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REVIEW ARTICLE

CONGENITAL RADIOULNAR SYNOSTOSIS

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Abstract

There are two different types of synostosis. In these cases the symptoms are mild because the compensate thein limitation in pronosupination with hypermobility of other joints of the upper limb. X-rays of upper limbs are the diagnostic procedures. The management will be conservative, but the treatment depends on the degree of functionality of the forearm.

Keywords: congenital radioulnar sinostosis, diagnostic radiology.

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1 | INTRODUCTION

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congenital radioulnar synostosis is an uncommon congenital anomaly, that causes a limitation of the forearm supination, often bilateral. The origin of this anomaly occurs when the differentiation of the cartilaginous contour of the radius and ulna does not take place during the embryonic period.

Objective: to show two congenital radioulnar synostosis cases analyzing variable about the topic.

Conclusions: proximal radioulnar synostosis is a non frequent affection, that brings about difficulties carrying out actions with the hands, Its diagnosis is relatively simple by means of the clinical findings andplain X-rays of the elbow.

The lack of separation of proximal and of the radio and ulnar is a non frequent disorder that causes rigidity in the pronation position of the forearm with variable degrees. It generally occurs due to the longitudinal stop of the segmentation process of the bone in the proximal part of the forearm. This rare congenital defect is detected between the four or five years of age when the functional requests increase. Frequently this discapability is compensated by the supination of the shoulder’s rotation (1) .

Supplementary information The online version of this article (10.15520/mcrr.v4i5.125) contains supplementary material, which is available to authorized users.

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The anatomic lesion is represented is the bone fusion between the ulna and the proximal radio**,** that causes fixed pronation of the limb, the trabecular framework between bones is visible. In other cases, this fision can be only fibrous, the ulna has a normal aspect, but the radius is arched on its axis; sometimes it has a prominent head, atrophic ori t does not exist. In relation to muscle lesions atrophy of pronators and supinators or theis abscence be seen (2) .

CASE PRESENTATION

Patient's information (I). 6 year old child with a family history of a maternal uncle and a mother´s uncle who had a history of decreased mobility of the forearm. The child presented difficulties when brushing his teeth and back handed position when placing bottler and toys. There is no previous history of trauma, pain or swelling.

Clinical findings. Child whith fixed pronation of both forearms in a prone position between 15- 20 degres. Moderate hand disability for everyday life, shortening and bending of forearms, hypermobility of the wrist and painless restriction of motion

Diagnostic evaluation. Plain radiology offers a safe diagnosis of affection in both elbow. Fusión of bone of forearm in proximal zone and also, the bending of radius´s diaphisis.

Therapeutic intervention. Suggested conservative management according to mild synostosis, ergonomics and activity modification was included.

Follow up and sequels . The patient was attended in consultation for more 15 years, keeping a life close to the normality.

Patient's information (II).

Clinical findings. 16- years old male student, that come to consultation presenting difficulty to perform certain motion with movements with the left upper limb. His mother said that he makes strange movements with the left upper limp since childhood but has never hindered him to have life.

Diagnostic evaluation. On examining it was notice a fixed prone position of the forearm, with a loss of the muscular contor when compared with the right upper limb. No pain.

When suggest to make some movements, it was found anchylosis in discreet pronation of the left-hand forearm. Simple X- rays of both elbows were takes and copared. There was a trabecular bone without articulation between the radius and compared. These was a trabecular bone without articulation between the radius and the proximal ulna, a vestige of the head of radius was appreciated.

Therapeutic intervention. Management was only rehabilitative to promote compensatory movement of the shoulder and to compensate for ankilosis in the radioulnar portion of the left elbow.

Follow up and sequels. The prognosis is good.

2 | DISCUSSION

The pathology presented and named as proximal radioulnar synostosis is also known as Sandifort-Lennoire deformity, the first autor recognized the pathology in 1793 and the second understood it as a cause of sickests.

The description of the development that the embryology of the upper limb bud arises from the unsegmented body wall at weeks four. The elbow becomes visible at weeks five, initially, the three cartilaginous buds of the humerus, radius, and ulna are connected before segmentation. Therefore, the radius and ulna share a common perichondrium. Abnormal events at that time may lead to a failure of segmentation. Duration and severity of the insult may determine the degree of subsequent synostosis.

Etiology of the forearm begins as a single cartilaginous but and divides from distal to proximal into the radius and ulna at week seven in the maternal uterus, so failure in differentiation results in synostosis in proximal aspect of the forearm. Frecuently there are other syndromes (30%), for example, Apert syndrome (acrocephalo syndactyly), Arthrogryposis and Carpenter's syndrome (acropoly syndactyly).

Acording to these pathogenies explanations, because the detaintment of development is contradictory due to the fact that in theinitial stage both bones are joined to thein cartilaginous outline, but in supination, while fusion can be seen in pronation.

Clinically, the elbow goes forward with a variable degree of pronation, totally blocked, in contrast,

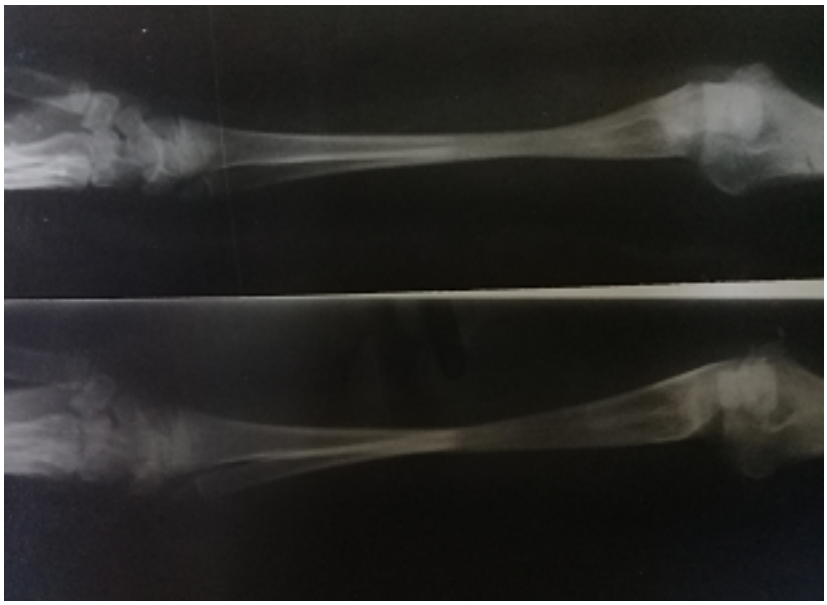

FIGURE 1: The non existence of radial cúpula in the antero lateral view.

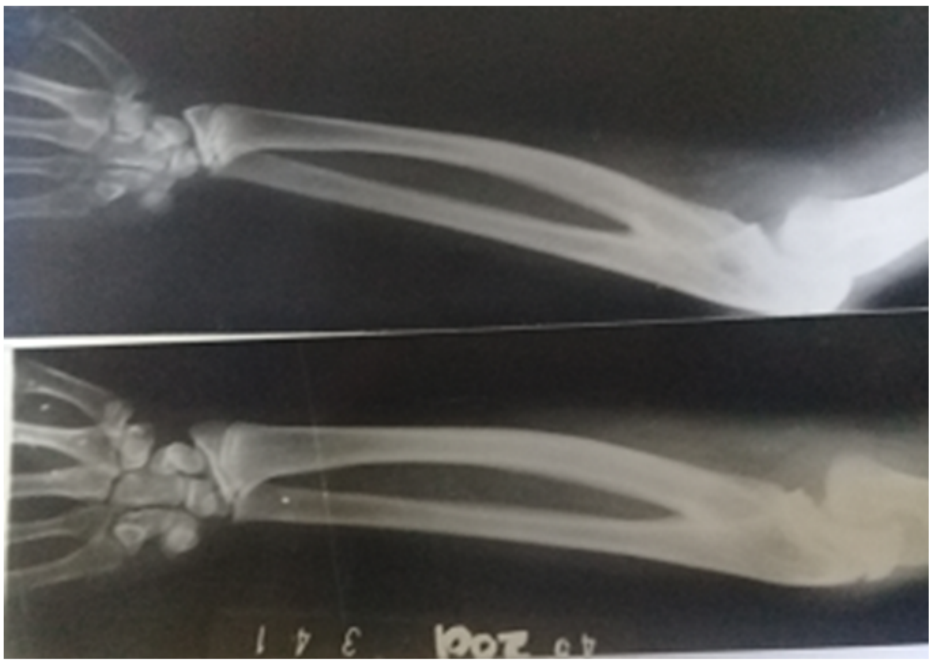

FIGURE 2: There is fusión of radius and ulna, tipe III sinostosis.

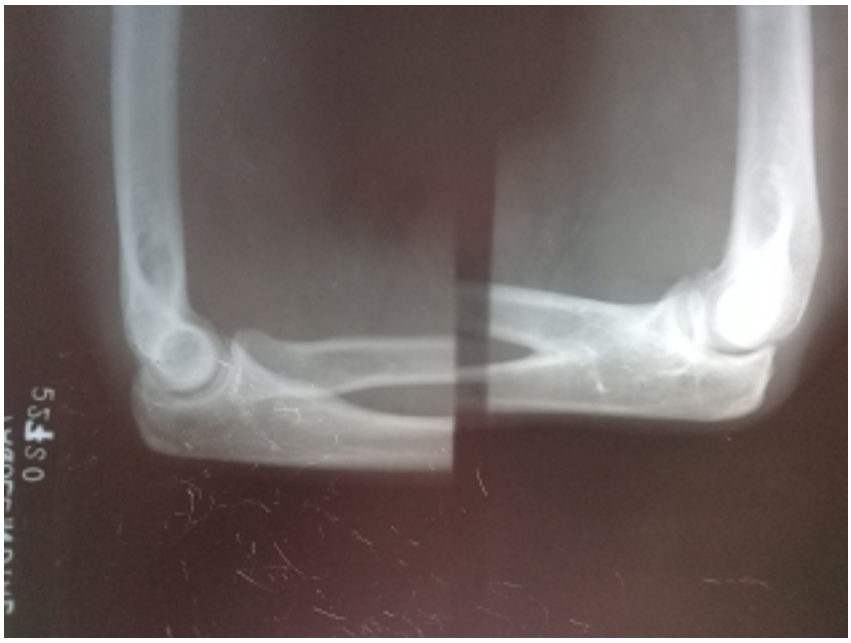

FIGURE 3: Note of theleft elbow proximal radioulnar synostosis, and observe the difference of a normal elbow in other X- rays.

there is integrity of the flexion and the extension. The differential diagnosis is among acquired, traumatic or infectious synostosis. [3]

There is no pain, it is commonly asymptomatic recognized by parents and teachers. The childsuffers from difficulties to perform specific tasks, like using a keyboard, failure for supine activities a deficient pronation, when eating, washing his face, catching a ball.

Standard age for clinical examination is at age 6 years , because in smaller children signs and symptoms may by unnoticed until early teenage, especially in unilateral cases. The motion of the elbow is usually reserved, there is fixed pronation of the forearm around 30° commonly; there is compensatory abduction, the motion of the shoulder compensates the loss of pronation with active abduction, and there may be also hypermobility of the wrist (4) .

Three kinds of congenital proximal radioulnar synostosis, in type I there is no head of the radius and there is completes and uniform fusion between the radius and ulna; it is the most severe lession. In type II the higher portion of the radius is present but more or less deformed and there is fission of both bones in the neck of the radius. In the type III, the head of the radius is deformed and subluxed and the fusion occurs in the proximal area of interosseous membrane.

Cleary and Omer Classification is based on appearance of the synostosis and radial head reduction, Type I- Lacks of bone involvement, reduced normal appearance of radial head, Type II- Distinct bone synostosis, but otherwise normal findings, Type III- Distinct bone synostosis with hypoplastic and posteriorly dislocated radial head and Type IV- Short osseous synostosis, anteriorly dislocated radial head, usually with a mushroom shaped deformity.

Wilkie Classification offers 2 types.

- Type I: Lack of proximal portion of radius, bone fusión of 3-6 cm, and radius and ulna are connected at medullary canal.
- Type II: Normal radius, synostosis is located just distal to proximal radial epiphysis and the radial head is dislocated anteriorly or posteriorly.

The treatment is under discussion because the need of practice some motion or procedure, is questionned because the lack of supination is compensated minimally with the shoulder's rotary motion (5) .

In children with bilateral congenital radioulnar synostosis, surgeons have traditionally recommended the reposition of the forearms in supination. How-

ever, the author considers that this position is not advisable nowadays because working with the computer´s keyboard stops a bilateral pronation. In general the surgery is not indicated, due to the adaptation that offers the abduction movement of the shoulder (6) .

The recommendation of resection the proximal portion of the radius must be accompanied by the resection of the bone membrane along the ulna, but it is also necessary to operate the soft parts. There are other surgical interventions as Galeazzi's operation, another surgical possibilities as Kelikian's intervention or Palagi's intervention (7) .

Pasupathy B, Tholgappiyan T, Sureshbabu M, assessed the functional outcome using double rotation osteotomy and osteotomy at synostosis site in congenital radio ulnar sinosthosis, but the double osteotomy at both radius and ulna should be reserved as a choice for older children with bilateral hyperpronation deformity (8) .

Surgical treatment would be indicated if there is a severe deformity in pronation that causes serious functional failures.

3 | CONCLUSIONS

Congenital proximal radioulnar synostosis is a nonfrequent affection, that causes restriction for the performance of actions with the hands. It´s diagnosis is very simple by means of clinical procedure and plain simple radiology of the elbow. There are several methods of treatment, but in the results are not satisfactory. It is a well tolerated condition therefore, the behaviour in both cases was conservative and expectant. the publication's principal contribution is presentation two - cases too little frequent.

Contribution of the authors: the authors assisted the cases and wrote the paper and analysis of the documents.

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