Madelung Deformity With Prior Distal Radius Fracture: A Case Report

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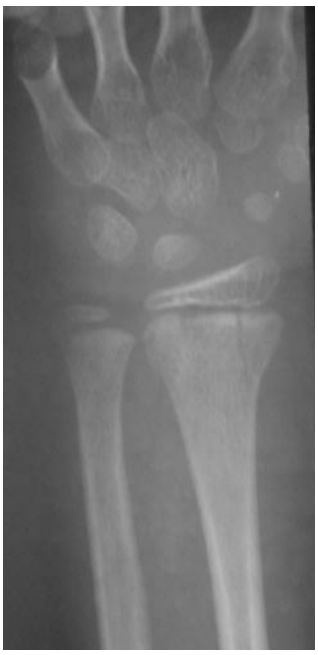
Madelung[1] first described the wrist deformity bearing his name in 1878. Although previous authors had described lesions that we might now term *Madelung deformity,* Madelung was the first to accurately describe it clinically and to propose both an etiology and a treatment. The deformity results from epiphyseal arrest on the ulnar and volar half of the distal radius, which causes the articular surface to be directed ulnarly and volarly. In 1992, Vickers and Nielsen[2] further described the abnormal physeal and ligamentous anatomy on the volar aspect of the radiocarpal joint. They identified a thick fibrous band spanning the radial metaphysis to the proximal carpal row (Vickers ligament) seen in congenital Madelung deformity.

The etiologies of Madelung deformity have been thoroughly described in the literature. Although confusion continues as to the exact etiology of this condition, most reports show the deformity to be a condition inherited by an autosomal-dominant trait with variable expressivity. Common to congenital Madelung deformity is the presence of Vickers ligament, which tethers the volar-ulnar distal radial physis. Other reports have described an acquired or pseudo-Madelung deformity wherein injury to the immature physis produces a bony bridge with progression to a deformity that may mimic that of a true congenital Madelung deformity. In this report, we present the case of an acquired Madelung deformity secondary to trauma with the presence of a Vickers ligament and absence of a bony bridge affecting the volar-ulnar distal radial physis. In the literature, presence of Vickers ligament associated with posttraumatic Madelung deformity is unique; it has been described only in the congenital variant.

Case Report

At age 7, a right-hand–dominant girl fell, landed on her hyperflexed left hand, and sustained a nondisplaced extra-articular left distal radius fracture that was treated with closed reduction and casting for 6 weeks (Figure 1). She healed uneventfully and did well until age 12, when she had increased pain and swelling, particularly of the ulnar side of the left wrist, as well as decreased wrist range of motion.



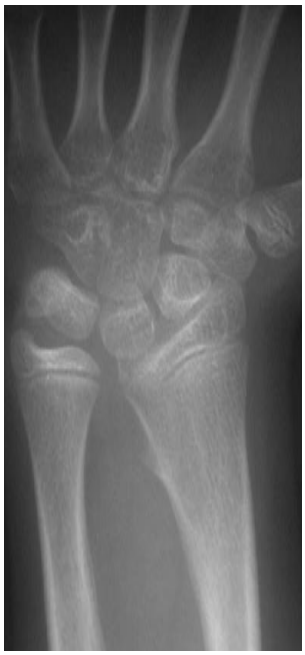


Figure1. (A, B) Left nondisplaced extra-articular distal radius fracture in 7-year-old girl.

Figure 2. Distal radius deformity in same child 5 years after injury.

On presentation, this now 12.5-year-old patient reported no significant past medical, surgical, or family history suggesting bony or soft-tissue abnormalities. She had no allergies and was not taking any medications. She actively participated in activities, including gymnastics, soccer, and softball.

The most recent preoperative plain films suggested a fused growth plate on the ulnar side of the distal radius (Figure 2) leading to a significant deformity, with the radial aspect of the radius continuing to grow with radial inclination increased well beyond normal, and also with the ulna continuing to grow and cause significant positive ulnar variance. The lateral plain film showed the ulna subluxating dorsally. Computed tomography with minor cuts did not reveal a significant growth plate anomaly, but a magnetic resonance (MR) image suggested a small physeal bar near the old fracture site.

The patient was premenarchal and of normal stature for her age. Physical examination revealed limitation of motion

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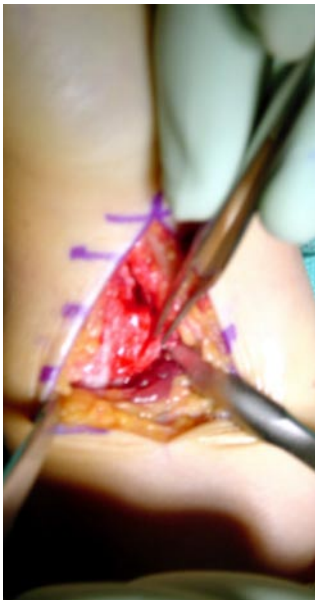

Figure 3. Vickers ligament.

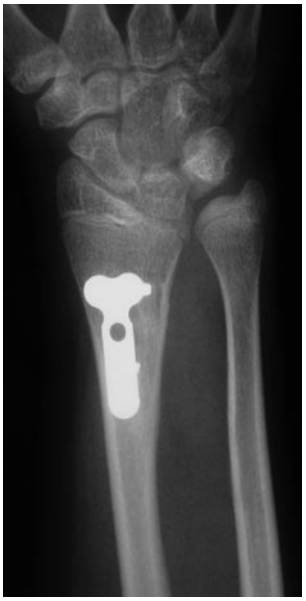

Figure 4. Two years postoperatively. Patient healed well, maintaining her position with normal resumption of radial physeal growth.

with pronation of 20° on the left versus 80° on the right side. Supination was 80° bilaterally. Wrist flexion was limited as well, to 45° on the left versus 60° on the right and with left wrist extension of 35° versus 70° on the right. Radial deviation and grip strength were significantly diminished on the left as well. The patient also had a prominent ulnar head, with pain and tenderness to palpation. Her contralateral wrist had no physical or radiologic abnormalities.

The surgical plan consisted of corrective osteotomy of the left distal radius with placement of structural allograft and plating secondary to continued symptoms. A modified Henry approach to the left distal radius was performed. Pronator quadratus was elevated off of the distal radius on the radial border, and Vickers ligament was identifiable in the ulnar aspect of the distal radius (Figure 3). This was an interesting finding, as the impression of this injury was that it was a malunited distal radius with a bony bridge. However, there was no evidence of a bony bridge on the ulnar aspect of the distal radius on direct visualization. These findings suggested that this was a unique posttraumatic Madelung deformity. Despite the past history of trauma, a Vickers ligament was present, not a bony bridge. Subsequently, the Vickers ligament was released off the distal radius, an osteotomy was made 3 cm proximal to the growth plate, allograft was inserted, deformity was corrected in all planes, and plate fixation was completed under fluoroscopy.

Two years after surgery, the patient was well healed and maintaining her position with normal resumption of radial physeal growth (Figure 4). She had a painless arc of motion and no ulnar prominence or tenderness. She could flex her left wrist 65° and extend it 80°. Grip strength, pronation/supination, and deviation improved to normal as well.

Discussion

The clinical manifestations of Madelung deformity are characterized by the insidious onset of pain in one or both wrists and increasing prominence of the dorsal ulnar head and bowing of the distal radius. Deformity progresses until the growth plate of the distal radius closes. Wrist motion, particularly extension and supination, is limited. Radiographically, there is an increase in the normal palmar and ulnar inclination of the distal radius. The ulna is unaffected and remains in its usual dorsal position.

Henry and Thorburn[3] classified Madelung deformity into 4 etiologies: dysplastic, chromosomal/genetic, idiopathic/primary, and posttraumatic.

Bone dysplasias associated with Madelung deformity include multiple hereditary osteochondromatosis, Ollier disease, achondroplasia, multiple epiphyseal dysplasias, and mucopolysaccharidoses. However, the most important associated dysplasia is dyschondrosteosis, a type of mesomelic dwarfism that should be suspected if the condition is bilateral or if there is a positive family history, short stature, or mesomelia.

One third of cases of Madelung deformity are transmitted in an autosomal-dominant fashion, with variable expression and 50% penetrance. The deformity is bilateral 50% of the time, and females are affected 4 times as often as males. Molecular genetic studies have shown that an X-chromosomal translocation is associated with Madelung deformity, Turner syndrome, and dyschondrosteosis.[4-6]

The posttraumatic deformity can occur from repetitive stress or from a single event that disrupts growth of the distal radial ulnar-volar physis. The literature includes many case reports of pseudo-Madelung deformity in gymnasts secondary to the sport's repetitive stress to the wrists. Vender and Watson[7] reported a case of a high-level gymnast with bilateral closure of the ulnar side of the distal radius physeal plate, with clinical and radiographic changes like those found in a congenital Madelung deformity. They concluded that cumulative microtrauma to the ulnar side of the distal radius physeal plate may cause premature closure leading to a Madelung-like deformity.

Conclusions

Thorough history-taking and physical examination indicated that the etiology of our patient's wrist deformity was unlikely to be genetics or a dysplastic disorder. The patient had an acquired Madelung deformity. Her diagnosis and etiology indicated that the initial injury to the physis caused the abnormal growth volarly and ulnarly at the distal radius. MI images suggested a physeal bridge. However, intraoperative findings included a Vickers ligament without evidence of a bony bridge. To our knowledge, there have been no other case reports citing a postfracture Madelung deformity with a Vickers ligament and absent a bony bridge. Whether there is a link between the initial fracture and the subsequent Madelung deformity with Vickers ligament remains unclear. In patients with past history of

fracture and continued wrist pain, an acquired Madelung deformity secondary to a bony bridge or a Vickers ligament should be considered as part of the differential diagnosis.

Author's Disclosure Statement and Acknowledgement

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