Case Report

A case report of congenital bilateral proximal radioulnar synostosis in a 22-month-old child

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Abstract

Congenital radioulnar synostosis is a skeletal abnormality present at birth which may appear clinically as a mild abnormality in the early years. The diagnosis is usually delayed until functional abnormality is noticed. This is a typical case of congenital bilateral proximal radioulnar synostosis diagnosed at about 2 years despite mild upper limb abnormality noticed at birth. The rarity of this condition prompted the report of this case.

Keywords: Abnormality, congenital, radioulnar synostosis

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INTRODUCTION

Radioulnar synostosis is a skeletal condition caused by failure of segmentation between the radius and ulna and was first described by Sandifort in 1793. Radioulnar synostosis could be congenital or posttraumatic. Congenital radioulnar synostosis is bilateral in 60% of case and have no sex predilection or apparent inheritance pattern.[1] Congenital synostosis is divided into two groups based on the proximal radioulnar junction. Posttraumatic radioulnar synostosis has been classified into the following three types, depending on location: Type 1: least common, occurs in the distal forearm; Type 2: occurs in the mid-forearm; and Type 3: occurs in the proximal forearm.[2]

CASE REPORT

A 22- month-old male child who was referred to the radiology department for X-ray of the upper limbs on account of inability to fully extend both upper limbs noticed when he was about 2 months of age. This was ignored until he was noticed to have difficulty in feeding himself. The child is the first of a 29-year-old housewife delivered through an emergency cesarean section at term. The pregnancy was uneventful. There was no history of birth asphyxia or birth trauma. He took all immunizations and had normal developmental milestone. There was no history of previous fracture or surgical intervention on both upper limbs. General examination revealed a limited extension at the elbow joint with limited forearm range in pronation and supination.

Radiographs of the upper forearm reveal fusion of the metaphyses of the radius and ulna with posterior dislocation of the proximal radius bilaterally [Figures 1 and 2].

DISCUSSION

This is a case of a bilateral congenital radioulnar synostosis. Congenital radioulnar synostosis is a skeletal condition caused by failure of segmentation between the radius and ulna and was first described by Sandifort in 1793. Congenital radioulnar

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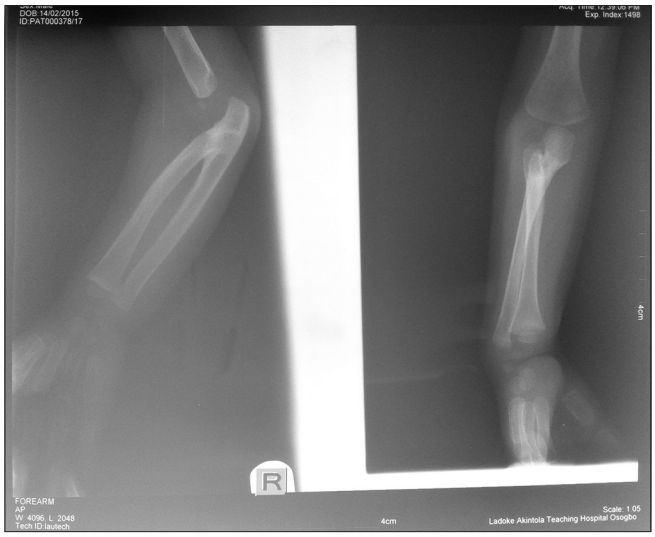

Figure 1: Anteroposterior and lateral view of the right forearm showing fusion of the proximal metaphysis of the ulnar and radius with posterior dislocation of the proximal radius

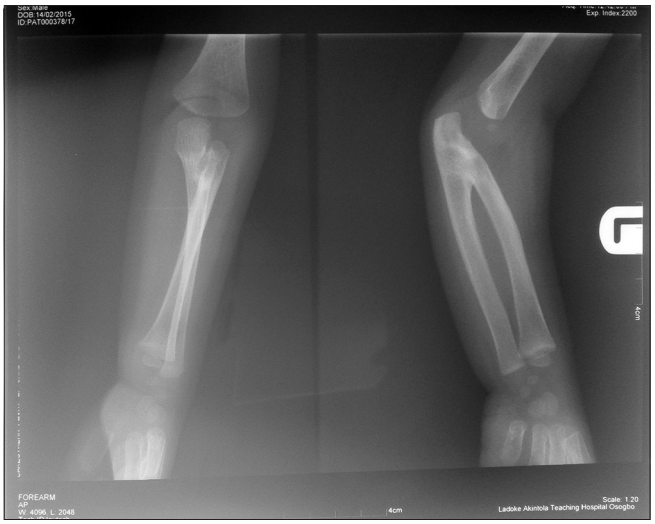

Figure 2: Anteroposterior and lateral view of the left forearm showing fusion of the proximal metaphysis of the ulnar and radius with posterior dislocation of the proximal radius

synostosis has no sex predilection and no apparent inheritance pattern and is bilateral in 60% of cases. Radioulnar synostosis could be congenital and posttraumatic.[1,2]

Congenital radioulnar synostosis occurs rarely, leading to a delayed clinical diagnosis. The average age at diagnosis is 6 years, with a range from 6 months to 22 years. This patient had mild deformity at 2 months, but it was ignored until 2 years when there was a functional limitation of both upper limbs. This implies the age at clinical diagnosis is determined by the severity. This case is bilateral with a bony bridge between the radius and ulna-bony synostosis with associated posterior dislocation of the radius.

Four types of congenital synostosis have been described by Cleary and Omer. These are (1) fibrous synostosis, (2) bony synostosis, (3) bony synostosis with associated posterior dislocation of the radius, and (4) associated anterior dislocation of the radius.[3] This index case is Type 3. Wilkie, however, reported two types of congenital synostosis, based on the proximal radioulnar junction. In Type 1, there is complete synostosis with a proximal fusion of the radius and ulna for a variable distance. Type 2 involves the fusion of the region just distal to the proximal radial epiphysis with associated radial head dislocation.[4]

Another study described congenital synostosis as a spectrum of anomalies in which the synostosis occurs in varying lengths, with or without the involvement of the radial head.[5]

Posttraumatic radioulnar synostosis has been classified into three types, depending on location: Type 1: least common, occurs in the distal forearm; Type 2: occurs in the mid-forearm, and Type 3: occurs in the proximal forearm. The features suggesting a congenital type, in this case, are the bilateralism, age at presentation, and absence of a history of trauma or surgical intervention. There is no history of trauma in this index case. Moreover, it is bilateral.

During embryological development, the upper limb bud arises from the unsegmented body wall at 25–28 days. The elbow develops at 34 days; and the humerus, radius, and ulna develop at 37 days. Initially, the three cartilaginous analogs of the humerus, radius, and ulna are connected before segmentation. Therefore, for a short period, the radius and ulna share a common perichondrium. Any insult at this time can lead to a failure of segmentation. The duration and severity of the insult determine the degree of subsequent synostosis.[6]

Endochondral ossification then proceeds, and the cartilaginous synostosis ossifies, either partially or completely, in the longitudinal or transverse plane. In the forearm, congenital radioulnar synostosis usually occurs between the proximal radius and the ulna as in this case.[7]

Although the condition is present at birth, congenital radioulnar synostosis is usually not discovered until early adolescence, when the patient presents with a lack of pronation and supination as seen in this case. Initially, the union may be cartilaginous but becomes osseous as skeleton matures and more radiographically apparent. There is an osseous union in this patient. Usually, motion between the two adjacent bones is minimal if present at all.[8]

Congenital radioulnar synostosis has associated abnormalities in about one-third of the cases. The associated skeletal

abnormalities are hip dislocation, knee anomalies, clubfoot, polydactyly, syndactyly, Madelung deformity, ligamentous laxity, thumb hypoplasia, carpal coalition, and problems of the cardiac, renal, neurologic, and gastrointestinal systems.[9,10] This patient has no associated anomaly or syndrome.

CONCLUSION

Congenital radioulnar synostosis is a rare congenital bone abnormality that is bilateral in 60% of cases and appears clinically as a mild deformity in early life but with a delayed presentation. Hence, children with mild or subtle deformity or abnormal movement of the upper limb should undergo a minimum of a radiograph of the affected limb to rule out any skeletal deformity.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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