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Case Report

Fortuitous enchondroma of hand in a traumatic fracture: a case report of Ollier's disease

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ABSTRACT

Enchondromas are benign cartilage forming tumors most commonly involving the marrow cavity of the long bones. Ollier disease is one of the subtypes of Enchondromatosis associated with lesions in the center of the bone. Most commonly presenting in 2nd and 3rd decade of life. Radiological evidence is adequate for the diagnosis. Exact etiology is unknown with increasing genetic predilection. Regular follow up with conservative management remains the mainstay of treatment.

Keywords: Enchondroma, India, Ollier's disease, Rare diseases

INTRODUCTION

Enchondromas are benign cartilage forming tumors most commonly involving the marrow cavity of the long bones, most commonly hand followed by Humerus and Femur.[1] The common subtype of Enchondromatosis is Ollier disease (Spranger Type 1) described by multiple Enchondromas with asymmetrical distribution.[2]

The estimated prevalence of Ollier disease is 1 in 1,00,0003. Clinical presentation includes pain, swelling, and restriction of movements, deformity and pathological fractures. Diagnosis is made by morphological and radiological evidence.[4] Treatment modality includes regular follow up with conservative management and may rarely require surgical excision or simple curettage.[2]

CASE REPORT

A 10-year-old pre-adolescent girl was brought to the emergency room with history of traumatic injury to the little finger of the right hand while doing her routine work at home 24 hours prior.

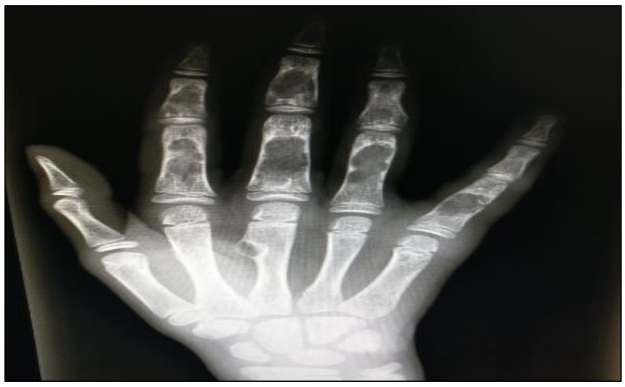

Figure 1: Frontal radiograph of right hand demonstrating multiple expanding lytic lesions involving metacarpals and phalange with thinning of the cortex. A pathological fracture is noted in the fifth metacarpal bone.

On clinical examination, vitals were stable, and no significant systemic findings noted. Local examination of the right hand was suggestive of swelling over the little finger, restriction of range of movements and no signs of inflammation. In view of suspected fracture of the little finger, frontal radiograph of right hand was obtained (Figure 1) with prior consent from the parent.

Child was further evaluated with ultrasonography of right hand, suggestive of bulky thenar muscles compared to left hand with no hemangiomas. A diagnosis of isolated Enchondroma - Ollier's disease was made based on the investigation findings of radiology and ultrasonography. Parents were advised for further evaluation but were lost to follow up.

DISCUSSION

Enchondromatosis being a benign cartilage tumor thought to be a precursor of secondary central Chondrosarcoma. The risk of malignant transformation to Chondrosarcoma is around 35% and there is no specific marker for early diagnosis. The exact etiology of Enchondromas is unknown and poorly understood.

Table 1: Spranger subtype classification of enchondroma.

Types	Conditions	Clinical features
Spranger type I	Ollier disease	Multiple Enchondroma of tubular and flat bones, predominantly unilateral.
Spranger type II	Maffucci syndrome	Same as Ollier disease, with hemangiomas.
Spranger type III	Metachondromatosis	Multiple Enchondromas and exostoses.
Spranger type IV	Spondyloenchondrodysplasia	Multiple Enchondroma with severe platyspondyly.
Spranger type V	Enchondromatosis with irregular spinal lesions	Multiple Enchondroma with dysplasia of vertebral bodies.
Spranger type VI	Cheirospondylo Enchondromatosis	Multiple Enchondroma, severe hand and foot involvement, mild platyspondyly, erosion of iliac crests.

Panasuriya TC et al in his genome analysis of Ollier disease presented with absence of DNA Copy Number Alterations (CNA) and Loss of Heterozygosity (LOH) in majority of subjects.[5] This suggests that point mutations or neutral structures changes such as inversions, insertions or deletions below the platform involving a single gene or axon have important role in pathogenesis, suggesting use of next generation sequencing approach.[5]

Spranger et al classified enchondromatosis based on radiographic appearance, anatomical site and mode of inheritance into 6 sub types (Table 1).[6]

Ollier disease must be differentiated from multiple hereditary exostosis and Mafucci syndrome. Osteochondroma are located on bony surface as compared to Ollier disease associated with lesions in the center of the bone. Mafucci syndrome are associated with hemangiomas. Treatment options include regular follow up with conservative management.[7] Early age of onset, malignant transformation, gross asymmetrical distribution and repeated surgeries are poor prognostic factors in Enchondromas.[8]

CONCLUSION

Our index case presenting with traumatic fracture was investigated with radiograph and diagnosed to have Enchondroma (Ollier disease). Differentiated by Mafucci syndrome by ruling out hemangiomas. Diagnosis of Ollier disease requires clinical and radiological evidence and does not require any special tests. Next generation sequencing approach is needed for evaluation of pathogenesis.

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