CASE REPORT

Osteofibrous Dysplasia Disguising as Chronic Osteomyelitis: A Diagnostic Dilemma

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ABSTRACT

Osteofibrous dysplasia is a rare benign fibro-osseous lesion with a strong predilection for involvement of tibia in the early childhood. Its biologic behaviour ranges from non-progressive to recurrent and more aggressive lesions however, most lesions become quiescent or even regress spontaneously as skeletal maturation is completed. We report a rare case of a 20 years old male patient admitted with a history of swelling in the left hand with complaints of pain and local signs of inflammation. Radiography and magnetic resonance imaging were done and indicated the involvement of the head and distal shaft of the second metacarpal with bone marrow edema, destruction of cortex and surrounding soft tissue edema. All the clinical features and radiological imaging were in favour of chronic osteomyelitis. Patient underwent surgery and biopsy indicated presence of osteofibrous dysplasia highlighting the importance of considering this differential diagnosis in such presentations.

INTRODUCTION

Osteofibrous dysplasia is a rare benign fibro-osseous lesion with a strong predilection for involvement of tibia in the early childhood. Histopathologically it is a benign fibro-osseous lesion composed of fibrous tissue with woven bone formation. It is most commonly found in the tibia and fibula, although a case with ulnar involvement has been reported[1].

Its biologic behaviour ranges from non-progressive to recurrent and more aggressive lesions however, most lesions become quiescent or even regress spontaneously as skeletal maturation is completed. Radiographic findings include eccentric, fairly well-marginated osteolytic lesion with a sclerotic border in the anterior cortex of the tibial diaphysis and can show longitudinal spread to metaphysis, cortical expansion, intramedullary extension and anterior bowing deformity.

Chronic osteomyelitis of the hand is uncommon and usually due to Staphylococcus aureus and affects a single bone. Involvement of the metacarpals occurs in only 3% of cases[2].

Differential diagnosis of osteolytic lesions of bone ranges from tumors to non-tumorous pathologies like infective lesions or metabolic problems[9]. Imaging can help narrow down the differentials however, biopsy is essential in most cases to identify the pathology, permitting a specific therapeutic approach. We report a rare case of osteofibrous dysplasia of metacarpal whose clinical and radiological findings were in favour of osteomyelitis which was turned out as osteofibrous dysplasia on histopathology.

CASE REPORT

A 20-year-old male presented with a swelling over the dorsum of left hand for four months with an increase in the size of swelling with development of pain over the past 15 days. He complained of feeling feverish on and off over the past 15 days but no definitive documentation of fever was present. On physical examination swelling and tenderness was present proximal to the left second metacarpophalangeal joint with evidence of local inflammation.

On X-ray examination, an osteolytic lesion with narrow zone of transition (Figure 1) was noted in the metaphysis of the left second metacarpal proximal to its head with suspicious break in cortex. Smooth periosteal reaction was noted. No associated expansion of bone or soft tissue component was seen. Other metacarpals were normal.

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On MRI examination, the lesion corresponded to altered marrow signal intensity appearing hypointense compared to surrounding marrow on T1 and T2 (Figure 2) and hyperintense on PD-FS image (Figure 3). Bone marrow edema was noted extending to the head and shaft of the metacarpal. Thinning of cortex with focal breach was noted near anteromedial aspect of distal metaphysis of the second metacarpal with surrounding circumferential periosteal reaction and soft tissue edema. The distal articular surface of the metacarpal appeared normal and intact. The lesion was categorized as osteomyelitis according to the clinical presentation and radiological findings.

CT guided biopsy of the lesion was performed which on histopathological examination (Figure 4) revealed irregular fragments of woven bone and lamellar bone lined by osteoblasts with fibrous tissue component consisting of spindle cells and infilterating collagen, with myxoid stroma. These findings were suggestive of presence of osteofibrous dysplasia lesion in the second metacarpal with an associated pathological fracture.

DISCUSSION

Osteolytic lesions of bone can have a wide range of differentials from infective to metabolic to neoplastic lesions. Chronic osteomyelitis is one of the differentials of an osteolytic lesion involving the metaphysis of metacarpal. More than two-thirds of patients with chronic osteomyelitis of the hand have involvement of a single bone[10], and young males are predominantly affected. It usually presents as an inflammatory swelling of the hand with functional incapacitation. A fever is uncommon in chronic cases and occurs chiefly in patients with an acute onset or a concomitant infection at another site[3]. Plain radiograph should be obtained routinely; they show bony destruction and a periosteal reaction, a combination highly suggestive of chronic osteomyelitis[9, 14]. Computed tomography is useful for detecting early evidence of bone destruction and periosteal reaction; however MRI is more sensitive and provides an accurate evaluation of the lesions, the bone marrow changes and changes in the surrounding tissues. The abnormal bone generates low signal on T1 images and high signal on T2 images[3].

Osteofibrous dysplasia, also referred to as ossifying fibroma of long bones, is a benign fibro-osseous lesion composed of fibrous tissue with woven bone formation. The lesions mostly occur in the tibia and fibula, although a case with ulnar involvement has been reported[6]. The features that differentiate it from fibrous dysplasia include the characteristic zonal phenomenon and osteoblastic rimming of bony trabeculae which are absent in fibrous dysplasia.

Earlier Osteofibrous dysplasia was referred to as ossifying fibroma of long bones. Although ossifying fibroma and osteofibrous dysplasia of the jaw have few common histological characteristics like the presence of cytokeratin-positive cells in the ossifying fibroma in the long bones and development of psammomatous calcification in the ossifying fibroma of the jaw are exclusive histopathological features that distinguish the two as separate diseases[4].

Osteofibrous dysplasia usually appears as a loculated osteolytic lesion circumscribed by a sclerotic border involving the anterior diaphyseal cortex of the tibia or fibula with adjacent cortical expansion. Intramedullary encroachment and anterior bowing deformity are commonly associated as the lesion progresses[1]. The signal intensity of osteofibrous dysplasia was intermediate on T1-weighted images and intermediate to high on T2-weighted images[1]. Superimposed haemorrhagic or cystic, myxoid change and even cartilaginous differentiation can modify the signal intensity and contribute to heterogeneous signal intensity on T2-weighted images[5]. In the study by Jung JY et al, complete medullary involvement was observed in 33% of osteofibrous dysplasia cases and soft tissue edema was generally absent or mild without pathologic fracture[1].

CONCLUSION

Although imaging can help narrow down the differential diagnosis, often it is not enough and histopathology is needed to come to an accurate diagnosis. We report a rare case of osteofibrous dysplasia affecting the metacarpal. This rare case suggests that Osteofibrous dysplasia can exhibit diverse imaging features with lesions with complete intramedullary involvement or peri-lesional marrow edema, pathological fractures and involvement of uncommon locations like metacarpals.

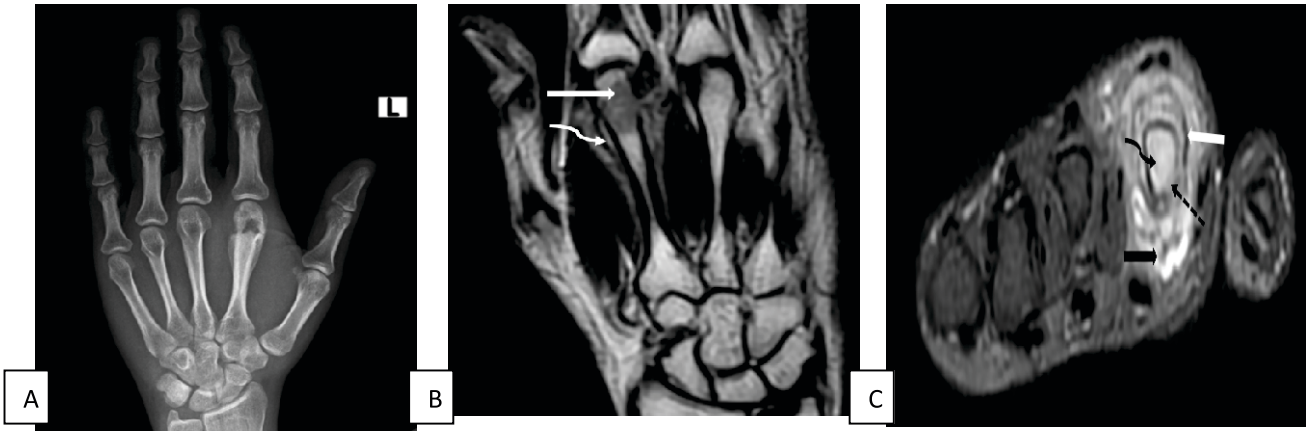

Figure 1: A - Radiograph of left hand (PA view) showing an osteolytic lesion with a narrow zone of transition, surrounding circumferential smooth periosteal reaction in distal metaphysis of second metacarpal. B - Coronal T2 W MRI image showing altered marrow signal intensity (white solid arrow) appearing hypointense compared to surrounding marrow in distal metaphysis of the second metacarpal with surrounding circumferential periosteal reaction (curved white arrow). C - Axial PD-FS MRI image showing marrow edema (curved black arrow) with discontinuity of cortex (dashed black arrow) near anteromedial aspect of distal metaphysis of the second metacarpal with surrounding circumferential periosteal reaction (white solid arrow) and soft tissue edema (solid black arrow).

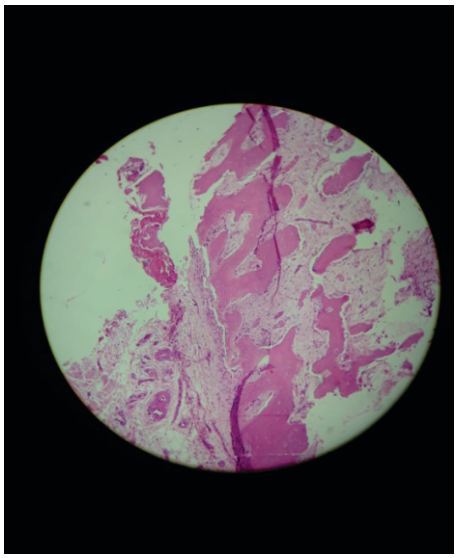

Figure 2: Irregular fragments of woven bone and lamellar bone lined by osteoblasts. Fibrous tissue component was also noted consisting of spindle cells and infilterating collagen, with myxoid stroma. Mitosis were rare.

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