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TOTAL HIP ARTHROPLASTY IN MULTIPLE EPIPHYSEAL DYSPLASIA- A CASE REPORT

Orthopedics

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ABSTRACT

Multiple epiphyseal dysplasia is a skeletal dysplasia characterised by abnormal maturation of the epiphyses, affecting the hips, knees and ankles. It is usually inherited as autosomal dominant or recessive type. This case report is a 32-year old female who came to the outpatient department of our tertiary care center with complaints of bilateral hip pain (R>L) and limp over right lower limb for 6 months. On examination, the patient was found to have dysmorphic dwarfism. Local examination of bilateral hips showed joint line tenderness and restriction in abduction and internal rotation. X-ray pelvis with both hips showed bilateral dysplastic shallow acetabuli and bilateral deformed femoral head with incongruent joint space reduction superior subluxation of right femoral head. The patient underwent uncemented total hip arthroplasty of right hip for the same. Intraoperatively, the patient was found to have shallow acetabulum which was augmented with a bone graft prior acetabular component implantation. The femur shaft was also found to be abnormally narrow intraoperatively which admitted only a pediatric CDH stem. Hence, we would like to emphasize the need for preoperative planning, CT imaging and templating in order to plan prosthesis sizing preoperatively in patients with dysplastic hips.

KEYWORDS

Multiple epiphyseal dysplasia, Total hip arthroplasty, Dysmorphic dwarfism

INTRODUCTION

Multiple epiphyseal dysplasia (MED) is a group of disorders affecting cartilage and bone development, primarily affecting the epiphysis of long bones.

Joint pain, particularly of the hips or knees, is a common and often develops during childhood but can also present in adolescents and young adults.

It may be inherited as an autosomal dominant Fairbank's disease (Mutation of COMP-MED genes) or an autosomal recessive Ribbing's disease (Mutation of MATN3-MED genes). Patients with multiple epiphyseal dysplasia usually present with predominant knee or hip necessitating surgical intervention. Beacuses of the anatomical deformities including a dysplastic acetabulum, a varus CCD, a short neck and a high-riding trochante, performing a THA can be challenging in these patients.

CASE REPORT

A 32-year old female patient presented to us with bilateral hip pain (Right>Left) for 3 years. Pain was insidious in onset, dull aching, gradually, progressive, aggravated in the last one year assocaited with a limp on the right side and difficulty in performing activities of daily living which involves squatting or sitting cross-legged.

Patient is known case of hypothyroidism on treatment. There was no family history or similar complaints in the family. On examination, the patient was found to have short stature with a moderately brachycephalic face with small spade like hands and feet. Bilateral Genu valgum and Hallux valgus deformities were also observed.

On local examination of bilateral hips, there was anterior and posterior joint line tenderness with restricted and painful movements. Axis deviation was present on the right side.

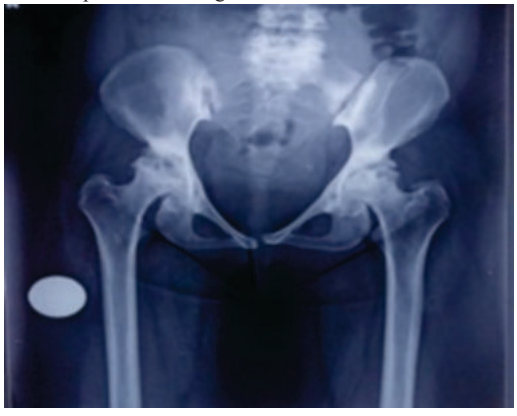

Fig 1: Xray Pelvis With Both Hips

Anteroposterior radiograph of pelvis with both hips shows bilateral dysplastic shallow acetabulum and bilateral deformed femoral head with incongruent joint space reduction with superior migration of femoral head. Sacral dysmorphism seen bilaterally.

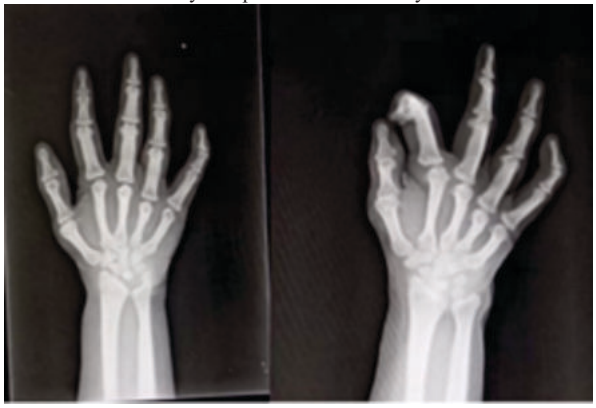

Fig 2: Xray Of Right Hand Anteroposterior And Lateral View: Small metacarpals and phalanges with flat distorted epiphysis and brachydactyly

MANAGEMENT

The patient underwent uncemented total hip arthroplasty for her right hip. Intraoperatively, acetabulum was found to be shallow and dysplastic so it was augmented with a bone graft over the superolateral aspect to accommodate the acetabular component.

The head of right femur was markedly deformed and sclerotic. The femoral canal was narrow and sclerotic and could only accommodate a 6size paediatric CORAIL stem.

Post operatively, the patient was advised in bed mobilization for one month to allow for the consolidation of the bone graft and started on weight bearing.

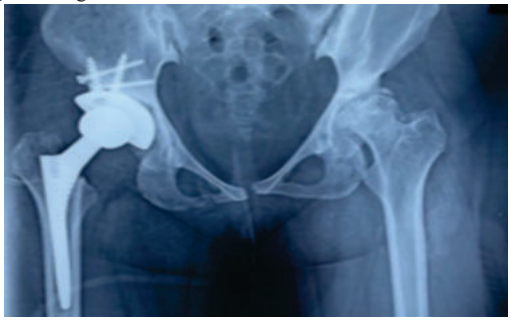

Fig 3: Immediate Postop X-ray Uncemented Prosthesis Insitu Right Hip.

Note the cortical screws on the superolateral aspect to fix the bone graft to accommodate the acetabular component.

CONCLUSION

Total hip arthroplasty is a viable treatment option in multiple epiphyseal dysplasia patients, with excellent mid-term clinical and radiographical outcomes but it needs proper preoperative planning like CT imaging and templating for ideal placement of prosthesis in dysplastic hips.

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