Madelung's Deformity

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Introduction

Madelung's deformity was first described by Malgaigne in 1855 however carries Madelung's name from his description in 1878 [1]. The deformity is caused by an abnormal growth arrest of the palmar-ulnar distal radius physis. In addition to the abnormal growth, there is an abnormal palmar ligament tethering the lunate to the palmar-ulnar radius epiphysis and metaphysis (Vickers' ligament) that is thought to contribute to the deformity [11]. The deformity is characterized by a prominent dorsal subluxation of the ulnar head and palmar sag of the hand and wrist with a shortened forearm. Patients present with wrist pain as well as concerns about the aesthetic deformity.

Clinical Features

Girls are more often affected more than boys. The disorder is usually present bilaterally with asymmetric severity and presents between the ages of 6 and 13 years. The deformity is likely present for years prior to diagnosis, but as it progresses

with growth, it becomes noticeable. It is most often diagnosed at the onset of adolescence when activity-related diffuse wrist pain or the abnormal appearance of the wrist prompts intervention. Studies have shown that either the pain or the appearance of the wrist can be the primary complaint. Patients also have decreased strength and range of motion in pronation, supination, and wrist extension in particular [4, 9, 11].

Imaging

Radiographically, there is an ulnar and volar curvature of the distal radius, increased inclination of the distal radius, and triangulation of the carpus as the lunate subsides proximally (Fig. 1). The distal radius epiphysis narrows progressively along its ulnar half. The deformity of the distal radius makes it difficult to clearly define radiographic measurements based on the radius. McCarroll et al. evaluated different methods for quantifying the deformity based on the ulna and the carpal bones rather than the radius. The two most reliable and reproducible measurements were found to be ulnar tilt, which measures the amount of increased distal radius inclination, and lunate subsidence, which measures the proximal migration of the lunate [6].

The role for MRI in diagnosis and treatment is not well defined. Vickers and Nielsen proposed using MRI to identify skeletally immature individuals with a family history of Madelung's deformity who may be candidates for release of Vickers' ligament with the goal of preventing progression of the deformity. Stehling et al. have shown that Vickers' ligament can be clearly visualized and defined by MRI [10]. However, the use of MRI is not routine in the work-up of our patients. Figure 2 shows an intraoperative photograph of Vickers' ligament.

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Fig. 1 Severe Madelung's deformity. **a** Note the *triangulation* of the carpus with proximal migration of the lunate. The *recess* on the ulnar volar aspect of the radius is the origin of Vickers Ligament. **b** Note the volar subluxation of the carpus and the hand in addition to the prominent dorsal ulna. Figure Copyright Charles A. Goldfarb, M.D

Genetics

All cases of Madelung's deformity are thought to have a hereditary component; however, the genetic origins are complex and our understanding is evolving. Madelung's deformity is known to be associated with various skeletal dysplasias and syndromes. In 1929, Leri and Weill described dyschondrosteosis as a dysplasia with three clinical features: mesomelia (short forearm), short stature, and Madelung's deformity. In 1997, Rappold discovered the short stature

homeobox (SHOX) gene deletion that causes short stature in Leri-Weill syndrome [7]. Haploinsufficiency of the SHOX gene leads to a range of phenotypes, even within the same family, from isolated short stature to Leri-Weill syndrome [3]. Madelung's deformity itself presents on a spectrum from involving the entire radius, which is typically a more severe deformity with a short radius and ulna, or only the distal radius [12]. Patients with Turner syndrome have a haploinsufficiency of SHOX and can present with short stature and Madelung's deformity [8]. We believe that so-called idiopathic cases will eventually be linked to a genetic anomaly.

Acquired Deformity

A Madelung's type of deformity can also result from trauma to the distal radius physis, infection, isolated osteochondroma, or multiple hereditary exostosis. In a review of 100 children with fractures involving the distal radius physis, ten developed growth arrest of the physis and subsequent deformity [5]. Repeated attempted reductions can traumatize the distal radius physis and should be avoided, especially in the "slipper" type fractures of the distal radius. Also, repetitive loading of the wrist, as has been reported in high-level gymnasts, can cause a physeal injury and result in a Madelung type of deformity [2]. Patients with an acquired Madelung type of deformity do not have an associated Vicker's ligament or the characteristic lunate subsidence, triangulation of the carpus, or palmar translation of the carpus to the degree seen in Madelung's due to a genetic predisposition.

Treatment

Treatment of Madelung's deformity begins with typical non-operative measures including activity modification, a trial of a resting splint, and nonsteroidal anti-inflammatory medications on a limited basis. These measures have been unsuccessful for most of our patients, and treatment is guided by the age of the patient at presentation. Ideally, the deformity is recognized prior to skeletal maturity, in which case physiolysis combined with release of the abnormal Vickers' ligament is the treatment of choice. In their series of 11 skeletally immature patients (15 wrists), Vickers and Nielsen report on their experience with physiolysis according to the Langenskiöld procedure using fat as an interposition and release of Vickers' ligament. Pain, which was the motivation for surgery in the majority of their study group, was reduced in all patients within 6 months of surgery [11]. Deformity was more difficult to quantify, but was reported as improving slightly in most cases. Range of motion improved as well, particularly in supination.

Most patients are closer to skeletal maturity at presentation in which case a physiolysis alone is not sufficient. We

Fig. 2 Intraoperative photograph of Vickers' ligament. The ligament, outlined here in a *white dashed line*, originates on the radius and inserts on the anterior lunate. Figure Copyright Charles A. Goldfarb, M.D

believe that these cases are best treated with an osteotomy of the radius alone or in conjunction with an ulnar shortening osteotomy. Harley et al. reviewed the technique and results of distal radius dome osteotomy in 18 patients (26 wrists) with a mean follow-up time of 23 months. The average age at the time of surgery was 13 years. In 11 of their patients, pain was the chief complaint, whereas deformity was the presenting complaint in 14 patients. The dome crescent-shaped osteotomy, concave distally, is made with K-wires to fenestrate the volar cortex and then using a curved osteotome to complete the osteotomy. The curve of the osteotomy allows for translation of the distal fragment dorsally, and longitudinal traction allows reduction of the hand to the ulna. The reduction is held with smooth Steinmann pins placed through radial styloid into proximal fragment. In some patients, an ulnar shortening or epiphysiodesis is performed for further correction. Harley et al. reported improved radiographic parameters (ulnar tilt and lunate subsidence) for all of their patients and increased range of motion particularly in supination and wrist extension. All patients with pain as the primary complaint preoperatively reported decreased pain postoperatively. Four of their patients did go on to have an ulnar-shortening osteotomy in the follow-up period for ulnar-sided wrist pain [4]. Ulnar-sided wrist pain, or perhaps more specifically dorsal-ulnar wrist pain, is common in Madelung deformity. We most commonly treat the painful, prominent dorsal distal ulna with an open ulnar shortening osteotomy. In the older patient, typically past skeletal maturity, this is our preferred primary treatment while in the younger patient (even with severe deformity of the radius), we prefer the dome osteotomy of the radius alone (Fig. 3).

Fig. 3 Madelung's deformity before and after dome osteotomy. **a** Note the *triangulation* of the carpus and lunate subsidence in the preoperative X-ray. **b** Note the correction of the radial inclination and decreased lunate subsidence after the osteotomy with the K-wires holding the reduction. Figure Copyright Charles A. Goldfarb, M.D

Conclusion

Madelung's deformity may cause pain and deformity, typically presenting in the adolescent population. Our treatment algorithm is based on age and skeletal maturity. For children less than 10 years of age, excision of Vickers' ligament and physiolyis with fat interposition is our treatment of choice. In the adolescent, a dome osteotomy of the distal radius is our primary treatment. And in patients older than 20 years, a distal radius dome osteotomy is combined with an ulnar shortening osteotomy for maximal benefit; on occasion, we perform an ulnar-shortening osteotomy alone.

Conflict of Interest Elisa J. Knutsen declares that she has no conflict of interest.

Charles A. Goldfarb declares that he has no conflict of interest.

Statement of Human and Animal Rights This article does not contain any studies with human or animal subjects.

Statement of Informed Consent Additional informed consent was obtained from all patients for which identifying information is included in this article.

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