Case Report

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Pathology Section

Unusual Presentations of a Benign Cartilaginous Tumour- An Interesting and Extremely Rare Case Report

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ABSTRACT

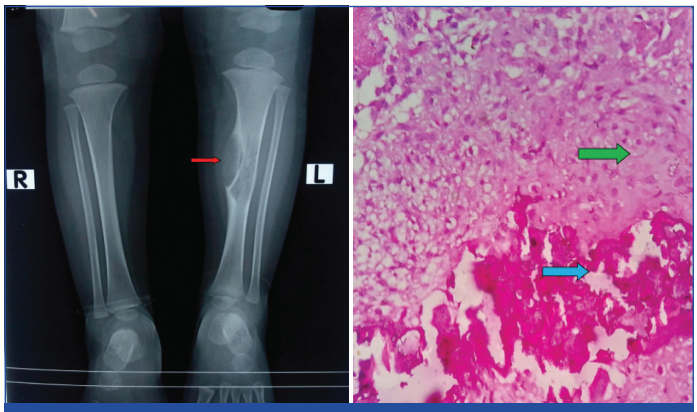
Chondroblastoma is one of the benign cartilaginous bone tumours. Location wise, it is mostly located in immature epiphysis of long bones. Along with epiphysis, many times it also involves metaphysis. Pure metaphyseal tumours are uncommon. Pure diaphyseal tumours are rarest with only very few cases reported in the literature. The most common age group for this tumour is 10-17 years. It is uncommon in patients less than 10 years and those more than 25 years of age. Diaphyseal chondroblastoma in less than 10 years of age group is extremely rare. Hereby, authors report a case of 14-month-old male child who presented to the paediatric orthopaedic outdoor with complaints of the swollen left leg and associated pain by the parents for the last 1 month. There was no history of trauma/weight loss/fever. No difficulty in walking was present. Local examinations of the left leg showed a well-defined globular swelling of size 3×3 cm in the middle part medial aspect. The underlying tibia was continuous with that of swelling having restricted painful movement. X-ray showed an osteolytic, expansile, cortical, eccentric lesion in the middle third (diaphysis) of the left tibia. Correlating the clinical and radiological findings, clinical diagnosis of osteofibrous dysplasia was considered. The intralesional curettage and bone grafting were done and tissue was sent for histopathology. Histopathology showed the characteristic findings of chondroblastoma. The final diagnosis of diaphyseal chondroblastoma of the left tibia was made. The postoperative events were unremarkable and the patient followed-up to eight months with a happy outcome without any residual disease or recurrence.

Keywords: Chondroblastoma, Diaphyseal, Histopathology, Paediatric, Tibia, Tumour

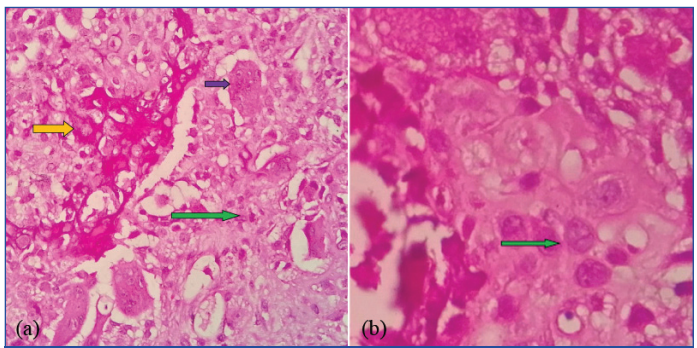
CASE REPORT

A 14-month-old male child presented to Paediatric Orthopaedic Outpatient Department (OPD) with complaints of the swollen left leg and associated pain by the parents for the last one month. The swelling was gradually increasing in size associated with mild pain as observed by parents. There was no history of trauma/weight loss/fever. No other swellings were present in the body. No difficulty in walking was observed. Family history was not significant. Developmental milestones were normal as per age. General examination was not significant with stable vitals. Local examination of the left leg showed a well-defined globular swelling of size 3×3 cm in the middle part medial aspect. The overlying skin was unremarkable. There were no associated features of inflammation like the local rise of temperature or redness. The swelling was firm in consistency with well-demarcated borders. The underlying tibia was continuous to that of swelling with restricted painful movement. The distal neural deficit was not present. Considering bony swelling X-ray was advised along with routine haematological and biochemical tests. Routine haematological and biochemical tests were within normal limits whereas X-ray showed an osteolytic, expansile, cortical, eccentric lesion in the middle third (diaphysis) of the left tibia [Table/Fig-1].

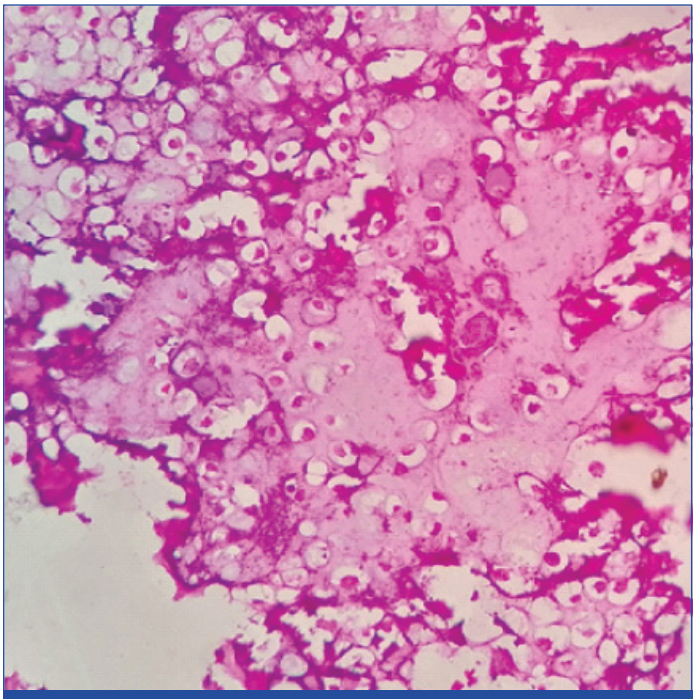
Correlating the clinical and radiological findings, clinical diagnosis of osteofibrous dysplasia was considered. The intralesional curettage and bone grafting were done as per standard protocol and tissue sent for histopathology. There were multiple bits of soft grey-brown tissue along with bone chips measuring 3.0×2.5×1.5 cm. Histopathology revealed a cellular neoplasm comprising of tumour cells having round to ovoid vesicular nuclei and some containing nuclear grooves. There were scattered multinucleated osteoclastic giant cells along with foci of cartilage. There were irregular areas of calcification along with chicken wire calcification [Table/Fig-2-4]. As



[Table/Fig-1]: X-ray anteroposterior view showing an osteolytic, expansile cortical, eccentric lesion in the middle third diaphyseal region of the left tibia.
[Table/Fig-2]: Microphotograph showing round to oval tumour cells (green arrow) and areas of calcification (blue arrow) (H&E,400X). (Images from left to right)



[Table/Fig-3]: a) Microphotograph showing tumour cells having pericellular chicken wire calcification (yellow arrow), osteoclastic giant cells (purple arrow), and round to ovoid to polygonal tumour cells (green arrow) (H&E, 400X); b) Microphotograph revealing some of the tumour cells showing nuclear grooving (green arrow) (H&E, 400X).



[Table/Fig-4]: Microphotograph showing benign cartilaginous tissue with chondrocytes and chondroblasts (H&E, 400X).

the histopathology showed a classical picture of chondroblastoma, final diagnosis of diaphyseal chondroblastoma of the left tibia was made. The postoperative events were unremarkable and the patient followed-up to eight months with a happy outcome without any residual disease or recurrence.

DISCUSSION

Chondroblastoma is one of the benign cartilaginous bone tumours. Location wise it is mostly in immature epiphysis of long bones. It is seen more commonly in males of age group 10-25 years [1]. Chondroblastoma belongs to a group of benign cartilaginous tumours in the 5th edition of the World Health Organisation (WHO) which was classified under intermediate biologic potential previously [2,3]. Characteristically, it is a lytic bone tumour with a site predilection for epiphysis. Along with epiphyses, many times it involves the metaphysis too. Pure metaphyseal lesions are uncommon. Pure diaphyseal tumours are rarest with only very few cases reported in the literature [4]. This benign chondroid neoplasm was first described by Kolodny. He called it a "cartilage containing Giant Cell Tumour (GCT)" in 1927. This tumour was afterwards renamed by Ewing as a "calcifying GCT" in the year 1928 [5-7]. Most common long bones affected by this tumour are the humerus in the upper limb and femur along with the tibia in the lower limb [5]. Mayo-Smith W et al., reviewed five large series and concluded that the bones surrounding the knee joint like the distal femur along with proximal tibia are most commonly affected and next is the proximal part of the humerus [8].

For knowing histogenesis and more affinity towards epiphyses of this tumour several studies were conducted and many types of researches are going on. The majority of them concluded that the major pathway is the active growth plate signaling pathway. Through this pathway, a committed mesenchymal cell for cartilage formation is the main culprit for the development of chondroblastoma. The above explanation may be a possible explanation of the association between chondroblastoma of epiphysis as well as epimetaphysis [9]. Present case is unique as we are describing a pure diaphyseal chondroblastoma. To know the possible pathogenesis in present case we searched literature extensively although could not get satisfactory answer. Brien EW et al., explained that multipotential mesenchymal cells located in tendon sheath may be a possible reason for this diaphyseal origin of chondroblastoma [10]. Ma X et al., published one case of diaphyseal chondroblastoma of the tibia in an 18-year-old male who thought that the origin may be from the embryonic rest of cartilage before enchondral ossification starts [4]. We are also thinking the same as that of Ma X et al., for possible cause in our case. The most common age group for this tumour is 10-17 years. It is uncommon in patients less than 10 years and those more than 25 years of age [11]. One less than 10 years of age and diaphyseal location of chondroblastoma are extremely rare. One case of diaphyseal chondroblastoma of radius in seven-year-old girl child was reported by Punit A et al., (2014) is the only reported case in less than 10 years [12].

The present case will be the first Indian reported case of diaphyseal chondroblastoma in such a young male child of fourteen months of age. Clinical presentations are often non specific. In present case, also there was pain and swelling at the local site. In a minority of cases, it may present as limp [13]. The radiographic apperance is usually suggestive of the diagnosis. It is an Intramedullary lesion with a sharp well-defined margin that may be sclerosed frequently. More often calcification is also seen inside [12]. In present case, it was a lesion with a clear distinct margin located at diaphysis but sclerosis and calcification were not well appreciated. So, we made a diagnosis of osteofibrous dysplasia clinically. Histopathology is confirmatory. It is a tumour comprising of chondroblastic cells with minimal pleomorphism, having a round to oval nuclei over the chondroid matrix. Some of the tumour cells contain nuclear grooves. A good number of osteoclastic giant cells are also noted. Chicken wire calcification (fine calcification surrounding tumour cells) is the most characteristic [4]. Present case was showing characteristic microscopic findings of chondroblastoma. Other giant cell containing lesions like GCT of bone may come as differential but in current case as it was diaphyseal location, containing chicken wire calcification GCT was ruled out. As there was nuclear grooving Langerhans Histiocytosis (LCH) may come as a differential but eosinophils were absent as well as chicken wire calcification was there along with cartilage strongly suggesting chondroblastoma. En-block resection and curettage are the preferred methods of treatment [14]. Chondroblastoma has a good surgical outcome but the major problem is a recurrence which is more common in curettage treated patients as well as in epiphyseal lesions compared to en-block dissection and other locations like diaphysis [12,14]. After curettage and bone grafting recurrence rate is still high varying from 10-35% [15].

CONCLUSION(S)

Diaphyseal chondroblastoma is a rare tumour and manifestations in less than ten years age group is extremely rare. This case we think is the first case of diaphyseal chondroblastoma having the earliest manifestation in 14 months of age. Although exact pathogenesis is not known, further researches may help to know the exact histogenesis. Early intervention results in a happy outcome.

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