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Case Report

Unilateral congenital proximal radioulnar synostosis in a 4-year-old boy: A case report ☆,☆☆

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ABSTRACT

Congenital radioulnar synostosis is a rare musculoskeletal disorder of the elbow, occurring as a result of variable degree and length of the congenital fusion of the proximal radioulnar joint. Patients presents early to the hospital depending on the severity of the synostosis and its effect on elbow function. It may have psychosocial effects on the affected individuals as they grow older especially when the deformity is dramatic. Treatment may be conservative, surgical (which may have a variable degree of success) and psychotherapy.

Introduction

Congenital radioulnar synostosis (CRUS) is an uncommon musculoskeletal condition in which the 2 bones of the forearm; the ulna and the radius, are abnormally connected or fused. This restricts the arm's ability to rotate, and it is associated with diverse symptom complex. In 1793, Sandifort gave the first account of the condition, and to date, CRUS has been reported in the literature in about 400 cases worldwide

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Fig. 1 – A picture of a 4-year-old right-sided dominant boy showing both upper limbs, with an outer angular deformity of the left elbow as shown with the blue arrow. The left upper limb looked comparatively shorter.

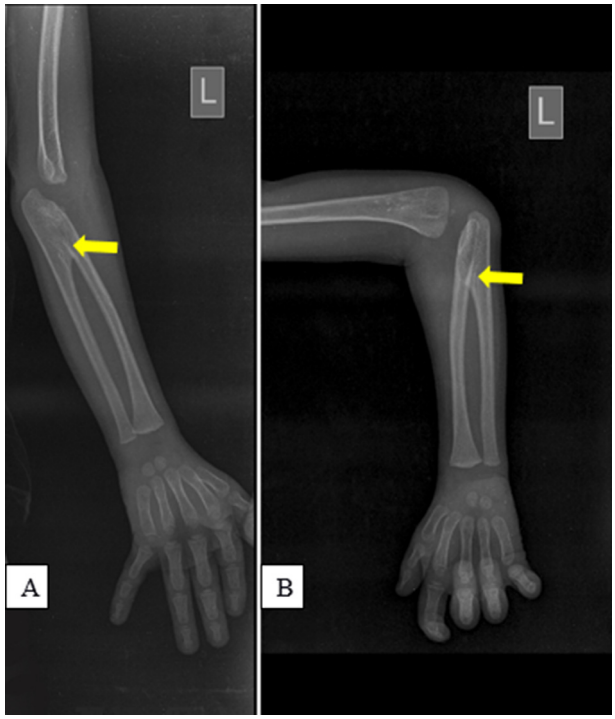

Fig. 2 – (A) AP and (B) lateral radiographs of the left elbow, radius, ulna, and hand bones showing fused radioulnar bones at the proximal fourth, an absent proximal radioulnar joint and a postero-medial orientation of the olecranon process (yellow arrows).

[1]. This X-linked dominant condition usually affects both elbows and has a paternal pattern of inheritance. However, the precise etiology is still unknown, and neither the diagnosis nor the course of treatment has been agreed upon hence, there is no gold standard for the treatment of congenital proximal radioulnar synostosis [2]. Treatment is usually conservative as surgical interventions rarely succeed. But when required, radioulnar rotation osteotomy is popular, but it comes with a high risk of ischemic muscle contracture, nerve damage, delayed union, and necessitates a lengthy period of forearm immobilization following surgery [3,4]. We present a case of a 4-year-old boy diagnosed with congenital left proximal radioulnar synostosis.

Case presentation

We report a case of a 4-year-old boy, who was brought to our radiology department by the mother for an upper limb radiograph on account of a left elbow joint deformity and inability to turn the left elbow joint to the side.

He has had this condition since birth, but it is more prominent as he grows older from the narration of the mother. There was no history of trauma and no family history of this condition. The mother said the contemporaries of her son had been teasing him with this deformity and thought this was reducing the self-esteem of the child at this age because he had been refusing to go out to play with his friends, even though he does not complain about the affected elbow to her.

On physical examination, he looked clinically well, with normal respiratory, cardiovascular, central nervous, and gastrointestinal systems. There was an obvious left elbow deformity compared to the dominant right side and an outer orientation of his left elbow during the examination of the musculoskeletal system. The affected forearm looked shorter than the dominant right one (Fig. 1). The child could not supinate and pronate but could flex and extend the ipsilateral elbow joint.

Radiographs of the left elbow, radius and ulna bones showed fused radioulnar bones at the proximal fourth, and an absent proximal radioulnar joint on the antero-posterior (AP) and lateral views, with a postero-medial orientation of the olecranon process (Figs. 2A and B). The rest of the left radius and ulna bones, distal to the defect and the distal radioulnar joint on the left looked normal but shorter compared to the right (Fig. 3). A diagnosis of a congenital left proximal radioulnar synostosis was made.

A comparative radiograph of the right upper limb showed no abnormality (Figs. 4A and B).

The child was then referred to the pediatric orthopedic unit of our hospital for further management, where a conservative management plan was started as the orthopedic surgeon decided that from literature surgical interventions are usually

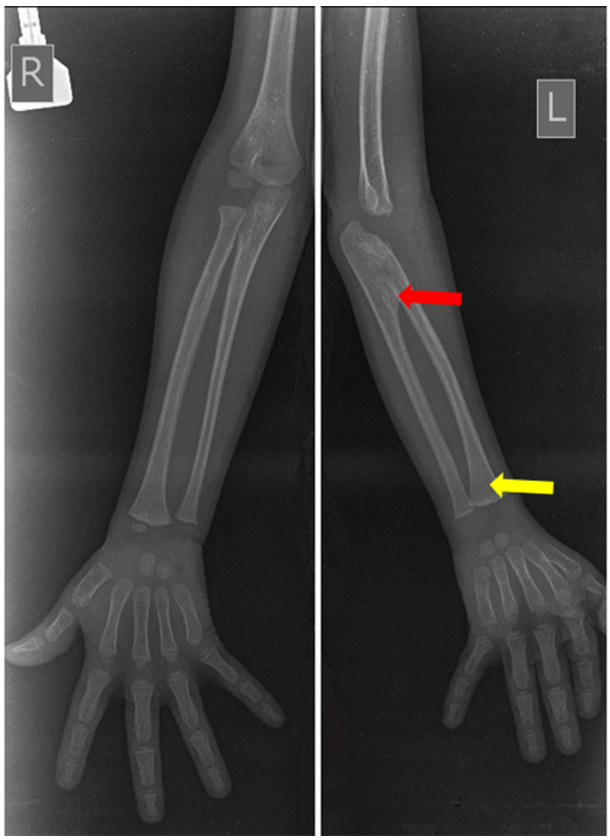

Fig. 3 – AP radiographs showing both elbows; radius and ulna bones; and both wrists and hands bones with a comparatively shorter left radius and ulna. It also shows fused proximal quarter (red arrow) but normal distal 3 quarters of the left radius and ulna bones with a normal left distal radioulnar joint (yellow arrow).

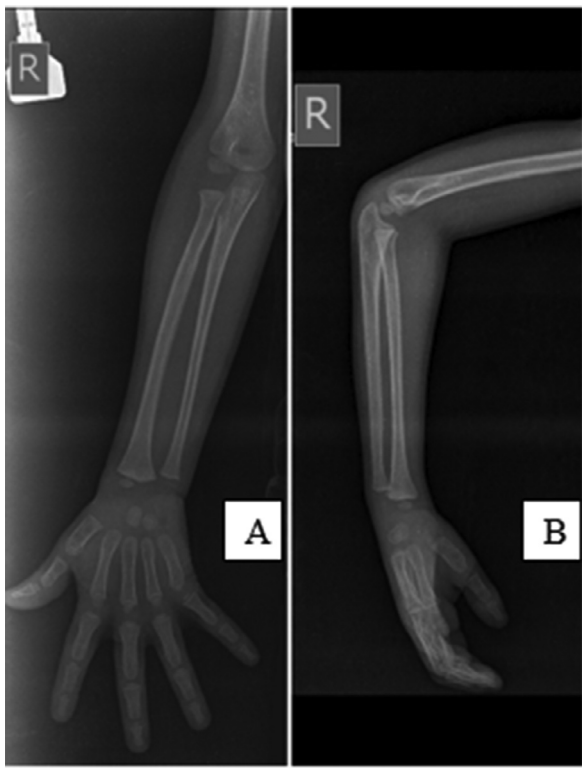

Fig. 4 – (A) AP and (B) lateral radiographs showing normal bones of the right elbow, radius and ulna, wrist and hand.

unsuccessful. The patient is also being seen by the clinical psychologist in order to assist him build self-confidence as he grows with this deformity. He is currently doing well. Permission was sought from the parents after thorough explanation of the condition to them, to write the case for publication, with the assurance of complete anonymity and confidentiality.

Discussion

Congenital proximal radioulnar synostosis is a rare disorder of the elbow which can be functionally tolerable by the patient in most cases when not severe. It can be associated with other musculoskeletal congenital conditions like syndactyly, polydactyly, congenital hip dislocation or clubfeet, which is not the condition of our patient. According to literature, CRUS is usually diagnosed before 5 years of age, and it is bilateral in about 60%-80% of cases, with a comparatively higher incidence in males [1,2]. In our case, even though the patient was less than 5 years, his condition was unilateral (Figs. 1 and 3).

CRUS causes variable degree of fusion of the proximal radioulnar joint and the fusion (synostosis) presents in varying lengths, which may involve the radial head or not. This congenital condition can be divided into types depending on its relation with the proximal radioulnar junction; Type I involves a complete fusion of the bones proximally with the fusion occurring for a variable distance, but Type II involves a partial union and it is related variably with the area distal to the proximal epiphysis of the radial head as described by Wilkie [5], and Simmons et al. [6]. CRUS was further described into 4 types by Cleary and Omer [7] as fibrous synostosis, bony synostosis, association with posterior dislocation of the radius and association with anterior dislocation of the radius. In our case, there was complete synostosis involving the bones and therefore classified as Wilkie Type I and affecting the proximal fourth of the left radius and ulnar (Fig. 2).

Our patient had a shorter left upper arm and forearm because of shortening of the left humerus and radius and ulna bones respectively (Figs. 1 and 5). This has also been reported in a study by Yammine et al. [8].

It can also be noted that radioulnar synostosis may occur as a result of trauma, termed as post traumatic radioulnar synostosis. Post-traumatic radioulnar synostosis may involve the distal forearm (the rarest), mid-forearm, and proximal forearm [3]. The current case report did not have any history of trauma and the deformity was noticed right from birth consistent with CRUS.

Literature has reported that CRUS may be associated with psychosocial problems as the child grows which may affect the patient's self-esteem resulting in depression, violent behavior, anxiety, and problems with relationships [9]. In this

Fig. 5 – Radiographs (AP) of both upper limbs showing a shorter left upper limb as a result of comparatively shorter humerus, radius and ulna bones with the osseous synostosis on the left.

patient, there was low self-esteem hence, the need for psychotherapy.

Conclusion

CRUS is rare but when present and severe can affect the mobility of the elbow and interfere with the daily activities of the affected individual. This therefore requires early diagnosis so that the appropriate multidisciplinary interventions are put in place to minimize the debilitating effects of CRUS.

Patient consent

Informed consent was obtained from the patient's parents and anonymity and confidentiality were ensured.

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