Case Report
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Humero-radio-ulnar synostosis: a case report with brief literature search

Abstract

Congenital humero-radio-ulnar synostosis is an extremely rare condition. So far approximately 30 cases of congenital humero-radio-ulnar synostosis have been reported worldwide. To the best of our knowledge, no such type of case has been reported in India as well as Asia. The case presented here is of a 7 month old male baby, born with right sided humero-radio-ulnar synostosis with no other musculoskeletal defect. In the right upper extremity the congenital defect was due to failure of cavitation during embryologic development. We believe that the case presented here deserves reporting not only because or rarity of the disease but also due to its uniqueness. There was neither any associated hypoplasia of upper extremity nor there was any familial or syndromic association. An attempt is also made to do brief literature search.

Keywords: congenital, humero-radio-ulnar synostosis, hypoplasia, familial

Volume 15 Issue 6 - 2023

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Received: October 30, 2023 | **Published:** November 14, 2023

Introduction

Congenital synostoses are defined as rare abnormal formations of primary anatomical structures.[1] Upper extremity synostoses are characterized by the fusion of upper extremity bones ensuing in the incapability to bend and/or rotate the forearm, subject upon the type of elbow synostosis. Congenital humero-radio-ulnar synostosis is an extremely rare condition. Way back in 1983, Gollop and Coates described bilateral developmental elbow synostosis with limb hypoplasia, followed by another similar report by Leroy and Speeckaert in 1984.[2,3] McIntyre and Benson reported that developmental elbow synostoses are anatomically divided on the basis of nature of bony ankylosis into humero-radial, humero-radio-ulnar and humero-ulnar ankyloses. The commonest are humero-radial synostoses followed by humero-radio-ulnar synostoses and humero-ulnar synostosis in that order.[4,5] McIntyre and Benson believed that the anatomical classification for developmental elbow synostoses may be ambiguous. Therefore they proposed an etiological classification for all congenital elbow synostoses.[4] Their current classification divides the elbow synostoses into class I (elbow fixed in extension with ulnar ray hypoplasia and sporadic) and class II (elbow fixed in flexion without hypoplasia and familial). Class II may be associated with multiple systemic anomalies.[4,6,7] We are reporting a case of 7 months, male baby with congenital right sided humero-radio-ulnar synostosis. The present case is unique in that there was neither any hypoplasia of upper limb nor there was any familial or syndromic association.The patients and their parents were informed that data concerning the case would be submitted for publication, and they consented.

Case study

A 7 months old male baby was brought to us by parents with complaints of inability to move right elbow joint. They further added that the child is also keeping his elbow in bent position since birth. The patient was their second child with one female sibling without any such complaints. There was no history of consanguinity among the parents. The baby was full term, delivered through vaginal route. There was no history suggestive of any maternal infection or any exposure to teratogenic agents throughout the antenatal period. There was no family history of congenital elbow anomalies in first degree lineages. There was no developmental delay. No history of other joint involvement or other systemic illnesses.

Clinical examination revealed that the right elbow was fixed at 90° of flexion with neutral forearm rotation (Figure 1). No movement was possible at the elbow and at radio-ulnar joints. There was no hypoplasia of the forearm, hand or fingers. The left upper extremity was absolutely normal. There were no other skeletal abnormalities. Furthermore, no definite syndromic features were noted. Systemic examination including cardiovascular system, central nervous system, respiratory system, abdomen and genitalia was unremarkable. Ultrasonography of abdomen and echocardiography were normal. Synostosis of humero-radio-ulnar joint was found on radiological examination (Figure 2). Radiograph revealed continuous bony trabeculae from humerus to radius and ulna with no evidence of any elbow joint, suggesting that the humero-radio-ulnar joint was never formed. The bones of the forearms were fused in a semipronated position in there proximal half, with no evidence of hypoplasia. According to current classification of McIntyre and Benson, it was class II developmental elbow synostosis. Radiological examination of left upper extremity was normal. In the present circumstance, we did not offer any operative procedure to the child and kept the child under vigilant observation. Parents were communicated about the future prospects as the child would grow, there will be limitations of activities which require coordinated movement of elbow and forearm rotations like feeding, hygiene, cleaning, writing and others.

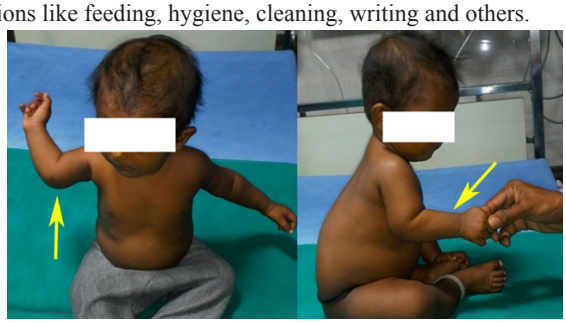

Figure 1 Clinical photograph of patient showed that the right elbow fixed in 90° of flexion with neutral forearm rotation (arrows). Also note there is no hypoplasia of the forearm, hand or fingers with absolutely normal left upper limb.

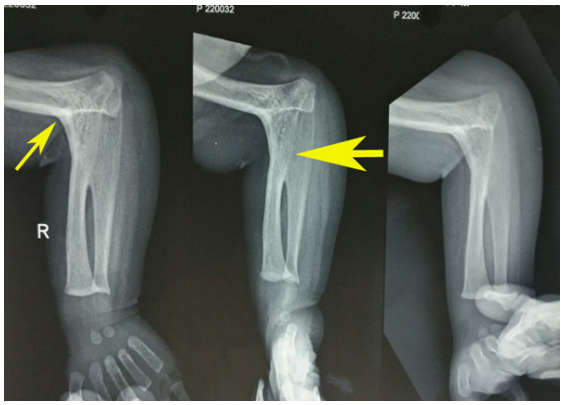

Figure 2 Radiograph revealed continuous bony trabeculae from humerus to radius and ulna with no evidence of any elbow joint (thin arrow), suggesting that the humero-radio-ulnar joint never formed. The bones of the forearms were fused in a semi-pronated position in there proximal half (thick arrow), with no evidence of hypoplasia. According to current classification of McIntyre and Benson, it was class II developmental elbow synostosis.

Discussion

Humero-radio-ulnar synostosis is an extremely rare condition. Approximately 30 cases of humero-radio-ulnar synostosis have been reported worldwide.[4,6,8] To the best of our knowledge, no such type of case has been reported in India as well as Asia. Subject of this case report is the 31st and the first case of humero-radio-ulnar synostosis being reported from India.[4,6,8] The condition presents with variable degree of limitation of elbow movement and forearm rotations. A review of the medical texts ascertains that congenital radio-ulnar synostosis is the most common form of congenital upper extremity abnormality.[4,6,9] Synostoses between the humerus and ulna, radius, or both have been recognized but are far less common. Although the incidence and exact cause of upper extremity synostosis is unknown, but familial[10] and sporadic[4,6] cases have been described. Synostoses may be congenital or secondary to trauma or disease process.[1]

During embryonic development a single mesodermal condensation appears along the long axis of the limb bud in the 3rd to 4th week of intra-uterine period.[4] As the upper extremity continues to develop, the cartilaginous precursors of the upper extremity are initially connected. By the 7th week, the elbows develop when chondrogenesis halts and cavitation commences.[11] However, if cavitation is interrupted due to any insult, the cartilaginous precursors fail to separate, and the connection of cartilaginous tissue remains. By the 12th week, endochondral ossification begins and eventually results in synostosis. In nutshell congenital upper extremity synostosis results from a failure of longitudinal separation and the retention of the cartilaginous anlage of the upper extremity bud during embryological development.[12] In our case cavitation failed to occur which resulted in synostosis.

Adjoining bones may also fuse together as a secondary complication of trauma or disease.[13] In delayed presenting cases one has to differentiate congenital from traumatic synostosis. Good clinical history and examination gives clue to the underlying etiology of the synostoses. Furthermore, role of radiological examination cannot be overemphasized in differentiating congenital from traumatic ones. Radiologically, traumatic synostoses are characterized by bone remodelling, sclerosis and ossification of soft tissues.[13,14] Individuals with congenital upper limb synostoses may have malformed skeletal structures. The types and severity of malformations vary and may include radial ray defects, congenital dislocation of the hips, clubfoot and other syndromes.[1,4,6,15,16]

Hersch et al. reviewed eight reports of humero-radio-ulnar synostosis, and presented another case in 1989.[17] All cases were sporadic and associated with limb hypoplasia. McIntyre JD and Benson MK in 2002, further identified 12 more reports of humero-radio-ulnar synostosis in literature. They also reported that cases follow a similar pattern of deformity and inheritance to cases of humero-radial and humero-ulnar synostosis, incidence may be familial or sporadic, hypoplasia may or may not be present, as in our case.[4] Our case was unique in the sense being class II elbow synostosis; it was not familial but sporadic.

Most of these patients do well if the elbow is ankylosed in a functional position provided that elbow synostosis is the only anomaly.[5] Unilateral cases perform far better than bilateral elbow synostoses patient. Literature recommends initial careful observation of the patient's function, if essential an osteotomy could be accomplished to obtain a more functional position of the elbows. Any surgical intervention performed in the future should address these needs by fetching at least one elbow to a functional position so that it can be transposed to facilitate self-feeding and ample hygiene procedures. There is a high recurrence rate of re-synostosis following surgical treatment.[4,5] We did not offer any operative procedure to the child and kept the child under vigilant observation, and to intervene accordingly in future.

Conclusion

Congenital humero-radio-ulnar synostosis is an extremely rare condition. The present case is unique in that there was neither any hypoplasia of upper limb nor there was any familial or syndromic association.Vigilant observation, soft tissue release, osteotomy and arthroplasty are the suggested treatment. This report will help the orthopaedic surgeons to diagnose the condition by systematic clinico-radiological examination of the patients. Further studies are required for proposing best treatment protocol for managing such patients, especially in rehabilitating them.

Acknowledgments

None.

Conflicts of interest

None.

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Citation: Sharique M, Rahman N, Khan AQ, et al. Humero-radio-ulnar synostosis: a case report with brief literature search. *MOJ Orthop Rheumatol.* 2023;15(6):210–212. DOI: 10.15406/mojor.2023.15.00647