Congenital proximal radioulnar synostosis

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Section: Musculoskeletal system
Area of Interest: Musculoskeletal bone
Imaging Technique: Conventional radiography
Special Focus: Congenital Case Type: Clinical Cases

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Patient: 20 years, male

Clinical History:

A 20-year-old male patient with no health complaints admitted to our hospital for obligatory health screening before military service. On physical examination, there was a fixed pronation of 30 degrees in the left arm, and the ability to supinate the left forearm and the hand was limited.

Imaging Findings:

Anteroposterior and lateral radiographs of the left elbow showed bony fusion between the proximal aspects of the radius and ulna.

Discussion:

Congenital proximal radioulnar synostosis (CPRUS) is a congenital deformity of the upper extremity characterised by bony fusion of the proximal ends of the radius and ulna. This rare abnormality is the result of the deficiency in the longitudinal segmentation of the radius and ulna during the 7th week of gestation. To date, approximately 350 cases of CPRUS have been reported [1,2]. The disorder may be detected either as an isolated abnormality or in association with various other musculoskeletal, heart (cardiac), neurologic, or gastrointestinal abnormalities. The other congenital musculoskeletal disorders with which CPRUS may occur are polydactyly, syndactyly, Madelung's deformity, carpal coalition, thumb aplasia, arthrogryposis, mandibulofascial dysostosis, congenital hip dislocation and clubfeet. It may also be detected as a component of some syndromes including Poland, Crouzon, Apert's, Carpenter's, Klinefelter, William's and Holt-Oram syndromes [3,4].

CPRUS is reported to be bilateral in nearly 60% of the cases. The most common complaint among the patients is the limitation in the forearm rotation with preserved flexion and extension functions of the elbow joint. The diagnosis is based on the depiction of the bony fusion of the proximal ends of radius and ulna on direct radiograms (Figures 1 and 2), and a detailed imaging may be performed by computerised tomography [2]. Cleary and Omer described four different radiographic patterns of CPRUS, depending on the presence of synostosis and the location of the radial head: type 1, no bony involvement and the radial head is in normal location; type 2, synostosis with normal radial head location; type 3, synostosis with posteriorly dislocated hypoplastic radial head; and type 4, synostosis with anteriorly dislocated radial head [5].
In the cases with mild deformity, the patients usually compensate with the adjusted functions of the shoulder and wrist joints. In order to avoid overloading these joints, occupational therapy and modification of the activities are recommended [2,6]. In most of these patients, functional adaptation has been reported to reach a good level [7]. However, in the cases with severe disability, surgical correction of synostosis by proximal derotational osteotomy is indicated [5-7]. But unfortunately, success rates of surgery in regaining the rotatory function is reported to be low [2,6].

Written informed patient consent for publication has been obtained.

**Differential Diagnosis List:** Congenital proximal radioulnar synostosis, Posttraumatic proximal radioulnar synostosis, Postsurgical proximal radioulnar synostosis

**Final Diagnosis:** Congenital proximal radioulnar synostosis

**References:**

Description: Bony fusion is seen between the proximal aspects of the radius and ulna. Origin: Department of Radiology, D??kap? Y?ld?r?m Beyaz?t Training and Research Hospital, University of Health Sciences, Ankara/ Turkey 2018.
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