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MANAGEMENT OF MADELUNG DEFORMITY IN LATE ADOLESCENT COMPUTER PROFESSIONAL: INTERESTING CASE REPORT WITH REVIEW OF LITERATURE

Orthopaedics

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ABSTRACT

Introduction: Madelung deformity is a developmental progressive deformity, usually bilateral, of the distalradioulnar joint and the radiocarpal joint in which the distal articular surface of the radius faces volarly and ulnarly secondary to a growth aberration in the corresponding distal radial physis with associated proximal subsidence and triangular deformation of the carpals. It becomes clinically evident in the post pubertal growth spurt in adolescence, more common in females. Although MRI is advocated for identifying associated thickened Vicker's ligament, clinically and plain radiograph examination usually suffices. Depending on age of presentation, degree of functional impairment and pain , numerous treatment options are described. **Case Report:** 20 year old gentleman presented to the out patient department with mechanical pain in the dominant right wrist with painful and restricted movements especially supination and dorsiflexion. Plain radiographs showed features consistent with Madelung deformity and a skeletal maturity. A Sauve-Kapandji procedure was performed. Two year follow-up shows good bony union between ulnar head and radius with improvement in functional range of motion and grip strength in a painless wrist. **Conclusion:** Madelung deformity is a rare deformity which causes considerable pain and functional limitation across the wrist joint. Management should to tailored to patients symptoms and age of presentation. Sauve-Kapandji is an easy and rewarding procedure if done in correctly selected patients.

KEYWORDS

Madelung Deformity, Sauve-Kapandji, Vicker's ligament, Ulna, Surgery

INTRODUCTION

Madelung deformity can be described a rare progressive developmental deformity of the wrist joint, which constitutes 1.7% of total congenital deformities occurring in children [1]. Otto Wilhelm Madelung was the first to make a complete description this condition in 1878, with Dupuytren (1834) and Malgaigne (1834) first to note this entity [1,3,4]. It results as consequence of asymmetric growth of the volar-ulnar segment of the distal radial physis. More commonly seen in females, it most commonly presents in adolescence coinciding with the pubertal growth spurt, rarely seen before the age of 7 years [2] with the complains of wrist pain, characteristic and aesthetically displeasing wrist deformity and decreased wrist range of motion particularly wrist extension and forearm supination.

Typical Madelung deformity is associated with a thickened ligament between lunate and distal radius on volar ulnar aspect called Vicker's ligament which crosses the physis and creates a tether, restricting its growth and causing the distal radius to deform with increased radial inclination and increased volar tilt. The lunate fossa is malformed causing subsidence of the lunate and subsequent adaptive deformation of the proximal row of carpal into an inverted pyramid. The distal radio-ulnar joint is incongruous and the ulna subluxates dorsally & laterally creating an aesthetically displeasing deformity. These features are easily described on a plain radiographs, with MRI helping in identifying the associated thickened Vicker's ligament and radiotriquetral ligament. Management depends upon the age of presentation and severity of symptoms and deformity, as the disease course is fairly unpredictable.

Here we present a case report of unilateral Madelung deformity entailing diagnosis, surgical management, two year follow-up and literature review of this rare entity.

CASE REPORT

A 20 year old gentleman presented to the outpatient department with mechanical wrist pain and limitation of supination with extension of dominant right side especially during sports activities since more than two years. He also complained of displeasing appearance of wrist. Physical examination revealed palmar subluxation and associated prominence of right ulnar styloid process, with marked pain on passive pronation. Piano sign for DRUJ instability was positive. Ulnar grind and scaphoid shift test was negative. Right wrist palmar- and dorsiflexion were 70 ° and 50 ° respectively; supination and pronation were 50° and 60° respectively. Plain radiography on anteroposterior view revealed increased radial inclination and radial height, V-shaped proximal carpal row, shorted ulna. Lateral view demonstrated dorsal subluxation of ulna and palmar shift of carpals (Figure 1). A Sauve-Kapandji procedure for arthrodesis of Distal Radio-UlnarJoint for creating a pseudopseudoarthrosis of distal ulna was planned.

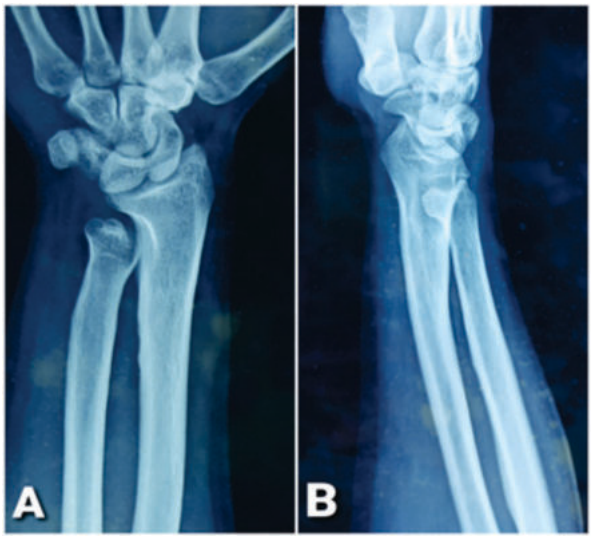

Figure 1: Preoperative plain radiograph of dominant right wrist. Anteroposterior view (A) showing increased radial inclination and radial height with V shaped wedging of proximal row of carpals. Lateral view (B) showing dorsal subluxation of ulna with palmar shift of proximal carpal row.

Under regional anaesthesia, a 10 cm incision (Figure 2.A) was taken cantered over the extensor digiti minimi. Dorsal sensory branch of ulnar nerve was identified and retracted. Extensor carpi ulnaris along with its sheath was identified and retracted ulnarly (Figure 2.B). Fifth extensor compartment was opened and EDM retracted laterally. The dorsal DRUJ capsule was opened longitudinally. With forearm in pronation the joint cartilage and subchondral bone of ulnar head facing the surgeon as well as the sigmoid notch of the radius was removed. The ulnar head is osteotomized just proximal to where the joint cartilage ends and fixed to the radius with 2 guide pins (Figure 2.C). A 5

mm segment of ulna proximal was osteotomized with an osteotome and excised. The ulnar head was fixed to the radius with two 4.5mm cancellous cannulated screws. Wrist was immobilised in a below elbow splint for 6 weeks. The patient was encouraged to do pronosupination and active cexercises of the wrist.

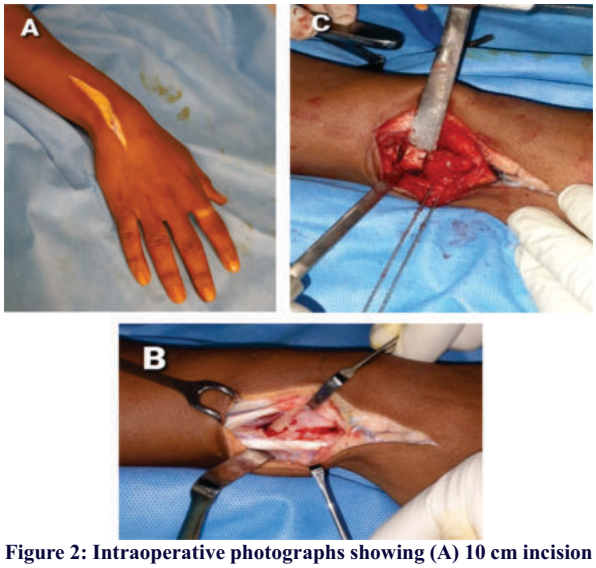

Figure 2: Intraoperative photographs showing (A) 10 cm incision centered over extensor digiti minimi (B) extensor carpi ulnar is retracted ulnarly and EDM retraced radially to exposed underlying distal ulna (C) 5 mm osteotomized ulnar segment proximal to DRUJ with ulnar head fixed to radius with two guidepins.

Two year follow up shows good union between ulnar head & radius with no evidence of instability (Figure 3) , significantly increased and painless range of motion at wrist (Figure 4) and improved aesthetics.

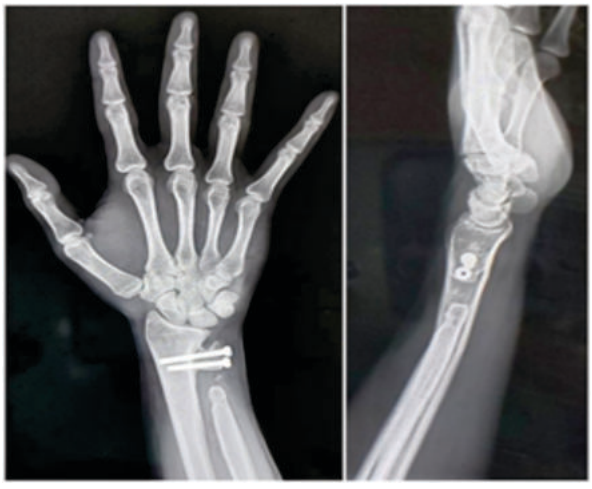

Figure 3: Two year follow up plain radiographs shows good union between ulnar head and radius evidenced by painless pronosupination and flexion-extension across the wrist.

Figure 4: Postoperative clinical photographs at two year follow up showing adequate functional range of motion at wrist.

DISCUSSION

Madelung deformity is a rare, complex, autosomal dominant entity with a incomplete penetrance of 40% and has also been described as dychondroplasia, osteochondrodystrophy and hemiatrophy of distal radial physis[1,7,8,9]It is typically bilateral (74%) [5], although in our case it presented as a unilateral deformity. With a female preponderance (M:F= 3-5:1) it presents in the age group of 6 – 13 years though some authors claim the presentation to be in mid to late adolescence linked to adolescent growth spurt [6].

Henry and Thorburn described four etiologic factors for MD: idiopathic, dysplasia, genetic, post traumatic [11]. The most dysplasia associated with MD is Leri-Weil dyschondrosteosis , which is characterised by mesomelia, short stature due to mutation in SHOX gene. Other dysplasias include Ollier's disease, achondroplasia, mucopolysaccharidosis, pseudohypothyroidism[10]. Turner's syndrome is frequently involved with genetic forms among others. Post traumatic MD results following multiple, repetitive insults to the physis over a prolonged period of time classically seen in gymnasts. Our case falls in the idiopathic group due to absence of family history, history of offending activity or trauma and no other developmental anomaly.

MD can be easily diagnosed by clinical and radiologic examination. MRI has been advocated by Vicker's and Nielsen [13] for identifying the offending Vicker's ligament and RT ligament, changes in TFCC and pronator quadratus but not necessary in every case. Increased radial inclination and volar tilt, short radius with dorsolateral bowing, positive ulnar variance with dorsolateral subluxation or dislocation of ulna, proximal migration of lunate with triangular arrangement of wedge shaped proximal carpals accompanied by volar suluxation [9]. Symptoms can stretch along the entire spectrum ranging from pain in wrist, decreased and painful movements of wrist including supination & pronation, cosmetically unappealing deformity, carpal tunnel syndrome, extensor tendinopathy and rupture. MD has a progressive and unpredictable course. Therefore treatment in most cases has to be individualised according to the age of presentation, severity of symptoms and deformity. Skeletally mature patients with mild pain and no functional limitation can be managed conservatively with analgesics, activity modification and physiotherapy with serial X-ray monitoring. Whereas surgical treatment is preferred in adolescents and young adults with severe pain, disabling functional impairment and bayonet shaped deformity of hand. Surgical techniques can be classified into one of the 3 groups: first group includes procedures involving radius alone like epiphysiolysis and corrective osteotomy; second group encompasses reconstruction of only the ulna which include epiphysiodesis, ulnar head excision, distal ulna resection with or without fusion of the DRUJ; the third group comprises combination of above techniques [12]. These techniques are often accompanied by soft tissue procedures like excision of Vicker's ligament, selective carpal denervation. Our case was a 20 year old skeletally mature, right dominant, computer professional involving with painful and reduced grip strength, restricted dorsiflexion and unappealing deformity. He was planned for a Sauve-Kapandji procedure for arthrodesing the DRUJ and creation of a pseudoarthosis in ulna.

Sauve-Kapandji is a relatively easy procedure to perform than radial osteotomy-which requires preoperative planning and surgical expertise along with a longer healing time. SK directly addressed the ulnar head deformity with stabilisation of DRUJ and radiocarpal joint to some extent thereby improving movement across the wrist joint and grip strength. A Eid et al [14] in his case series of 13 wrists with idiopathic Madelung treated with Sauve-Kapandji reported a good rate of union of ulnar head to radius with a mean period of 10.3 weeks and complete alleviation of pain at the end of a mean follow up period of 16.2 months. L C Angelini et al [15] over a period of nine years managed 15 wrist with MD in adolescents in the age group of 16-23 years and presented a favourable outcome with improved grip strength and aesthetics with elimination of pain.

Our patient at a two year follow up, had significant improvement in wrist motion, particularly in degree of extension and pronation which increased to 80°, which greatly benefitted his vocation as a computer professional. He had a painless wrist, improvement in daily activities like holding a glass, eating and was satisfied with aesthetic appearance of his wrist.

CONCLUSION

Madelung deformity should be managed surgically in young

adolescent growing age group especially if associated with significant pain, functional impairment along with appealing cosmesis. In skeletally mature patients with very less or completed growth potential, Sauve-Kapandji procedure provides a technically easy and rewarding alternative to osteotomy with adequate clinical benefits.

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Declaration
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Ethical Approval: Ethical clearance was given by the Ethical committee of Government Medical College and Hospital, Nagpur, Maharashtra, India

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