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Case Report

Chondromyxoid fibroma of distal phalanx of great toe: a rare case report with literature review

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Abstract: Chondromyxoid fibroma (CMF) in the foot is a rare condition. We report a case of CMF in a 7-year-old girl, affecting the distal phalanx of the great toe. Radiographs revealed a lytic lesion involving the entire distal phalanx, with destruction of both the medial and lateral cortices, while the articular surfaces remained intact. The diagnosis was confirmed through histopathological examination. The patient underwent extensive curettage followed by bone grafting. After a six-month follow-up, no signs of recurrence were observed. This case report aims to present and underscore the uncommon occurrence of CMF in the distal phalanx of the toe in a paediatric patient, drawing attention to its atypical location and age of presentation.

Keywords: Chondromyxoid fibroma, foot, distal phalanx, great toe, benign cartilaginous tumour

Introduction

Chondromyxoid fibroma (CMF) is a rare benign cartilaginous tumour, representing 2% of benign bone tumours and 0.5% of all primary bone tumours [1]. While CMF can occur at any age, it is more frequently seen in males during adolescence and early adulthood (10-30 years) [1]. It tends to affect the bones of the lower limbs, most commonly the metaphysis of the proximal tibia, followed by the ilium, ribs, and distal femur [1]. Pain is the most common symptom, often accompanied by localized swelling [8]. A reduction in range of motion (ROM), joint effusion, and pathological fractures are rare manifestations. The typical radiographic appearance of CMF is a well-demarcated, eccentrically located radiolucent lesion in the metaphysis of long bones. In rare cases, it can extend into the epiphyseal and diaphyseal regions [8]. The definitive diagnosis of CMF is established through histopathological examination, with the presence of a myxoid component being the hallmark feature of the tumour [9]. Surgical intervention is the mainstay of treatment, though there is no universal agreement on the optimal approach [8]. Treatment options include intralesional curettage alone, curettage combined with bone grafting or cementing, wide resection, or en-bloc segmental resection. Postoperative recurrence is relatively common, occurring in approximately 25% of cases, particularly in those treated with intralesional curettage and bone grafting. Notably, recurrence rates as high as 80% have been reported in young children [10]. Additionally, malignant transformation has been observed following radiation therapy.

Involvement of the short tubular bones of the hands and feet is less common, with the toes accounting for fewer than 5% of CMF cases [2]. To date, only 20 documented cases of CMF in the great toe have been reported [2]. We present a rare case of CMF originating in the great toe of a 7-year-old girl, highlighting its unusual location and age of presentation. This report also discusses the clinical, radiological, and histopathological characteristics, along with the challenges in diagnosing and treating this tumour in a young child, thereby contributing to the existing knowledge of this rare tumour in paediatric patients.

Case report

A 7-year-old girl presented to our outpatient clinic with swelling and aching in her big toe,

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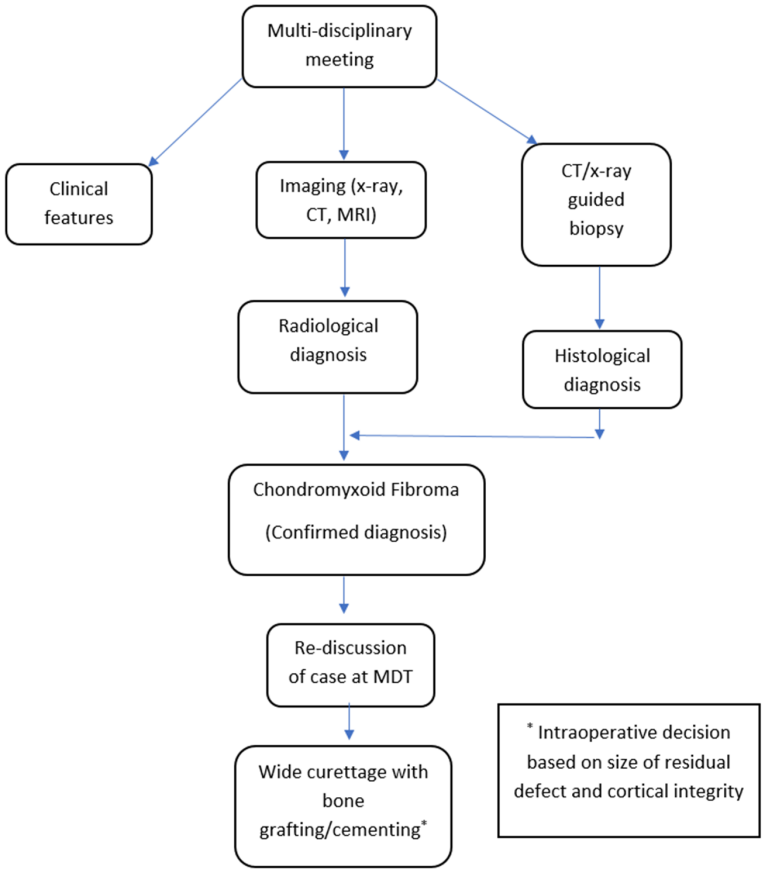

Figure 1. Algorithm for the standardised approach for the treatment of Chondromyxoid Fibroma. MDT, multi-disciplinary team; CT, computed tomography; MRI, magnetic resonance imaging.

which had persisted for the past 8-9 months (**Figure 2**). The pain was dull and aching, with mild to moderate intensity, and showed no diurnal variation. It was not associated with fever, loss of appetite, or weight loss. Initially, the swelling gradually increased in size, but over the past three to four months, it had remained stable. On clinical examination, there was no localized increase in temperature, though the area was tender to palpation. There were no skin changes overlying the swelling, and the lump varied in consistency from firm to hard. The range of motion at the distal interphalangeal (DIP) joint was severely restricted. Standard blood and biochemical tests returned results within normal limits. An X-ray of the foot revealed a lytic lesion in the right distal phalanx of the great toe, involving nearly the entire shaft, eroding the medial and lateral cortices, and extending towards the growth plate. The articular surfaces were spared, and the lesion's borders were well-defined, with no periosteal reaction noted. An MRI of the distal phalanx showed an expansile lytic lesion with areas of cortical erosion, internal septations, and extracortical extension into the surrounding soft tissue. No signs of calcification were observed. The lesion appeared hypointense on T1 and T2 images and hyperintense on PDFS sequences. The patient underwent an open biopsy, wide curettage, and bone grafting. The distal phalanx was accessed via a medial approach, and intraoperatively, soft tissue involvement was noted. The lesion and the cavity wall were thoroughly excised until healthy tissue was visible, and the defect was filled with G-bone (modified hydroxyapatite granules). The curetted specimen consisted of multiple irregular tissue fragments, pearly white in colour, with a consistency varying from firm to soft. The specimen was sent for histopathological examination (HPE). HPE revealed areas of haemorrhage and a lobulated architecture composed of spindle-shaped cells with hyperchromatic nuclei, along with a few multinucleated giant cells, all within a myxoid to chondroid stroma. The diagnosis was confirmed as chondromyxoid fibroma.

At the 6-month follow-up, the patient demonstrated evidence of new bone formation and graft integration, with no reported symptoms. The surgical scar had healed well.

Discussion

Incomplete cartilage maturation is a hallmark of CMF [3]. Jaffe and Lichtenstein first described the condition in 1948 [4]. CMF is typically found eccentrically in the medullary cavity of the metaphysis of long bones, particularly the proximal tibia and distal femur. However, it can also arise in atypical locations, such as the mandible, ilium, ribs, skull, toes, and metatarsals, which can complicate diagnosis [5].

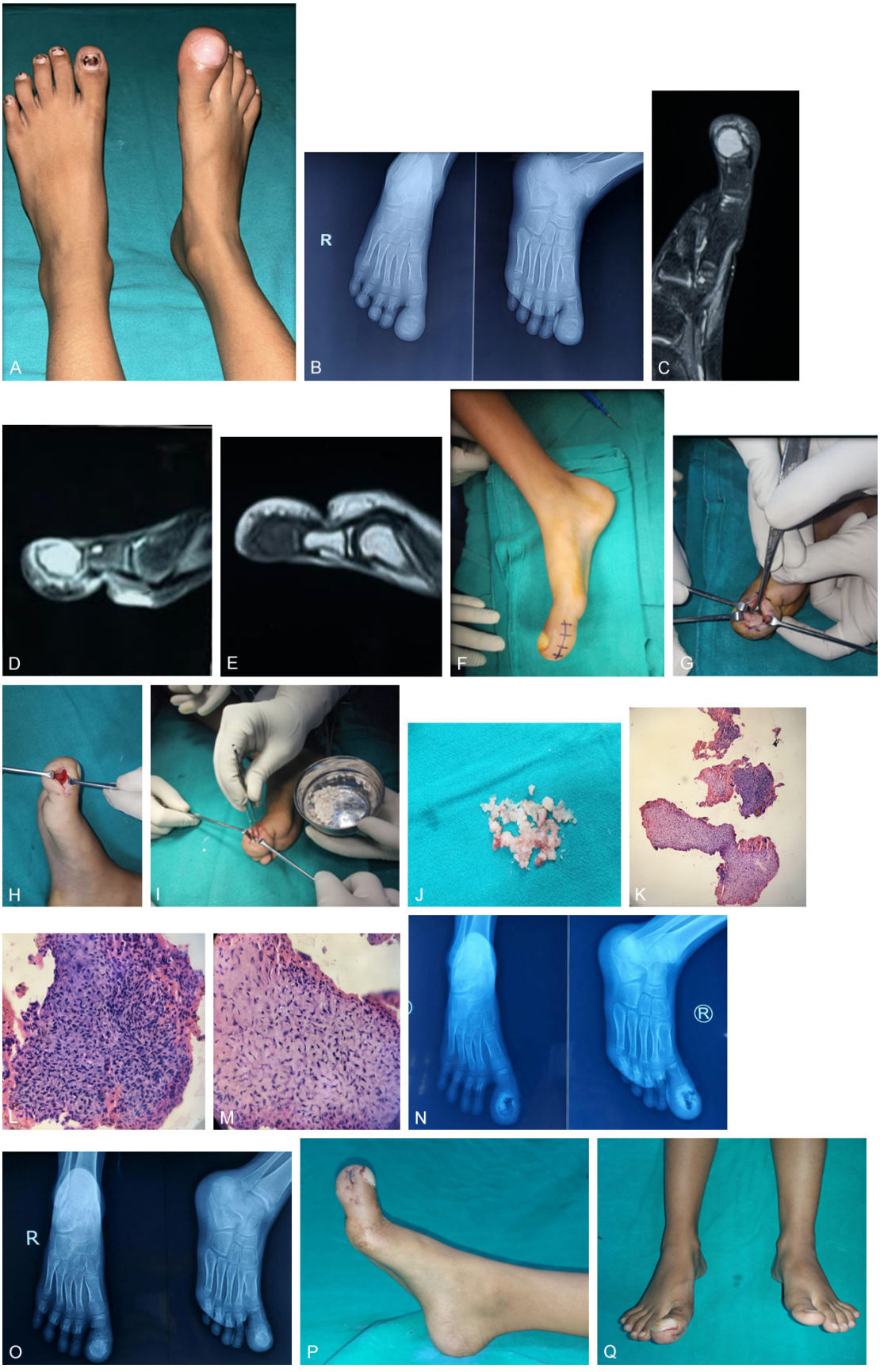
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Figure 2. 7-year female child with swelling of great toe right foot (A), X-ray showing lytic lesion in the distal phalanx of the great toe eroding medial and lateral cortex and approaching growth plate. No periosteal reaction seen (B), well defined expansile lesion with multiple cortical breach and extension into adjacent soft tissue appearing hypointense on T1 and T2 and hyperintense bright on PDFS (C-E), intra-operative image showing marked incision for medial approach (F), curettage of tumour tissue after subcutaneous tissue dissection (G), dead space created after extended curettage (H), dead space filled with modified hydroxyapatite granules (I), gross image of tumour tissues showing several uneven tissue fragments. Its consistency ranged from firm to soft (J), HPE Showing lobulated architecture consisting of spindle shaped cells with hyperchromatic nuclei along with few multinucleated giant cells in a myxoid to chondroid stroma along with areas of haemorrhage (K-M), follow-up x-ray at 3 months and 6 months showing complete uptake of graft (N, O), Clinical image at 6 months of follow-up showing healthy scar tissue with no recurrence (P, Q).

Patients often report persistent localized discomfort, with gradual enlargement of the affected area, as observed in our patient. Clinical examination usually reveals a firm soft tissue mass and restricted motion in the involved joint, consistent with our case. Additionally, pathological fractures may occur [5]. CMF may occasionally be detected incidentally on routine X-rays. On radiographs, it appears as a spherical, lucent lesion that is eccentrically located, with a sclerotic margin and a geographic pattern of bone destruction. Sporadic ridges and septations may also be seen. Calcifications and cortical erosions are rarely observed [6]. MRI is valuable for characterizing the tumour and assessing its extent. CMF typically shows low signal intensity on T1-weighted sequences and high intensity on T2-weighted sequences, consistent with our case. While a CT scan can be used to evaluate cortical integrity and expansion, it was not performed in our case due to the evident cortical breach seen on X-ray. The radiological differential diagnosis includes conditions such as non-ossifying fibroma, fibrous dysplasia, adamantinoma, osteofibrous dysplasia, giant cell tumour, and Brodie abscess [1].

The diagnosis of CMF primarily relies on its characteristic histological features. Histopathological examination (HPE) reveals spindle-shaped or stellate cells dispersed within an extracellular matrix that is either myxoid or chondroid, divided by septa that may contain varying numbers of large cells. Anaplasia has been observed in CMF [7]. The differential diagnosis of CMF based on HPE includes chondroblastoma, enchondroma, and myxoid chondrosarcoma [1]. The diagnosis of CMF in our case was established by integrating radiological, histological (confirmed via needle biopsy), and clinical findings in a multidisciplinary meeting. Treatment options for CMF include curettage, with or without bone grafting or cementation, en-bloc excision, and amputation. The recurrence rate for curettage alone ranges from 20% to 80%, which may be attributed to inadequate excision of pseudopod projections from the tumour into skeletal fissures [2]. In contrast, wide curettage combined with cementation or bone grafting significantly reduces the risk of fracture, facilitates early weight bearing, and has a recurrence rate of only 7% [2]. In our case, the patient was treated with wide curettage and bone grafting (G-bone), which integrated successfully, and the lesion has shown no signs of recurrence. Recurrence rates are higher in children under the age of 15, likely due to their lower resistance to tumour growth [2]. Given the rarity of CMF, particularly in the distal phalanx of the great toe, only a few case reports have been published on this topic. Therefore, we present a standardized approach to the treatment of CMF (**Figure 1**). This condition poses significant diagnostic challenges, especially when it occurs in atypical locations (**Table 1**). Histopathological examination remains the gold standard for definitive diagnosis, as radiological findings can sometimes be misleading.

Conclusion

Chondromyxoid fibroma (CMF) is a rare benign bone tumour that should be considered as a potential differential diagnosis for a solitary, well-defined lytic bone lesion. Due to its high likelihood of recurrence, accurate diagnosis, thorough surgical curettage, and diligent follow-up are essential. Our case of CMF in the distal phalanx of the great toe is only the second reported instance in the literature occurring prior to physeal closure. Although CMF typically affects adults, it can also involve short tubular bones in paediatric patients.

Table 1. Literature review of chondromyxoid fibroma of the big toe

Study	Age (years)/ sex/side	Anatomical site	Duration of symptoms	Clinical features	Radiological features	Surgery	BG	Outcome
Kashyap J et al [11] (2023)	35/M/L	Distal phalanx great toe	-	Gradually progressive pain and swelling	X ray-lytic lesion MRI-lobulated expansile lesion with geographical bone destruction. Iso to hypointense on T1 and hyperintense on T2	Curettage	-	Excellent at 6 months follow-up
Vasudeva N et al [2] (2020)	11/F/R	Distal phalanx great toe	6 months	Gradually progressive swelling and diffuse pain	X ray-lytic lesion abutting the growth plate and eroding the medial cortex. No articular involvement. No periosteal reaction and calcification	Open biopsy and curettage	+ (2^{nd} surgery)	Recurrence at 6 months Wide curettage and synthetic bone grafting (2^{nd} surgery) At 6 months after 2^{nd} surgery, no recurrence
Mallya SP et al [12] (2018)	24/M/L	Mid shaft 1^{st} MT to distal phalanx	8 years	Pain and swelling left great toe. Diagnosed to have CMF on biopsy and was treated. Presented again with pain and swelling in the same toe after 6 years	X ray-expansile, lytic, radiolucent and medullary lesion. The cortex was thinned out with soft tissue involvement MRI-lobulated lesion in the proximal phalanx with predominantly soft tissue component	1^{st} surgery-Curettage and fibular grafting 2^{nd} surgery-toe amputation	+ (1^{st} surgery)	No recurrence at 2 years follow-up
Chang KB et al [13] (2010)	40/F/R	Distal phalanx big toe	-	Gradually enlarged swelling	X ray-expansile lesion with sclerotic border affecting base of distal phalanx. There was cortical erosion and calcification MRI-T1 hypointense and T2 heterogenous mildly hyperintense mass in the mid portion of the distal phalanx of the big toe	Amputation at the base of proximal phalanx	-	No recurrence at 6 years follow-up, mild pain

M - male, F - female, R - right, L - left, (+) - yes, (-) - no, BG - bone grafting.

Disclosure of conflict of interest

None.

Abbreviations

CMF, Chondromyxoid Fibroma; DIP, Distal Interphalangeal; HPE, Histopathological Examination; MRI, Magnetic Resonance Imaging; PDFS, Proton density-weighted fast spin-echo sequence.

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