

A case of medelung deformity in a 13-year-old Iranian girl

Abstract

Objective: a case of a 13-year-old left-handed Fars female with acute left wrist pain was presented.

Background: Madelung deformity is a wrist developmental anomaly which results in palmar and ulnar wrist subluxation with a more tendency to happen bilaterally and in female. This deformity usually appears clinically between the ages of 6 to 13 years.

Conclusion: Although rare, Madelung deformity is typically corrected surgically in with chronic pain and disability. This case demonstrates an example of successful surgical management.

Keywords: triangular fibrocartilage complex, madelung, Iran

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Case presentation

A 13-years old left-handed Fars female, pre-menarche, presented with wrist mechanical pain and supination and extension limitation of her right forearm lasting for more than 1 year. Physical examination revealed ulnar head prominence, volar tilting of the carpus and hand, and 13 and 25 degree tender limitation of extension and supination of the wrist, respectively.

Blood cell count and the blood chemistry analysis were normal. Her familiar history was positive for grandmother's deformity similar to her own, with being less symptomatic. The postero-anterior wrist X-rays (Figure 1) illustrated typical Madelung deformity changes on her both wrists which is characterized by shortening of radius bone, in comparison with the ulna, radially curved radius, a disparity of the distal radioulnar joint plus a triangular-shaped carpus. The lateral X-ray (Figure 2) showed major changes on the right wrist: dorsal

dislocation of the ulnar and an exaggerated volar tilt of the radioulnar joint in a way which caused palmar carpal displacement and an anterior translation of the hand and the wrist. She was candidate for closed wedge osteotomy performed on the right side of left distal radius with Vicker's ligament release, and an ulnar shortening osteotomy with plate fixation (Figure 3). No intra and postoperative complications were occurred. Osteotomies were protected with dorsal forearm plaster splint for 3 weeks. Postoperative management depends on what is done. Casting immobilization might be necessary for some weeks after the osteotomy of the radius or ulna. Physiotherapy might be needed in children not able to regain range of motion on their own after 2-3 months without restrictions. In our case no physiotherapy was needed. After 2 months, there was no pain, a good aesthetic and mobility improvement of the right wrist mainly in prone-supination. The radiographic images (Figure 3) illustrated improving alignment and healing of the bone.



Figure 1 Posteroanterior and Lateral X-rays at presentation.



Figure 2 instant postoperative radiographic images.



Figure 3 Radiographic images after 2 months of surgery.

Discussion

The Madelung deformity is a rare condition, representing 1.7% of the congenital diseases¹ associated with Vicker's ligament which creates a tether across the volar-ulnar radial physis, resulting in early growth plate arrest at the medial volar aspect of the distal radius. As the remaining physis grows, the radius bone deforms with ever-increasing radial inclination and volar tilt; the unaffected ulna continues to elongate, resulting in a progressive ulnar positive variance. Finally, the distal radioulnar joint fails to develop normally and the distal ulna subluxates or dislocates dorsally.² Congenital Madelung deformity may arise as a part of Turner syndrome and is thought to be a result of a mutation or absence of the short stature homeobox gene (SHOX). Madelung-like deformity can also be acquired as the result of repetitive traumatic pressure. The disorder frequently occurs in females, with a ratio of 3 to 5:1.³ Madelung is evident clinically between six to thirteen years old.⁵ Patients have usually complaint of wrist pain, impaired grip strength, limited range of motion, commonly in supination, and disfigurement based on their prominent ulna.^{1,3} Madelung deformity could be found in anteroposterior and lateral wrist x-ray. Specific thresholds have been determined accurately detect Madelung deformity. These limits are at least 33° of ulnar tilt, 4 mm of lunate subsidence, lunate fossa angle of 40° or more and 20° ≤ mm palmar carpal displacement.⁴⁻⁶ Radiological deformity does not systematically correspond to functional disorder or pain; functional impairment found in certain cases necessitates complementary analysis, preferably by MRI, of structural lesions.³ Treatment depends on the age of patient at presentation and distal ulnar growth potential, magnitude of symptoms and the degree of deformity.^{2,7} Progressive deformities in children with considerable remaining growth potential, advanced or symptomatic deformity are indications for operation to avoid continuing worsening.² There is no universally favored surgical approach strategy; the standard treatment is a radial dome osteotomy plus Vicker's ligament release.⁵ Ulnar approach may be chosen in

severe cases including increased lunate subsidence, ulnar variance and volar carpal displacement,⁵ to achieve more suitable length symmetry between the radius and ulna. Although the techniques vary significantly, each can be categorized into 1 of 3 groups: the 1th group applied to the radius alone includes epiphysiodesis and corrective osteotomy; the 2nd group involves repair and reconstruction limited to the ulna including epiphysiodesis, ulnar head excision, shortening osteotomy, distal ulnar resection and creation of a pseudarthrosis with or without fusion to the radius; the 3rd group involves some combination of the above mentioned techniques.⁷ Mild asymptomatic deformity needs a period of nonsurgical management with serial x-ray examinations control because the natural history is unpredictable; generally most patients not at all require surgical intervention.² The technique we described aims to restore the anatomy of the wrist to as normal as possible, in order to relieve pain, acquire esthetic correction and improve joint motion. We joined both bones osteotomies, to restore immediately the anatomy, to Vicker's ligament release to avoid loss of correction in the future. Our result after one year follow up show that grip strength and range of movement improved notably; there is no pain and the patient is satisfied with the esthetic appearance.

Conclusion

It is a rare condition that, when diagnosed and treated properly may improve the mobility and function of the wrist, and so the adolescent's quality of life. Our technique introduced acceptable results for the treatment of the Madelung deformity.

Acknowledgment

None.

Conflicts of interest

None.

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