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Madelung Deformity: A Case Report of a Young Nigerian Man

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Abstract
The purpose of this article is to emphasise the influence of cultural orientation as a factor that could lead to delays in the diagnosis of some wrist deformities. This is important in improving the need for detailed clinical evaluation and judicious radiological investigation of wrist deformities so as to prevent undue physical, economic, and artistic losses among patients with such conditions. Madelung deformity involves the arrest of growth at the medial distal end of the radius, resulting in abnormal wrist alignment and angulation. It occurs more frequently in women than in men, and cases remain underreported despite the possibility of diagnosis using conventional radiography. A 27-year-old male patient presented with recurrent wrist pain and inability to use the right wrist. Clinical examination indicated the presence of a deformity affecting the right wrist. A right forearm X-ray, including the elbow and wrist joints in anteroposterior and lateral views, demonstrated lateral bowing of the radial shaft, widening of the interosseous space, a radial inclination angle of 34.68°, and positive ulnar variance. These findings were consistent with Madelung deformity. This case report highlights the value of obtaining a comprehensive clinical history, including relevant cultural considerations, alongside a thorough physical examination when diagnosing Madelung deformity. It also highlights the value of the early and judicious use of X-ray imaging as a diagnostic tool, even in low-resource settings. Early diagnosis facilitates timely conservative or surgical management, which is likely to result in better clinical outcomes while minimising economic and artistic losses for the patient. Furthermore, this report adds to the existing body of knowledge and contributes to promoting the early diagnosis and specialist management of Madelung deformity in low-resource settings through the effective use of conventional radiography.

Key terms: Artistic losses, case report, interosseous space, Madelung deformity, Roentgen, ulnar variance.

1.0 INTRODUCTION

Madelung deformity is an abnormal deformity of the wrist joint with increased angle of inclination, volar tilt of the distal radius, and positive ulnar variance. The underlying deformity could affect the economic well-being of such patients. It is under-reported in developing countries, of which Nigeria is one. This could be attributed to a lot of factors, which include poor data keeping, dearth of medical specialists, and limited availability of diagnostic equipment, especially in rural areas. The poor availability of data has affected the incidence and prevalence of the deformity in low-resource settings. Therefore, there is a need to improve the authenticity of data on such deformities, even in low-resource settings. This case report would undoubtedly add to the body of knowledge on this type of deformity.

The aetiology of wrist deformity is generally accepted to arise from premature closure of the medial volar aspect of the distal radial physis (Ghatan & Hanel, 2013). The consequence of premature closure of the distal medial aspect of the radius would lead to an increased angle of inclination, volar tilt of the distal radius, and positive ulnar variance. The palmar inclination angle is obtained on the lateral radiograph by the horizontal line drawn from the styloid process of the radial shaft, which is perpendicular to the long axis of the radius. The second line is then drawn by joining the volar and dorsal aspects of the radius along its articular surface. The angle between the horizontal and the line connecting the volar and dorsal aspects of the radius is the palmar inclination angle. The angle is 10°–25°. Variance is the relationship between the articular surfaces of the radius and ulna at the radiolunate articulation. It is categorised into positive ulnar variance when the ulnar articular surface is distal to the radius, negative when it is proximal to the radius, and neutral when both are at the same level. It is accepted that the radial styloid process is 9–12 mm distal to the articular surface of the ulna on a posteroanterior radiograph.

The official description of the wrist abnormality was first provided by Otto Madelung in 1878 (Arora & Chung, 2006). There is poor documentation of the deformity in developing countries. The diagnosis could easily be missed, especially in poor economic settings where documentation and clinical and radiological features were not reliably available. It is more common in adolescent females, showing a predominance of 4:1 and representing less than 2 per cent of all paediatric hand deformities (Ali et al., 2015). It usually affects both wrists (Schmidt-Rohlfing et al., 2001) but can also be unilateral. About eighty per cent of deformities appear between the ages of 11 and 15 years (Golding & Blackburne, 1976).

2.0 CASE PRESENTATION

A 28-year-old Nigerian male presented with an inability to use the right upper limb, particularly the hand. The patient attributed the onset of symptoms to a perceived spiritual attack at the age of eight years. A spiritual solution has been sought by the parents from the onset of the symptoms. Recent recurrent pain led to the need for orthodox treatment by the patient.

Examination revealed a well-hydrated young man, orientated in time, place and person. Musculoskeletal examination revealed deformity of the right wrist joint and abnormal contracture deformity of the hand. An X-ray of the right forearm, including the wrist and hand, was done on anteroposterior and lateral views. There is a deformity of the wrist joint. There is lateral bowing of the radial shaft with widening of the interosseous space. The radial inclination angle is increased, measuring 34.68° (normal 10-25°). The radial length measures 14.1 mm (normal 9- 12 mm). There is positive ulnar variance. The proximal and volar

migration of the lunate showed the characteristic triangulation of the fused proximal carpal bones. An assessment of Madelung deformity was made for further orthopaedic specialist review and management.

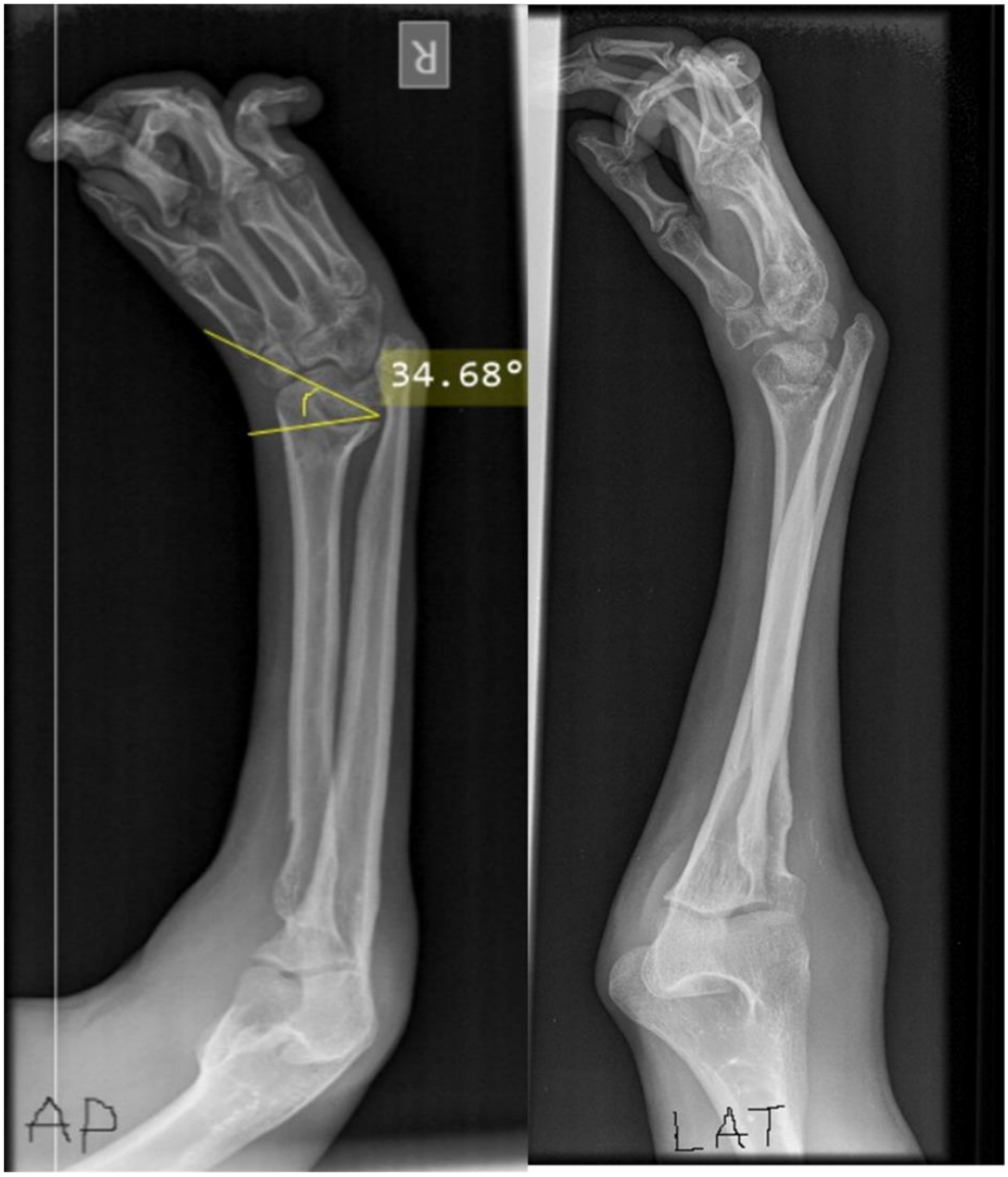

Figure 1: AP and Lateral Radiograph Showing the Right Forearm and the Wrist

Figure 1 shows a deformity of the wrist joint. On the anteroposterior view, the distal end of the radial diaphysis shows a volar inclination of the articular end, widening of the interosseous space, and radial inclination of the bones. The lateral radiograph shows the increase in radial inclination, bayonet deformity and positive ulnar variance. The proximal and volar migration of the lunate showed the characteristic triangulation of the fused proximal carpal bones. There is a flexion deformity of the fingers.

Verbal consent was taken for the usage of the images.

3.0 DISCUSSION

The index case is a twenty-eight-year-old young adult Nigerian male with limited ability to use the right hand, which had been noticed since he was eight years old. The recent recurrent pain eventually prompted the patient to seek orthodox care after years of unorthodox management. Clinical examination revealed a young adult male with deformity of the right wrist and hand. Radiological evaluation with X-ray demonstrated lateral bowing of the radial shaft with consequent widening of the interosseous space, increased inclination angle, and positive ulnar variance. The radiological features are indicative of Madelung deformity of the wrist.

The wrist is one of the joints of the human body that contributes to functional and daily activities. It has simple mechanical and mobility functions, being a synovial ellipsoid joint that articulates with several bones, including the distal radius, the scaphoid, the lunate, and the triquetrum carpal bones.

Some important anatomical measurements are relevant to determining the anatomical alignment of the wrist joint. Among these are the anatomical variations of the ulnar and radial lengths. The articular ends of the radius and ulna should be at the same level of the distal radioulnar joint. The variation in the articular surfaces of the distal radius and ulna gives rise to ulnar variance. The distal extension of the ulnar articular surface beyond the radial surface refers to positive ulnar variance, whereas when the ulnar surface is proximal to the radial articular surface, it is negative ulnar variance. The styloid process of the radius is distal to the ulnar articular surface by 9–12 mm on a postero-anterior radiograph. Ulnar slant of the radius is the angle made by the distal articular surface of the radius with respect to a postero-anterior radiograph, which is 15°–25°. The ulnar slant of the radius increases in Madelung deformity, and the minimum angle of 33° is a criterion described by (McCarroll et al., 2005).

The premature closure of the distal aspect of the radial physis, specifically the volar and medial part, is widely accepted as the cause of Madelung deformity. There is a resultant increase in the curvature of the distal radius with subsequent palmar subluxation of the hand in relation to the distal radioulnar joint (Samuel et al., 2019).

Recent extensive studies on genetic and associated deformities have widened knowledge of the aetiopathogenesis of this deformity. Congenital Madelung deformity is now believed to be associated with a mutation or absence of the short stature homeobox (SHOX) gene in the pseudoautosomal region of the sex chromosomes (Benito-Sanz et al., 2005). The mutation is known to be associated with other syndromes and deformities, including Leri-Weill dyschondrosteosis, Turner syndrome, and some cases of idiopathic skeletal abnormality (Plafki et al., 2000). Extensive studies have also shown that there is a ligament called the *Vickers ligament*, located on the volar aspect, which tethers the lunate to the radius and is hypothesised to hinder growth by compressing the epiphyseal plate (Vickers & Nielsen, 1992).

The summary of the aetiological factors was put together into four categories: post-traumatic, dysplastic, genetic, and primary (Henry & Thorburn, 1967). The residual progressive deformity gave rise to complaints by the patients, including pain, poor range of movement, deformity, and even affected the artistic and physical activities of the patients.

The case study underscores the role of X-ray imaging in diagnosing this deformity in low-resource settings. The management of Madelung deformity is multivariate in most instances. Conservative and

surgical management depend on the severity of the deformity. Conservative management includes the use of drugs, especially non-steroidal anti-inflammatory agents (NSAIDs), and modification of the patient's activities. Surgical intervention is recommended for patients with severe pain, restriction of movement, and cosmetic reasons.

Conclusion: A case of Madelung deformity in a young adult male is presented, in which delayed diagnosis and intervention contributed to functional disability. The case underscores the importance of early detection and timely management in improving patient outcomes.

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