Short report: Madelung deformity presenting clinically as radioulnar dislocation

Anand Damodaran and Alexandra Thomas
Department of Emergency Medicine
University Hospital of North Tees
Hardwick Road, Stockton-on-Tees
TS198PE. UK
alex.thomas@nth.nhs.uk

A 28 year old woman presented to the Emergency Department after a fall. Examination showed a tender deformity over the dorsal aspect of the right distal ulna suspicious of radio-ulnar dislocation. The patient was of below average height with short forearms and lower legs.

X-rays revealed a hitherto undiagnosed Madelung deformity (Figure 1).

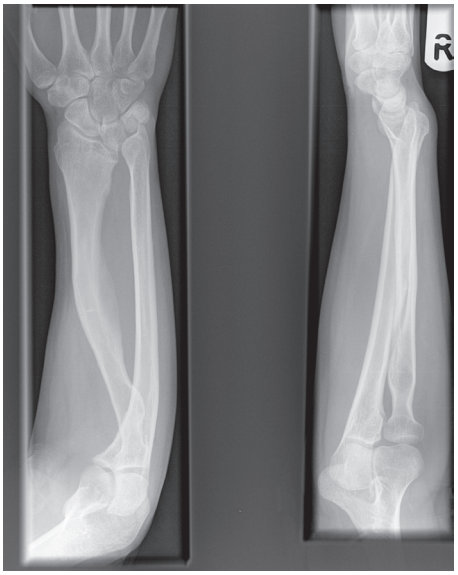

Figure 1. *Radiograph of forearm showing shortening of the ulnar margin of the distal radius, proximal migration of the proximal carpal row and dorsal dislocation of the ulnar head.*

Discussion

Madelung deformity (MD) occurs due to growth disturbance in the epiphyseal growth plate of the radius resulting in a volar curvature and a tilt towards the ulna. The ulna continues to grow straight resulting in a dorsal and distal prominence. It is almost always noted in adolescent females who present with pain and reduced mobility at the wrist. It may go unrecognized and present much later, as in this case.

Otto W. Madelung described the wrist deformity bearing his name at the seventh German Surgical Congress of 1878 as a case of "spontaneous forward subluxation of the hand". He described it clinically and proposed both an etiology and treatment. Henry and Thorburn classified MD into four different etiologic groups:

1. post-traumatic
2. dysplastic
3. chromosomal or genetic (Turner syndrome),
4. idiopathic/ primary.

It is bilateral in 50% of cases.

Although there are many bone dysplasias that may cause Madelung deformity (eg multiple hereditary exostoses, Ollier's disease, achondroplasia and mucopolysaccharidosis), the most important dysplasia associated with this condition is dyschondrosteosis, a form of dwarfism that is characterized by shortened forearms and lower legs (mesomelic dwarfism). Our patient had features highly suggestive of this musculoskeletal condition.

First described by Andre Leri and Jean Weill in 1929, dyschondrosteosis is characterized by variable short stature, short forearms, and tibial/ fibular shortening. It is often caused by mutations in the SHOX (short stature homeobox) gene found in the pseudoautosomal region (PAR1) of the X and Y chromosomes and follows autosomal dominant inheritance. Homeobox genes normally help regulate development.

The diagnosis is confirmed radiographically with PA and lateral views of the forearm and

wrist. Shortening of the ulnar margin of the distal radius, proximal migration of the proximal carpal row and dorsal dislocation of the ulnar head were noted on radiographs in our patient as shown in figure 1.

Treatment : Application of a splint may be helpful in skeletally mature individuals with MD and mild-to-moderate short-term wrist pain. Goals for surgical correction of MD consist primarily of pain relief and correction of the cosmetic deformity. A secondary goal is to increase range of motion.

Some of the procedures used to correct the condition include:

1. Release of Vicker's ligament – a thick fibrous band spanning the radial metaphysis to the proximal carpal row.
2 Osteotomy of the radius.
3. Radioulnar length adjustment.
4. Ulnar resection or Darrach procedure.

Our patient had no problems with wrist joint movement and was not worried about the cosmetic deformity either. She was discharged on analgesics .

References:

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