys of right distal femur with a wide zone of transition and permeative bone destruction and associated lamellated as well as Codman's periosteal reaction. Large soft tissue opacity associated with the lesion. No matrix mineralisation was noted. A provisional diagnosis of primary bone tumor ,Ewing's sarcoma was made.Subsequently plain radiograph,contrast enhanced MRI of the local part and a core needle biopsy were advised (Figure 1).

MRI of the right knee showed a long segmental involvement of the marrow of distal meta-diaphysis of the right femur (approximately 16 cm in length), which was hypointense on T1W images, no signal drop on the opposed phase images or on fat suppressed STIR images thus ruling out the presence of intracellular and macroscopic fat respectively. Associated extraosseous soft tissue was noted in the distal diaphysis and metaphysis, infiltrating

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the marrow and destroying the cortex and measuring 6.5 cm in longitudinal extent. The lesion shows restricted diffusion with marked ADC hypointensity and heterogenous post contrast enhancement with areas of necrosis and a type 3 kinetic curve pattern on dynamic contrast enhanced MRI images. Mass effect and displacement of the surrounding muscles noted without frank infiltration. No neurovascular bundle encasement was observed. Distal femoral epiphysis was spared and right knee joint space was uninvolved(Figure 1).

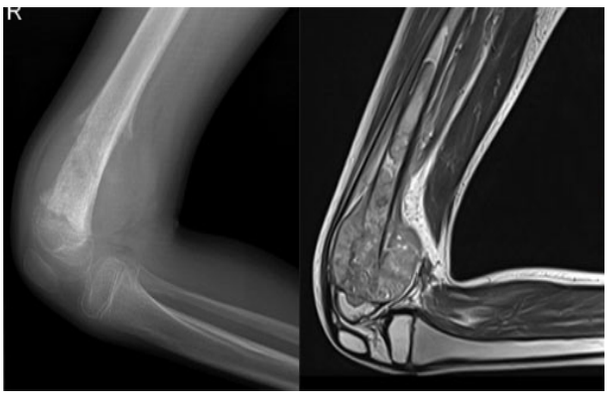

Figure 1. X-ray lateral right knee and corresponding MRI T2 sagittal images showing bone destruction, lamellated periosteal reaction and associated soft tissue component in metadiaphysis of right femur. Presumptive diagnosis of Ewing's sarcoma was made.

In view of the radiograph and MRI findings of a long segmental lesion centered within the meta-diaphysis of right distal femur with extra osseous soft tissue, a presumptive diagnosis of Ewing's sarcoma was made.

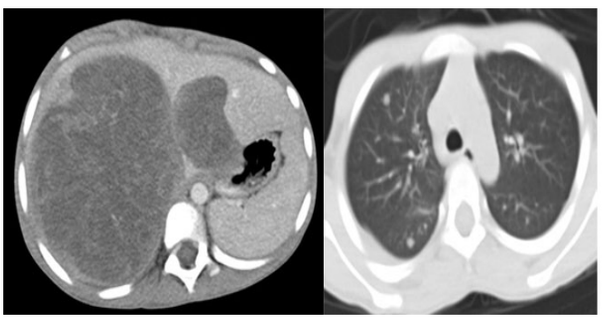

Figure 2. CECT Whole Abdomen and thoracic axial scans show fairly well circumscribed hypodense lesion with internal linear and patchy enhancement in the pre-caval region extending superiorly up to the porta hepatitis and infiltrating right hepatic segments V, VII and VIII and compression IVC and bilateral pulmonary nodules.Retroperitoneal sarcoma with pulmonary metastases was made a possible diagnosis.

On evaluation of the cause of right shoulder pain, a chest X-ray was ordered which showed small bilateral pulmonary nodules and a raised right hemi-diaphragm, which led to the child undergoing a CT scan of the thorax and whole abdomen(Figure 2). There was a large 17 cm circumscribed hypodense lesion (without macroscopic fat) in the retroperitoneum and extending upto the porta hepatis with infiltration right hepatic segments V, VI, VII and VIII. The lesion showed patchy and heterogenous areas of enhancement. Another lesion with similar characteristics was seen in segment IVA of liver measuring five centimeters representing a hepatic metastatic lesion. This large intra-abdominal lesion was the cause of elevated right hemidiaphragm and right shoulder pain in the patient (Figure 2). Multiple bilateral pulmonary nodules were noted on the CT thorax (Figure 2).

Trucut biopsy from the bone lesion revealed a tumour composed of sheets of highly pleomorphic spindled to oval cells with hyperchromatic nuclei, variably prominent nucleoli and ill-defined cytoplasm—favouring the diagnosis of liposarcoma. Immunohistochemistry was done which showed pleomorphic lipoblasts showing strong positivity for Calretinin stain, focal positivity for Desmin stain in the spindled tumour cells and negative for S100, Cytokeratin (AE1/AE3), STAT6 and CD34 (Figure 3 a-f). MDM2-FISH and FUS Breakapart FISH genetic study was suggested and done to rule out De-differentiated and Myxoid / Round cell liposarcoma respectively and was found to be negative for both, thus establishing the final diagnosis of pleomorphic metastatic liposarcoma.

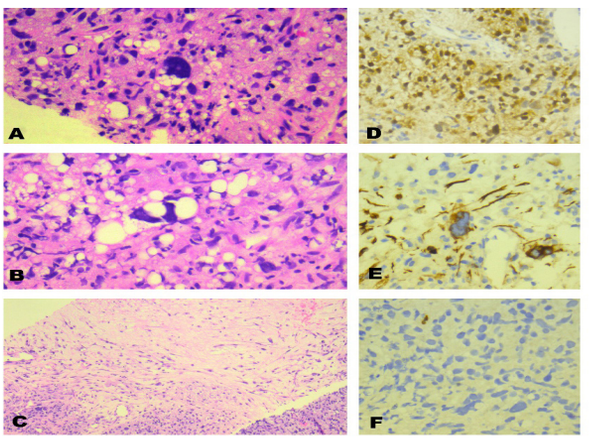

Figure 3. (a-f) H&E, 40x, shows numerous univacuolated (A) and multivacuolated (B) pleomorphic lipoblasts.(c) H&E, 20x, shows sheets of spindled to oval cells with very focal myxoid areas.(d-f) Immunohistochemistry done at TMC shows the pleomorphic cells to be positive for Calretinin (D), Desmin (E) and negative for STAT6 (F), 40x.

In view of poor prognosis and advanced stage of the disease treatment with palliative intent was initiated.

So far, the child has been receiving doxorubicin and ifosfamide chemotherapy. The abdominal pain and right shoulder pain have reduced. The child is non-ambulatory and moves about on a wheelchair, but has been otherwise well and on routine clinical follow up since three months just until this report was written.

DISCUSSION

Soft tissue sarcomas uncommon in children. The incidence of soft tissue sarcomas in children younger than 20 years of age is reported to be 11.0 per million, representing 7% of cancer cases in this age group.[4,5] Amongst soft tissue sarcomas, liposarcoma forms the major bulk with approximately 45% of the cases in children.[6] The peak incidence is reported in the second decade of life, which makes this an unusual case since our patient is only seven years old. Pediatric myxoid liposarcomas are the most common entity while pleomorphic liposarcoma is the least common entity,[7,8] The thigh and retroperitoneum are the commonest sites of involvement. To the best of our knowledge, 12 cases of pleomorphic sarcoma have been reported in children upto 21 years of age[5,7] The age ranging from three years to 21 years, including three in the neck, two in the shoulder, one in the orbit, one in the leg, one intrathoracic tumor, one in the mesenteric root, one in the thigh, one in the back and one in the groin[5,7] A few rare points in our case are as follows; first the child presented with a lesion mimicking a primary bone tumor due to the long length involvement of femur, associated soft tissue and periosteal reactions and no matrix mineralisation, all favouring a radiological diagnosis of Ewing's sarcoma. Metastasis to the appendicular and axial skeleton in all varieties of liposarcomas are rare[9] Second, our patient had a primary retroperitoneal liposarcoma which was of the pleomorphic variety as confirmed on classical histopathological and FISH diagnosis as supported by studies in literature.[10,11] Lastly, the pattern of metastases to the liver and skeleton are rare, since the lungs are the most common site for liposarcomas to metastasise thus demonstrating the poor prognosis of these lesions.

CONCLUSIONS

Our case contradicts to popular belief that long lesions of the limbs in childhood usually favour a primary bone tumor. Even metastases from rarely reported lesions such as a pleomorphic retroperitoneal liposarcoma in our case is a possibility and a multi modality approach including imaging, pathology and cytogenetics is required to come to the most accurate diagnosis.

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