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Growth modulation in Genu Valgum secondary to Multiple Hereditary Exostosis

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ABSTRACT

Objective: Hereditary Multiple Exostoses (HME) is a rare disease with a prevalence of 1:50,000, which can manifest in various ways. This cartilaginous tumor can appear in a spectrum, ranging from mobility restriction to chronic pain and alignment disorders. Literature often describes forearm deformities such as excessive radial bowing, ulnar shortening, and radial head dislocation. Factors resulting in genu valgus and specific treatment strategies are not frequently studied. Considerations specific to the disease, such as the morphology of the lesion, time taken for deformity correction, and timing of exostosis excision, should be taken into account when treating genu valgum in patients with MHE.

Case: Our case report highlights a teenager with underlying MHE who presented to us with "knock knees" since childhood. Clinically, it was observed that the patient had bilateral genu valgum and multiple bony swellings overlying the proximal tibia and distal femur. The deformity was subsequently addressed with bilateral proximal tibia hemi-epiphysiodesis, exostoses excision of bilateral proximal tibia, and left distal tibia. The radiographs and surgical technique are detailed in the case report. Our literature review indicates that distal femur exostoses play a significant role in causing genu valgus in these patients. The morphology of the exostoses also affects the development of genu valgus, with sessile lesions contributing more bony deformity compared to pedunculated lesions. Hemi-epiphysiodesis, as performed in our patient, often achieves outcomes similar to idiopathic genu valgum. However, the correction would take about 6 months longer, so in cases of MHE, correction should start earlier. Another dilemma in cases of pathological genu valgus is whether to remove exostosis during corrective surgery. Literature advocates delaying the excision of proximal tibia exostoses as long as possible to reduce the risk of accelerating deformity worsening.

Conclusion: In conclusion, factors influencing genu valgus progression include the morphology of the lesion (sessile), presence of metaphyseal flaring, and a history of prior exostosis excision. Early detection of the pathology would yield better outcomes, as the rate of deformity correction in children with MHE is slower due to abnormal physis. Meticulous surgical technique to prevent injury to the physis and knee immobilization post-excision of exostosis are important factors in preventing the development of genu valgus in children with MHE.

Keyword: Genu valgum, multiple hereditary exostosis, osteochondroma, tension band plate, epiphysiodesis

Case Report Article

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INTRODUCTION

Multiple hereditary exostosis (MHE) is a rare genetic disorder characterized by the development of benign cartilaginous tumors originating from the perichondrium, flanking the cartilage growth. With a prevalence of approximately 1:50,000 in the Western world, MHE predominantly affects males (1). Also known as multiple hereditary osteochondromas and diaphyseal aclasis, this autosomal dominant disorder can be inherited from an affected parent in half of the cases. Typically impacting the growth plate of bones such as the ribs, pelvis, vertebrae, and the meta-diaphyseal segment of long bones, the disease manifests in various ways, including chronic pain syndromes, restricted range of motion, limb deformity, short stature, and scoliosis.

The underlying cause of MHE is attributed to mutations and functional loss of the EXT1 and EXT2 genes, which encode enzymes involved in heparan sulfate synthesis (2). This genetic condition significantly influences the quality of life and physical activity levels from childhood.

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From an orthopedic standpoint, treatment options are limited to resection, limb shortening, and surgical deformity correction, recommended when cases are symptomatic or when malignant transformation is suspected. However, there is scarce literature addressing factors leading to genu valgus and specific considerations in managing this deformity in children with MHE.

CASE REPORT

A 13-year-old boy, diagnosed with multiple hereditary exostoses, presented with a history of multiple bony swellings on both upper and lower limbs since childhood. The patient experienced difficulty walking, accompanied by progressively worsening 'knock knees.' Clinical examination revealed bilateral genu valgum, and the patient walked with knee flexion. Palpation indicated multiple bony swellings overlying bilateral distal femur and proximal tibia. Active knee range of motion was limited from 10 to 90 degrees, with intact neurovascular status in both lower limbs.

Plain radiographs revealed sessile exostoses on the medial aspect of the distal femur and proximal tibia physes. A long limb view assessed the degree of genu valgum. The anatomical lateral distal femoral angle showed 80.2 degrees on the right side and 82 degrees on the left side, within the normal anatomical variation of 80-85 degrees (Figure1). However, the anatomical medial proximal tibia angle was 98 degrees bilaterally, significantly higher than the normal range of 80-85 degrees, contributing to the genu valgum (Figure2).

The deformity was addressed through bilateral proximal tibia hemi-epiphysiodesis, and exostoses were excised from bilateral proximal tibia and the left distal tibia. The patient was positioned supine, and bilateral tourniquets were applied preoperatively.

A medial incision overlying the bony exostosis on the proximal tibia exposed the periosteum, and the exostosis was removed with a chisel. The periosteum was repaired, and the medial proximal tibial physis was identified using C-arm fluoroscopy. A Kirschner wire was inserted parallel to the joint line in the mid-sagittal plane. Once the desired position was achieved, a guided growth '8' plate was slid over it.

Guide pins were inserted first into the epiphyses and then into the metaphysis, followed by the use of a cannulated drill. Screws were inserted into both guide pins, tightened sequentially, and the pins were removed.

Postoperative radiographs confirmed that the screws were parallel to the joint line in the AP view and in the mid-sagittal plane in the lateral view (**Figure 3 and 4**). The patient was discharged on day 3 postoperatively, able to walk with a walking frame.

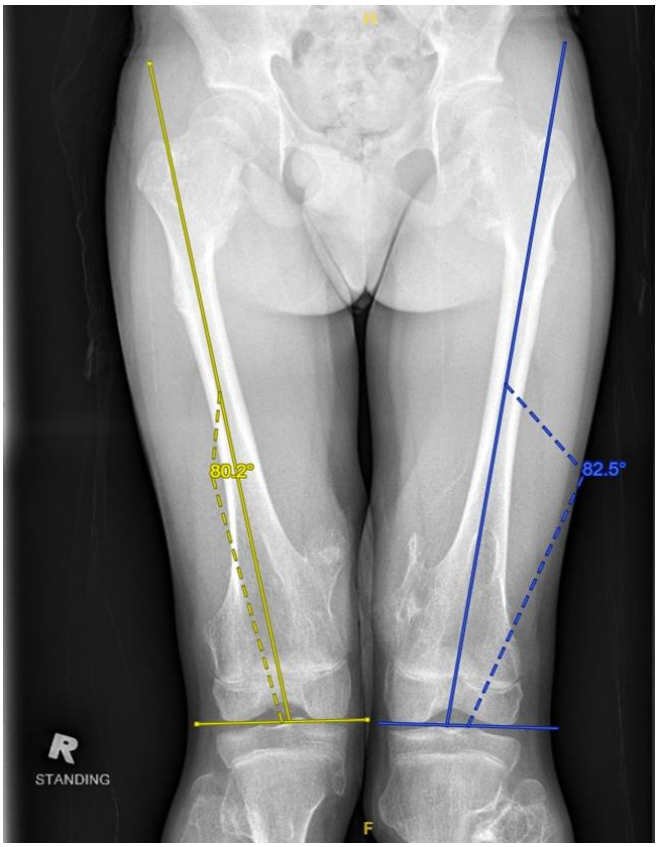

Figure 1: Long limb view of bilateral femur with LDFA within normal values.

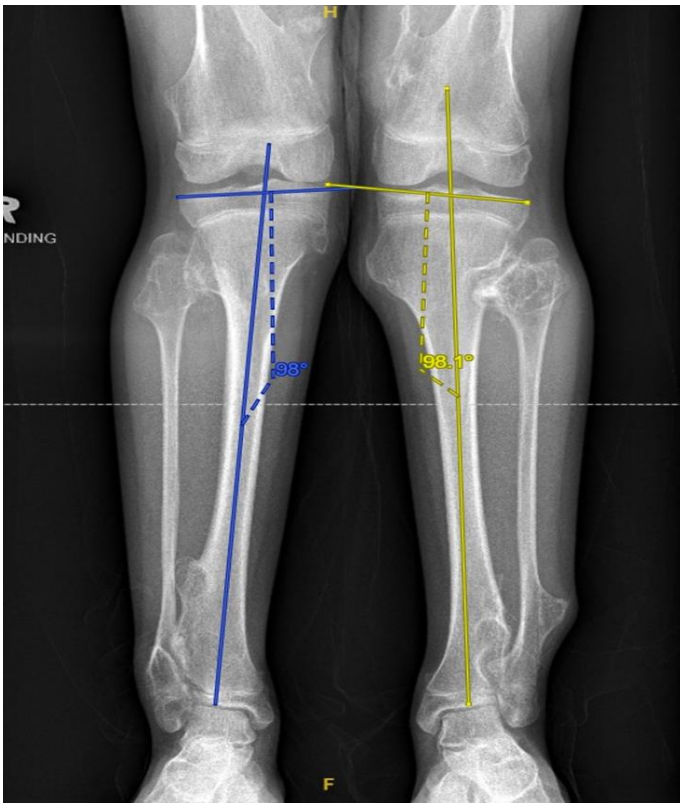

Figure 2: Long limb view of bilateral tibia with MPTA of 98 degrees.

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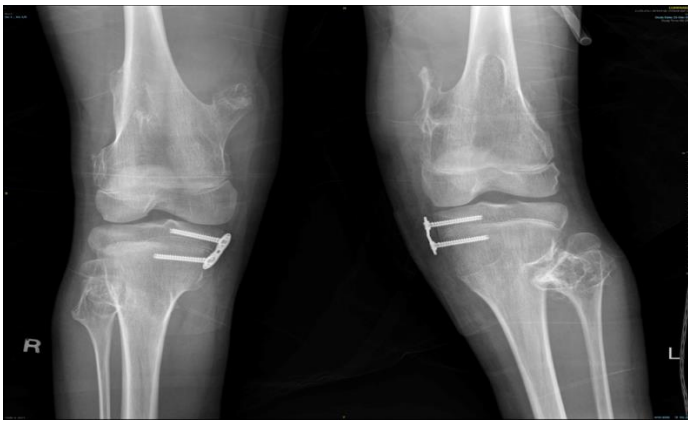

Figure 3: (AP View) Showing both screws parallel to the knee joint line.

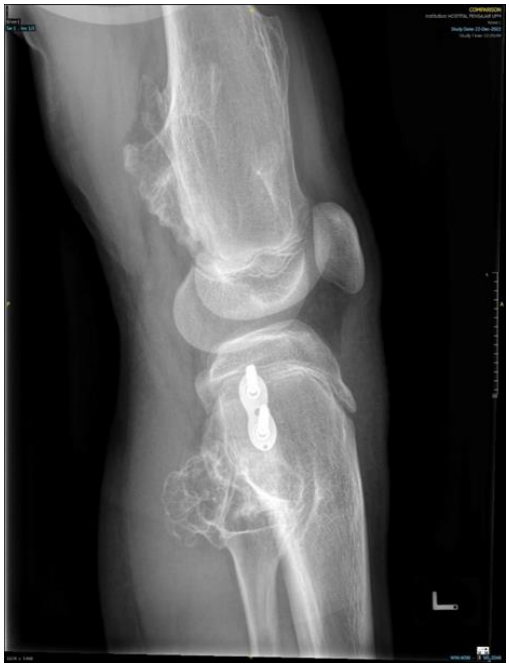

Figure 4: (Lateral View) showing both plates placed in mid sagittal plane.

DISCUSSION

Genu valgum, commonly known as knock knees, is a coronal plane deformity frequently observed in children. While often asymptomatic and part of the normal physiological development of lower limbs in children, understanding the normal developmental pattern and acceptable variations is crucial for assessing any underlying medical conditions. According to Salenius and Vanka, the tibiofemoral angle in children follows a familiar pattern as they develop (3). Genu varum is evident at birth until about 18 months, followed by a significant genu valgum at 2-3 years. The valgus nature gradually resolves, leaving a physiological genu valgum of 5-7 degrees as the skeletal matures. Any variation of +/- 8 degrees is considered pathological and warrants investigation (3).

Various causes contribute to genu valgum, including rickets, skeletal dysplasias, trauma, infection, and, in extremely rare cases, Multiple Hereditary Exostoses (MHE), as in our case. Approximately one-fourth of MHE patients develop genu valgum (2). Liu et al attempted to study the factors causing genu valgus in MHE patients (4). The severity of MHE, indicated by the number of exostoses or the presence of concomitant forearm deformity, did not influence the development of genu valgum. However, the debate continues on whether the deformity is driven by the proximal femur or the distal femur. Clement et al suggested that exostoses of the distal femur play a crucial role in knee disorder development (1). Madoki et al reported concurrent ankle valgus in 44 percent of MHE patients and limb length discrepancy of 2 cm in 19 percent (1).

Lesion morphology is also a crucial factor in causing genu valgum (4). Sessile lesions usually have a larger base, contributing to more bone deformity and a higher possibility of limb malalignment. Pedunculated lesions, on the other hand, may extend further into surrounding tissues, causing compression rather than alignment deformities. Metaphyseal flaring, along with sessile lesions, appears to be significant causes for genu valgus in terms of lesion morphology.

Hemi-epiphysiodesis, as performed in our patient, often achieves outcomes similar to idiopathic genu valgum, but correction takes about 6 months longer. In cases of MHE, correction should commence earlier (4). Pathologic genu valgus in MHE patients typically occurs during peak growth ages of 10 to 14 years, as observed in our thirteen-year-old patient (4). The condition is thought to arise due to the exostosis mass affecting bone growth.

A dilemma in cases of pathological genu valgus is whether to remove exostosis during corrective surgery. Alexandra et al reported two cases of rapidly progressing genu valgum post-excision of exostosis overlying the distal femur and proximal tibia, necessitating hemi-epiphysiodesis (5). They attributed the progression to the procedure rather than the pathology itself. The inferred mechanism suggests that increased blood flow to the injured area, as a natural response to trauma, stimulates surrounding physis, leading to asymmetric bone growth. Hence, Madoki et al advocate removing exostosis at the proximal medial tibia as delayed as possible, if feasible, to reduce the risk of worsening valgus deformity (1).

CONCLUSION

Genu valgum is a common condition encountered in pediatric orthopedics. Distinguishing between pathological genu valgum and variations in normal development requires a comprehensive understanding of the physiological progression of lower limb development. A less common but significant cause of genu valgum is observed in patients with Multiple Hereditary Exostoses (MHE). Factors influencing the progression of genu valgus in MHE patients encompass the morphology of lesions (particularly sessile types), the presence of metaphyseal flaring, and a history of prior exostosis excision.

Early detection of the pathology is crucial for achieving optimal outcomes, as the rate of deformity correction in children with MHE is inherently slower due to abnormal physis. Employing meticulous surgical techniques to prevent physis injury and implementing knee immobilization post-excision of exostosis emerge as critical factors in preventing the development of genu valgus in children afflicted with MHE. A proactive approach towards understanding and managing these factors is essential for the effective treatment and prevention of genu valgum in this specific patient population.

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Author Contributions: ST, NK, IIBI: Designed and directed the study, Literature search, Data collection, **ST:** Article writing, Final revisions. All authors reviewed the results and approved the final version of the manuscript.

Ethical approval: The present study was conducted in strict accordance with the principles outlined in the Declaration of Helsinki. Ethical approval for the study was obtained from the appropriate ethics committee.

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