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Case Report

Type 4 congenital proximal radio-ulnar synostosis mimicking as malunited radial neck and deformed radial head with insidious onset extension block: a case report and results after excision

Chandan Arora*, Satyajeet Jagtap, Prateek Jain, Nilesh Sakharkar

Government Medical College and Hospital, Chandrapur, Nagpur, Maharashtra, India

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***Correspondence:**
Dr. Chandan Arora,
E-mail: rr.chandan@gmail.com

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ABSTRACT

Proximal radio ulnar synostosis is a rare entity presenting with restriction of supination and pronation. Among the various types, type 4 variety is even more-rare and its presentation, can pose a diagnostic challenge specially, if with history of trauma. Radiologically, it is usually overlooked as a malunited radial neck and therefore needs thorough evaluation. We presented a case of a 15-year-old male who complains of new onset extension block following trauma, with chronically restricted supination and pronation. History suggested the restriction in supination and pronation since birth, and the extension block occurring de novo. Radiographs and CT scan of the elbow revealed proximal radio-ulnar synostosis and anteriorly dislocated, mushroom-shaped radial head, misleading and mimicking like an old, neglected and malunited radial neck fracture with a deformed radial head. The family insisted to not address the restriction of supination pronation but, asked for solution to more disabling extension block. Thus, patient was managed with open procedure, involving radial head-neck excision and removal of malformed radial head. At 1-year follow-up, the child has a well-reduced and stable elbow joint with a functional range of flexion and extension movements with restricted supination and pronation. Type 4 congenital radio ulnar synostosis not only leads to conventionally known restriction of supination pronation but also give rise to extension block de novo as presented in this case. Excision of the deformed head and neck can treat the restriction in extension, however the synostosis will require various other augmented procedures.

Keywords: Synostosis, Congenital, Radial head, Excision

INTRODUCTION

Congenital radial ulnar synostosis is a congenital condition due to failure of differentiation characterized by the presence of a bony bridge between the proximal radius and ulna.

Clinical presentation is that of restricted supino-pronation, however radiography is relied upon for definitive diagnosis showing proximal radial ulnar bony connection. Male(s) appear to be suffering more than females with a ratio of 3:2 with 60% showing b/l involvement.[1] There are various types, among which, type four is a rare entity and the diagnosis can be challenging because of its clinical presentation with lack of knowledge about various types and superimposing history, like that of trauma.[2]

All these factors miss guide to a different diagnosis, for instance, a malunited radial head neck fracture. Asymptomatic patients are usually managed conservatively whereas, Surgical management is indicated in patients with disabling sympotms.[3] Due to the rarity of

this complex condition, in the absence of keen observation and meticulous attention, the correct diagnosis might be missed leading to unsatisfactory management and related complications. This is a case of type four proximal radio-ulnar synostosis with dislocated and deformed radial head which mis leaded and mimicked a malunited radial neck fracture with de novo development of extension block.

CASE REPORT

15-year-old male presented with complains of new onset restriction in elbow extension with a known history of inability to perform supination and pronation since childhood.

On history and evaluation, it was found that the he had an episode of trauma at the age of seven years which was followed by spontaneous resolution of symptoms and did not require any intervention. Patient now complains of de novo insidious onset of restriction of his left elbow extension since 3 months along with restricted supination and pronation (Figure 1).

On examination, the range of motion with respect to flexion-extension was 100 to 130 degrees only with a bony end block. The supination pronation arc was about 50 degress with a bony end point. The three point bony relationship was maintained and there was no varus-valgus instability noted and a negative pivot shift test.

On radiological evaluation (Figure 2)- reduced and maintained ulno humeral joint; and anteriorly dislocated radius with a deformed and mushroom shaped radial head depicting a cleary type four 4 proximal radioulnar synostosis.

The following surgical procedures were performed: proximal radius osteotomy.

The Kaplans elbow approach was used. During surgery, dislocation of the radial head, proximal synostosis of the radius and ulna, and absence of the annular ligament were confirmed (Figure 3). Rotation of the forearm was severely limited. The anteriorly dislocated radial neck and deformed head were identified and using a drill-saw, osteotomy was performed proximal to synostosis to remove the deformed radial head (Figure 4).

The synostosis was not addressed as the family did not consent to it. Bone wax was applied to the surfaces of the osteotomy sites for haemostasis. The incised LUCL was repaired and augmented using double loaded suture anchor (inserted in the lateral epicondyle of humerus) (Figure 6) to increase stability. The incision was closed after haemostasis and douching.

The elbow was immobilized in the flexion and supination position via a plaster slab for 1 week. Non-steroidal anti-inflammatory drugs were administered to prevent heterotopic ossification.

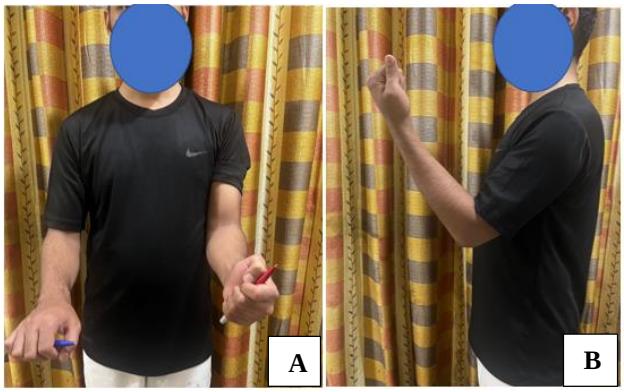

Figure 1: Pre-operative photograph of the left elbow range of motion- (A) restricted supination; and (B) restricted elbow extension.

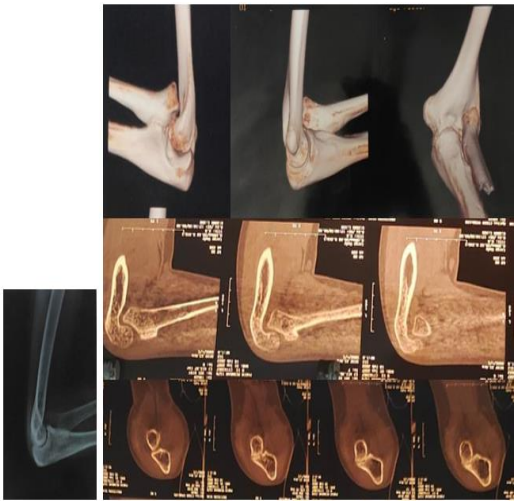

Figure 2: CT scan images of the left elbow illustrating- (A) lateral view of affected elbow showing dislocated radial head; and (B) 3D images showing the proximal radio-ulnar synostosis with dislocated and deformed radial head. Sagittal view of the CT scan depicts the bony outgrowth which might be the cause of restriction in extension. Axial images depict the synostosis between radius and ulna.

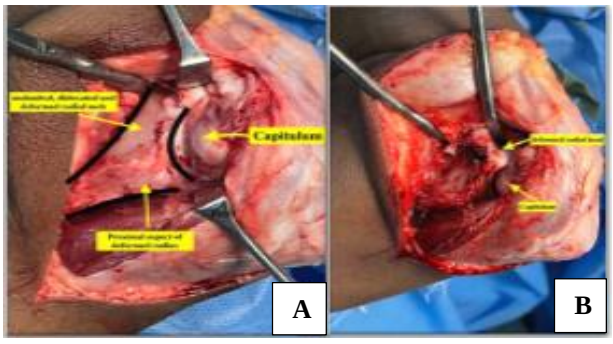

Figure 3: (A) Intra-operative images depicting cleary type 4 proximal synostosis with dislocated and deformed radial head; and (B) note the relationship of radial head with capitullum.

Figure 4: Intra-operative image after excision of bony bridge and deformed radial head.

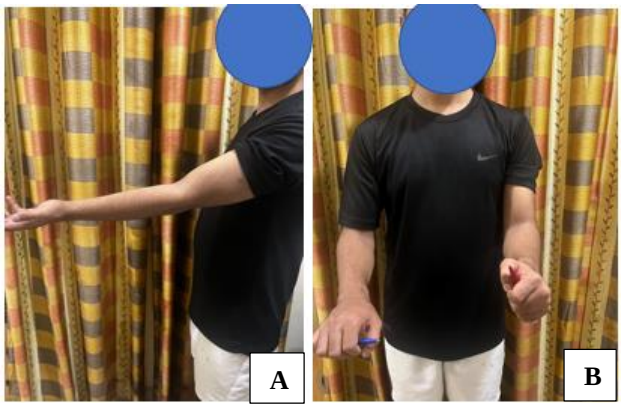

Figure 5: Post-operative one year follow up of the same case showing (A) restoration of elbow extension; (B) illustrates partially restored supination; as the synostosis was not removed completely (patients demand), the supination was still restricted, however slightly improved to an arc range of 90 degree.

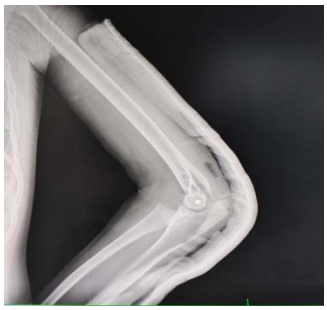

Figure 6: Post-operative X-ray radiography depicting suture anchor inserted at the distal humerus. The fibre wires were used to augment the LUCL which increased elbow stability. The deformed and dislocated radial head was excised however, the synostosis was not addressed, and a bony connection can still be appreciated at the proximal radio ulnar joint.

DISCUSSION

Cleary classified proximal radio ulnar synostosis in various types.[1] In type I, there is a decreased size of the radial bone, the fusion does not involve bones; in type II there is a radioulnar synostosis, and the remaining bone structures do not reveal any other changes. In type III, there is a radioulnar synostosis, hypoplastic head of the radial bone, and posterior subluxation of the radial head. In type IV, there is a short radioulnar synostosis, mushroom-shaped malformation of the head of the radial bone, and anterior subluxation of the radial head. The present case was diagnosed as with congenital proximal radioulnar synostosis of the right elbow presenting with extension block.

Meticulous examination of this case suggested that the synostosis site involved only the proximal epiphysis of the radius, which may be mostly cartilaginous, as this bridge provided small extent of movement when the patient rotated the forearm.

As per Guma et al some rotation of the forearm are partly compensated by the distal radioulnar joint.[4] The patient's symptoms may only have emerged after 6 years of age because this is typically when ossification of the epiphysis of the radius occurs.[5] This ossification limits the development of the radius along the longitudinal axis of the forearm.[6] In the presence of a malformation, the closing of the epiphysis also may have contributed to the occurrence of the valgus deformity as well as dislocation of the radial head observed in this patient. In support of this theory, preoperative DR showed that the fusion site was at the original location of the proximal radialis epiphysis. During surgery, this bony fusion was confirmed at the site of the original epiphysis of the radius. Thus, it is not astonishing that the final diagnosis of congenital proximal radioulnar synostosis was initially missed in this patient, given that there was no family history and its initial presentation with extension block. Furthermore, the cartilaginous bridge would not have been clearly visible on an X-ray. This case suggests that a patient younger than 6 years old who reports long-term issues with forearm rotation, but no family history or DR abnormality, should be considered for congenital proximal radioulnar synostosis. Soft tissue abnormalities concomitant with bony malformations are common with synostosis; although no preoperative MRI examination was conducted to assess muscle or ligament abnormalities in this case, surgery confirmed the absence of the annular ligament.[7]

Tsai et al recognize that bony malformations and soft tissue abnormalities usually coexist in this condition, and that surgical reconstruction of the bone alone cannot completely restore the rotation function of the forearm.[8] However, as the family and the patient himself did not consent to address the restricted supination-pronation; they were concerned with the more disabling extension block and demanded the extension block to be treated only. We failed to convince even after multiple rounds of

counselling and thus a decision for excision of the bony bridge and the deformed head was taken (Figure 4) without removal of the synostosis. According to Guma et al the range of motion is restricted while the synostosis is developing however, in our case, the elbow flexion-extension function had regressed over last one year such that the patient was no longer able to place his elbow in a fully extended position. These symptoms seriously affected the quality of the patient's daily life.[9] Surgery to excise the bony bar and the deformed radial head with reconstruction of the the annular ligament relieved these symptoms and restored an appropriate forearm rotation arc (Figure 5). Although the proximal radioulnar joint synostosis was not addressed, within 6 months post procedure, the patient's forearm rotation function was improved, elbow flexion-extension was completely restored and the patient could perform most activities of daily living. These results suggest that in Cleary type four even surgical excision of the bony bridge and the deformed head may be the best treatment for this disease, as synostosis are already known to reoccur and fuse again.[10,11]

CONCLUSION

The findings from this case suggest that we should carefully monitor all patients who report long-term issues with forearm rotation. This case also highlights that synostosis which is formerly known to cause restriction in supination pronation, can also present with de novo block in elbow extension due to a bony outgrowth, signifying the importance to assess other components of range of motion as in this case. Not all cases require surgical treatment, but when surgery is needed, a suitable method should be selected according to the individual needs of the patient. Any surgery performed should treat both the bony malformations and soft tissue abnormalities to maximize the therapeutic effect and reduce complications during and after surgery.

Clinical message

Congenital proximal radioulnar synostosis is a rare genetic disease. The findings from our case suggest that patients with congenital proximal radioulnar synostosis may also present with restriction in extension with no prior family history. The surgical methods performed should be selected according to the individual needs of the patient, and should treat both the bony malformation and the soft tissue abnormality with an appropriate fixation to reduce complications and maximize the therapeutic effect. We believe that treatment of congenital proximal radioulnar synostosis will continue to be improved as orthopaedic technology continues to develop.

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REFERENCES

1. Cleary JE, Omer GE. Congenital proximal radio-ulnar synostosis. Natural history and functional assessment. J Bone Joint Surg Am. 1985;67(4):539-45.
2. Sachar K, Akelman E, Ehrlich MG. Radioulnar synostosis. Hand Clin. 1994;10(3):399-404.
3. Radhakrishna R, Babu BR, Prashanth V. A case of osteochondroma of distal radius mimick-ing distal radio: Ulnar synostosis: Rare presentation-with review of literature. J Evol Med Dent Sci. 2015;4:11918-21.
4. Guma M, Teitel AD. Adolescent presentation of congenital radioulnar synostosis. Clin Pediatr (Phila). 1996;35(4):215-7.
5. Jupiter JB, Ring D. Operative treatment of post-traumatic proximal radioulnar synostosis. J Bone Joint Surg Am. 1998;80(2):248-57.
6. Shoham Y, Gurfinkel R, Sagi A. Idiopathic distal radioulnar synostosis. J Plast Surg Hand Surg. 2014;48(1):89-90.
7. Failla JM, Amadio PC, Morrey BF. Post-traumatic proximal radio-ulnar synostosis. Results of surgical treatment. J Bone Joint Surg Am. 1989;71(8):1208-13.
8. Tsai J. Congenital radioulnar synostosis. Radiol Case Rep. 2017;12(3):552-4.
9. Wurapa R. Radioulnar Synostosis, 2009. Available at: http://www.emedicine.medscape.com. Available at: 01 September 2022.
10. Osterman AL, Arief MS. Optimal management of post-traumatic radioulnar synostosis. Orthop Res Rev. 2017;9:101.
11. Pfanner S, Bigazzi P, Casini C, Angelis C, Ceruso M. Surgical Treatment of Posttraumatic Radioulnar Synostosis. Case Rep Orthop. 2016;2016:5956304.

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