Bangladesh Journal of Neuroscience 2011; Vol. 27 (2) : 101-104

CASE REPORTS

Madelung Deformity - A Case Report with Literature Review.

MD. RAFIQUL ISLAM [1], MOHAMMAD NAJIM UDDIN[2], HASAN ZAHIDUR RAHMAN[3]

Abstract:

Madelung deformity (MD) is a rare autosomaldominant disordercharacterized by epiphyseal growth plate disturbance in distal ulnar sidewhereina progressive ulnar and volar tilt of itsarticular surface occurs in associationwith a dorsal subluxation of the distalulna.Diagnosis of the diseaseis by correlation of clinical data with radiological findings and genetic transmission.In this study, radiographic findings ofa 16 years old female patient progressive bilateral wrist joint deformity and bowing of distal end of forearm without significant functional impairment and systemic complaints and diagnosed as Primary Madelung deformity, is reported.

Keyword: *Wrist Joint, Deformity, Bilateral, Madelung*

Introduction:

Madelung deformity (MD) is a rare inherited disorder involving the epiphysealgrowth plate of distal radial end and consequent deformity as unaffected radial and dorsalportions of the growth plate continue to grow, accounting for 1.7% of all congenital hand anomalies.[1]It can be bilateral in 50 - 66% of patients[2]. It often occurs as rare congenital deformity and does not usually manifest until 10 - 14 yearsand isprimarily found in females[2-4]. One third of cases of MD are transmitted in an autosomal dominant fashionwith variable penetrance. It may also be seen as an acquired consequence of trauma to the growth plate (e.g. Salter V fracture). Henry and Thorburn classifiedMD into 4 different etiologic groups: Posttraumatic, Dysplastic, Chromosomal or genetic (Turner syndrome), andIdiopathic or primary[4]. The underlying cause of this is unclear, with possibilities including local vascular insufficiency trauma, infection (osteomyelitis)or muscular disorders. And sometimes MD associated with Leri-Weill syndrome(autosomal dominant dyschondrosteosis & mesometric dwarfism), Turner syndrome, nail-patella syndrome, diaphyseal aclasis (hereditary multiple exostosis), Hurler mucopolysaccharidosis, achondroplasia.Madelung deformity was first described Malgaigne in 1885 and later in 1878 by OttoWilhelm Madelung, German surgeon (1846 - 1926) as “Spontaneous forward subluxation of the hand”[5-9]. It was defined in terms of radiological findings andgenetic transmission. Prompt and correct diagnosis is very important in orderto ensure correct treatment and to provide the necessary consultationservices to families. Herein, we present a case of MD in a teenage girlpresented with progressivebilateral wrist joint deformity and bowing of distal end of forearm without significant functional impairment and systemic complaints and that was diagnosedwith radiological and clinic findings, and typed by literature review.

Case Report:

A 16-year-old Muslim girl, second of two siblings of consanguineous birth to normal parents, hailing from Korban Ali’s Bari, Jamalpur, ShenbaghThana, Noakhaliand presented withshort stature and progressive bilateral wrist deformities.

She was alright 1 year back, then she developed gradual onset, progressive, painless dinner fork disfiguration of right wrist with associated outward bowing of right forearm and little bits difficulties in writing& gripping.Now for last 5 monthsit’s also involved her left hand with sparing spine & lower limb.

She had no joint pain, morningstiffness, sensory complains and no history of preceding trauma or surgery or delay in achieving mile stone of development.

1. Professor, Department of Neurology, Bangabandhu Sheikh Mujib Medical University, Dhaka.
2. Resident, Department of Neurology, Bangabandhu Sheikh Mujib Medical University, Dhaka.
3. Associate Professor, Department of Neurology, Bangabandhu Sheikh Mujib Medical University, Dhaka.

Fig.-1: *Showing mesomelic shortening of both upper limbs with lumbar lordosis.*

She was well cooperative and mentally & emotionally stable with normal vital signs. She was 141 cm tall with US: LSratio of 0.9 and total arm span 113 cm & OFC 53 cm.

She had mesomelic shortening of both upper limbs with lumbarlordosis (Fig. 1). The limbs showedwideningof wrist with garden spade deformity, fixed pronator deformity & bowing andbilateral cubitus valgus with normal warmth, bulk of muscle& no reddening & tenderness on palpation (Fig. 2). There was no limitation inmovement of joints& spine andgait with normal other systemic examination without any organomegaly.

X-ray of hand and wrist were diagnostic of Madelung deformity with lunate subsidence 22 mm (normal < 4 mm), lunate fossa angle 45^0 (normal < 40^0), palmar carpal distance 21 mm (normal < 21 mm) (Fig. 3,4). The present case had ulnar tilt 11^0 which is not fit radiologic criteria of Madelung deformity (e"33^0 in Madelung deformity).

All routine investigations were within normal limit. Some second line investigations also performed and the findings were within normal limit like Serum

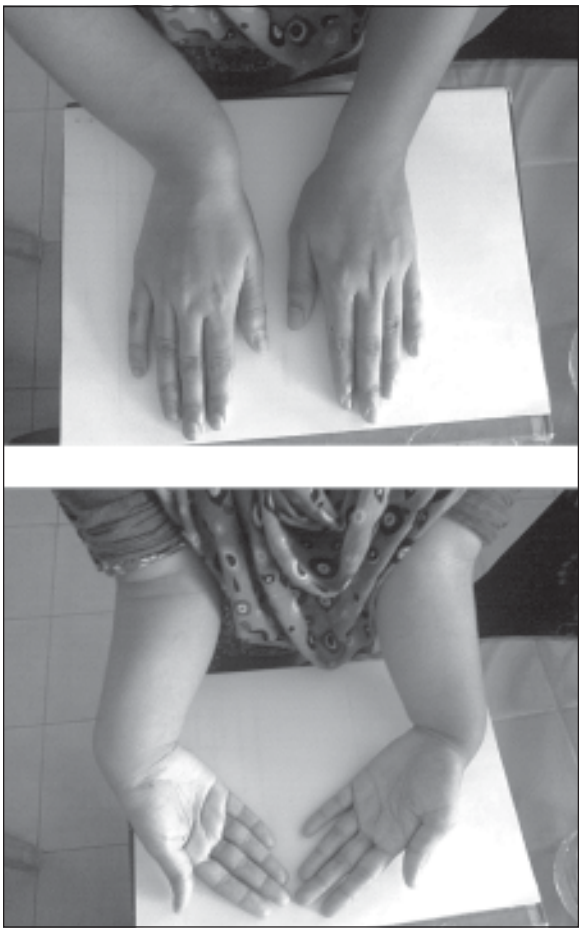

Fig.-2: *Showing widening of wrist with garden spade deformity with radial bowing &bilateral cubitus valguspredominantly in right than left.*

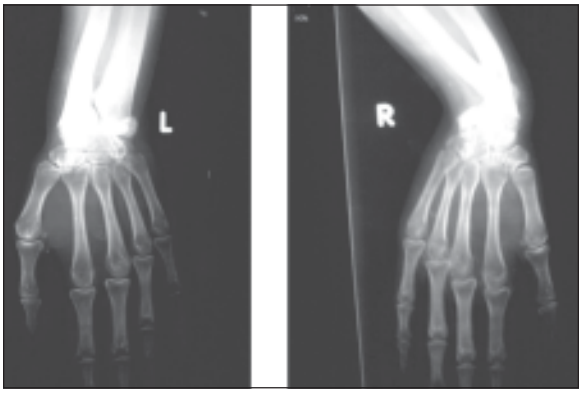

Fig.-3: *X-ray of hand and wrist shows lunate subsidence 22 mm, lunate fossa angle 450 which are diagnostic for Madelung deformity.*

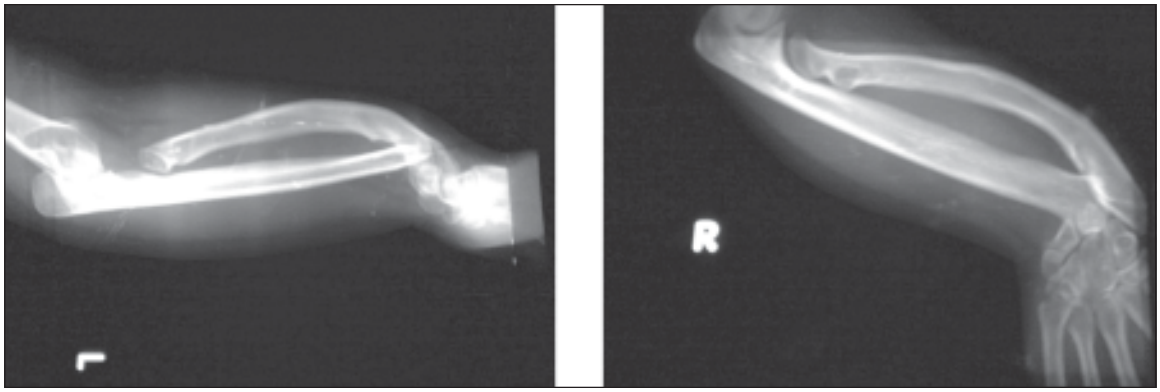

Fig.-4: *X-ray of hand and wrist shows palmar carpal distance 21 mm and dorsal & radial bowing of radius with exaggerated radial inclination.*

calcium 8.7 mg/dl, SerumInorganic Phosphate 4.5 mg/dl, Serum Alkaline Phosphatase 70 U/L, Serum PTH 51.4 pg/ml, normal Ultrasonography of whole abdomen without any organomegaly and normal thyroid status with Serum TSH 1.51 µIU/ml, F T_4 1.18 ng/dl & F T_3 3.14 pg/ml.

She was diagnosed as a case of Madelung deformity of rightwrist and initiation of similar deformity in left wrist.Orthopedics consultation was advised and preparation of surgery was undertaken. But the patients and attendant didnot give consent for operative procedure anddischargedfrom hospital with her own deformity.

Discussion:

The Madelung deformity is relatively uncommon, with prevalence less than 2% and female predominance with female: male ratio is 3 to 5[10,11].

Typically, the deformity is presentbilaterally and seldom manifests clinically before the age of7 years and typically middleto late adolescent onset of the disorder may be linked to theadolescent growth spurt[2-5].

The long-standing and progressive radial deformity graduallyworsens until it is suddenly exacerbated by the increasedgrowth rate, often occurring concurrently with a prematurephysical fusion.The present case had support the above statements.

Radiographic features of MD are characterized by[12,13]

1. dorsal and radial bowing of the radius
2. exaggerated palmar (up to 35°) and ulnar tilt (up to 60°) of the radio carpal articulation 3
3. failure of ossification of the ulnar side of the distal radial epiphysis
4. exaggerated radial inclination
5. Decreased carpal angle below 118°; normal from 118° to 139°.
6. carpal subluxation in a palmar and ulnar direction
7. lunate is gradually forced to the apex of the V-shaped radioulnaocarpal joint
8. "V-shaped" proximal carpal row = herniated proximal carpal row.
9. dorsal subluxation of the distal ulnar and positive ulnar variance
10. wedging of the carpus between the radius and ulna

Management is usually conservative.Persistent pain and/or severe deformitycall for orthopedic surgery involving radialosteotomy. In addition,ulnar shorteningin skeletally immature patients orexcision of distal ulnar head in the skeletallymature is done. Surgical prophylaxisby resection of the abnormal part ofdistal radial epiphysis and its replacementby autologous fat (also known asphysiolysis) have recently been shown torestore growth and minimize deformity.[14-17]In our patients she was asymptomatic but herdeformity was rapidly developing and also creates socialstigma. That's may be the reason why orthopedicsdepartment wanted to operate on her.

Conclusion:

Madelung deformity is a rare condition that affects thestructure and function of the wrist.one third of disease is transmitted as autosomal dominant fashion and is commonly linked toseveral heritable factors& sometime local traumamay also be responsible. Conservative approach in asymptomaticpatient is the key but improved surgical technique forsymptomatic patients with mature bone are rewarding now a days.

Conflict of Interest: None

References:

1. Jobe MT, Wright PE. Congenitalanomalies of hand. In: Compbell's OperativeOrthopedics, 8th Edn. Ed.Crenshaw AH. New York, Mosby YearBook, 1992, pp 3419-3421.
2. Anton JI, Reitz GB, Speigel MB. Madelung's deformity.Ann Surg. 1938;108(3):411-39.
3. Wheeless' Textbook of Orthopaedics, Clifford R. Wheeless,III, MD.
4. Henry A, Thorburn MJ. Madelung's deformity. A clinicaland cytogenetic study. J Bone Joint Surg Br. Feb 1967;49(1): 66-73.
5. O. W. Madelung "Die spontane Subluxation der HandnachVorne." Verhandlungen derdeutschenGesellschaftfürChirurgie, Berlin, 1878, 7: 259-276.
6. Otto Wilhelm Madelung fromwhonamedit.com, thedictionary of medical eponyms. Otto Wilhelm Madelung
7. Feliciani C, Amerio P. Madelung's disease: inherited froman ancient Mediterranean population? N Engl J Med1999;140:1481
8. Lamb D. Madelung deformity. J HandSurg 1988,13-1:3-4.
9. Dannenberg M, Anton JI, Spiegel MB.Madelung's deformity: Considerationof its roentgenological diagnostic criteria.Am J Roentgenol Rad Therapy1939,42: 671-676.
10. Casford B. Madelung's deformity. Available at: http://brighmrad.harvard.edu/Cases/mcr/hcache/205/full.html.Accessed April 19, 2000.
11. Brashear HRJr, Raney RB Sr. Handbook of OrthopaedicSurgery. 10th ed. St Louis, MO: CV Mosby Co; 1986: 496–7.
12. http://radiopaedia.org/articles/madelung-deformity
13. UW Radiology Main Online Teaching File. Roentgenographicabnormalitie in Madelung's deformity. Available at: http://www.rad.washington.edu/ aintf/cases/unk53/answers.html. Accessed April 19, 2000.
14. Vickers D, Neilsen G. Madelung deformity:Surgical prophylaxis(physiolysis) during late growth periodby resection ofdyschondrosteosislesion. J Hand Surg (British) 1992,17B:401-7.
15. Lovell WW, Winter RB, MorrissyRT et-al. Lovell and Winter's pediatric orthopaedics. Lippincott Williams & Wilkins. (2006) ISBN:0781753589.
16. Yang JH, SohnYH, KoSY et-al. Anthropological analysis of Koreans using HLA class II diversity among East Asians. 2010
17. Golding JS, BlackburneJS. Madelung's disease of the wrist and dyschondrosteosis. J Bone Joint Surg Br. 1976;58 (3): 350-2.