Madelung Deformity—the Hallmark of Dyschondrosteosis

M. Bhat
R. Chetan
K.A. Shivashankar
M. Jayaram

Madelung deformity is an abnormality of the distal radial epiphysis wherein a progressive ulnar and volar tilt of its articular surface occurs in association with a dorsal subluxation of the distal ulna. It is a rare inherited disorder accounting for 1.7% of all congenital hand anomalies. The inheritance pattern is autosomal dominant with variable penetrance (l).

Madelung-like deformity has been reported following trauma to the distal radial epiphysis as also infections and neoplasia. However, the term 'Madelung deformity' should preferably be confined to the congenital disorder which is also known as Dyschondrosteosis.(2). Dyschondrosteosis is an inherited dysplasia of the bone characterized by bilateral Madelung deformity with mesomelic dwarfism of both upper and lower limbs. It is. the commonest variety of mesomelic dwarfism(3).

Madelung deformity may also be seen associated with diverse disorders such as mucopolysaccharidosis, Turner's syndrome, achondroplasia, dyschondroplasia (Ollier's disease), multiple exostosis and multiple epiphyseal dysplasia. Madelung's original description of the lesion befits its restriction to Dyschondrosteosis (2).

Case Report

An 11-year-old Muslim girl, second of five siblings of non-consanguienous birth to normal parents presented with short stature and progressive bony deformities. She was 98 cm tall with US: LS ratio of 0.9. She had mesomelic shortening of all limbs with a short neck, pectus carinatum, kyphoscoliosis and lumbar lordosis (*Fig. 1).* The limbs showed widening of wrist, elbow and ankle joints, bilateral cubitus valgus, coxa valga and genu varum. There was limitation in movement at hip and knee joints and gait was of waddling type. X-ray of hand and wrist were diagnostic of Madelung deformity *(Fig. 2).*

Discussion

Dyschondrosteosis (Syn. Madelung's disease(3), Leri-Weill's syndrome) is the commonest type of mesomelic dwarfism and is associated with bilateral Madelung deformity. It is often seen in girls and becomes clinically apparent by late childhood or early adolescene(4). Madelung deformity was first described by Malgaigne in 1885 and later by Madelung in 1878 as "Spontaneous forward subluxation of the hand."(2).

From the Departments of Pediatrics Sri Devaraj Urs Medical College, Tamaka, Kolar 563 101, Karnataka

Reprint requests: Dr. Ravi Chetan, Associate Professor of Pediatrics, 613, 11th cross 2nd Main, J.P. Nagar-Phase III, Bangalore 560 078

Received for publication: August 1,1994;
Accepted: November 28,1994

Danneberg *et al.(5)* reviewed 172 cases and outlined 12 radiological criteria for the diagnosis of Madelung deformity: (a) Lateral and dorsal bowing of the radial diaphysis; *(b)* Widening of the interosseous space due to *(a)* above; (c) Shortening of the radius in comparison to other bones and to normal standards for age; *(d)* Articular surfaces of distal radius is altered to face in ulnar and palmar direction; *(e)* Premature fusion of the ulnar half of the lower radial epiphysis, accentuating the deformity in early adolescence; (f) Dislocation/ subluxation of the inferior radio-ulnar joint shifting the distal ulna dorsally; *(g)* Increased bone density proximal to the abnormal radio-ulnar fusion line; *(h)* Exostosis along the inferior ulnar border of the radius: (i) Triangularization of the normally quadrangular shaped outline of the distal radial epiphysis with the apex pointing medially; (j) Hypercondensation of trabeculations of the ulnar head; *(k)* Wedging of the carpal bones between the abnormal distal radioulnar joint giving them a triangular configuration with the lunate at its apex; and (l) An arched curvature of the carpal bones in continuation with the arch of the dorsally bowed radial diaphysis. Criterias (c), (i) and *(k)* are essential for diagnosis whereas *(g), (h)* and (j) are secondary phenomena(5). The present case had all criteria except *(h)* and (j).

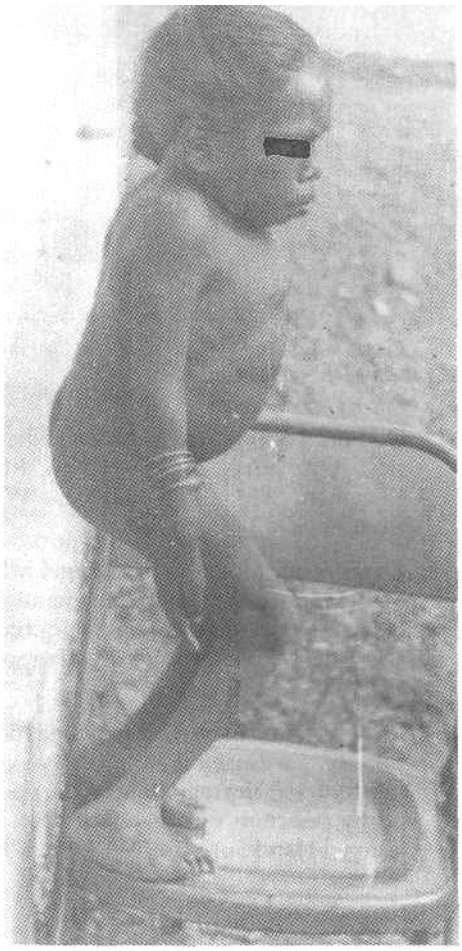

Fig. 1. Clinical photograph of the patient

Management is usually conservative. Persistent pain and/or severe deformity call for orthopedic surgery involving radial osteotomy. In addition, ulnar shortening in skeletally immature patients or excision of distal ulnar head in the skeletally mature is done. Surgical prophylaxis by resection of the abnormal part of distal radial epiphysis and its replacement by autologous fat (also known as physiolysis) has recently been shown to restore growth and minimize deformity(6).

Acknowlegement

We thank the Distric Surgeon, S.N.R. Hospital, Kolar for allowing us to publish this case report.

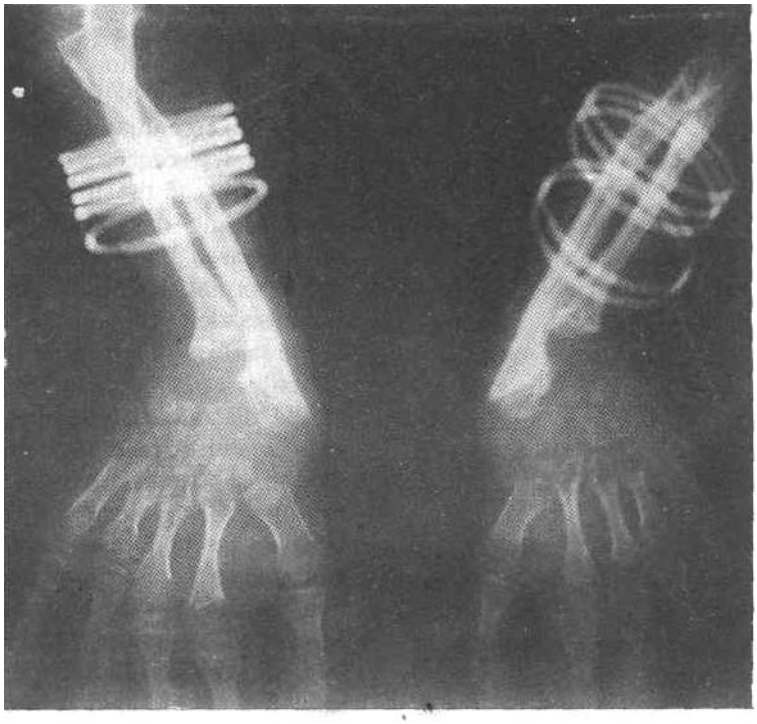

Fig. 2. X-ray of wrist PA view showing the madelung deformity.

REFERENCES

1. Jobe MT, Wright PE. Congenital anomalies of hand. *In:* Compbell's Operative Orthopedics, 8th Edn. Ed. Crenshaw AH. New York, Mosby Year Book, 1992, pp 3419-3421.
2. Lamb D. Madelung deformity. J Hand Surg 1988,13-1:3-4.
3. Langer LO. Dyschondrosteosis, a heritable bone dysplasia with characteristic roentgenographic features. Am J Roentgenol Radio Therapy 1965, 95: 178-188.
4. Sillence DO. Genetic skeletal dysplasias. *In:* Nelson Text Book of Pediatrics, 14th edn. Eds. Behrman RE. Kliegman RM. Philadelphia, WB Saunders Co, 1992, pp 738.
5. Dannenberg M, Anton JI, Spiegel MB. Madelung's deformity: Consideration of its roentgenological diagnostic criteria. Am J Roentgenol Rad Therapy 1939,42: 671-676.
6. Vickers D, Neilsen G. Madelung deformity: Surgical prophylaxis (physiolysis) during late growth period by resection of dyschondrosteosis lesion. J Hand Surg (British) 1992,17B: 401-407.