CONGENITAL BILATERAL RADIOULNAR SYNOSTOSIS: a case report

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Abstract: Congenital RadioulnarSynostosisis a bony connection between the radius and ulna. In 1793 for the first time Sandifort explained the initial description of congenital radialulnarsynostosis[1,2]. The condition may be unilateral or bilateral that is more common. Functional defects associated with the anomaly depend on the severity of the deformity and on whether or not it is bilateral[3,4,5]. The condition may associate with other abnormalities and syndromes but we didn't find in our patient any of them[6-11]. Important view of this manuscript was rarity of this anomaly and also in patient opinion is unimportant disturbance that caused him didn't refer to specialist since long past.

Keywords: factors; Radioulnar Synostosis; Congenital; bony connection

CASE REPORT:

A 22 years old Iranian male student referred to outpatient orthopedic clinic at jun 10, 2012 because of limitation in range of motion of his arms that he said he have this problem since childhood and its not recent problem. At present he has no weakness or loss of feeling or tingling and also pain in his hands or arms. He denies major elbow trauma or fractures or dislocations and also denies any similar problem in his family. The patient states that movements are difficult especially elbow pronation. He is right hand dominant and has no significant past medical history and has normal growth. We examined the patient. Both of his arms, forearms and grip have Strength 5/5. In neurovascular he is healthy person. His elbows have limited range of motion such as elbows in supination were completely fixed in both active and passive movement but both of his elbows have fixed in 20 degrees of pronation (figures 1,2,3). Range of motion in flexion of both elbows from0-100degrees. Wrists motion have 70 degrees range in pronation and 20-30 degrees in supination and in flexion was near normal (figures 2,3). radiographies demonstrate that both elbows have an osseous synostosis in proximal borders of radius and ulna (figures 5,6).

Comment:

RadioulnarSynostosis is fusion of the radius and ulna at their proximal margins. On the other hand in this condition a bony connection exist between these two bones [1,2]. This anomaly includes two types: congenital (is present at birth) and post traumatic (occur between these two bones along the length of the interosseous membrane) that are separate entity from others [12]. For the first time Sandifort explained the initial description of congenital radialulnarsynostosis in 1793[1]. This condition is caused by failure in Separation between radius and ulna between the 5th and 8th week of gestational age, because we know that upper limb bud arises from the un segmented body wall at 25-28 days and radius and ulna are paired in this time but should be separate and any disturbance in segmentation in this period cause synostosis of these two bones[2,3,13]. Initially we have cartilaginous synostosis that changes to ossified tissue gradually with bone maturation and time passes. The disease usually is not discovered until early adolescence, when the patient presents with Limited movement in forearm[14].2 types of congenital synostosisexist. In type 1, the radius and ulna fusion are in proximal margin but with complete synostosisand a variable distance. Type 2 is so milder and two bones union is partially and this type is



Figure 1:Photograph of the patient's forearms at 22 years of age. Forearms in nuetr position.



Figure 2:Photograph of the patient's forearms in a pronation position.



Figure 3:Photograph of the patient's forearms in a supination position. Limited range of motion is clearly obvious.



Figure 4: Anteroposterior and lateral radiographies of right upper limb. These radiographies shows bony connection between the radius and ulna at proximal border.



Figure 5: Anteroposterior and lateral radiographies of left upper limb. These radiographies shows bony connection between the radius and ulna at proximal border.

associated with radial head dislocation that caused anterior or posterior dislocation and is lower than proximal radial epiphysis [6,7,12].

This condition occurs rarely and almost 350 cases reported in the presented manuscripts. One problem in this condition is delayed diagnosis that caused by the rarity of the disease. Range of patient age at diagnosis time is 6 months to 22 yearsold. We don't know the accurate incidence radioulnarsynostosisbut there is no preference in sex of congenital synostosis [6,8].In 50-60% of patients are bilateral. Generally this synostosis occurs sporadic but in some cases has autosomal dominant inheritance[14]. This condition may be an an isolated abnormality or associate with abnormalities and syndromes because of embryonic nature of it. This anomalies include DDH, clubfoot, polydactyly, syndactyly, Madelung deformity, laxity of ligament, thumb hypoplasia, carpal coalition, cardiac, renal, neurologic and GI disease and Apert syndrome, multiple exostoses, arthrogryposis, mandibulofacialdysostosis, acrocephalosyndactyly, William syndrome, Klinefelter syndrome, Carpenter syndrome, microcephaly[6-11,13].

studv we reported a congenital radioulnarsynostosis in a 22 years old Iranian male student that referred to outpatient orthopedic clinic at jun 10, 2012 because of limitated range of motion of his both arms. Thispatients was young and had long bone maturation and growth plate radiological closure was observed. The radiologic findingrevealeda bilateral bony synostosis in the proximal border of the radius and ulna that cause decrease in his range of motion. He has no significant past medical history and has normal growth. This condition is caused by failure in Separation between radius and ulna and may associate with abnormalities and syndromes that we did not find any similar problems in this patient and also he denied any equivalent problem in his family. One of important point in this manuscript is that we must find similar cases and find their concomitant functional problem and if is possible solve them before to be late.

Consent:

Written informed consent was obtained from our patient for publication of this manuscript and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Conflict of Interests: None of the authors of the paper have declared any conflict of interests within the last years, which may arise from being named as an author of the manuscript.

Authors' contributions:

Mohamadhosseinkariminasab ,MasoudShayeste-Azar Seyedmohamadmehdidaneshpoor interpreted the patients data and was a major contributor to the writing of the paper and gave approval for the final paper. shadiShayeste-Azar and collected the clinical data. Seyedmohamadmehdidaneshpoor and MajidSajjadisaraviobtained and interpreted radiological studies. Mohamadhosseinkariminasabreviewed the literature and substantial contributions to conception and design. All authors read and approved the final paper.

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References:

- 1. Wang E, Wenger DR, Zhang L, Zhao Q, Ji S, Li J. The mechanism of acute elbow flexion contracture in children with congenital proximal radioulnarsynostosis. *J PediatrOrthop*. Apr-May 2010;30(3):277-81.
- 2. Shinohara T, Horii E, Tatebe M, Yamamoto M, Okui N, Hirata H. Painful snapping elbow in patients with congenital radioulnarsynostosis: report of two cases. *J Hand Surg Am*. Aug 2010;35(8):1336-9.
- 3.Bayne LG, Costas B, L,, Lourie GM. The upper limb. In: Morrissy RT, Weinstein SL, editors. Pediatric Orthopaedics. Philadelphia: Lippincott-Raven; 1996. p. 781-847.
- 4.Bolano LE. Congenital proximal radioulnarsynostosis: treatment with the Ilizarov method. Journal of Hand Surgery -American Volume 1994; 19(6): 977-8.
- 5.Lescault E, Mulligan J, Williams G. Congenital radioulnarsynostosis in an active duty soldier: case report and literature review. Military Medicine 2000; 165(5): 425-8.

- 6.Cleary JE, Omer GE. Congenital proximal radioulnar synostosis.Natural history and functional assessment. *J Bone Joint Surg Am*. Apr 1985;67(4):539-45.
- 7. Dohn P, Khiami F, Rolland E, Goubier JN. Adult post-traumatic radioulnar synostosis. Orthop Traumatol Surg Res. 2012;98(6):709-14.
- 8. Wierer M, Huber-Wagner S, Mutschler W. Post-traumatic proximal radioulnar synostosis. Surgical technique and review of the literature. Unfallchirurg. 2012;115(5):451-6.
- 9.Giuffre L, Corsello G, Giuffre M, et al. New syndrome: autosomal dominant microcephaly and radio-ulnar synostosis. *Am J Med Genet*. Jul 1 1994;51(3):266-9.
- 10.Selicorni A, Ferrarini A, Cagnoli G, Fratoni A, Bottigelli M, Milani D.Additional case of Tsukahara's syndrome or new syndrome: further delineation of the association of microcephaly and radio-ulnar synostosis. Am J Med Genet A. 2005 Jan 15;132A(2):189-90.
- 11.Gaspar H, Albermann K, Baumer A, Schinzel A. Clinical delineation of Giuffrè-Tsukahara syndrome: another case with microcephaly and radio-ulnar synostosis with apparent X-linked semi-dominant inheritance. Am J Med Genet A. 2008 Jun 1;146A(11):1453-7.
- 12._Siemianowicz A, Wawrzynek W, Besler k. Congenital radioulnar synostosis case report. Pol J Radiol. 2010; 75(4): 51–54.
- 13.Koç A, Kaymak AO, Karaer K, Ergün MA, Aksu T, Perçin EF.A case with bilateral radio ulnar synostosis.Genet Couns. 2008;19(2):193-8.
- 14.Miura T, Nakamura R, Suzuki M. Congenital radio-ulnar synostosis. *J Hand Surg*[Br]. Jun 1984;9(2):153-5.