



RESEARCH ARTICLE

BILATERAL PROXIMAL RADIOULNAR SYNOSTOSIS-A CASE REPORT

*Ashwini, N. S. and Venkateshu, K. V.

Department of Anatomy, Sri Devaraj Urs Medical College, Tamaka, Kolar

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ABSTRACT

Synostosis or osseous union of any 2 adjacent bones can involve any part of the body. Synostosis between radius and ulna can take two forms, congenital and post-traumatic. Proximal Radioulnar Synostosis is a very rare congenital anomaly and most cases are sporadic, congenital and less often post-traumatic, bilateral in 60% and more common in males. It is often part of syndromes such as Crouzon, Apert's and Poland's, 4p- syndrome, Williams syndrome, acrofacial dysostosis, Antley-Bixler syndrome, Genito- patellar syndrome, Greig Cephalo- polysyndactyly syndrome, hereditary multiple osteochondromas (hereditary multiple exostoses), limb-body wall complex, and Nievergelt syndrome. Although the exact etiology is not clear, it has been documented that there is a genetic basis for the failure of differentiation between the radius and ulna. We came across such rare specimen of bilateral proximal radioulnar synostosis in Department of Anatomy, Sri Devaraj Urs Medical College, Kolar, Karnataka. The specimen was incidentally obtained from disarticulated upper limbs of a male cadaver aged 70yrs which were buried under the soil to obtain bones for study purpose. The procured bone specimen of proximal radioulnar synostosis was cleaned and later relevant segmental measurements were taken. The rarity of this condition often leads to the delayed clinical diagnosis. The rationale for surgery is dictated more by individual functional limitations rather than on absolute forearm position.

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INTRODUCTION

Synostosis or osseous union, of any 2 adjacent bones can involve any part of the body. Synostosis between radius and ulna can take two forms, congenital and post-traumatic. Congenital radioulnar synostosis is caused by failure of segmentation between radius and ulna. Congenital proximal radio-ulnar synostosis is a rare anomaly and is often part of syndromes such as Crouzon, Apert's and Poland's 4p-syndrome, Williams syndrome, acrofacial dysostosis, Antley-Bixler syndrome, genitopatellar syndrome, Greig cephalopolysyndactyly syndrome, hereditary multiple osteochondromas (hereditary multiple exostoses), limb-body wall complex, and Nievergelt syndrome. Approximately one-third of patients with radio-ulnar synostosis have associated anomalies involving the cardiovascular, genitourinary, gastrointestinal, central nervous, and musculoskeletal systems. This malformation is commonly connected with chromosome X aberrations. However, there are reports on cases of congenital radioulnar synostosis in chromosome Y aberrations.

Post-traumatic radioulnar synostosis is a rare complication of forearm fracture resulting in loss of forearm axial rotation with incidence of 2% according to Vince and Miller. Incidence is elevated in case of neurological brain lesion.

Literature Survey/Problem Definition

Proximal Radio-ulnar Synostosis is a very rare congenital anomaly and most cases are sporadic, congenital and less often post-traumatic, bilateral in 60% and more common in males. Although the exact etiology is not clear, it has been documented that there is genetic basis for the failure of differentiation between the radius and ulna. During development the radius and ulna divide from distal to proximal therefore the synostosis is usually observed in the proximal half. Familial cases with autosomal dominant inheritance has been reported. Patients frequently have duplication in sex-chromosome.

MATERIALS AND METHODOLOGY

This rare case of bilateral proximal radioulnar synostosis was reported in Department of Anatomy, Sri Devaraj Urs Medical college, Kolar, Karnataka. The specimen was incidentally obtained from disarticulated upperlimbs of a male cadaver aged

*Corresponding author: Ashwini, N.S.

Department of Anatomy, Sri Devaraj Urs Medical College, Tamaka, Kolar

70yrs which was buried under soil to obtain bones for study purpose. The procured bone specimen of proximal radioulnar synostosis was cleaned and relevant segmental measurements of radius and ulna were taken using osteometric board and vernier calipers.

CASE REPORT/RESULTS

We report a rare case of bilateral proximal radioulnar synostosis in Department of Anatomy, Sri Devaraj Urs Medical college, Kolar, Karnataka. Relevant segmental measurements of radius and ulna were taken using osteometric board and vernier calipers. The maximum length of ulna from most superior point of olecranon process to most inferior point on styloid process was 23.4cm on right and 23.1cm on left side. The physiological length of ulna from inferior point of coronoid process to inferior surface of distal end of ulna was 20cm on right and 19.6cm on left side. The transverse diameter of ulna (distance between medial and lateral surface at crest) was 1cm on both sides. The antero-posterior diameter of ulna at crest was 0.6cm on both sides. The length of radius (distance between radial tuberosity to styloid process) was 24.2cm on right and 24cm on left. The antero-posterior diameter of radius at midshaft was 1cm on both sides. The transverse diameter of radius at mid-shaft was 1.4cm on both sides. The transverse diameter of radioulnar synostosis on both sides was 2.8cm and 2.7cm respectively. Considering the above measurements slight shaft shortening of radius and ulna was observed on the left side compared to right. Radial shaft bowing was present bilaterally (FIGURE 1-3). Vince had classified radioulnar synostosis based on location. As per his classification this cadaver had type 3 deformity involving the proximal 3rd of radius and ulna bilaterally. In 1985 Cleary and Omer had observed distinct radiographic patterns based on presence of osseous synostosis of radius and ulna. Accordingly if radiographs were taken our report fits into type 3 classification with osseous synostosis with hypoplastic and posteriorly dislocated radius bilaterally which was quite obvious in the specimen. Wilkie had also classified radioulnar synostosis based on proximal radioulnar junction. Accordingly the bilateral radioulnar synostosis fits type 1 complete synostosis with radius and ulna fusing proximally for a variable distance.



DISCUSSION

Radioulnar synostosis is a rare complication of forearm trauma. Botting described a case of post-traumatic synostosis without fracture, secondary to a knife-wound in the upper third of the forearm. These account for 2 to 5% of synostoses in adults (Dohna *et al.*, 2012).

Congenital radioulnar synostosis was reported in a 4-years-old male child, born after 30 years of non-consanguineous marriage. There was duplication of left index finger left and middle finger on right side. No other congenital anomalies were detected either clinically or radiologically. X ray both elbow joints revealed proximal osseous radio ulnar synostosis with diminutive radial head (Dogra *et al.*, 2003). A case of 10-year-old child with pain and restricted mobility of the elbow joint was reported wherein radiological examinations revealed congenital radioulnar synostosis (Anna Siemianowicz *et al.*, 2010). In 1793, Sandifort provided the initial description of congenital radioulnar synostosis. The synostosis was supposed to be caused by failure of segmentation between the radius and ulna. During development the upper limb bud arises from the unsegmented body wall at 25-28 days. The elbow is visible at 34 days, and the humerus, radius, and ulna is visible at 37 days. The 3 cartilaginous analogs of the upper limb bones i.e humerus, radius, and ulna are connected before segmentation. For short time the radius and ulna share a common perichondrium. Any abnormal events in course of development can lead to a failure of segmentation. Endochondral ossification then proceeds, and the cartilaginous synostosis ossifies, either partially or completely, in the longitudinal or transverse plane. In 20% of their patients, Cleary and Omer found a genetic basis for an autosomal dominant form (with variable penetrance) of congenital radioulnar synostosis (Raymond Wurapa *et al.*, 2014). A case of right proximal radioulnar synostosis in a 5 year old child was observed. The elder child did not suffer radioulnar synostosis. Her 39 year old father had suffered proximal bilateral radioulnar synostosis (Mohammad Fakoor, 2006).

A 17 year old boy presented with limited range of motion of both the forearms. Radiographs of both the arms demonstrated a bony synostosis of proximal radius and ulna and spectrum of anatomic variation, absence of radial head, forearm shortening and radial shaft bowing (Ramakrishna Avadhani *et al.*, 2011) A case of 25 yr old male patient came with bilateral congenital radioulnar synostosis. Pain was not a presenting symptom until teenage years while progressive and symptomatic radial head subluxation was noted. The disability is most significant in bilateral cases with severe problems in pronation. Indications for surgical interventions in congenital radioulnar synostosis still remain somewhat controversial but are related to bilaterality and to the degree of deformity. Wrist hypermobility allows further functional compensation. Surgery is performed in childhood before school-age and the rationale for surgery is dictated more on the individual functional limitations rather than on absolute forearm position. Surgery can be performed either to remove the abnormal soft tissue or bony connection or to re-position the forearm to a better functionally acceptable position (Bhatt *et al.*, 2011). MacFellxnder reported a case of synostosis in the distal radio-ulnar joint. In 1985 Cleary and Omer observed 4 distinct radiographic patterns based on osseous synostosis and position of radial head. Type 1 fibrous synostosis, type 2 osseous synostosis, reduced radial head, type 3 osseous synostosis, posteriorly dislocated radial head, type 4 osseous synostosis, anteriorly dislocated radial head. In 1987 Vince classified 3 types based on location. Type 1 distal forearm (least common), type 2 includes middle 3rd or non-articular part of distal 3rd of radius and ulna, type 3 includes proximal 3rd of radius and ulna. Radioulnar synostosis is not noted until late childhood, as function may be normal especially in unilateral cases. Increased wrist motion may compensate for the absent forearm motion. Pain is generally

not a problem, unless radial head dislocation should occur. Individuals whose forearms are fixed in greater amounts of pronation (> 60 degrees) face more problems than those with around 20 degrees. In bilateral cases where function is impaired, recommendations for the optimum position of the non-dominant arm range from neutral to 30-40 degrees of supination. After osteotomy through the synostosis, neurovascular complications are not rare. Strategies have been planned to reduce complications which include resection of a portion of the synostosis with external fixation and Osteotomy is performed in the distal radius instead of at the synostosis site. Most attempts to divide the synostosis and insert some type of interpositional material have failed. Success has been reported following vascularized fascio-fatty graft. Nonoperative observation is usually preferred treatment, especially if deformity is unilateral. Osteotomy with fusion is indicated to obtain functional degree of pronation. The technique should be performed at 5yrs of age and cannot recreate proximal radial-ulnar joint as it will reossify and recur.

Conclusion

Proximal Radio-ulnar Synostosis is a very rare congenital anomaly and most cases are sporadic, congenital and less often post-traumatic, bilateral in 60% and more common in males. Although the exact etiology is not clear, it has been documented that there is genetic basis for the failure of differentiation between the radius and ulna.

Future Scope

The rarity of proximal radio-ulnar synostosis often leads to delayed clinical diagnosis and thereby clinicians should be aware of this clinical condition to avoid any future disability. It is recommended that surgery be performed in childhood before patients are school-aged. Appropriate workup includes plain radiography performed in orthogonal planes. The indication for surgery in posttraumatic radioulnar synostosis is functional

limitation of forearm rotation. This limitation must be assessed on an individual basis. Therefore rationale for surgery is dictated more on individual functional limitations rather than on absolute forearm position.

REFERENCES

- Anna Siemianowicz, Wojciech Wawrzynek, Krzysztof Besler. 2010. Congenital radioulnar synostosis – case report. *Pol J Radiol.*, 75(4),pp.51–54.
- Bhatt, C R., C D Mehta, 2011. Case Report: Congenital Radioulnar Synostosis and its embryological correlation and functional assessment”, *J. Anat. Soc. India*, 60(2), pp. 236-238.
- Dogra B B, Singh M, Malik A. 2003. Congenital proximal radioulnar synostosis. *Indian J Plast Surg.*, 36,pp.36-8.
- Dohna, P., F, Khiamia, E *et al.* 2012. Adult post-traumatic radioulnar synostosis. *Orthopaedics & Traumatology. Surgery & Research*, 98(6), pp. 709–714.
<http://emedicine.medscape.com/article/1240467-overview>, Raymond Wurapa, Harris Gellman, Francisco Talavera. 2014. Radioulnarsynostosis.
<http://en.wikipedia.org/wiki/Synostosis>
- Mac Fellxnder. A case of synostosis in the distal radioulnar joint.
- Mohammad Fakoor, 2006. Radioulnar Synostosis in a Father and his 5 year old Daughter. *Pak J Med Sci.*, 22 (2), pp.191 – 193.
- Nick G L, Nikolaos K, Kanakaris, Peter V Giannondis. Trauma & Orthopaedic classifications: A comprehensive overview. Chap 38, Post-traumatic radioulnar synostosis, pp.175-78.
- Ramakrishna Avadhani, Bindhu S *et al.* 2011. Radio Ulnar Synostosis: A Case Report. *NUJHS*, 1(4),pp.43-45.
www.orthobullets.com/hand/6071/congenitalradioulnar-synostosis
www.Posna.org/education/studyguide/Radioulnar-Synostosis.asp
