Monostotic fibrous dysplasia of the distal radius metaphysis: case report

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Abstract We report the case of a 56-year-old male with distal radius monostotic fibrous dysplasia that was detected when the patient suffered a pathologic fracture. This case is unusual because of the location of the tumor and the age of the patient.

Introduction

In 1938, Lichtenstein first coined the term “fibrous dysplasia” to describe a subset of benign bone tumors previously classified as osteitis fibrosa [14]. He described this pathology as filling involved bones with fibrous tissue containing trabeculae of immature bone and postulated that it resulted from aberrant activity of bone-forming mesenchyme; he determined that it generally presented by early adult life, predominantly in females, as pain, deformity, and a predisposition to pathologic fracture. In 1942, Lichtenstein and Jaffe described two types of fibrous dysplasia: monostotic, occurring at one locus in one bone, and polyostotic, occurring at multiple loci in one or multiple bones [15]. Monostotic fibrous dysplasia is the more common of the two and generally portends a better outcome [11, 19, 20]. It is commonly found in the tibia, fibula, ribs, and the bones of the jaw and skull but may occur in any bone [1, 8, 10, 15]. When it arises in the upper extremity, the humerus is most often affected [9, 13, 20]. It is reportedly a rare tumor of the hand; however, several case reports describe monostotic fibrous dysplasia presenting in the metacarpals [2, 11, 13, 19, 21]. We present a unique case of monostotic fibrous dysplasia in the distal radius of an adult male.

Case Report

A 56-year-old male gardener presented to an outside clinic with dull wrist pain. He fell on an outstretched arm 1 month prior and visited an outside physician because the pain was not resolving. Plain radiographs demonstrated a mass in the distal radius metaphysis, and a subsequent MRI demonstrated an expansile lesion in the distal radius with cortical erosion and surrounding soft tissue swelling. The patient was then referred to our hand surgery clinic. Repeat radiographs revealed an expansile, lytic lesion in the distal right radial metaphysis with a slightly radiopaque central nidus and surrounding lucency (Fig. 1). The lucency extended into the subchondral portion of the articular surface, which was well aligned. The cortex showed some expansion, both radially and ulnarly. On the volar surface, there was a small, non-displaced fracture in the region of cortical thinning. Computed tomography (CT) revealed some unusual trabeculation (Fig. 2). Complete blood cell count and serum alkaline phosphatase were within normal limits. The patient’s chest X-ray was normal.

An incisional biopsy of the distal radial mass was performed, which showed a predominantly fibroblastic proliferation and irregular woven bone formation. The bone formation was consistent with metaplastic bone formation as seen in fibrous dysplasia. Cytogenetic analysis of the specimen revealed a normal karyotype.

At this point, the patient was counseled that, while this was a benign lesion, he was at risk for further pathologic fracture of the distal radius, especially given his work as a gardener. He was advised to undergo curettage and grafting of the lesion. The patient then returned for curettage from a dorsal approach. The tumor was accessed via cortical window in the floor of the fourth dorsal compartment. The white, rubbery mass was fully removed from the metaphysis (Fig. 3). It was found that the cortical bone was solid on all sides

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Fig. 1 a, b PA and lateral radiographs of the affected wrist demonstrating a lytic lesion in the metaphysis of the distal radius

except for a small perforation on the ulnar side. In order to provide additional stability to torsional and bending forces, two crossed 0.062-in. Kirschner wires were placed across the metaphysis, and then the metaphyseal void was filled with carbonated apatite bone cement (Norian SRS, Synthes North America, West Chester, PA) (Fig. 4). Pathologic analysis of excised material confirmed the diagnosis of fibrous dysplasia.

It was our intent to leave the Kirschner wires in the radius, but one was prominent and was irritating to the patient. Therefore, the pins were removed after 2 months (Fig. 5).

The patient is currently at 17 months following tumor excision and grafting. He is back to work and reports no

Fig. 3 Fibrous dysplasia of the distal radius following curettage. The mass was light tan in color, rubbery in texture, and was easily removed as an intact single large mass

pain. He has full pronation and supination. Wrist flexion is 65°, and extension is slightly limited at 30°. Radiographs demonstrate fracture healing, and there is no evidence of recurrence of the tumor.

Discussion

Monostotic fibrous dysplasia is a benign, expansile lesion that rarely affects the radius. Although some authors report polyostotic fibrous dysplasia of the radius to be relatively common, a review of the literature revealed only 14 cases of fibrous dysplasia occurring in the radius; it is unclear if the proximal or distal radius was affected in these cases [8, 11, 14, 18, 20]. There are, to date, no published reports of monostotic fibrous dysplasia of the distal radius.

Radiographic findings in fibrous dysplasia are non-specific [18, 19]. Lesions in long bones generally appear at the metaphysis as an intramedullary expansion or broadening of the bone with thinning of the cortex [16].

Fig. 2 a, b Axial sections of CT scan of the wrist demonstrating lytic lesion of the distal radius

Fig. 4 Intraoperative radiograph demonstrating distal radius with crossed Kirschner wires, and the metaphysis filled with cement

They are typically said to have a “ground-glass” appearance, but depending on the extent of the dysplastic fibrous tissue and on the amount of bone and degree of calcification, they may range from radiolucent to sclerotic [2, 7, 17]. Radiographically, the differential diagnosis may include Paget’s disease, solitary bone cysts, aneurysmal bone cysts, enchondroma, adamantinoma, low-grade intramedullary osteosarcoma, osteofibrous dysplasia, and giant cell tumor [5, 19]. In the presented case, as in all bone tumor cases, a biopsy was required to make a definitive diagnosis.

The etiology of the tumor remains unclear, but fibrous dysplasia appears to be linked to a single nucleotide mutation in the $G_s\alpha$ gene on the long arm of chromosome 20 (at 20q13.2–3) that results in a disturbance of the tissue differentiation process [5, 6, 17]. This mutation occurs in somatic cells sometime after fertilization and therefore is not inherited [17, 19]. Chromosome 12 has also been implicated in the pathogenesis of fibrous dysplasia, but so far, no chromosomal changes have been demonstrated consistently [4, 17].

Proper treatment of monostotic fibrous dysplasia in long bones of the upper extremity depends on the presentation. Asymptomatic cases may simply be monitored. When symptomatic, particularly in cases of pathologic fracture, treatment may involve closed fixation, curettage alone, curettage and bone grafting, or internal fixation [12, 20, 22]. More extensive cases may actually require vascularized bone grafting [13, 21].

In this particular case, the tumor involved the entire metaphysis of the distal radius. It was our estimation that curettage of this mass would leave the patient with a very large void of the radius metaphysis. Despite the void, the radius was stable. We did have concerns about using either autogeneic or allogeneic bone grafts due to the potential for recurrence of tumor and/or fracture in a large area [17, 20]. In one study, 52% of autologous bone grafts and 12% of

allografts were unsuccessful at treating fibrous dysplasia [20]. It was for this reason that we chose to utilize Norian as the graft material. Other authors have speculated that bone cement can give immediate mechanical strength and possibly kills residual tumor cells [22]. Norian is an injectable apatite cement that has been demonstrated to be safe and effective in other uses in the distal radius, specifically distal radial fractures [3]. We also chose to supplement the Norian with two Kirschner wires. The reasoning for this is that the wires would serve as a sort of “re-bar” within the cement, providing added stability to torsional and bending forces. With intact cortex on the radial, ulnar, and volar aspects of the radius, these Kirschner wires were likely not needed. In retrospect, the mistake made with the Kirschner wires is that they should have been cut shorter and tamped into the cortex so that both ends of the wires were sub-cortical. This was not done, and it was then necessary to remove the pins due to irritation. Despite this misstep on our part, the removal of the pins has no detrimental effect on the patient’s recovery and radial stability.

In conclusion, this bone tumor of the distal radius was unusual for two reasons: it was a monostotic fibrous dysplasia of the distal radial metaphysis, and it occurred in a middle-aged male. While not a common tumor of the hand, fibrous dysplasia should be included in the differential diagnosis for masses of the adult distal radius.

Fig. 5 Radiograph of the wrist demonstrating appearance of the radius following pin removal

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