

Madelung disease: a case report

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ABSTRACT

Madelung's deformity is an anomaly characterized by subluxation of the carpus in relation to the forearm, that is, a progressive volar inclination of the ulna in relation to the distal articular surface of the radius. It commonly affects women and is bilateral, manifesting at the end of childhood. It can be caused by genetic changes, or after trauma, infection or neoplasia. The present report addresses a 13-year-old patient who complained of pain in the wrist with painful irradiation to the forearm. On radiography, abnormalities were identified in the radius, ulna and carpal bones. Due to its low complexity, a conservative treatment for symptom relief was chosen, with no provision for surgical procedures. Nevertheless, surgery can be an alternative to correct severe cases and relieve persistent pain. It consists of the dome osteotomy, which provides better volar coverage to the lunate and has a high success rate, mainly in the resolution of pain.

Keywords: orthopedics, Madelung disease, Madelung deformity, wrist

INTRODUCTION

Madelung's deformity is an anomaly at the palmar and ulnar region of the distal physis of the radius bone, characterized by a progressive ulnar and palmar inclination that gets developed at the distal articular surface of the radius and also by the dorsal subluxation of the distal ulna [1]. This disease was initially described by Malgaigne in 1855 and afterwards by Madelung in 1878 [2]. It is believed to be a congenital disorder, although is rare to notice until the late childhood or youth [3].

This anomaly is considered rare, as it represents only 1.7% of hand anomalies. Most of the cases are asymptomatic, but the wrist pain and the stiffness can lead the affected people to medical services. It is more prevalent in females [4]. The cause is still uncertain; nevertheless, it can be transmitted by an autosomal dominant pattern. Vickers has once described an abnormal ligament that connects semilunar to the distal radius next to the physis [5]. It is usual to believe that this ligament prevents the growth of the palmar-ulnar segment of the distal radius, commonly known as the Vickers ligament.

Trauma, neoplasm or infections can also lead to the occurrence of similar Madelung deformities ("Madelung like"). There is no definitive method to distinguish these deformities from the Madelung idiopathic deformity [6].

Vender and Watson classified Madelung deformities and "Madelung like" in four groups: post-traumatic, dysplastic (dyschondrosteosis or diaphyseal aclasis), genetical (i.e., Turner Syndrome) and idiopathic. They had suggested that the acquired deformities are usually distinguished by a lack of appropriate physical findings, as the unilaterality, less severe deformities and a proper history of repetitive lesion or stress [7].

We aim to report a case of Madelung's deformity found in a 13-year-old girl.

CASE REPORT

A 13 years old female reached the emergency sector with a complaint of wrist pain during sports practice. This pain evolved with painful irradiation to the forearm (volar surface). She denied trauma or previous infections. The physical

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examination revealed painful limitation to the extreme range of motion of the wrist and joint instability. Distal neurovascular bundle was preserved. A deformity of the distal epiphysis of radius was observed in a radiographic image (Fig. 1). The patient showed a better symptomatic reaction and continues orthopedic follow-up service, as there was no indication of surgery at the moment.



Figure 1: Wrist radiography (anteroposterior view) displaying the distal radius deformity.

DISCUSSION

The wrist deformity similar to Madelung's deformity is often associated to dyschondrosteosis, the most usual type of mesomelic dwarfism. This disturb consists in stature reduction, decrease of the medium segment of the superior and inferior extremities and Madelung's deformity. Recently, mutations in the homeobox SHOX gene, located in the pseudoautosomal 1 region of both X and Y chromosome, were demonstrated as the causers. Another associated condition includes Turner Syndrome, achondroplasia, mucopolysaccharidosis, multiple exostosis, multiple epiphyseal dysplasias and dyschondrosteosis (Ollier disease) [8].

Madelung's deformity typically consists in a volar subluxation of the hand, containing a distal

ulna prominence and volar and ulnar angulation of the distal radius. Usually, it is bilateral and affects more often girls than boys. A family history is commonly present. The deformity is generally noted in late childhood or premature adolescence with reduction of the movement and minimum pain. As the growth occurs, the deformity gets worse in the appearance. The radiographic anomalies are observed in the radius, ulna and carpal bones (Fig. 1). The radius gets incurved in the dorsal and radial convexity, also there is a similar angulation of the distal radius articular surface. The forearm is relatively short [3].

The distal epiphysis of the radius is triangular because of the growth failure of the ulnar and volar aspects of the physis, and the premature closure of these epiphysis aspects is also common. The ulna is dorsally subluxated, the ulnar head is enlarged and the overall length is reduced. In the distal radioulnar joint, which usually moves apart, the carpus seems to have been ulnar and palmar subluxated.

Children with Madelung's deformity usually possess slight pain and good wrist function, what normally guarantees an initial conservative approach. The surgery must be considered in severe deformities or persistent pain and when there is ulnocarpal impact of the carpus [9]. Vickers and Nielson had reported some success in the resection of the abnormal portion of radial physis and fat insertion as a kind of prophylactic surgery. In their group, all the 17 patients had pain relief or no progression [10].

Carter and Ezaki had recommended the isolated excision of the Vickers' ligament in such young patients or in combination with a distal radius osteotomy in cupola in case there is a considerable deformity [11]. The cupola osteotomy tends to supply a better volar covering to the semilunar and corrects some of the ulnar positive variations. The ulnar shortening can be posteriorly necessary, whether the pain at the ulnar side of wrist persists in association with a ulnar positive variant. In their group of 23 wrists, they had noticed a Vickers' ligament in 91%, in which 10 needed ulnar shortening to relief the persistent pain at ulnar side of the wrist. Furthermore, the cupola osteotomy had

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been applied in 16 wrists with pain relief in all of them.

CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

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None.

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RESUMO

Abertura do olécrano: um achado acidental

A deformidade de Madelung é uma anomalia caracterizada pela subluxação do carpo em relação ao antebraço, isto é, uma inclinação volar progressiva da ulna em relação à superfície articular distal do rádio. Comumente, acomete mulheres e é bilateral, manifestando-se ao fim da infância. Ela pode ser causada por alterações genéticas, ou após trauma, infecção ou neoplasia. O presente relato aborda uma paciente de 13 anos que apresentou queixa de dor no punho com irradiação dolorosa para antebraço. Na radiografia, foram identificadas anomalias no rádio, na ulna e nos ossos do carpo. Em razão de sua baixa complexidade, foi optado por um tratamento conservador para alívio dos sintomas, sem previsão de procedimentos cirúrgicos. Não obstante, a cirurgia pode ser uma alternativa para correção de casos graves e alívio da dor persistente. Ela consiste na osteotomia de cúpula, a qual fornece melhor cobertura volar ao semilunar e possui elevada taxa de sucesso principalmente na resolução da dor.

Palavras-chave: ortopedia, doença de Madelung, deformidade de Madelung, punho