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Case Report

A case report of Leri-Weill dyschondrosteosis with bilateral Madelung deformity managed by Vicker's ligament release and dome osteotomy

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ABSTRACT

Leri-Weill dyschondrosteosis (LWD) is an uncommon skeletal dysplasia marked by mesomelic limb shortening and the typical wrist abnormality known as Madelung deformity. It results from reduced activity of the SHOX gene. We report a 9-year-old girl with a 4-year history of progressive, painless deformities affecting both forearms. Examination revealed short stature (116 cm), bilateral forearm shortening, dorsal prominence of the ulna, and restricted ulnar deviation and elbow extension. Genetic evaluation confirmed a pathogenic SHOX mutation. Surgical treatment with bilateral distal radius dome osteotomy combined with Vicker's ligament release was performed. Postoperatively, the patient achieved good cosmetic correction and marked functional improvement. This case emphasizes the importance of evaluating SHOX-related disorders in children presenting with short stature and wrist deformity. Combined ligament release with corrective osteotomy offers both functional and cosmetic benefit in LWD patients.

Keywords: Leri-Weill dyschondrosteosis, Madelung deformity, SHOX gene, Mesomelic short stature, Dome osteotomy, Vicker's ligament

INTRODUCTION

Short stature is a frequent concern in pediatric clinics and may result from a wide range of causes, including endocrine disorders, skeletal dysplasias, congenital syndromes, and malabsorption problems.[1-6] In female patients, Turner syndrome is often the first differential diagnosis and can be excluded by karyotype testing. Another key genetic condition to consider is Leri-Weill dyschondrosteosis (LWD), a pseudoautosomal dominant disorder caused by the insufficient expression of the short stature homeobox (SHOX) gene.[7]

LWD typically presents with mesomelic shortening of the extremities, especially involving the forearms and lower legs, along with the pathognomonic Madelung deformity of the wrist. Madelung deformity is characterized by abnormal curvature of the distal radius, early closure of the growth plate, and dorsal subluxation of the ulna. An abnormal fibrous band, known as Vicker's ligament, is frequently implicated in restricting radial growth and worsening deformity.[2] This report describes a classic case of LWD with bilateral Madelung deformity successfully managed using Vicker's ligament release and a distal radius dome osteotomy.[3]

CASE REPORT

A 9-year-old female presented to the orthopedic outpatient department with visibly abnormal deformities of both forearms. Her parents first noticed an abnormal prominence on the ulnar aspect of her forearms approximately four years prior.

The deformity was progressive and was associated with a gradual limitation of wrist and elbow movements. There

was no history of trauma, infection, or any significant birth, past, or family history.

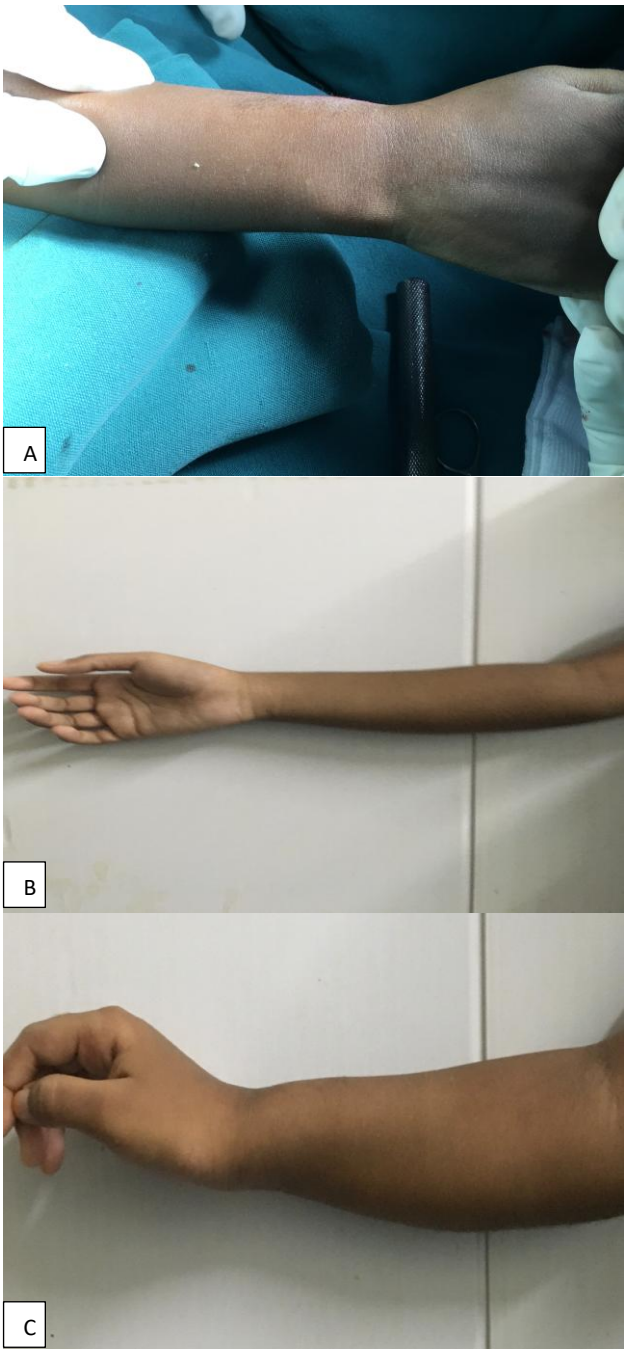

Figure 1 (A-C): Abnormal prominence on the ulnar aspect of both forearms, progressive curvature or bowing of the radius and ulna and limited forearm and wrist movement.

On examination, her height was 116 cm, which was significantly below the expected height of 132 cm for her age. The upper segment to lower segment (US:LS) ratio was 56:60 cm, and her arm span was 110 cm, consistent with mesomelic shortening. Head circumference was normal at 51 cm. Local examination of the forearms revealed bilateral shortening, a dorsal prominence of the distal ulna, and a positive "ulnar styloid sign" (where the ulnar styloid process is palpated distal to the radial styloid). Bilateral mild manus valgus was present. Range of motion assessment showed significant limitation in ulnar deviation and elbow extension.

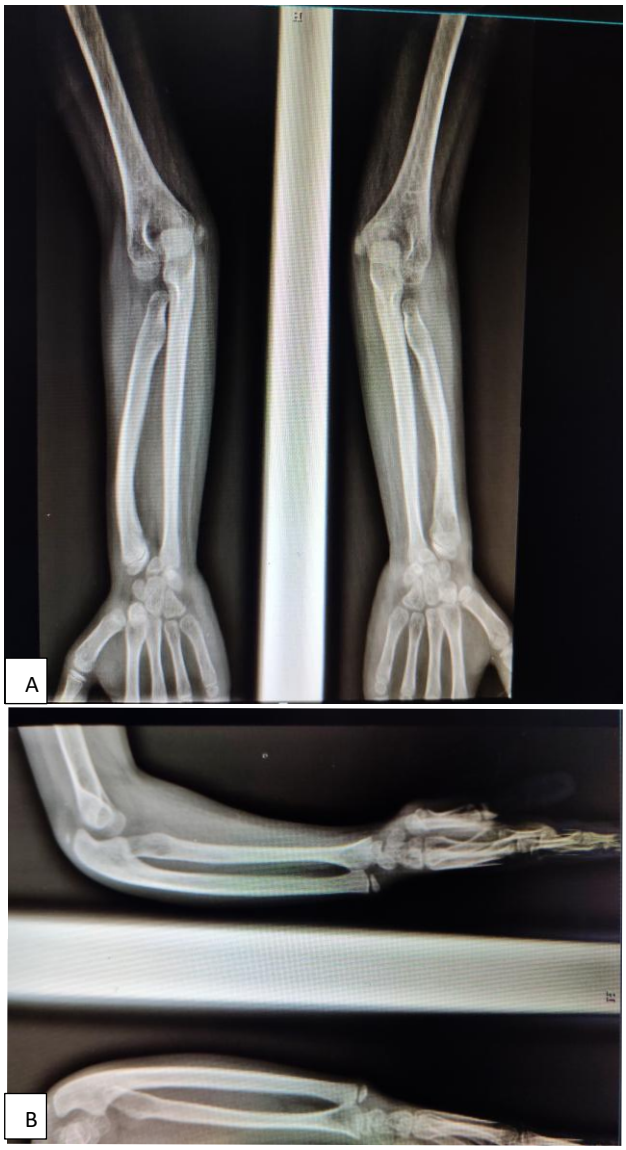

Figure 2 (A and B): AP and lateral X-ray of bilateral forearms showing distal radial bowing with relative ulnar overgrowth. Classical Madelung deformity with the abnormal volar-ulnar tilt of the distal radius is noted.

A clinical suspicion of LWD was raised. Genetic testing was performed and identified a pathogenic mutation in the SHOX gene, confirming the diagnosis.[2]

Management

The patient was planned for surgical correction to address the deforming forces and the bony deformity.

The procedure performed was a dome osteotomy of the distal radius to correct the angular deformity, combined with a release of Vicker's ligament to eliminate the pathological tether preventing normal radial growth.[3,8]

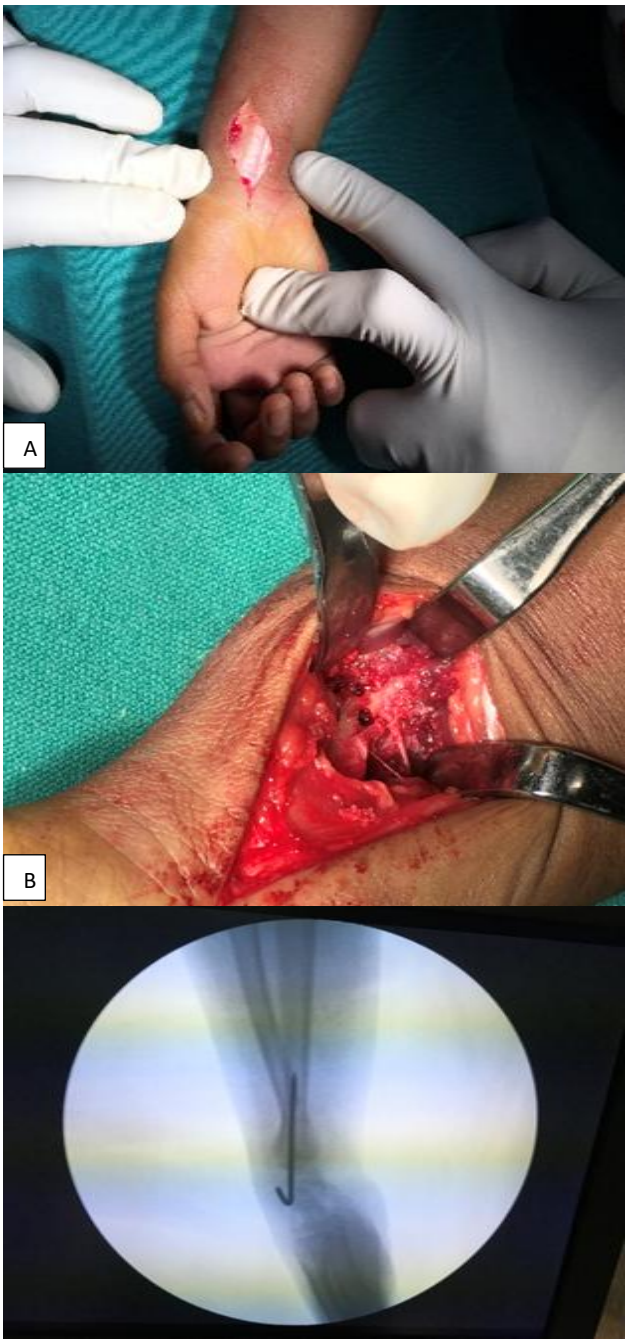

Figure 3 (A-C): Intraoperative images of dome osteotomy of distal radius with Vicker's ligament release.

Outcome

Post-operatively, the patient showed excellent results. The cosmetic deformity of the forearms was significantly corrected. More importantly, there was a marked improvement in the functional range of motion at the wrist and elbow joints.[5,10]

DISCUSSION

LWD is an important but under-recognized cause of disproportionate short stature, particularly in girls when Turner syndrome has been ruled out.[7] The hallmark feature is bilateral Madelung deformity, which arises due to a combination of SHOX gene deficiency affecting endochondral ossification and mechanical tethering by Vicker's ligament.[8]

Treatment strategies should consider both the pathological ligament and the osseous deformity. Mild or non-progressive cases may be observed conservatively, whereas progressive deformities with pain or functional limitation benefit from surgery.[11] The combined approach of releasing Vicker's ligament and performing a corrective dome osteotomy of the radius is well established.[2]

This dual procedure addresses both the tethering effect and the angular deformity, restoring alignment, motion, and cosmetic appearance. The excellent outcome in our patient supports the effectiveness of this method, consistent with previously published experience.[10,11]

CONCLUSION

This case underscores the necessity of a genetic workup for SHOX mutations in children presenting with mesomelic short stature and Madelung deformity. A timely and correct diagnosis allows for appropriate surgical planning. A combined approach of Vicker's ligament release and dome osteotomy of the distal radius is an effective treatment for correcting the deformity and restoring function in patients with LWD.

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