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Tumour excision and hip morphology correction in multiple hereditary exostoses by ganz safe surgical dislocation-A case report

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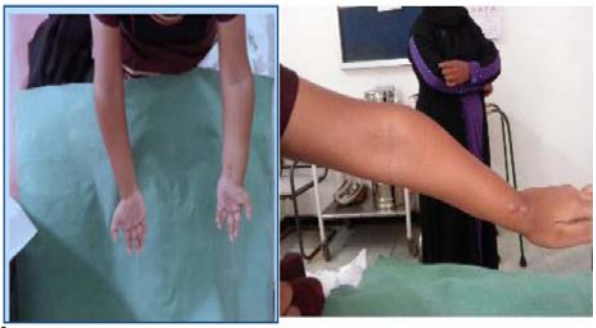
ABSTRACT:
Multiple hereditary exostoses also known as diaphyseal aclasias, most common benign bone disorder in children, is an autosomal dominant genetic disorder with complete penetrance, with an incidence of 1:50,000 individuals comm only occurring along the metaphysic of the long bones1. Proximal femur is affected in approximately 90% of individuals associated with growth disturbance, acetabular dysplasia and possible subluxation10. We report a case of multiple hereditary exostoses with proximal femur lesion treated by ganz surgical dislocation and have analysed the effectiveness of this procedure in restoring hip morphology.

Keywords:
Autosomal dominance, multiple hereditary exostoses, safe surgical dislocation.

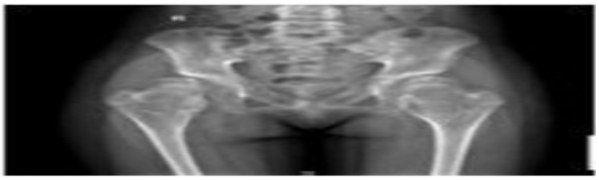
INTRODUCTION:
Multiple hereditary exostoses is characterised by the outward growth of cartilage capped bone tumours1. They usually develop in the first decade of life and start ceasing once the child attains skeletal maturity. Although exostoses are benign lesions, they are often associated with characteristic skeletal deformities and may cause clinical symptoms1-4. The most common deformities include short stature, limb length discrepancies, valgus deformities of the knee and ankle, asymmetry of the pectoral and pelvic girdles, bowing of the radius with ulnar deviation of the wrist and subluxation of the radiocarpal joints1-6. Linkage analysis has implicated mutations in the EXT gene family, resulting in an error in the regulation of normal chondrocyte proliferation and maturation that leads to abnormal bone growth6.

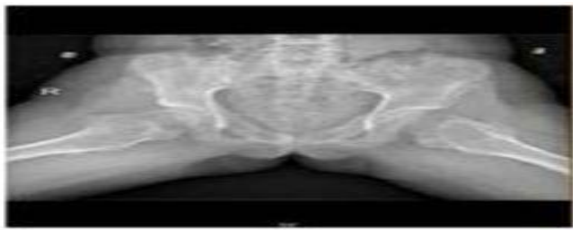
CASE REPORT:
14 year old girl presented with multiple swellings of her bilateral elbows, wrists and knee joints, for the past 9 years. Swellings were insidious in onset, gradually progressed to the present size. Swelling was not associated with any pain, had difficulty in walking for a long distance, difficulty in squatting. There was no history of any associated constitutional symptoms. There was a positive family history with similar complaints in her father and elder brother. Birth history was noted normal. On examination, patient had a normal gait with a palpable bony hard swellings on bilateral shoulders, medial aspect of both knees and bilateral distal end of ulna. Bilateral valgus deformity of knee and varus deformity of elbow were noted (as in FIGURE1), there was no restriction of any movements, no neurological deficits and examination of spine was normal. Xrays of bilateral knee and wrist were taken and findings of abnormal pedunculated bony projections of distal femur and sessile lesions in proximal tibia were noted.Xray of bilateral forearm showed gross shortening of the ulna with a lesion over distal radius and bilateral radial bowing were noted (as in FIGURE 2).XRAY pelvis showed multiple bony lesions around the femoral neck (metaphyseal trumpeting) with mild acetabular dysplasia and subluxation of both the hip joints(as in FIGURE 3) XRAY right shoulder also showed pedunculated bony lesions in metaphyseal region and xray dorso lumbar spine was found normal(as in FIGURE4). Clinicoradiologically, patient was diagnosed as a case of MULTIPLE HEREDITARY EXOSTOSES. We opted to perform exicision osteoplasty of the proximal femur and ganz safe surgical dislocation of the hip (as in FIGURE 5) in order to improve the range of movements and relieve pain around the hip joint. A sample was sent for histopathological analysis to confirm the diagnosis and rule out malignancy, it was confirmed as a case of exostoses (as in FIGURE 6).



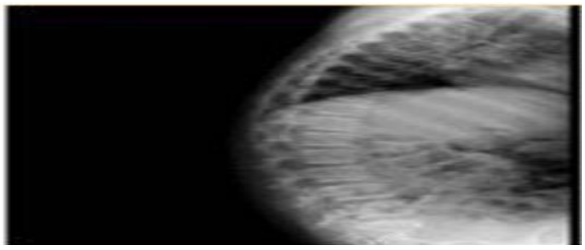
Clinical images(figure 1)

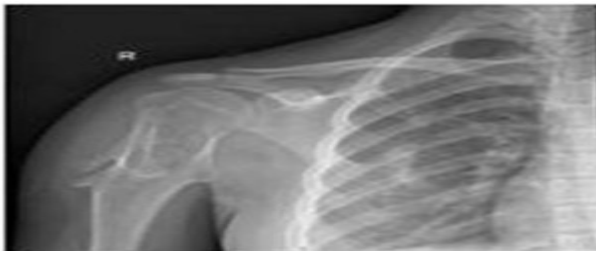
Xray images of bilateral knee and forearm(figure 3)



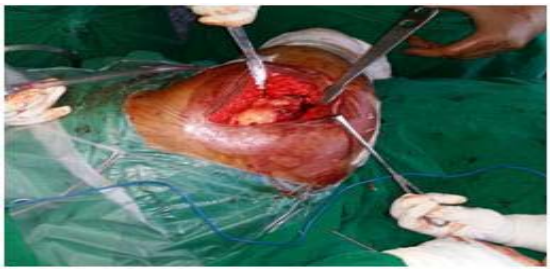


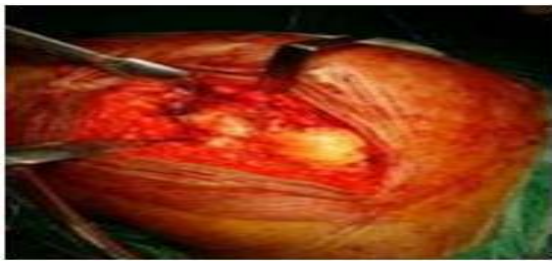
Xray images of pelvis and frog lateral views(figure3).

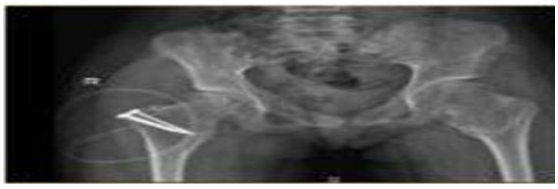




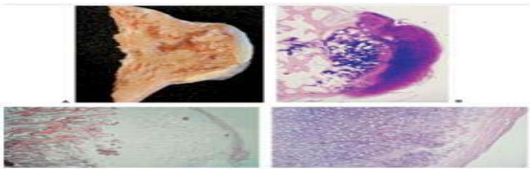
xray dorso lumbar spine lateral and xray right shoulder(figure 4)







Ganz safesurgical dislocation and excision osteoplasty (figure 5)



Postop xray and histopathological images(figure 6)

DISCUSSION:

Multiple osteochondromas is characterised by development of two or more cartilage capped bony outgrowth of the long bones1. .Diagnosis of multiple osteochondromas can be made when radiologically atleast two or more osteochondromas of the juxta-epiphyseal region of long bones are observed7. The suspicion of secondary chondrosarcoma is indicated by growth of the tumour after puberty, presence of pain or thickness over 1cm of cartilaginous cap in adults8. It mostly affects the long bones and can lead to growth disturbances especially disproportionate short stature due to local effect on growth plates and the systemic influence of the gene disorder on growth mechanism1-5. EXT gene disorder affects the heparin synthase synthesis which could have a systemic effect or influence on growth plate closure6.

Patients with MHE most frequently report pain and cosmetic concerns. Pain may be the result of variety of problems associated with the exostoses; such as bursa formation or repeated soft tissue trauma over a prominent osteochon droma8,9. Restricted range of movements is a common report of individuals with severe involvement of the proximal femur or forearm. There will be disproportionate shortening of the ulna, because the distal ulnar physis responsible for greater longitudinal growth relative to that of the distal radius, consequently radial bowing was thought to be caused by a tethering effect of relative ulnar shortening8,9. Limb length discrepancy is more commonly seen in MHE. A clinically notable inequality >2cm has been reported in 10 to 50%of the affected individuals. Shortening can occur in the femur as twice as tibia10. Femoral anteversion and valgus deformity have been associated with exostoses located in proximity of the lesser trochanter10.

Osteochondromas occurs as solitary lesion in the proximal femur and these typically are not associated with acetabular dysplasia or coxa valga; however many problems can arise from these lesions such as labral tears, nerve compressions, hip dislocation, external snapping hip and malignant transformation. A variety of surgical techniques have been reported in the literature for these solitary lesions without dysplasia. The main concerns for surgical resection of femoral neck and peritrochanteric osteochondromas are exposure and femoral head vascularity11.We performed a ganz safe surgical dislocation of hip followed by excision osteoplasty of the lesion in the femur neck of right hip. Postoperatively, patient was immobilised for a period of 3 weeks and range of movements notably improved after surgery. But there was neither a observable change in the gait pattern nor the hip morphology.

CONCLUSION:

There is still not a conclusive uniform surgical treatment for patients with hip joint osteochondromas with or without dysplasia, but from the various literature reviews, it has been noted that an additional femoral varus derotational osteotomy combined with ganz surgical dislocation and excision osteoplasty of the lesion , could substantially restore the normal biomechanics of the hip joint and hip morphology.

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