ORIGINAL RESEARCH PAPER

Radiology

OLLIER DISEASE (MULTIPLE ENCHONDROMATOSIS) – A CASE REPORT

KEY WORDS:

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ABSTRACT

Ollier disease is called multiple enchondromatosis with unilateral predominance and rare congenital disease. These are benign growths of cartilage that develop within the bones. Few of them can develop into chondrosarcoma. These lesions are mostly located in the metaphyseal region.1 Herein, we report a case of Ollier Disease in an 8-year-old male patient.

INTRODUCTION

Ollier disease is characterized by multiple, unilateral limb endogenous chondromas associated with limb deformities. [2] Clinical symptoms include local pain, palpable bone mass, bone swelling, and bone deformity. The risk of transformation into chondrosarcoma is high. Diagnosis of disease mainly depends on clinical presentation, imaging (location of lesions), and histological examination. [2]

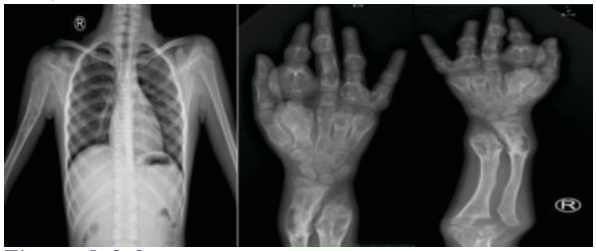
CASE REPORT

An 8-year-old male patient presented to our hospital with complaints of obvious mass on his right index, middle, ring, and little fingers. The patient initially presented 2 years back in a private hospital at the age of 6 years. The patient did not take any treatment at that time. The patient then noted the increase in the size of the lesions. There is disproportionate growth of upper limbs, the normal length of left side but there is a shortening of the right upper limb, especially forearm. The patient complains of pain, limited function, and movement. Moreover, the left hand, right shoulder, right knee, and right foot also get minimally swollen.

After clinical evaluation, the orthopedic surgeon prescribed a skeletal survey for this patient, which shows multiple osteopathies of the hands, right shoulder, right leg, and right foot. The clinical diagnosis of Ollier disease was made. However, as a definitive diagnosis was made clinically, the patient did not undergo a computed tomography (CT) scan.

Imaging findings

In this case, the skeletal survey was done. There is the presence of multiple expansile, lytic lesions with sharp margins, and a narrow zone of transition, with endosteal scalloping, noted in multiple bones of the body. These types of lesions are present in the right humerus, scapula, radius, ulna and few phalanges and metacarpal bones in both hands, left humerus, right fibula, few phalanges and metatarsals of both feet. Larger lesions show cartilage calcifications. (Figure 1,2,3) [3]



Figures 1, 2, 3

There are vertical streaks of radiolucency are noted in the meta-diaphysis of long bones like the lower ends of both femurs and tibia, the upper end of the left tibia, and the upper end of the left radius. (Figure 4,5) [3]

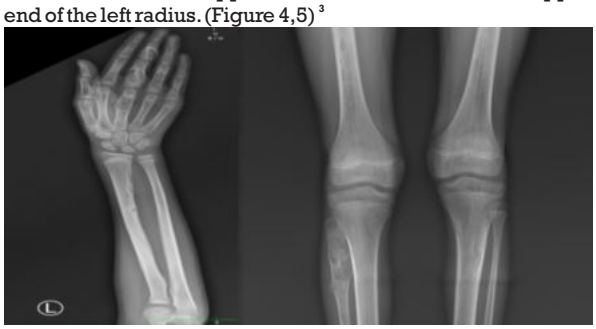


Figure 4,5

Lesions mostly appear to involve the shaft and metaphysis of a short or long bone. Lesions are oval in shape. In short bone, lesions show scalloping due to limited space, the cortex becomes thinner. Due to the large growth space, no characteristic scalloping is seen in the long shaft, and the cortex does not become thinner.

Ultimately, X-rays can diagnose correctly typical cases. A CT scan can reveal calcification foci, by which we can predict malignant alteration. MRI can visualize the cartilage components in the lesions for improved accuracy.

DISCUSSION

Multiple enchondromatosis is a rare heterogeneous skeletal disease. There are six subtypes are noted: 1. Ollier disease, 2. Maffucci syndrome, 3. metaphyseal chondromatosis, 4. metatarsal chondromatosis, 5. spinal interstitial chondrodysplasia and 6. spinal interstitial chondromatosis. [5] The prevalence of the disease is 1 in 100,000. [4] Diagnosis of Ollier disease is based on clinical manifestations and radiological evaluation. Histological analysis is mainly for malignant transformation. The first onset of Ollier disease is usually within the first 10 years of life. There appear as single or multiple bone lesions. With increasing age, limb deformities appear. So, deformities can lead to affects the growth and development of children. Ollier disease is characterized by at least 3 enchondromas. [6] No marker is there for the prediction of malignant transformation.

Treatment of patients with Ollier disease without any significant deformities go for long-term follow-up. Treatment of complications remains predominantly surgical. Bone

lengthening surgery can correct deformities. [6]

CONCLUSION

Ollier disease is characterized by multiple benign enchondromas with a limb deformity. They occur in proximity to growth plate cartilage. X-rays are sufficient for the diagnosis of Ollier disease. Due to the high risk of malignant transformation, careful follow-up is beneficial for the patient.

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