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Bilateral Both Bone Leg Fracture in a Case of Hereditary Multiple Exostoses with Bilateral Proximal and Distal Tibiofibular Synostosis

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Abstract
The Hereditary Multiple Exostoses (HME) is a neoplastic disorder affecting multiple skeletal sites in the form of bony protuberances of varying sizes and shapes . The clinical features are site specific and mostly relate to the effect of swelling on the adjacent tissues . Associated abnormalities like bowing deformities of bones, shortening and mechanical axes deviations may lead to increased risk of stresses over the bones . Associated tibiofibular synostosis if present can further complicate the aforementioned problems .We present a rare and previously unreported case of bilateral both bone leg fracture following low energy shear and torsion mechanism in the setting of bilateral tibiofibular synostosis . The condition was satisfactorily managed conservatively owing to minimal displaced fracture patterns and patient compliance to the treatment
Key Words - *hereditary multiple exostoses , tibial fracture , synostosis*

INTRODUCTION

Tibiofibular synostosis is reported as an associated anomaly with HME, mostly proximal one . The presented case has bilateral tibiofibular synostoses at proximal and distal level. This arrangement has a theoretical basis for the leg working as monobloc thus failing at both bones as a result of torsion low energy trauma. This unique injury pattern however can be managed successfully with conservative measures.

CASE REPOT

We present a case of bilateral both bone of leg fracture in a case of Hereditary multiple exostosis . A young 19 years old boy presented in the casuality with the history of trivial accidental trauma to both of his legs .The injury was a low energy tripping while walking down the street . The patient could not stand on his own and was taken to the hospital by his friends. On clinical examination localized pain and crepitus was found over both legs in junction of middle and distal third region . Radiographic evaluation revealed a diagnosis of both bone bilateral spiral and minimally displaced fractures . Apart from it, there were multiple bony protuberances over multiple sites in the body including bilateral lower and upper leg, upper thigh, bilateral wrist and upper forearm , chest and shoulder region . The provisional diagnosis of Hereditary Multiple Exostoses was made which was further confirmed after positive family history of similar affectation to his brother, father and his uncle. There was associated synostosis of both bones of his leg at proximal and distal level. This association is not a common occurrence.

RESULT

The patient was treated conservatively with bilateral above knee plaster of paris splintage. Splintage was modified as to avoid direct bony contact with protuberances to check local complication of skin irritation and breakdown. The fracture united under the course of treatment in thirteen weeks uneventfully. Gradual weight

bearing to tolerance was started and soon the patient was ambulatory and joined his pre injury functionality . He was asked to visit for regular checkup for bony lesions and future intervention as per the complaints. He was also asked for regular follow up in view of clinic-radiological screening for the any malignant transformations .

The natural course of the disease and treatment options were discussed with the patient and the guardian. The patient had functional deficit in the form of limited pronation and supination in bilateral forearm but he preferred the current status to continue and regular checkup in the future .

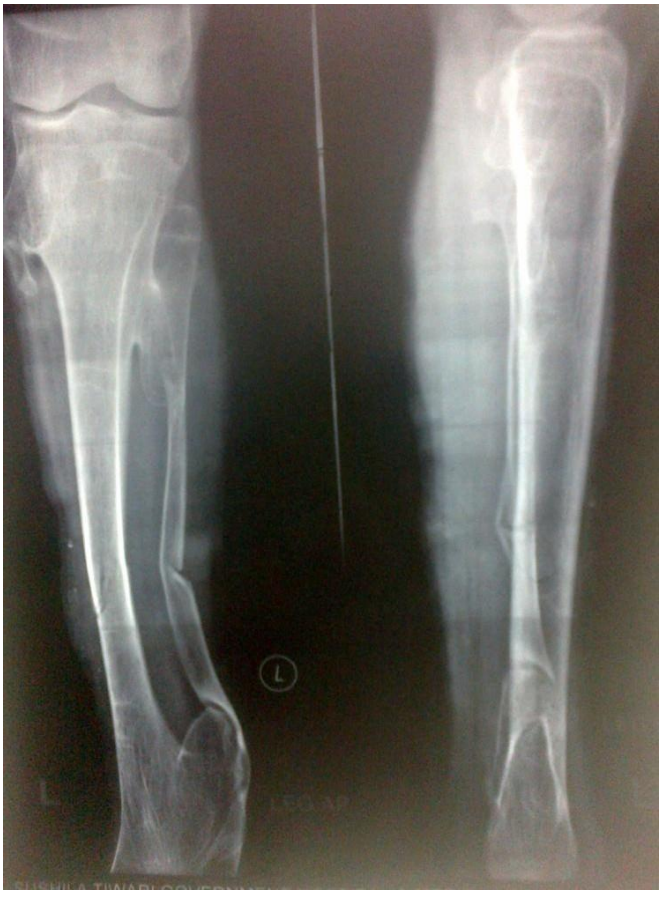

Fig 1 – Left leg radiograph showing fractures and exostoses with synostoses

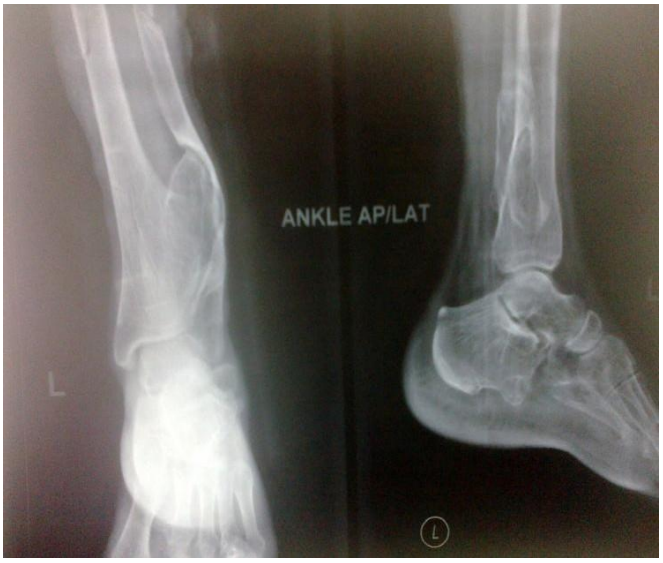

Fig 2 – Left ankle with leg radiograph showing fracture extent and synostosis

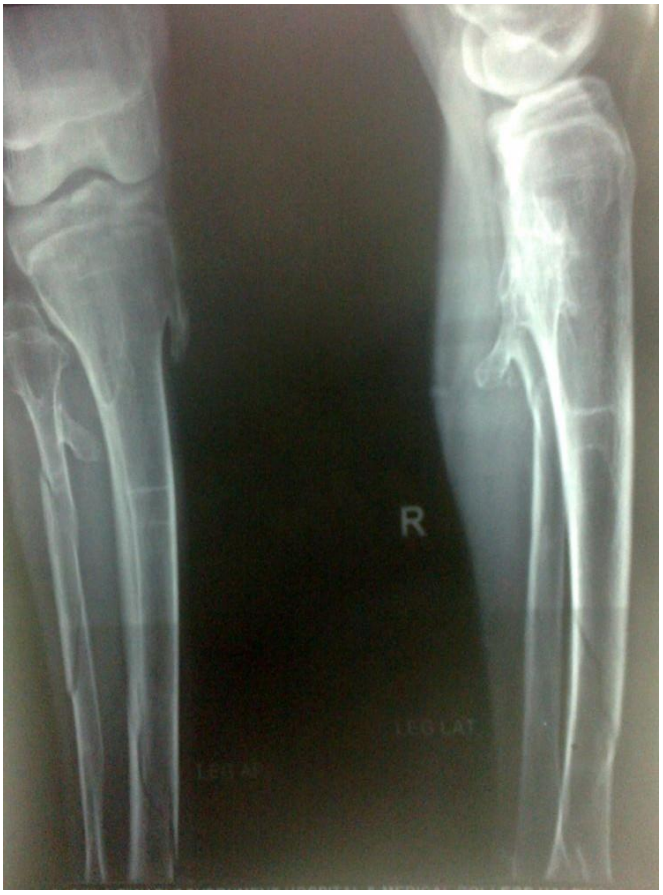

Fig 3 – Right leg radiograph showing fractures and synostoses

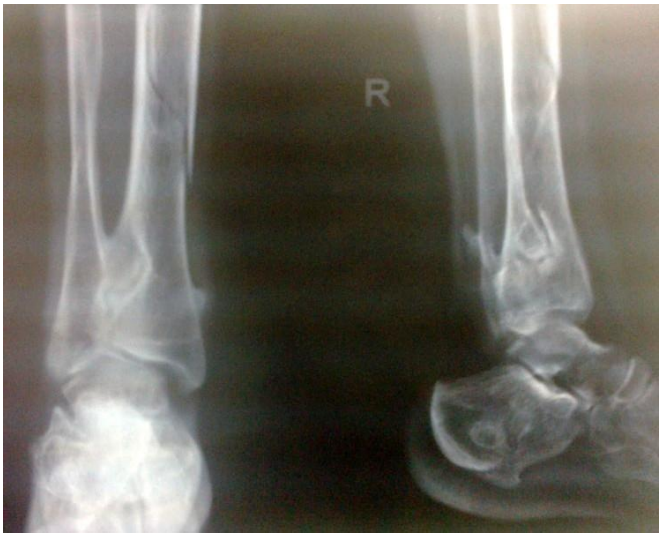

Fig 4 – Right ankle with leg radiograph showing extent of fractures and synostosis

DISCUSSION

Hereditary Multiple Exostoses (HME) is a benign bone lesions presenting as bony swellings at multiple sites with variable site specific clinical features . The disorder has autosomal dominant inheritance and males bear the brunt of it . [1] Other synonyms of the condition are Mutiple Osteocartilagenous exostosis and Diaphyseal Aclasis . There has been an association reported with linkage to EXT 1,2 and 3 gene locus . Key clinical features of the disease are knobby protuberances over multiple skeletal sites, short stature, characteristic pattern of forearm deformities , genu valgum , coxa valga and tibiofibular synostosis . The multiple exostoses are reported in various locations in the body but mahority of them are found around the knee .[2] Radiographic pattern may reveal multiple exostoses of varying sizes over the bone with associated bony deformities . Most of the exostoses are sessile in nature.[3]

Abnormal shape and bowing deformities in the long bones may itself be a cause of increased stresses over the bone to decrease its yield strength . Proximity of osteochondroma at either end of the bones of leg can result in tibiofibular synostosis . [4] Usually the synostosis is proximal with distal synostosis reported rarely. [5] Concomitant tibiofibular synostosis compounds the problem by making the leg a monobloc that is constantly battling cyclic loads . Coxa valga , a common associated abnormality may have its implications on altered mechanical axis of the limb especially when coupled with abnormal leg axis . In cases of HME, where there is bilateral affectations of the lower limb , serious altered biomechanics of lower extremities may increase the risk of fractures . Our reported case had no singnificant effect on activities of daily living as of now, despite radiological deterioration of mechanical axes and joint mal-positioning . However the patient was explained the natural history of the disease and possible future degenerative changes in all the affected joints .There are reports of cases of multiple fractures in cases with HME .[6] A low energy simultaneous , bilateral both bone leg fracture in the settings of tibiofibular synostosis at both levels is not reported as per the literature search by the authors.

CONCLUSION

This case report highlights rare co -existing skeletal abnormalities in the setting of HME, complicated by multiple fractures . Radiological evaluation and anticipation of the similar anomalies should be kept in mind when dealing with HME . Furthermore , a due explanation about future degenerative joint diseases and importance of regular follow up is warranted for better and holistic patient care .

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