Chondromyxoid Fibroma

Adam M. Budny, DPM,1 Ameera Ismail, MD,2 and Lawrence Osher, DPM3

Chondromyxoid fibroma is an uncommon benign cartilaginous tumor accounting for less than 1% of all bone tumors. The classic site of involvement is the metaphyseal region of long tubular bones, usually presenting in the second to third decades of life with nonspecific pain and swelling of the affected part. This case is an interesting incidental finding in a juvenile hallux abductovalgus deformity with no prodrome, eccentrically located in the diaphysis of the first metatarsal. Early detection and complete surgical resection are valuable in preventing recurrence in the affected bone. ACFAS Level of Clinical Evidence: 4. (The Journal of Foot & Ankle Surgery 47(2):153–159, 2008)

Key Words: condromyxoid fibroma, benign neoplasm, cartilaginous tumor

Chondromyxoid fibroma (CMF), first described by Jaffe and Lichtensein in 1948 (1), is generally accepted to be the least common neoplasm derived from cartilage and represents less than 1% of all bone tumors (approximately 0.5%) (2–4). CMFs arise most frequently in the metaphysis of long tubular bones; however, they may involve virtually any bone of the body (5–10). Roughly 75% of cases involve the lower extremity, with 40% involving the knee; nonetheless, some 17% to 20% occur in the foot, making this the second most common location (11, 12). A number of authors have opined that the metatarsals are the most commonly affected (6, 7, 13).

Clinically, these tumors present with an insidious onset of pain and swelling, which is tender on palpation but not aggravated with range of motion of adjacent joints unless it is in a juxta-articular location (3, 6, 14–16). Occasionally, the tumor may be asymptomatic and discovered on routine radiographs; however, pathologic fracture is also a distinct possibility if development has gone unnoticed. Most cases are found in the second and third decades of life and demonstrate a slight bias toward male sex with a prevalence (male:female) of 1.5:1 (3, 4, 12, 14, 17).

Radiographic appearance of CMF can be variable; however, typical features include those associated with a benign, slow-growing tumor. The classical appearance is that of a well-circumscribed, eccentric geographic pattern of bone destruction with a round to ovoid shape (1, 3, 13, 14, 18–20). In addition, a well-defined, narrow sclerotic zone of transition is seen, and the lesions are typically in the metaphyseal region. Periosteal new bone formation and cortical erosion are uncommon.

Advanced imaging modalities including computed tomography, magnetic resonance imaging (MRI), angiography, and bone scans have all been used as adjuncts in diagnosis (3, 12, 14, 17). In addition to the aforementioned radiographic features, specific findings discerned from MRI and computed tomography include excellent spatial resolution and delineation of the anatomic boundaries of the tumor. The MRI features associated with CMF are varied, and there is considerable overlap with other neoplasms. It has been described as homogenous and isointense with muscle on T1-weighted images but hyperintense on T2 images (12, 14, 20). In addition, peritumoral marrow edema may be identified. Tc-99 bone scans and angiography characteristically demonstrate increased uptake, and neovascularization may be present (12, 20).

Definitive diagnosis of CMF is based on histological analysis via biopsy or curettage. Microscopic pathology demonstrates variable proportions of chondroid, myxoid, and fibroid tissues (1, 3, 4, 6, 12, 16, 17). The cartilaginous portion is in different stages of maturity, ranging from chondroblasts to chondrocytes with stellate or spindle-
shaped cells, whereas the myxoid areas have a loose, vac-

The lateral view demonstrated obvious involvement of

Fibroblasts, histiocytes, and multinucleated giant cells are found in the fibrous regions.

Treatment for this neoplasm includes curettage and pack-

Curettage and packing are usually successful with a low rate of reoccur-

Most recurrences are within the first 2 to 3 years after the index procedure (3, 4, 15).

Case Presentation

In January 2005, a 10-year-old girl, active in athletics, presented with complaints of painful bilateral bunion defor-

The orthopedic examination was consistent with juvenile hallux abducto valgus, which was painful over the medial osseous prominence, and there was mild crepitus through range of motion.

Apart from an obvious hallux valgus deformity, antero-

The typical radiographic picture of a chondromyxoid fibroma is that of a sharply defined, eccentric, round, or, somewhat less commonly, oval geographic lytic lesion centered in the metaphysis that is usually less than 5 cm
in its greatest diameter. Almost all CMFs in long bones are eccentric, whereas centrally placed lesions are more common in thin bones such as the ribs, fibulae, and small tubular bones. Contours are commonly scalloped, and the endosteal margin is usually well demarcated by a rim of host bone sclerosis (type A margin). Cortical thinning and/or replacement (“expansion”) is quite frequent along with a trabeculated appearance that largely results from ridging of the inner border (surrounding host bone). Matrix/intralesional calcification, periosteal new bone production, and pathological fracture rarely occur (10, 12, 21). Periosteal reactions, when present, are thought to largely be the result of pathologic fracture, although this is not universally true (3).

There are several characteristics that make our case presentation unique, apart from the obvious fact that CMFs are generally quite rare: 1) the lesion was asymptomatic and discovered incidentally; 2) the lesion is eccentrically located, even though the involved bone is a short tubular foot bone, and calculation of the lesion epicenter strongly suggests a juxtacortical origin; 3) unlike most CMFs of the small tubular foot bones, the lesion did not occupy the entire medullary cavity; 4) the lesion is centered in the diaphysis and without evidence of metaphyseal extension; and 5) the

FIGURE 2  (A) Anteroposterior radiograph. (B) Close-up detail of first metatarsal. (C) Lateral radiograph.

FIGURE 3  Coronal T1 MRI.
lesion is oval in shape (atypical when situated in short tubular bones).

In his textbook, Mirra states that more than 95% of CMFs are centered in the metaphysis (13). Indeed, in our case, the diagnosis of CMF was originally discounted because of the lesion being centered in the diaphysis. In addition, “extreme eccentricity” is an unusual presentation of CMFs that is frequently accompanied by a periosteal buttress (13). When small to moderate in size (1–4 cm), these lesions can resemble another rare tumor—the parosteal chondroma. Although Mirra does not quantify “extreme eccentricity,” the comparison suggests that these lesions are largely juxtacortical without a significant medullary component. However, in our case, the lesion demonstrated a striking tendency toward early planter eccentricity with cortical replacement and demonstrates that eccentricity of CMFs can indeed occur in small tubular bones.

We postulate that eccentricity typifies the early picture of these slow-growing lesions in short tubular bones, and its importance should outweigh considerations of epiphyseal, metaphyseal, or diaphyseal origin. Unfortunately, the vast majority are discovered when symptomatic and much further developed. In Wilson et al’s review, cortical expansion was one of the most consistent radiographic features and was present in 89% of the cases. In addition only, 4 out of 30 CMFs involving long and short tubular bones were solely confined to the diaphysis (11). Indeed, CMF should always be considered when evaluating a solitary geographic bone lesion that has lobulated margins, septations, cortical expansion, and/or a sclerotic rim in a patient who is in the second or third decade of life (11).

The significance of this early eccentricity should not be discounted when one considers that it is quite likely to be the defining radiographic feature of our case (eg, early, eccentric growth with cortical replacement), even though the anatomic epicenter in the diaphysis favored other lesions (ie, fibrous). Chondromyxoid fibroma is not unique, how-

**FIGURE 4** (A-D) Coronal T1-weighted images demonstrating markedly eccentric plantar-lateral location of lesion. The lesion is intermediate in signal intensity and isointense with muscle. Tracings outlining contour of lesion demonstrate approximately equal intraosseous and extraosseous components with respect to the first metatarsal bone.
ever, in its tendency to be situated eccentrically within the bone, and several other primary neoplasms of bone do so (Table 1). However, with respect to these other lesions, we are unaware of any other studies that demonstrate this early predilection (as indicated by the MRI evaluation), whereby the extraosseous and intraosseous proportions are approximately equal. According to Schajowicz and Gallardo, in short or small tubular bones such as the metatarsals, phalanges, fibula, or the ribs, CMFs will generally occupy the entire width of the affected bone, producing fusiform expansion along with thinning of both cortices (15). This notion is echoed in a number of other textbooks and case reports where it has indeed been noted that, at the time of discovery, CMFs tend to occupy the entire bone when short tubular bones are involved (2, 5, 15, 21). Timely identification may be difficult because of the long period of latency before symptoms manifest. In contradistinction, our case presents a unique opportunity to observe the early behavior of CMFs before clinical complaints of deforming enlargement or symptomatic discovery.

In addition, we explored the plausibility of a hypothesis that pedal chondromyxoid fibromas demonstrate preferential eccentric plantar bony cortical extension, as occurred in our case study. To this end, we reevaluated much of the available pedal CMF literature. Although a few studies have noted this with reviews of case photos of CMFs involving

FIGURE 5  (A) Coronal T2. (B) Coronal STIR. (C) Oblique-axial short tau inversion recovery clearly demonstrates midshaft location within first metatarsal bone.
the os calcis, much to our surprise, our initial findings did indeed support this contention for other major weightbearing foot bones (14, 22–25). In most instances, it is interesting to note that the involved bones are not symmetrically shaped but demonstrate increased external plantar concavity (inner convexity) primarily in the sagittal plane. Inasmuch as the plantar surfaces of the metatarsal and calcaneal bones are subject to tensile forces, this structural geometry is presumably best suited to oppose the stress of weightbearing. Conversely, those CMFs in small tubular bones, which demonstrate metaphyseal to epiphyseal extension, where the inner cortical surfaces become biconcave, were much more likely to be concentrically situated. If this observation proves to be correct, it is enticing to theorize that bioelectric principles may be involved. It should also be noted that no such trends could be identified in CMFs arising in phalan-

FIGURE 6 Intraoperative photos. (A) Creation of triangular window. (B) Cortical window removed exposing lesion.

FIGURE 7 Photomicrographs. (A) 10×. (B) 20×.

FIGURE 8 Radiographs at 8-month follow-up. (A) Anteroposterior. (B) Lateral.
Conclusion

Herein, we present an interesting case of chondromyxoid fibroma as an incidental finding during management of juvenile hallux valgus. The rarity of this early, asymptomatic discovery affords an opportunity for a noteworthy discussion of potentially discriminating radiographic clues for a lesion otherwise thought to be radiographically similar to a host of other benign neoplasms. Diagnostic clinical and radiographic characteristics for differentiation are presented along with a comparative review of the literature.

References