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Congenital mesenchymoma of tibia: Case report and review of literature☆,☆☆

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Abstract Fibrocartilaginous mesenchymoma is a rare bone tumor arising from long bones in children and adolescents. With only 21 cases reported in the literature, it is often not included in the differential diagnosis in patients with a bone tumor. Previously, the youngest case reported was in a 19-month-old boy, and a congenital variant had not been reported.

We describe a case of congenital fibrocartilaginous mesenchymoma of the proximal tibia that presented shortly after birth. Biopsy of a suspected malignancy on imaging confirmed the diagnosis. The child was temporarily lost to follow up and presented again with genu varum and limb shortening but surprisingly, the tumor had completely resolved without surgical intervention.

Fibrocartilaginous mesenchymomas are rare and unique tumors of bone which were first reported by Dahlin et al. [1]. These tumors are known to occur in children and adolescents. As of April 2011, fibrocartilaginous mesenchymoma (FCM) has been reported only 21 times in the literature. The reported sites of occurrences include the long bones (femur, tibia and humerus), vertebrae, pubic bones and metatarsals [2]. Common presenting symptoms include pain, localized swelling, tenderness and erythema. [3]. The methods of diagnosis include radiological investigations and histological confirmation. This condition was previously classified as a low grade malignancy by Dahlin et al. [1] but in view of the recent evidences it is now not being considered as malignant [4].

The youngest patient reported with FCM in the current literature is a 19 month old male infant. [5]. We describe a case of congenital FCM of the proximal tibia in a female infant noted shortly after birth making our case the youngest reported case of FCM.

1. Case report

A three month old female child with a localized swelling over the right knee was brought to the pediatric orthopedics outpatient clinic by her parents. The swelling was first noticed by the parents a few days after birth and there was a gradual increase in its size. The child was born of a term

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☆☆ Location: The case reported is of a patient who was treated in the Department of Orthopedics, Post Graduate Institute of Medical Education and Research, Chandigarh, India.

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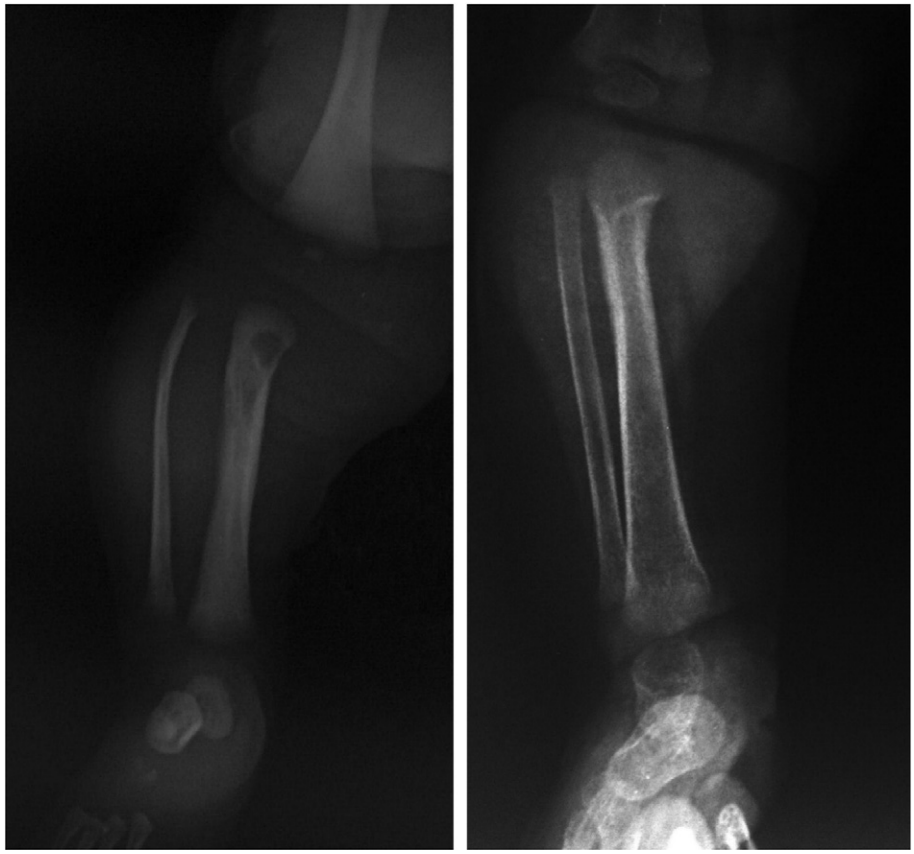

Fig. 1 Anteroposterior radiograph of right proximal tibia at presentation (3 months of age) (left) as compared to that at 17 months of age (right).

pregnancy. On physical examination the child was noticed to have achieved the developmental milestones as per age and was active. A 4 cm×2 cm×0.5 cm localized swelling of was observed around the right proximal tibia which was firm on palpation, non tender and had no accompanying skin erythema. A plain radiograph of the right knee was obtained focusing on the proximal tibia (Figs. 1 and 2), which showed a well demarcated lytic lesion with sclerotic margins suggesting a benign neoplastic condition. Aneurysmal bone cyst, simple bone cyst and Langerhans cell histiocytosis were considered as the probable diagnoses and a magnetic resonance image (MRI) of the affected limb was obtained (Fig. 3). MRI findings showed a large infiltrative mass lesion in the region of epiphysis and metaphysis of the right proximal tibia which was hypointense on T1 and T2 images and showed mild to moderate enhancement post contrast. The lesion extended into the nearby muscles including the tibialis anterior, tibialis posterior and gastrocnemius and encased the popliteal vessels. A diagnosis of malignant soft tissue tumor was made and a biopsy was obtained from the lesion but no other surgical procedure was performed. Histopathological findings showed oval to spindle shaped cells with hyperchromatic nuclei and conspicuous nucleoli embedded in dense fibrocollagenous tissue along with the fascicular arrangement of the cells, anisonucleosis and low mitotic activity consistent with a diagnosis of fibrocartilaginous mesenchymoma. Curettage of the lesion was recommended but the patient's parents refused further surgery. The child was lost to follow up for a period of 14 months. The parents returned with the child (now 17 months of age), complaining of bowing and shortening of the right leg. Surprisingly, the swelling of the proximal tibia had completely resolved and radiographs obtained also corroborated the finding of resolution of the lytic lesion. A plan was set for conservative management of the mild varus of knee with regular and frequent follow-up visits and to intervene surgically if the deformity did not correct on its own. Considering that the tumor may recur, the child will be kept under surveillance and followed periodically.

2. Discussion

Fibrocartilaginous mesenchymoma (FCM) was first identified as a distinct entity in 1984 in the case series of five patients by Dahlin et al. [1]. On the basis of their findings of unusual histologic appearance and local recurrence they considered it as a low grade malignant condition of bones. The gross appearance is characteristic of a locally aggressive lesion that tends to invade the surrounding soft tissues and may break through cortex of the bones.