Case Report

Case report - A case of Ollier's disease of the hand

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Enchondroma is a benign growth of cartilage arising in the bone metaphysis as a solitary or multiple primary lesions. This form of multiple enchondromatosis with unilateral predominance is termed Ollier's disease. We have recently treated a case of Ollier's disease with the chief complaint of pain and swelling in the right hand. The patient was a 22-year-old female. Radiographic examination showed honeycombed clear spaces in the metaphyses of the middle and proximal phalanges of the 3rd and 4th digits as well as of the 3rd, 4th metacarpals, and thinning of the cortex of these bones, with pathological fracture of 4th proximal phalynx. With regard to the prognosis of Ollier's disease, malignant transformation into chondrosarcoma or osteosarcoma has been reported of the chondroma. Since Ollier's disease is self-limiting disease so we fixed the pathological fracture with K wire and extended curettage with bone grafting.

KEY WORDS: Bone grafting and pathological fractures, Enchondroma, Olliers disease

INTRODUCTION

Enchondroma is a benign growth of cartilage arising in the bone metaphysis as a solitary lesion or multiple primary lesions. This form of multiple enchondromatosis with unilateral predominance is termed as Ollier's disease.[1] We have recently treated a case of Ollier's disease with the chief complaint of pain in the right hand due to pathological fracture of 4th proximal phalynx.

CASE REPORT

The patient was a 22-year-old female that came to the outpatient department with the chief complaint of pain and swelling in the 3rd and 4th metacarpal and incidental finding of fracture of fourth proximal phalynx. The history and family history were not remarkable, nor were there any abnormalities on routine examination. On first examination, a painful mass was noted with protrusion from the radial aspect of the middle phalanx of the right ring finger and another mass was protruding from the volar side of the proximal phalanx to the metacarpophalangeal joint (MPJ) of the fourth digit of right hand. There was no discrepancy of the upper and lower limb length or detectable venous mal formations of the skin. The range of motion was 0–90° for the proximal interphalangeal joint (PIPJ) and 0–60° for MPJ of the 3rd finger, and 0–90° for the PIPJ and 0–15° for MPJ of the 4th finger. Radiographic examination showed honeycombed clear spaces and popcorn calcifications in the metaphyses of the middle and proximal phalanges of the right ring and middle fingers as well as of the third, fourth metacarpals, and thinning of the cortices of these bones, but with evidence of pathological fracture of 4 proximal phalynx [Figure 3].

Magnetic resonance imaging of the right hand was done which revealed multifocal variable sized lesions in the third and 4 digits phalynx with pathological fractures of 4th proximal phalynx likely enchondroma [Figure 2]. The patient was diagnosed as a case of multiple enchondroma in 4th proximal phalynx and head of 3rd metatarsal and was treated surgically. To prevent circulatory

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disturbance of the affected digits, the surgery was performed as a single stage operations which comprised of curettage of the tumor and fixation of the fracture with 2 cross K wires. The tumors were removed, and the curettage was done and the defect was filled with the bone graft and the fracture of 4th proximal phalynx was fixed by two cross k wires. A plaster splint was applied for a period of 4 weeks after surgery. Biopsy revealed that the lesion was composed of small masses of hyaline cartilage with densely populated, small proliferating chondrocytes with slight atypia. The microscopic features were compatible with diagnosis of enchondroma [Figure 3].

Similar large lytic, eccentrically located lesion with central calcification present in the metaphyseal region of 3rd metacarpal distally.

DISCUSSION

Ollier in 1899, termed a condition that is characterized by unilateral occurrence of multiple enchondromas as Ollier's disease.[1,2] Based on a careful scrutiny of similar cases reported since then, it is customary to categorize as Ollier's disease all those enchondromatous conditions that manifest a unilateral predominance of lesions, even if there is bilateral involvement.[2]

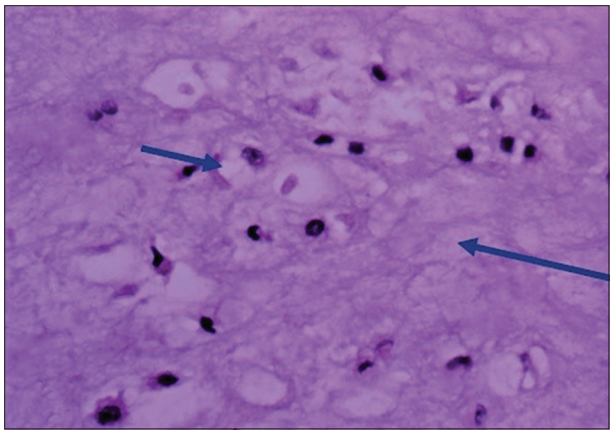

Figure 1: Histopathology slide showing small masses of hyaline cartilage with densely populated chondrocytes with slight atypia suggestive of enchondroma

According to Hunter and Wiles the following are pathognomonic of Ollier's disease:

1. The onset is in early childhood
2. Radiograms show changes limited to the ends of long bones. There is striping of rarefied areas.[3-5] The epiphysis are involved only secondarily [Figure 3]. Speckling appears in the affected metaphysis and epiphyses as growth proceeds
3. A portion of tissue corresponding to a pale area in the radiogram is found, when excised and examined histologically, to consist of cartilage.[6,7]

In our patient, the enchondromatous lesions were localized to the right hand, with radiographic evidence, but with no involvement of any other extremities or the pelvis, and met all of Hunter's criteria for diagnosis of Ollier's disease. To the best of our knowledge, a total of 16 cases of Ollier's disease affecting the hand bones predominantly unilaterally, including the present case, have been reported during the past decade. It consisted of 7 males, 3 females, and 5 other patients (sex unrecorded) and 10 left and 11 right hands, with the left ring and little fingers or right middle and ring fingers being involved more frequently.[8-11]

The surgical indication is predominantly pathological fracture angulation and/or rotation in both upper and lower limbs which is similar to our study. Curettage of lesions followed by bone grafting is the most commonly used therapeutic procedure and has been generally reported to have a good outcome [Figures 4 and 5].[11,12] In the present case, surgical resection of tumors was performed with bone grafting and fixation of pathological fracture with k wire. The K wire was removed after 6 weeks [Figures 6 and 7]. The postoperative course was uneventful, and bone formation was achieved without fracture recurrence and with no progressive deformities. This suggests the necessity of curetting enchondromatous lesions before they infiltrate the epiphyseal line.

With regard to the prognosis of Ollier's disease not only in the hand but also in both upper and lower limbs, malignant transformation into chondrosarcoma or osteosarcoma has been reported.[13] A condition, known as Maffucci's syndrome in which Ollier's disease is accompanied by venous malformation

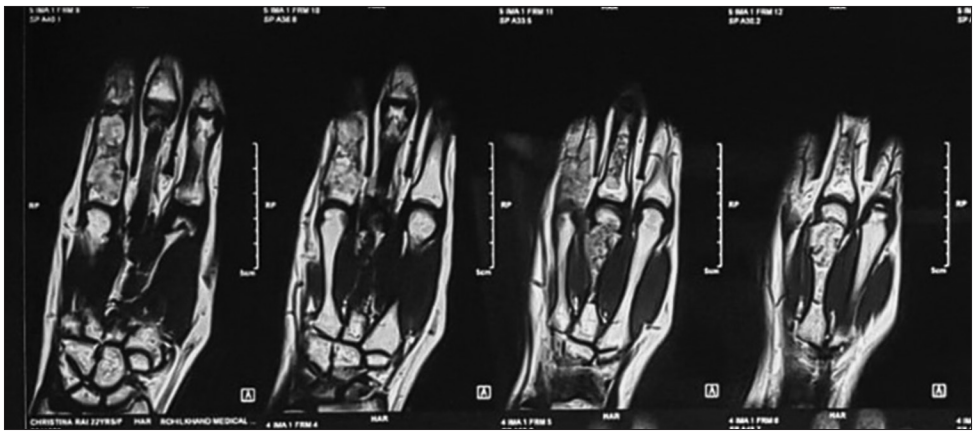

Figure 2: MRI of the right hand showing multifocal variable sized lesions in 3rd and 4th digits and phalanges and in 3rd metacarpal with pathological fracture of proximal phalynx of 4th digit

Figure 3: (Pre-op X-ray image) multiple lytic and expansile, lesions with rings and arcs central calcification and "popcorn" like appearance in the metaphyseal region of the proximal and middle phalynx of the 3rd and 4th digit along with a pathological fracture of 4th proximal phalynx.

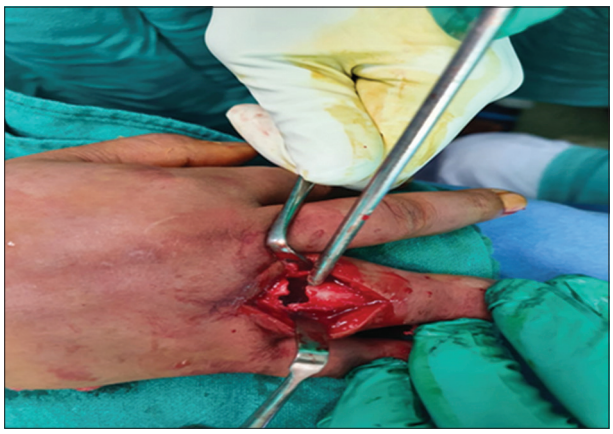

Figure 4: Intraoperative big cavity left behind after performing extensive curettage

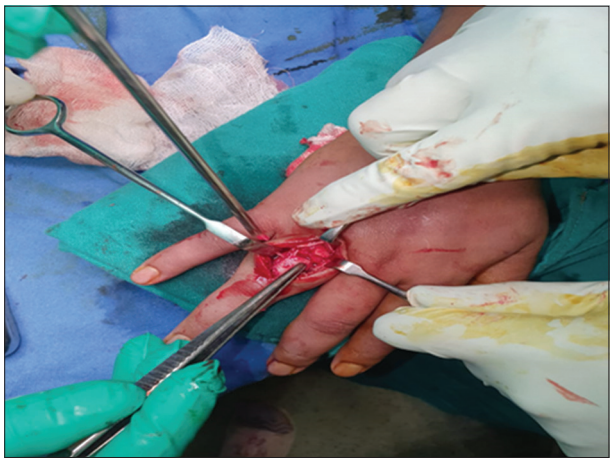

Figure 5: Cavity filled with bone graft

and retarded growth, is thought to have greater potential for malignant transformation.[7] Prediction of whether and when actively proliferating chondrocytes will become malignant is quite difficult on the basis of radiographic and laboratory examinations.[8] It requires a long-term follow-up study. Since Ollier's disease is self-limited, in that it usually stops spontaneously as the patient grows and cartilag- inous lesions in occasional cases may regress or even disappear, any, cartilaginous lesions that are still active or painful after

Figure 6: X-ray image 4 week follow-up

Figure 7: X-ray image of right hand on 6 weeks follow-up showing healed fracture

termination of the growth period should be examined thoroughly for suspicion of undergoing malignant transformation.[9] Overall incidence of the development of chondrosarcoma is 40%, but may, due to age-dependency, increase when considered as a lifelong risk.[14,15] Patients with enchondromas located in long bones or axial skeleton, especially the pelvis, have a seriously increased risk of developing chondrosarcoma, and are identified as the population that needs regular screening on early detection of malignant transformation.

CONCLUSION

The study emphasis on the possible treatment of pathological fracture in Ollier's Disease where one can go for debridemnet of the disease followed by bone grafting and k-wire fixation as one of the most commonly followed procedure.

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