PATHOLOGIC FRACTURE THROUGH A SOLITARY ENCHONDROMA OF THE RADIAL DIAPHYSIS- A RARE CASE REPORT

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ABSTRACT

BACKGROUND

Solitary enchondromas are rare but are well recognised benign bone tumours. They are extremely rare in the radius. We report a case of solitary enchondroma in the distal radial diaphysis which was incidentally detected when patient presented with recurrent pathological fractures. Typical enchondroma is an incidental finding, while atypical variant is associated with pain. We are presenting a rare case of typical incidentally detected solitary enchondroma of radius (not associated with pain).

KEYWORDS

Enchondroma, Distal Radial Diaphysis, Pathological Fracture.

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BACKGROUND

It is very uncommon to find solitary enchondroma in distal radius and a very few such cases have been reported in literature. We report such a case presenting with deformity after fall and initial radiological investigations suggesting other differential diagnoses. The final diagnosis was made based on histopathology.

Case Report

A 19-year-old male presented with deformity of the right forearm after he sustained fall 4 weeks back for which immobilisation of the limb was done. Pain started after lifting weights which was aggravated by physical work. There was no history of night pain. There were four similar episodes 4-years back, which gradually resulted in the deformity of the right forearm; this was treated by immobilisation of the limb. In between the episodes, the patient was relatively symptom-free.

On local examination, there was an ill-defined swelling measuring 4 x 3 cm, with tenderness on palpation. There were no other swellings or discharging sinuses or engorged veins. Systemic examination was nil remarkable. A diagnosis of pathological fracture of right radius was made clinically.

Routine blood examination, calcium, phosphorus, alkaline phosphatase and PTH (Parathormone) assay were within normal limits.

(Figure-1) - Triple Phase bone scan with Tc–99m MDP showed hot spot in right radius with irregularity of bone – suggestive of a fracture.

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(Figure - 2A) Radiographs of the right forearm (AP and lateral views) showed a long segment, mildly expansile lytic lesion involving the mid and distal radial diaphysis, partially extending into metaphysis, with areas of cortical thinning and endosteal scalloping. Multiple internal areas of internal matrix calcifications noted (in the form of flecks and arcs). There is a dorsally displaced pathological fracture across the lesion, in the distal radial shaft; with mild sclerosis along the fracture margins.

Figure 2B shows post-op radiographs of the right forearm orthopaedic internal fixation devices across the pathological fracture.

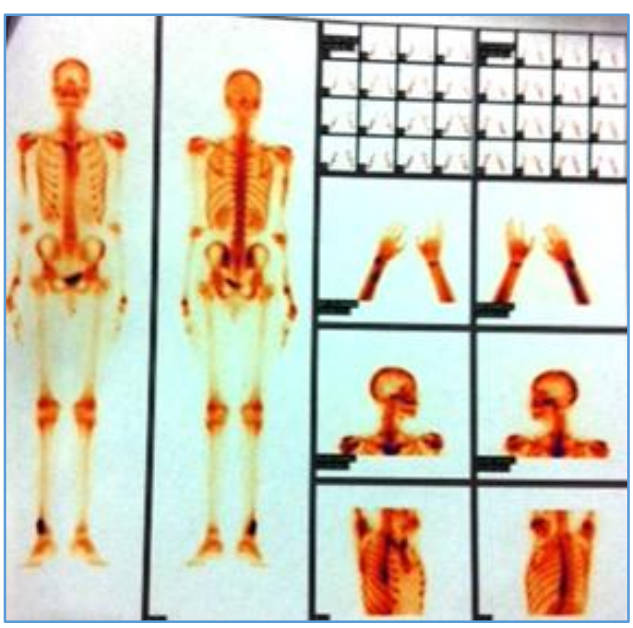

Figure 1

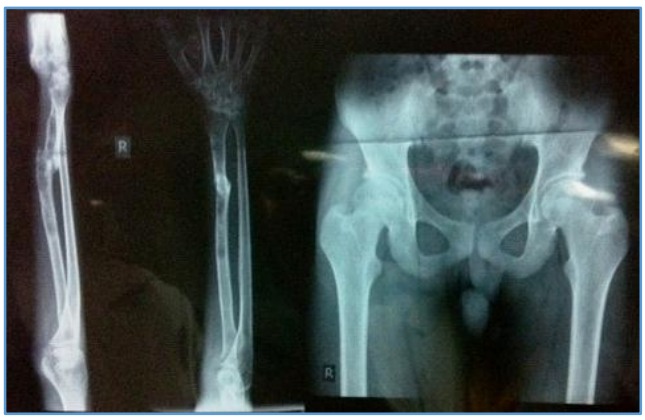

Figure 2A

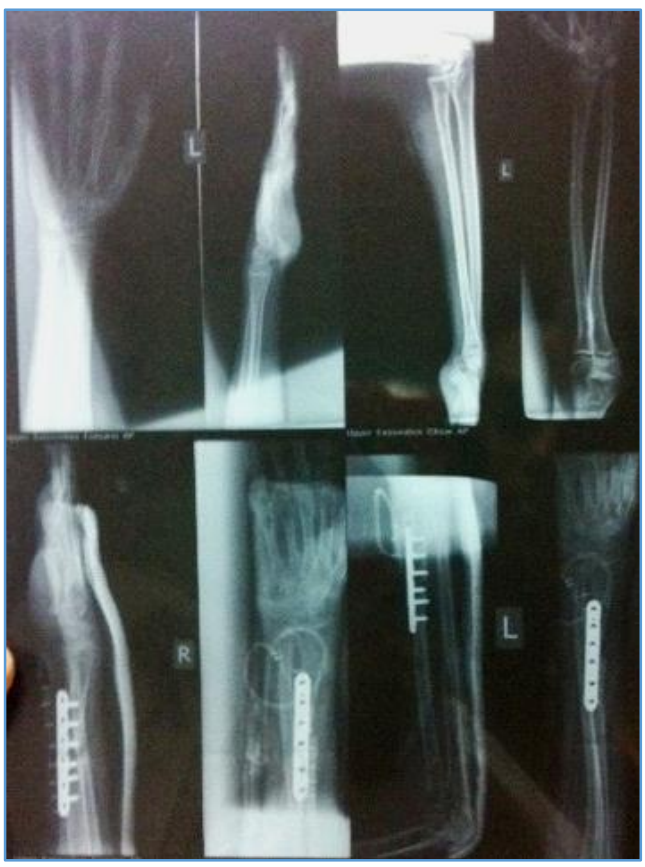

Figure 2B

Computed Tomography (CT) of right forearm confirmed the lytic lesion seen on radiograph, with internal chondroid matrix calcifications and a pathological fracture of the lower 1/3rd of radial shaft.

On MRI (Magnetic Resonance Imaging)

The right radial lesion appeared T1 hypointense, T2 heterointense and STIR hyperintense, with mild marrow oedema along the fracture margins, and no periosteal reaction or soft tissue component- findings suggestive of a benign skeletal lesion in the distal radial shaft.

A pre-operative differential diagnosis of fibrous dysplasia was considered.

Intraoperatively, a malunited fracture site with a 6 cm long and 2 cm wide lytic lesion was seen. An intra-operative biopsy was done which was sent to our laboratory for histopathological examination. Postoperatively, the patient was immobilised with cast for 8 weeks.

We received multiple bony bits and grey white soft tissue bits altogether measuring 1.2 cc. The bony bits were subjected to short decalcification and soft tissue bits were routinely processed. The bits were submitted for histopathological examination.

Histopathology showed mature bony lamellae with lobules of cartilaginous areas and mature chondrocytes without pleomorphism and with fibrocollagenous stroma (Figure - 3).

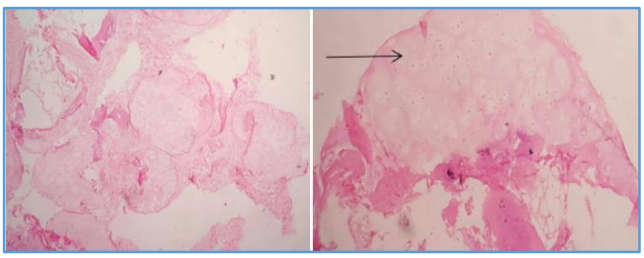

Figure 3

DISCUSSION

The term enchondroma is usually reserved for intramedullary chondromas as opposed to eccentrically located periosteal chondromas or soft-tissue chondromas.[1] Enchondromas are second most benign cartilaginous tumour, after osteochondromas.[2]

Approximately, 35% of all enchondromas arise in the hand[3] followed by femur, humerus and tibia. Solitary enchondroma of radius is rare.

Enchondromas are relatively common benign medullary cartilaginous neoplasms, usually found in children and young adults which can lead to pathological fractures or may rarely undergo malignant transformation. Mostly these are incidental findings with benign imaging features.

Location

Almost all enchondromas are located centrally within the medullary cavity of tubular bones. This is the most common benign tumour of small bones of hands and feet (50%); other locations such as long tubular bones (proximal humerus, distal femur, proximal tibia) are rare. References for exact incidence of solitary enchondroma in radius are not found, although 1% incidence is seen radius and ulna involvement.[4] Only 1 out of 136 cases in Huvos series,[5] occurred in the radius (metaphyseal region).

Typical enchondroma shows benign biologic behaviour, whereas atypical one is an ominous condition and may be associated with a malignant change (low-grade chondrosarcoma).[6] If missed, chondrosarcomas usually recur within 2 years and need followup for 10 years.

Enchondromas are frequently diagnosed in childhood or early adulthood with a peak incidence of 10-30 years. They account for ~5% (range 3-10%) of all bone tumours and ~17.5% (range 12-24%) of benign bone tumours.

Pain can be caused by bone weakening, and there can be pathological fractures after strenuous activities. Any patient experiencing sudden pain after physical stress is likely to have an enchondroma rather than a chondrosarcoma.[1] In chondrosarcoma, pain is usually insidious and progressive. Soft tissue mass and night pain is present in >20% cases.[5] It is

important to note that if an enchondroma is painful in the absence of a fracture, it should be considered malignant.

Enchondromas arise from rests of growth plate[7] cartilage/chondrocytes that subsequently proliferate and slowly enlarge and are composed of mature hyaline cartilage. Hence, they are seen in any bone originating from cartilage. Lesions are usually <3 cm, translucent, nodular, and are grossly grey-blue.

Associations

Two Syndromes are associated with Multiple Enchondromas-

- Ollier disease.
- Maffucci syndrome.

Rarely an enchondroma may extend through the cortex and demonstrate an exophytic growth pattern. This is known as an enchondroma protuberans. It occurs in either the typical phalangeal location or the proximal humerus.[8] They may either be seen sporadically or as part of Ollier disease.

Radiological features such as cortical destruction, periosteal reaction, and soft tissue mass strongly suggest the diagnosis of chondrosarcoma. The differentiation between enchondroma and grade 1 chondrosarcoma can be quite difficult on histology. The cytological features often overlap too.

Treatment includes excision of the tumour with a wide margin to prevent recurrence.

CONCLUSION

Lytic bone lesions on radiology have many overlapping features. Solitary enchondroma involving distal radial diaphysis is rare, but it should be included in the differential diagnosis; and histopathology helps in arriving at a definitive diagnosis, as notable from our case. The combination of clinical, radiological, and histopathological findings are essential to provide correct management for the patient.

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