Case Report

Congenital proximal radioulnar synostosis—a case report✩✩✩✩

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A B S T R A C T
Congenital radioulnar synostosis is a rare anomaly of the forearm with restrictions of full movement of the affected limbs. It is often seen in early childhood when they present with functional impairments. A 10-year-old female with progressive difficulty with supination in both upper limbs seen in the OPD had bilateral forearm x-ray which showed bilateral proximal radioulnar synostosis. She was then scheduled for a corrective osteotomy. Diagnostic imaging helps in evaluation and treatment planning for congenital radioulnar synostosis as seen in this index case.

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Introduction

Congenital radioulnar synostosis was first described by Sandifort in 1793. It is a skeletal abnormality caused by a failure of segmentation between the radius and ulnar. 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 and ulna develop at 37 days. The cartilaginous analogues of the humerus, ulnar 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 will determine the degree of subsequent synostosis [1–4].

Case report

A 10-year-old female was referred to the radiology department for an X-ray of both forearms (anteroposterior and lateral) on account of progressive difficulty in supination, inability to touch her back with her fingers for 9 years. Physical

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radiograph of both forearms showing proximal radioulnar synostosis.

examination showed a valgus deformity of both elbows and restricted supination range to 120°. The child was in a generally good state of health and no other concomitant malformations was seen.

AP and lateral radiographs of both forearms including the elbows was done (Figs. 1-4) and these show approximately 5 cm bony synostosis of the proximal radius and ulnar bilaterally with no elbow dislocation or radial head abnormality seen.

She was subsequently referred to the orthopedic surgeon for corrective osteotomy.

Discussion

Congenital radioulnar synostosis is rare leading to delayed clinical diagnosis. Approximately 400 cases of congenital radioulnar synostosis have been documented in literature [4]. The average age at diagnosis is 6 years with a range from 6 months to 22 years [1]—our case being 10-year old at presentation.

Four types of congenital radioulnar synostosis have been described by Cleary and Omar [5], the classification is based on the morphological appearance of the synostosis and radial head reduction with no significant prognostic value in contrast to the classification of acquired(post-traumatic) radioulnar synostosis. These are type I—fibrous synostosis, does not involve bone and is associated with a normal, articulating radial head, type II—osseous synostosis, associated with a normal, articulating radial head, Type II—osseous synostosis, with a hypoplastic and posteriorly dislocated radial head, Type IV—short osseous synostosis with an anteriorly dislocated, mushroom shaped radial head. By this classification the index case is classified as type II proximal radioulnar synostosis.

The management of congenital radioulnar synostosis could be conservative (non-operative) or operative. According to Cleary and Omar non operative is the preferred treatment.
Fig. 2 – Lateral radiograph of both forearms; normal axial relationship.

Fig. 3 – Magnified AP view of the left elbow joint demonstrating proximal radioulnar osseous synostosis.

when asymptomatic and unilateral and operative intervention is rarely indicated and based on functional deficits [5]. However, Simmons et al stated that a pronation over 60° is an absolute indication for surgery [6]. The index case is symptomatic with bilateral involvement.

Operative management include surgical separation and reconstruction techniques which was considered the ideal treatment but the final outcomes were not satisfactory [7,8]. Derotational osteotomy currently remains the most commonly performed procedure in patients with congenital radioulnar synostosis. These include derotational osteotomy at the synostosis followed by fixation with k wire and cast immobilization [9]; derotational osteotomy at the synostosis site followed by fixation with crossing k wires [10] and derotational osteotomy at the radial diaphysis and fixation with a cast only [11]. A recent approach with less postoperative morbidity is derotational osteotomy at the synostosis site followed by the plate for rigid internal fixation in case of correction loss, and plaster splint was used for external immobilization which enabled convenient close observation to monitor edema and peripheral circulation as well as ease of release [12].

The reason for late presentation despite noticeable functional impairment with difficulty in supination and radiologically evident osseous bridge might be attributed to lack of sig-
significant cosmetic concerns although no specific reason was given by the caregivers.

**Conclusion**

The rare incidence of congenital proximal radioulnar synostosis could lead to a delay in clinical diagnosis. However, diagnostic imaging plays a role in evaluation and planning for treatment by the orthopedic surgeons.

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