

Case Report Orthopaedics

CONCOMITANT FRACTURE NECK FEMUR AND SLIPPED CAPITAL FEMORAL EPIPHYSIS IN A CHILD WITH OSTEOPETROSIS - A RARE CASE REPORT

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Abstract:

The osteopetrosis is uncommon and debilitating disorder and clinical profile range from fatality to normal life span with morbidity. The dense and sclerosed bones are fractured especially at areas of higher stress concentration like proximal femur. The neck femur fracture is uncommon association reported in few studies. Concomitant, ipsilateral slipped capital femoral epiphysis in the presence of fracture neck femur in such patients is a rare feature and we report a 12 year old male child with this association. The child, however, did not live longer to evaluate and manage him to a longer follow up.

Key-words: Osteopetrosis, Slipped capital femoral epiphysis, Fracture, Neck femur, Hip, Child, Congenital, Diagnosis

Introduction:

Osteopetrosis is a disorder with abnormal bone resorption and remodeling owing to osteoclast dysfunction. This heterogeneous group of skeletal dysplasia is debilitating with variable inheritance pattern.¹ There are three reported variants of osteopetrosis, autosomal recessive type, autosomal dominant type and malignant type. Malignant form has fatal course and autosomal dominant ones have better life span of the three.²

The fractures in the presence of osteopetrosis are difficult to treat and are reported chiefly of proximal femur. Very few studies about these cases are present and thus no validated treatment guidelines exist. Neck femur fractures have previously been reported with osteopetrosis but are related to potential complications including suboptimal healing.³ The thickened, coarse bone with varying degree of medullary cavity obliteration and associated co-morbidities play obstacles in the operative treatment. There have been, however, attempts of internal fixations of fractures in diverse cases with variable outcomes.^{4,5} The associated constellation of symptoms related to hematological, neurological and renal disorders compound the treatment with complexity and guarded outcome.

Case Report:

A 12 year old male child was brought to us with complaints of painful weight bearing and limp for last two weeks as noticed by mother. It was initially considered as transient phenomenon and neglected till the problem persisted. There was history of frequent hospital visit of the child for various complaints. There was failure to thrive and the child was of short stature and underweight as compared to his peers. He was first

and only child of parents at that time. The child appeared like a five year old kid. There was vision problem for which spectacles were advised in the past along with hearing problem under evaluation and treatment. There had, however, been no history of previous fractures following trauma or spontaneous. The child was anemic and had frequent bouts of upper respiratory infections as per medical records. The radiograph of the pelvis and both hips showed normal hip joint in the left side but abnormal one in the right side. There was apparent inferior subluxation of capital epiphysis on right side along with cortical irregularity along neck region (two arrows in Fig.1 a). Slight oblique view delineated and confirmed the features (Fig.1 b). The picture was suggestive of slipped capital femoral epiphysis along with fracture neck femur. The findings were confirmed on magnetic resonance imaging (MRI) sections (Fig.1 c,d). The femorae were denser with faint medullary cavity. A provisional diagnosis of osteopetrosis was made on basis of clinic-radiological assessment. The child was advised rest while the probable diagnosis was explained to parents. A treatment protocol including conservative and operative options was explained in detail with prognosis. The parents chose conservative option and wished to review after two weeks. The radiograph done after two week showed decreased neck shaft angle suggestive of instability and displacement (Fig.2 a) The operative intervention to stabilize the neck and slipping with in situ pinning or other procedure was declined by parents. The other radiographs of spine showed increased sclerosis of end plates and that of skull showed dense and sclerosed calvaria with sclerosis

of base of skull (Fig.2 b and c). On evaluation of clinic-radiological features a diagnosis of osteopetrosis was made with probable recessive variant. The further investigations and genetic evaluation was declined by parents. The child was advised one and half hip spica but parents refused and promised supervised bed rest. The child was lost to follow up for more than four months. On home visit of the case, it was known that the child succumbed to serious chest infection two weeks later of our visit.

Fig.1 –
The radiograph of pelvis and both hip showing right capital epiphysis subluxation and cortical irregularity in neck region (a,b) along with dense bone with faint medullary outline. MRI sections confirming the diagnosis (c,d)



Fig.2 –
The worsening of neck shaft angle (a) and radiograph of spine (b) with end plate sclerosis and that of skull (c) showing sclerosed, dense calvarium.



Discussion:

There are reports describing association of slipped capital femoral epiphysis (SCFE) and osteopetrosis as concomitant disorders. One case report of a 9 year old male child was managed by in-situ screw fixation.⁶ The same article described one previous report similar to theirs.⁷ Some report the combination in the setting of endocrine disorders like hypothyroidism.⁸ Thus SCFE and fracture neck femur is independently reported with osteopetrosis but the reports of any case with all the three as concomitant features is unlikely. The association of SCFE and fracture neck femur is thus rare entity. One unique observation, however was made by Joseph et al highlighting a case of delayed slipping following trans-cervical fracture in a normal child.⁹ The presence of similar pattern might be present in our case as a probable mechanism. Decreased femoral anteversion and neck shaft angle are linked to increase slipping in one study by Kordellee et al.¹⁰ The very presence of decrease neck shaft angle may also increase chances of neck fracture due to higher strain and shear force concentration. Our report of the case of unilateral concomitant fracture neck femur and SCFE might be first report of its kind. The fracture of the neck was not clear in routine radiographs and was confirmed on MRI scan of the hip.

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