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OLLIER DISEASE – A CASE REPORT

Orthopaedics

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ABSTRACT

Ollier's disease is unusual presentation of enchondroma includes multiple bones. We report the case of 7-year-old male child who is diagnosed with Ollier's disease involving multiple bones of right hand. His diagnosis was delayed due to this rare entity. This report examines the diagnostic difficulties faced by clinician in arriving to a correct diagnosis. The patient enchondroma was quiescent during the follow-up of two years.

KEYWORDS

Ollier's disease, hand, enchondroma

INTRODUCTION

Enchondroma is a common primary bone tumor. Which develop from the growth plate. The multiple enchondroma when present is known as Ollier's disease. It is rare disease usually present in first decade of life. It is characterized by asymmetrical involvement with massive variability in site, size, shape, location, progression and management that they behave nearly independent of each other and need to be monitored individually. Diagnosis is based on clinical examination and radiological investigation only. There is no definitive management of the Ollier's disease. The treatment is largely surgical and involves the management of complications only.

CASE REPORT

A 7-year-old boy presented with the multiple swelling in right hand which were painless bony, appear embedded to underlying bone with no associated skin changes and no history of trauma.

On x rays' examination, multiple lytic expansile, homogenous lesions with elongated to oval shape with endosteal scalloping well with no cortical breech involving multiple metacarpals, proximal phalynges, middle phalynges. The widening of metaphysis, bowing of the diaphysis is also seen.

The patient was subjected to scan for any associated haemagioma, visceral malignancies.

The patient was followed up for four consecutive years. There was no complication observed including pathological fracture or malignant transformation to chondrosarcoma.

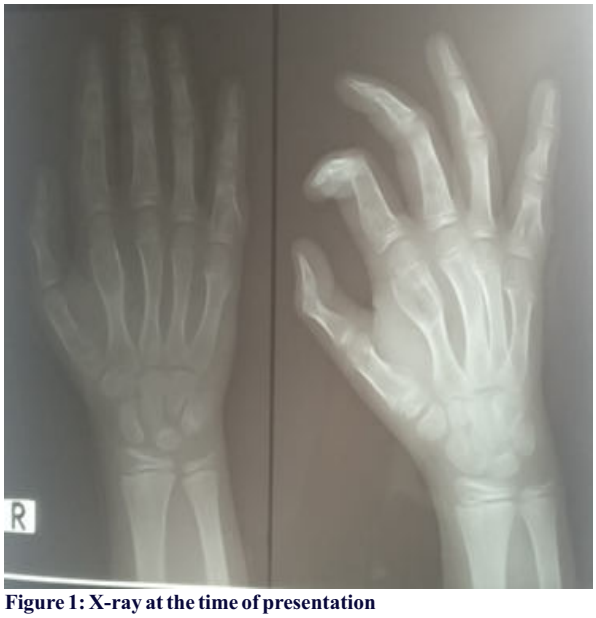

Figure 1: X-ray at the time of presentation

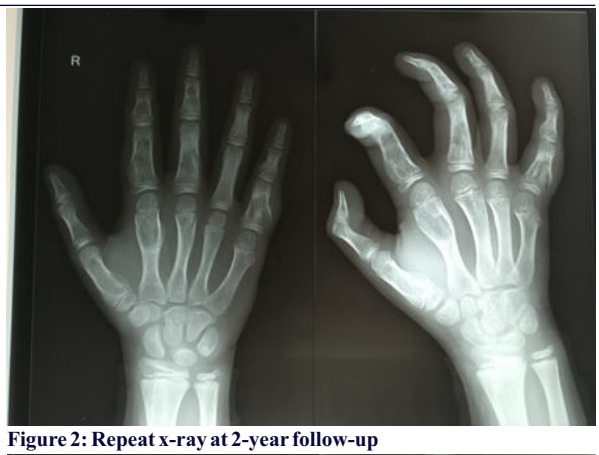

Figure 2: Repeat x-ray at 2-year follow-up

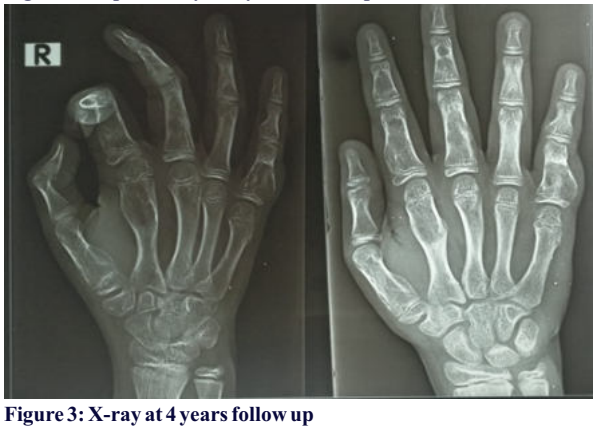

Figure 3: X-ray at 4 years follow up

Differential Diagnosis

Maffucci syndrome
Hereditary Multiple Exostosis
Chondrosarcoma
Metachondromatosis
spondyloenchondroplasia

DISCUSSION

Clinical manifestations in Ollier's disease often appear in the first decade of life and usually start with the appearance of palpable bony masses on a finger or a toe, an asymmetric shortening of an extremity with limping, osseous deformities associated or not with pathologic fractures [1-3]. Physical examination is nonspecific for diagnosis except for painless swelling. Roentgenograms typically show multiple, radiolucent, homogenous lesions with an oval or elongated shape and well defined slightly thickened bony margin [1-3]. On histopathological examination, picture is almost same as chondrosarcoma grade I, which is its most common complication, so cannot be relied upon. There is no medical treatment for Ollier's disease. scintigraphy and visceral scan

should be done to differentiate it from the more sever Maffucci's syndrome which has hemangiomas associated with multiple enchondromas Surgery is indicated in case of complications (pathological fractures, growth defect, malignant transformation). The prognosis of Ollier's disease is difficult to assess [1]. Prompt treatment is encouraged as soon as there is pain or pathologic fracture to prevent damage to the bone [4]. Patient with numerous quiescent lesions may have a better prognosis than patients with localized cartilaginous changes or limb shortening, Enchondromas in Ollier's disease present a risk of malignant transformation of enchondromas into chondrosarcomas, which usually occurs in young adults, and thus at an earlier age than observed in patients with chondrosarcoma[5]. The reported incidence of malignant transformation is variable and estimated to occur in 5–50% of the cases [3,6-8]. It is higher in Maffucci's syndrome, the prognosis of which is more severe than that of Ollier's disease [1,2]. Association of Ollier's disease with other tumors has been reported [1,9-11].

CONCLUSION

Ollier's disease is different and rare entity. As all the enchondromas are totally different from each other and does not follow any order. So, a follow-up is required to check for any complication like pathological fractures or malignant transformation, The treatment is mainly for the complications and there is no definite treatment for Ollier's disease.

Consent

As per international standard informed and written participant consent has been collected and preserved by author.

Ethical Approval

No ethical Issues were involved in the manuscript.

Competing interest

Authors have declared that no competing interests exist.

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