



Proximal Radio-Ulnar Synostosis-Two case reports

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ABSTRACT

Radioulnar Synostosis, which is caused by fusion between the proximal ends of the radius and ulna, is an uncommon deformity of the upper extremity, caused by a failure of segmentation between the radius and ulna. During routine demonstration classes for the MBBS students we found proximal fusion between radius and ulna of two macerated bones. The proximal ends of radius and ulna were completely fused in two cases and the radial head was completely fused with radial notch of the ulna. The processes and notches of ulna were incomplete. Such anomalous fusion may lead to difficulties in daily activities, such as writing, eating, and accepting objects in an open palm. Knowledge of proximal radioulnar Synostosis is important for present day surgeon orthopedician and radiologist in interpretation of CT-scans and MRI.

Key words: synostosis, segmentation, malformation.

INTRODUCTION

Synostosis is the union of any adjacent bones, can involve any part of the body. Radioulnar Synostosis is a rare malformation that affects the elbow and forearm. It is caused by the fusion of the proximal radius and the ulna with the presentation of forearm fixed in some degrees of pronation. Loss of rotation of the forearm substantially decreases the functions of the upper limb and can have profound restriction on patient's daily activities such as eating, writing, washing, and dressing. The first anatomical description was given by Sandi fort in 1793 [1].

According to Lewis (1901) description on the development of upper extremities in man, the embryology of upper limb bud arises from the unsegmented body wall at 25-28 days. The elbow becomes visible at 34 days, and the humerus, radius, and ulna become visible at 37 days [2]. Initially, the 3 cartilaginous analogs of the humerus, radius, and ulna are connected before segmentation. Therefore, for a short time, the radius and ulna share a common perichondrium. Abnormal events at this time can lead to a failure of segmentation. The duration and severity of the insult can determine the degree of subsequent synostosis. Endochondral ossification then proceeds, and the cartilaginous synostosis ossifies, either partially or completely, in the longitudinal or transverse plane. In the forearm, congenital radioulnar synostosis usually occurs between the proximal radius and the ulna. Although the condition is present at birth, it usually is not discovered until early adolescence, when the patient presents with a lack of pronation and supination. Initially, the union may be more of a synchondrosis, but as the skeleton matures, the osseous bridge between the radius and ulna becomes more radiographically apparent.

Case Reports:

During routine osteological demonstration for MBBS students we found rarely fused proximal ends of radius and ulna bone in two cases. In the first case (figure-1) the head of radius bone was disfigured and completely ossified with radial notch of the ulna, the fusion extended up to the proximal part of the shaft of the ulna. The measurement of fusion from anterior to posterior was 4.0 cm. The coronoid notch and two processes (olecranon and coronoid) of ulna were appears to be normal. In second case (figure-2) the anatomy of the coronoid notch and the two processes of ulna were disfigured. The fused part measured 3.8 cm.

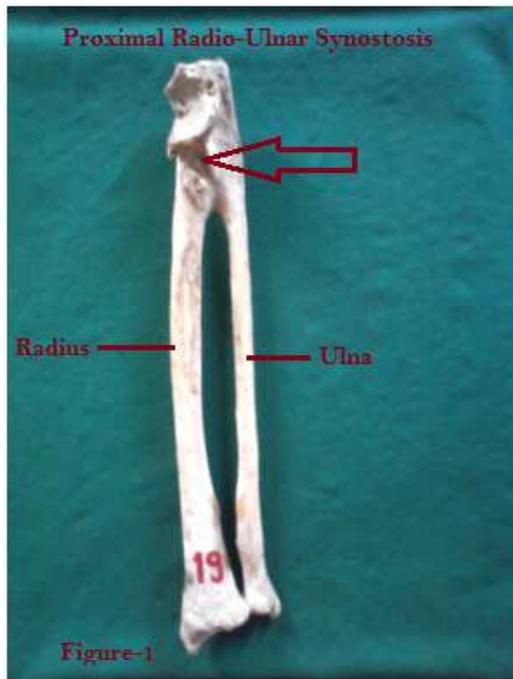


Figure1: shows proximal radioulnar fusion with olecranon & coronoid processes and radial notch of ulna

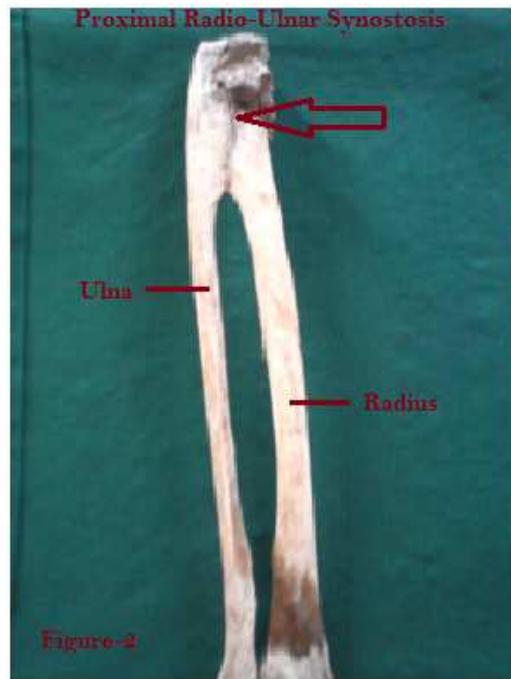
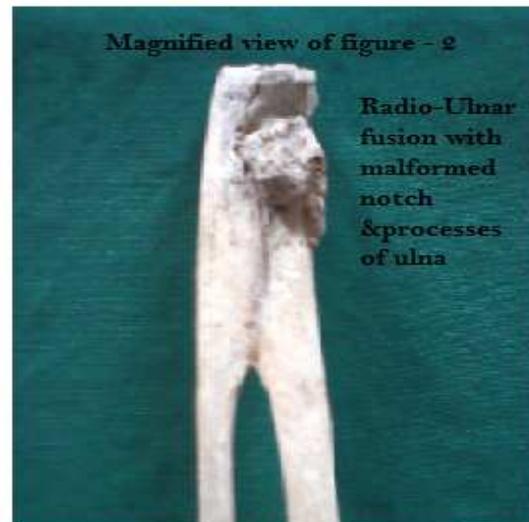
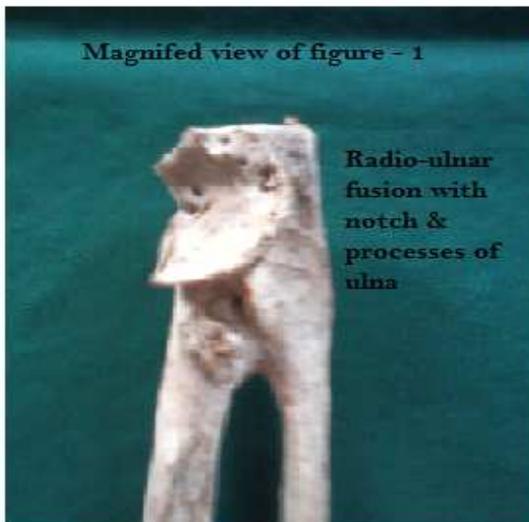


Figure 2: shows radioulnar fusion with disfigured processes and notch



Magnified view of figure 1&2 shows synostosed parts of the radius and ulna

DISCUSSION

Radioulnar synostosis is regarded as an anomaly of longitudinal segmentation. It is presumed that some factor inhibits the interzonal mesenchyma between the cartilage anlage of the developing radius and ulna from undergoing dislocation during the seventh week of intra uterine life thus the inter zonal mesenchyma persist and undergo chondrification ossification and eventually synostosis [3]. Synostosis is, however, probably no mere accident, but due to some alterations of the chromosomes. There is probably a gene mutation that alters the various developmental impulses that direct the formation of the proximal ends of the bones of the forearm [4]. Congenital radioulnar synostosis may be associated with some musculoskeletal anomalies and congenital syndromes such as polydactyly, syndactyly, Madelung's deformity, carpal coalition, thumb aplasia, Apert's syndrome, Carpenter's syndrome, arthrogryposis, mandibulofascial dysostosis, Klinefelter syndrome and William's syndrome. This condition occurs equally in males and females with bilateral involvement in 60% to 80% of patients [5,6]. Several classification systems based on the radiological findings have been proposed. Cleary and Omer described four types. Type I is a fibrous synostosis with a normal and reduced radial head. Type II is a visible osseous synostosis with a normal radius. In type III, a bony synostosis exists with the radial head hypoplasia and dislocated posteriorly. In type IV, the radial head is dislocated anteriorly. However, there is little clinical value in this classification because the operation depends on the severity of the functional deficit and on whether or not it is bilateral [7].

CONCLUSION

Radioulnar synostosis occurs as either a congenital or a posttraumatic condition. Congenital radioulnar synostosis is a rare problem and is thought to be a failure of longitudinal segmentation between the radius and ulna at the 7th week of development. Because it is due to a defect in utero or it may be associated with some musculoskeletal anomalies and congenital syndromes. Congenital radioulnar synostosis may lead to difficulties in daily activities. Many operative procedures have been developed to mobilize the fixed forearm but the long-term results are still unsatisfactory.

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