

## AN UNUSUAL BONE TUMOUR - EWING'S SARCOMA AT ELEVEN MONTHS OF AGE

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### **Abstract**

#### **Keywords:**

*Bone tumors, Ewing's sarcoma, X-ray, Magnetic Resonance Imaging (MRI), Malignant small round blue cells.*

Ewing's sarcoma below the age of 5 is extremely rare and radiological features of young Ewing's is not established and can be ambiguous. We present the radiological features of a 11 months old infant that is pathologically proven as Ewing's sarcoma. Ewing's sarcoma can be included in the differential diagnosis list in the presence of sunburst appearance in X-ray and hair on end low signal striations in MRI.

### **Introduction**

Ewing's sarcoma is a bone tumor that is commonly seen among male and female in the second decade and its occurrence is very rare in the first decade. It is the second most common malignant long bone tumor occurring in children and young adults. It has a slight male predilection with a male to female ratio of 2: 1 (1).

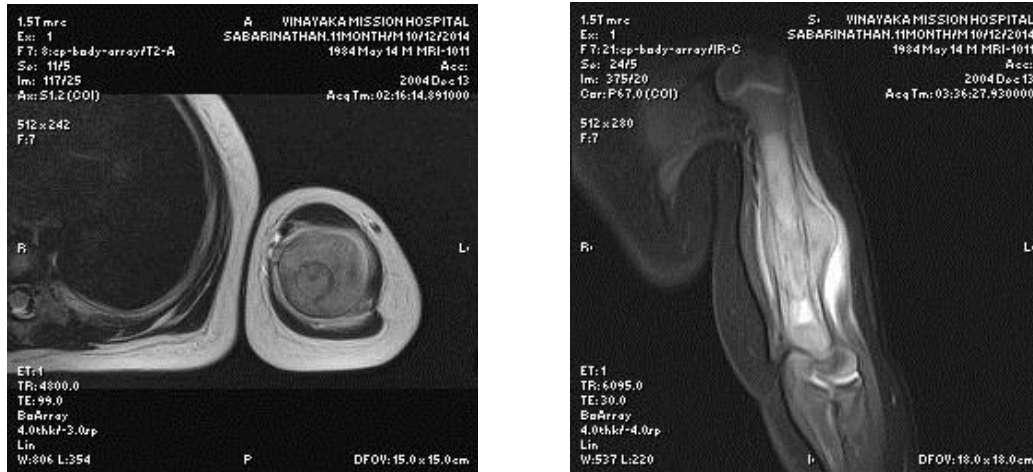
### **Case report**

A 11 months old male child of Indian origin presented with complaints of swelling and pain in the left arm since 6 months. Swelling increasing in size gradually and painful on touch. No history of fever, draining sinus or fall and no constitutional symptoms. Child was treated on and off with antibiotics and anti-inflammatory medication but the symptoms did not subside. On examination a solitary diffuse, warm, tender and non-pulsatile swelling at left mid and lower arm with a smooth and regular margin, hard in consistency and fixed to the bone with no sign of emptying and no sign of indentation was noted. No evidence of discharging sinus from the swelling and no involvement of regional lymph-nodes was seen. Blood investigations revealed elevated white blood cell (WBCs) count and increased erythrocyte sedimentation rate (ESR).



X-ray of left arm shows thick periosteal reaction giving a typical sunburst appearance with bone expansion, cortical thickening and endosteal scalloping.

MRI of left humerus in T2 sequence shows an ill-defined marrow hyperintensity in the humerus shaft (diaphysis) with a transverse fracture at proximal shaft. Extensive mildly hyperintense soft tissue surrounding the shaft with elevation of periosteum is observed. Focal hyperintense fluid noted at the metaphysis extending to the epiphysis. High signal with hair on end low signal striations of the periosteum spotted in the T2 weighted images. A hyperintense soft tissue with moderate signal surrounding the shaft and no extension into the joint space is also seen.



Histopathological examination of the bone revealed an infiltrating neoplasm composed of cells arranged in diffuse sheets. Cells are small with scanty cytoplasm and irregular vesicular nuclei with inconspicuous nucleoli and some cells showed moderate amount of cytoplasm. Extensive areas of necrosis with few viable cells were noted around the vessels. The osteoid is bluish, curvilinear with abortive lacunae formation. No rosettes or calcifications were seen. Malignant small round blue cells witnessed in between trabeculae. One foci of malignant cartilage was present.

Fig. 1

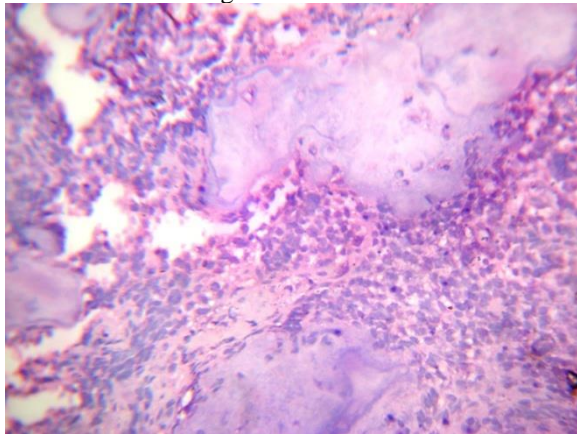


Fig. 1 – High power ( 400 x) – malignant small round blue cells in between the bony trabeculae.

Fig.2

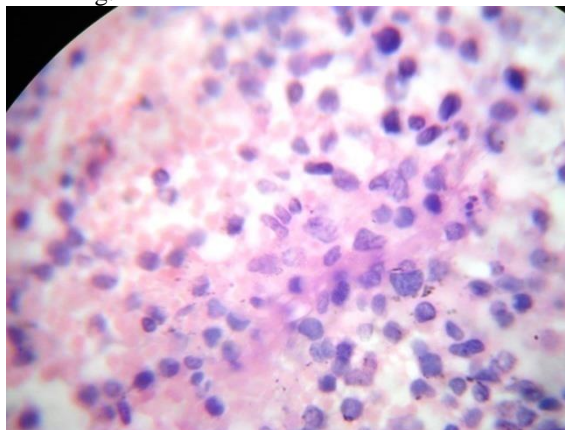


Fig .2 – Oil immersion (1000 x) - Malignant cells

## Discussion

Ewing's sarcoma is the second most common childhood primary bone malignancy and it accounts for 10 – 15% of all primary malignant bone tumours (7). Most of the cases clinically present with a localized pain, with or without a palpable soft tissue swelling. It is caused by translocation between chromosomes 11 and 22 (2). Incidence of Ewing sarcoma in the body is commonly seen in femur (25%) then pelvis (14%) and tibia (11%), in humerus, scapula, and ribs it is (10%,8% and 6 % ) respectively. In long bones it commonly affects the diaphysis (33 – 44%), metaphysis (15%) and epiphysis (1 – 2%). (3)

Bone tumours are rare in pre-school children under the age of 6 (5). Park DY, et.al reported Ewing's sarcoma in a 3 year old boy (6), similarly Maygarden SJ , et.al reported nineteen patients with Ewing sarcoma of bone, each younger than 3 years of age (4). Adding to the former views, this case report of Ewing's Sarcoma in 11 months old baby shows that there is a greater possibility of this tumor occurrence at any age group, even in infants.

## Conclusion

Ewing's sarcoma can occur in infants and it can be included in the differential diagnosis when there is a sunburst appearance in X-ray and hair on end striations in MRI.

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