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Case Study

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UNILATERAL CONGENITAL RADIO-ULNAR SYNOSTOSIS: A CASE REPORT

***Dr. Monika Negi, Dr. Prateek Madaan, Dr. Uzma Khadtija and Dr. Akhilesh Negi**

Venue: Department of Radiodiagnosis and Interventional Radiology, Vardhman Mahavir Medical College and Safdarjung Hospital, New Delhi.

***Corresponding Author: Dr. Monika Negi**

Venue: Department of Radiodiagnosis and Interventional Radiology, Vardhman Mahavir Medical College and Safdarjung Hospital, New Delhi.

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INTRODUCTION

Congenital radioulnar synostosis is a rare entity that manifests in children later on in life, however a few cases have also been reported in adults. It develops in utero due to failure of segmentation of radius and ulna due to some insult antenatally. It causes limited rotational movements of the forearm. About 350 cases have been reported till now in the literature. In 60-80% of the cases it is bilateral.[1,2] However, this is the most common congenital functional disorder of the elbow joint.

CASE REPORT

An eight year old male child presented with the complaint of inability to supinate the right forearm since 1 year. No history of trauma or neurological deficit. On examination, there was limited supination of the right forearm with prominence of the ulnar head at the elbow joint. Rest of the movements were preserved. Bilateral elbow joint AP & lateral radiographs and non contrast computed tomography (NCCT) bilateral elbow joint were done.

The x-ray showed sclerosis and increased joint space of the proximal right radio-ulnar joint with few fine linear densities between radius and ulna on the superior aspect (Figure 1 & 2). The radial tuberosity was less prominent compared to the contralateral side. It was associated with malalignment of the radiocapitellar line suggesting radial head dislocation(Figure 2).

Non contrast enhanced CT was done for further evaluation, and there was evidence of partial bony radioulnar fusion for a distance of 1.2cm, 0.69 cm distal to the coronoid process (Figure 3).

It confirmed radioulnar synostosis of type III by Cleary-Omer classification and type II by Wilkie classification which is osseous fusion between radius and ulna with posterior dislocation of radial head.

Final diagnosis in our case was right proximal radioulnar synostosis (Type II by Cleary-Omer classification) based on clinic-radiological evidence and the patient was managed conservatively with physiotherapy.

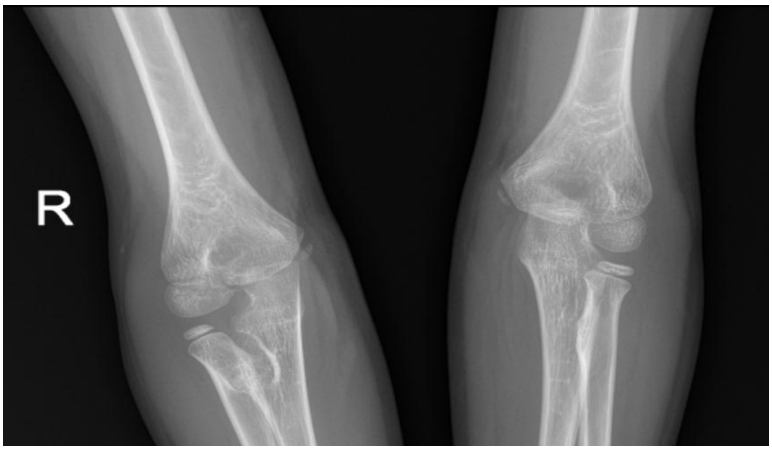
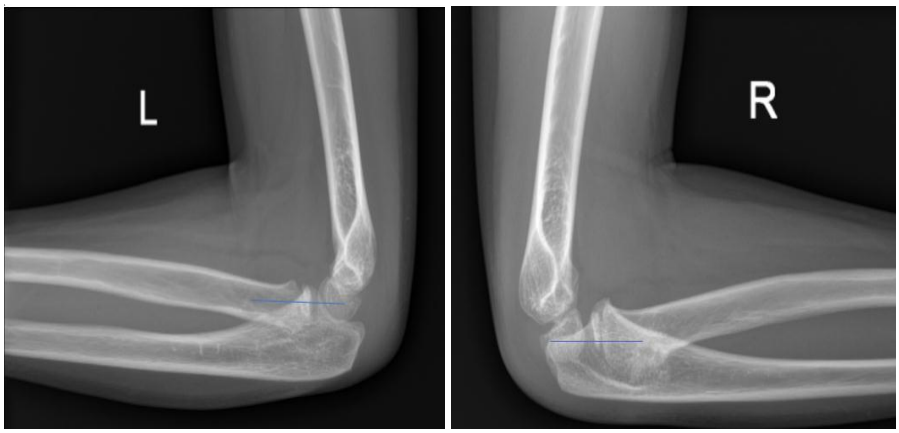


Figure 1. X ray(AP Radiograph) shows sclerosis in the proximal parts of radius and ulna and the margins of proximal ulna and radius cannot be differentiated in the region . Left elbow radiograph appears normal.



2(a) 2(b)

Figure 2. 2(a) shows the normal radiocapitellar line (marked with blue line) on the lateral radiograph of left elbow joint which is seen passing through the middle of the radial neck to intersect the capitellum in midline. Figue 2(b) shows normal radiocapitellar line (marked with blue line) in the right elbow joint suggesting no radial head dislocation with associated sclerotic changes and possible fusion in the region of proximal radius and ulna.

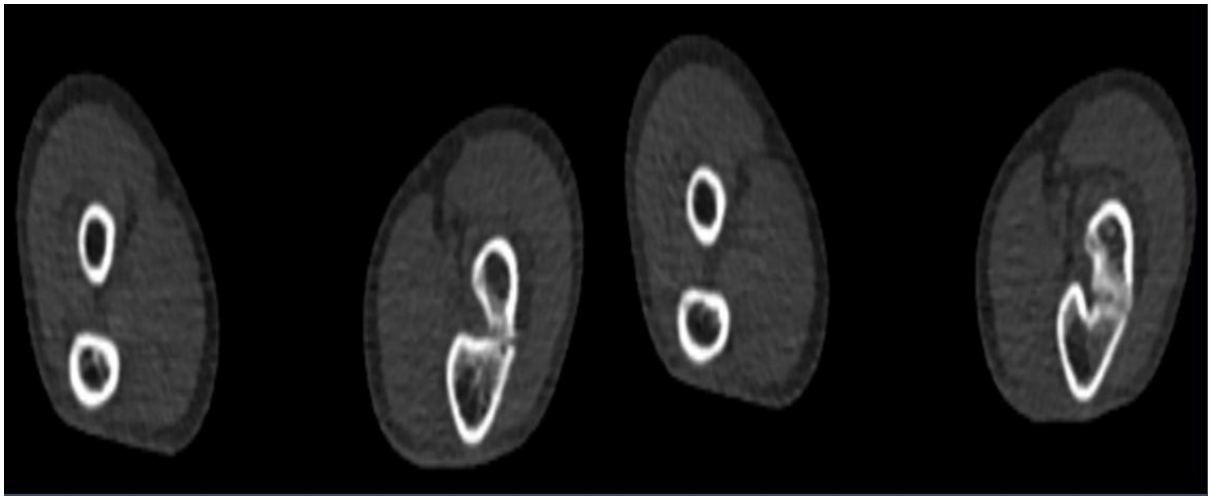
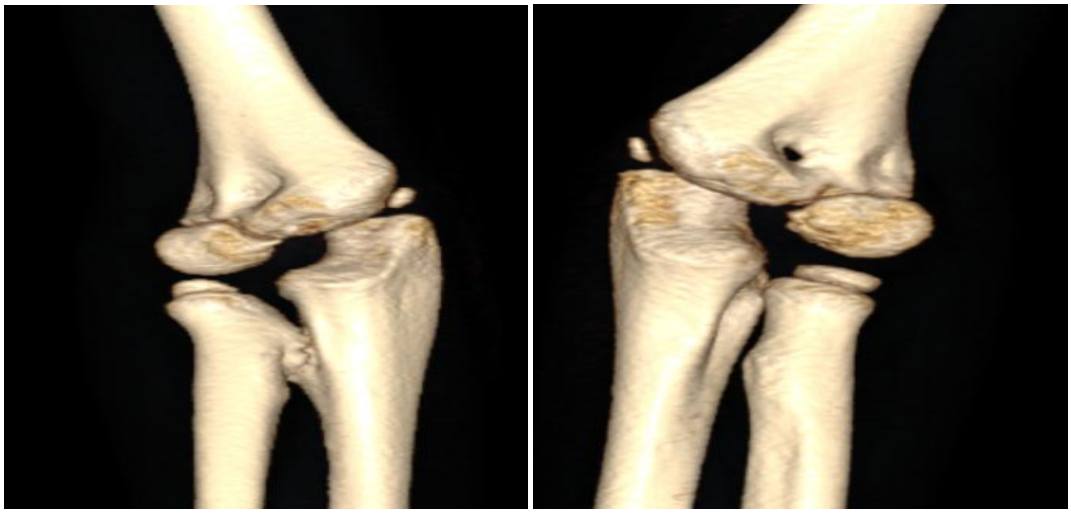


Figure 3. NCCT scan of both elbow joint. The scan was suggestive of partial bony radioulnar fusion on right side for a distance of 1.2cm, 0.69 cm distal to the coronoid process.



4(a) 4(b)

Figure 4 shows 3D imaging of both elbow joints. 4(a) shows bony fusion of right proximal radius and ulna Figure 4(b) shows normal left proximal radius and ulna.

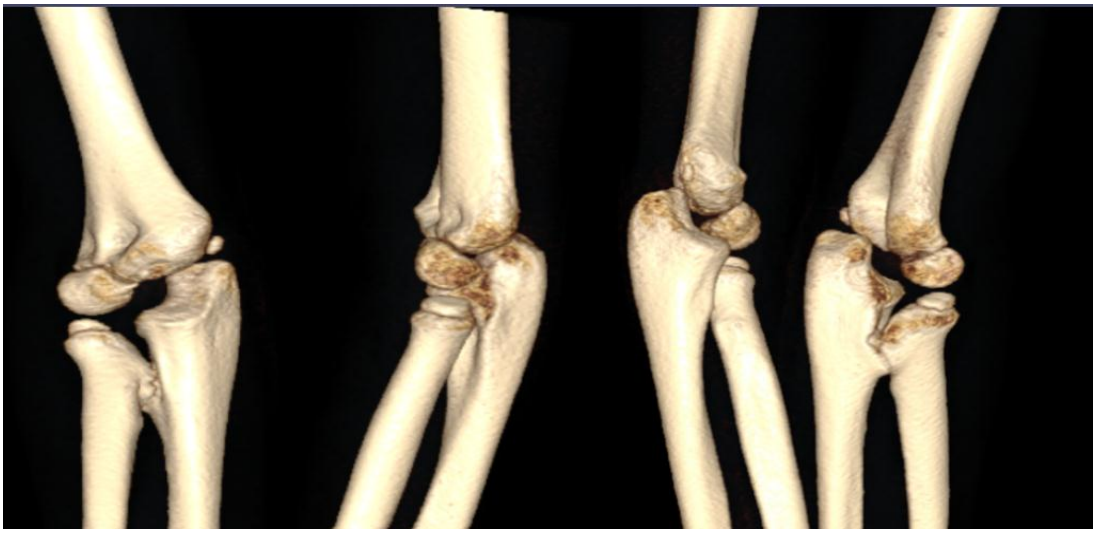


Figure 5. 3D imaging of both elbow joints showing bony fusion in different sections.

DISCUSSION

Congenital radioulnar synostosis is a rare anomaly of the upper extremity that manifests with abnormal fusion of proximal radius and ulna at birth. It is predominant in males with more than 60% cases showing bilateral involvement and is usually diagnosed by the age of 5.[1,2]

It occurs due to in utero defect in the longitudinal segmentation of the embryologically connected radius and ulna. Normally the segmentation begins distally and progresses proximally, therefore most common site of synostosis is proximal.[1,2] It may be isolated, be familial, or be loosely associated with a variety of congenital syndromes. MSK disorders associated are polydactyly, syndactyly, mandibulofascial dysostosis, Madelung's deformity, thumb aplasia, arthrogryposis, congenital hip dislocation, carpal coalition and clubfeet. Syndromic association include Apert's, Crouzon, Carpenter's, Holt-Oram, Poland, Klinefelter, and William's syndromes.[3,4]

It causes limited rotational movements of the forearm with preserved flexion and extension movements. In most cases, there is no associated shoulder or distal radioulnar joint abnormalities.[5] Diagnosis is based on the depiction of fusion of proximal radius and ulna on direct radiographs followed by supportive imaging features on CT scan. Four radiologic patterns of proximal congenital radio-ulnar synostosis have been observed by Cleary and Omer based on the presence of synostosis and the location of the radial head.

Patients with mild deformity usually compensates with the adjusted functions of the shoulder and wrist joints. Occupational therapy and modification of the activities are recommended. For severe deformity, surgical correction of synostosis by proximal derotational osteotomy is indicated. However, success rates are low with surgical correction.[2,6]

CONCLUSION

Congenital radio-ulnar synostosis is a rare entity involving upper extremities causing functional limitation. This case was found to be unique in a way that it was unilateral and presented beyond the usual age of presentation.

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