Case Report
Chondromyxoid fibroma of the distal phalanx of the great toe: A tumor with unusual histological findings

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Chondromyxoid fibroma (CMF) rarely arises in the distal phalanx of the foot and less than 20 cases have been reported in the literature. It has also been known to show a wide spectrum of histology mimicking other primary bone tumors. An unusual case of CMF arising in the distal phalanx of the left great toe is reported because of its unique anatomic site of origin and histology. A 53-year-old female presented with a slow growing, painful great toe of the left foot which she had had for 3 years. She had first noticed the mass 25 years ago. On admission, plain X-ray revealed an osteolytic mass with a sclerotic margin expanding to the distal phalanx of the great toe. Interestingly, the lesion was microscopically composed of hypercellular chondromyxoid lobules separated by hypocellular fibrous tissue, which is in contrast to the typical histology of CMF. In addition, the lesion showed an aggregate of tumor cells with pleomorphic multinucleate or giant nuclei within the chondromyxoid matrix, which were not similar to the osteoclast-like type. Perhaps these unusual histological findings may be associated with its long duration and presenting location.

Key words: chondromyxoid fibroma, distal phalanx, foot, histology

Chondromyxoid fibroma (CMF) is an uncommon benign primary tumor of the bone that was first described by Jaffe and Lichtenstein in 1948.1 It occurs most frequently in young adults and usually develops in the metaphyses of the long bones of the lower extremities.2,4 Rarely does CMF arise in the distal phalanx of the foot, which has been reported in less than 20 cases in the English literature.4-7 Chondromyxoid fibroma has also been known to show a wide spectrum of histology mimicking other primary bone tumors. We report a case of chondromyxoid fibroma arising in the distal phalanx of the left great toe because of its unusual anatomical site of origin and histology.

CLINICAL SUMMARY
A 53-year-old female presented with a slow growing, painful great toe of the left foot which she had had for 3 years. She had first noticed the mass 25 years ago (Fig. 1a). On admission, plain radiograph revealed an osteolytic mass with a sclerotic margin expanding to the distal phalanx of the great toe (Fig. 1b). The lesion showed low-signal intensity on T1-weighted magnetic resonance imaging (MRI) while a heterogeneous high-signal intensity with focal low-signal was observed on T2-weighted MRI (Fig. 1c). Two days after admission a metatarsophalangeal disarticulation of the left great toe was performed.

PATHOLOGICAL FINDINGS
The tumor, which measured 3 x 2.2 cm across, was relatively well demarcated, gray-tan in color, and solid. It replaced the entire distal phalanx of the left great toe but did not invade the articular cartilage of the metatarsophalangeal joint or adjacent soft tissue. Focal myxoid areas within the tumor were noted (Fig. 2).

Microscopically, the tumor consisted of well-circumscribed hypercellular lobules of myxoid or cartilaginous tissue that were delineated hypocellular septae (Fig. 3a). The lobules were composed of stellate-, spindle- or oval-shaped tumor cells residing within the lacunae. More cartilaginous foci characterized by oval or spindle tumor cells residing within the lacunae were merged with the myxoid areas. In some areas, atypical giant cells with abundant eosinophilic cytoplasm and aggregates of foam cells were seen. These giant cells were not similar to those of the osteoclast type (Fig. 3b). The tumor cells were not arranged in cohesive nests or linear cords. Additionally, no mitoses or
Figure 1  (a) The patient had a slow-growing, painful great toe of the left foot for about 25 years. (b) Plain radiographs of the left foot showing an osteolytic mass with a sclerotic margin. (c) T2-weighted magnetic resonance imaging of a well-defined, lobulated, high-signal intensity mass mixed with several hypointense signal areas.

areas of necroses were identified. On immunohistochemical staining, the tumor cells were stained strongly with high- and low-molecular weight cytokeratins (AE1/AE3) and vimentin, but no staining was demonstrated for S-100 protein, CAM5.2 or epithelial membrane antigen (EMA) (Fig. 4; Table 1).

DISCUSSION

Chondromyxoid fibroma is an unusual tumor that occurs mostly in the metaphysis of the long bones near the knee of young adults. It rarely arises within the distal phalanx of the foot (close to 4% of cases). Some 75% of patients are younger than 30 years of age, most being between 10 and 20 years and the age ranges from 4 to 79 years. It has been reported that cases arising in the hands and feet occur in older patients compared with CMF of the long bones. The typical signs and symptoms are: pain of varying duration, ranging from 2 weeks to 20 years; a palpable mass or swelling; and infrequently pathological fracture. In the present case, the patient had clinical symptoms of a longer than usual duration. Radiographically, most CMF of the long bones have manifested as well-demarcated, metaphysically centered, eccentric osteolytic lesions bordered by a thin rim of sclerosis. In contrast, CMF of the short tubular bones have been known to show a centrally located symmetric expansion. Therefore, given these radiological features and location, prime diagnoses are either enchondroma or CMF, the former being favored because it is the most common benign tumor occurring in the bones of hands and feet.

Most CMF do not recur after simple curettage but local recurrence and malignant transformation have been
Chondromyxoid fibroma of the great toe

Figure 3  (a) The tumor was composed of well-circumscribed hypercellular lobules of myxoid or cartilaginous tissue with hypocellular septae (HE). (b) In some areas, atypical giant cells not similar to those of the osteoclast type, are seen (HE).

Figure 4 Immunohistochemical staining of (a) keratin (AE1/AE3) and (b) vimentin demonstrating a strong positive reaction in the tumor cells (avidin-biotin peroxidase method).
reported. In rare cases, CMF can also recur in the soft tissue. The cause of recurrences is considered to be inadequate removal with limited growth potential, rather than an inherently aggressive or potentially malignant property of the lesion.10,11

Chondromyxoid fibromas have highly variable histological patterns, perhaps second only to osteosarcoma. Typically, CMF are characterized by hypocellular chondromyxoid lobules surrounded by dense peripheral bands or condensations of fibroid elements. Rarely, varied sized foci of osteoclast-like giant cells; degenerative changes such as foamy macrophages and cholesterol clefts; osteoid and/or woven bone production; and giant nuclei of tumor cells are described.12 Interestingly, the lesion of the present case was composed of hypercellular chondromyxoid lobules separated by hypocellular fibrous tissue, which is in contrast to the typical histology of CMF. In addition, this case showed an aggregate of tumor cells with pleomorphic multinucleate or giant nuclei within a chondromyxoid matrix, which were not similar to those of the osteoclast- like type. Foci of foamy macrophages admixed with plasma cells were noted but mitoses or necroses were not seen. Perhaps these unusual histological findings may be associated with the long duration of 25 years compared with conventional cases of CMF (average duration, 22 months) and the presenting location of this case, which is susceptible to trauma.

The important neoplasms in the histological differential diagnosis of CMF of the present case chiefly include high-grade conventional chondrosarcoma, myxoid chondrosarcoma and enchondroma with hypercellularity. Some chondrosarcomas may demonstrate a lobular growth pattern similar to CMF, while some CMF can destroy the cortex and expand into the soft tissue and often show cytological pleomorphism. However, chondrosarcoma can be distinguished from CMF by its permissive growth pattern, the presence of well-formed hyaline cartilage and mitoses, and the absence of a fibrous component. In addition, most high-grade chondrosarcomas exhibit more aggressive features on radiographs, such as extensive and irregular cortical destruction and extension into surrounding soft tissues, which is different from those of the present case.5 The majority of myxoid chondrosarcomas arise from the extraskeletal soft tissue, and the tumor cells are frequently arranged in cords surrounded by myxoid stroma, which is not commonly seen in CMF.12,13 Immunohistochemical studies were not helpful in separating chondrosarcoma from CMF because both have been known to express vimentin or S-100 protein.14-16 Enchondromas are predominantly characterized by paucicellular cartilage, although hypercellular areas are commonly seen. The nuclei of enchondromas maintain a round to oval configuration, even in areas of increased cellularity. Moreover, typical findings of CMF, which demonstrate lobular arrangements of myxomatous and chondroid stroma surrounded by fibrous tissue with spindle cells, are not seen in enchondromas.5

Immunohistochemically, CMF usually demonstrated vimentin or S-100 protein but not keratins or EMA.13-16 However, in the present case, the tumor cells were positive for keratins (AE1/AE3) although they expressed vimentin. Keratins (AE1/AE3) are used for detecting high- and low-molecular weight keratins in the form of cocktails while CAM5.2 is only for low-molecular weight keratins.17 This suggests that the tumor cells may be positive only for high-molecular weight keratins. S-100 protein is identified in chondrocytes, chondromas, a varying percentage of CMF and chondrosarcomas.13,14-16 Thus, it indicates that the expression of vimentin in conjunction with keratins and the absence of S-100 protein, may be associated more with the anaplastic morphology of the tumor losing its chondroblastic activity.

REFERENCES
