CASE REPORT: CONGENITAL RADIOULNAR SYNOSTOSIS AND ITS EMBRYOLOGICAL CORRELATION AND FUNCTIONAL ASSESSMENT

C R Bhatt, C D Mehta
Department of anatomy, Govt. Medical College, Surat.

ABSTRACT

Congenital radioulnar synostosis(CRS) is a rare anomaly and approximately 400 cases were reported worldwide so far. CRS is the failure of the longitudinal segmentation and the persistence of the cartilaginous anlage between the radius and ulna during the seventh week of development that results in a persistent bridge of tissue. Here we are discussing on a case of 25yrs old, male patient with bilateral congenital synostosis. On the left hand the pronation and supination movements are restricted completely where as on right side 10 degree of supination and 20 degree of pronation is possible.Radiologically in our patient synostosis is classified as type II variety by Wilkie(1914)[1] classification and type IV by the Cleary and Omer classification(1985). The position of forearm was not found to be related to subjective functional limitation, or employment status. Main line of treatment is surgical mainly rotational osteotomy but is rarely indicated. Our patient is not able to rotate his forearm especially on the left side still he has no functional limitation so he has refused for the operative treatment. No study has objectively compared the preoperative functional limitation of the patients with their postoperative functional improvement in order to justify surgical intervention In the authors opinion the only major factor that is to be taken into consideration of operative treatment is functional limitation to the patient.

KEY WORDS: Congenital Radioulnar Synostosis, Embryological basis

INTRODUCTION

Congenital synostosis of proximal radius and ulna is a rare malformation. The malformation caused by failure of normal prenatal separation of the radius and ulna.

The persistent connection between the two bones is nearly always proximal; while distal radioulnar synostosis is extremely rare. The connection is initially cartilaginous and is not diagnosed until it ossifies, forming a bony synostosis. CRS often results in functional, cosmetic, and cultural limitations. CRS is often part of syndromes such as Crouzon, Apert's and Poland's . Although 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 embryonic period the forearm is in pronation and the same position is found in almost all radio-ulnar synostosis.[1] Proximal one third of forearm is the most common site of involvement, with male predilection. Patient generally present around the age of 3 years with functional problems such as difficulty in holding objects with both hands and in dressing and feeding themselves. Patients are usually seen holding objects with a backhanded posture.

Correspondence
Dr. Chintan R Bhatt
A-19/ Nitinagar Row House, Opp. Partibha Farm House
Near Sai Ashish Society, University Road,
Surat -17, Gujrat
Mob. : 09825649578

DEVELOPMENT

The development of the upper extremity has been well reviewed. Lewis[2](1901) described the humerus, radius and ulna as being continuous with each other and joined by a common perichondrium, at five weeks of gestation. By six weeks the cartilaginous anlage of the three bones are separated by condensations of tissue, and no joint cavitiy is yet visible. The forearm is in a neutral position at this time, although rotation into pronation occurs by eight weeks due to growth discrepancy between the arterial tree and the radius.[3]

It is the failure of the longitudinal segmentation and the persistence of the cartilaginous anlage between the radius and ulna during the seventh week of development that results in a persistent bridge of tissue.[4] Usually this will ossify into an osseous synostosis although fibrous synostosis are well recognized also. A fixed, pronated forearm is thought to reflect developmental arrest at this specific time of the fetal development. The frequently associated deformity of the radial head may be due to early interference with joint formation that results in a complete proximal coalition[5], or to limited fusion distal to the epiphysis that results in unequal growth of the radial head.

Genetic basis has been reported by numbers of

authors in the CRS. Evidence for etiology includes the common family history and the frequent association with other congenital syndromes. In a number of cases it is associated with chromosomal abnormalities, more especially multiple X-Y syndromes[6]

CASE REPORT

A 25 year old left handed person diagnosed with bilateral congenital radioulnar synostosis. The patient was diagnosed at the age of 2 years when his mother noticed that he could not supinate his hands. His nearby pediatrician diagnosed him as a case of CRS. (Figure: 1. Elbow joint; 2. proximal part of ulna; 3.shaft of radius 4. Wrist joint). There was no history of drug intake or viral fever during pregnancy. He has no other congenital anomaly. The rest of his physical examination was essentially normal with the exception of his extremities.

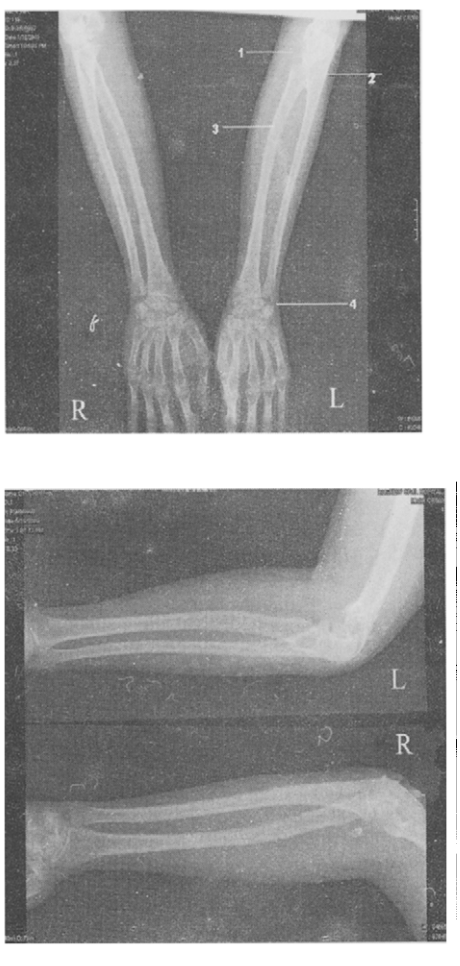

Figure: 1. Elbow joint; 2. proximal part of ulna; 3.shaft of radius 4. Wrist joint

He had full active range of motion of both his shoulder and wrist bilaterally. Additionally his elbow showed full active range of movement in flexion and extension. Both of his forearms were noted to be semi fixated in an anatomically neutral position. The Left forearm demonstrated no movement of pronation or supination whereas the right forearm demonstrates 10 degree of supination and 20 degree of pronation. There was no objectively observable muscle atrophy. However, this individual functionally increases range of movement through compensatory motions at the shoulder and wrist.

DISCUSSION:

Sandifort originally described CRS in 1793 as a rare congenital deformity. Till now fewer than 400 cases of CRS had been reported. However it is the most common congenital anomaly that functionally impairs the elbow.

Radio-ulnar synostosis is regarded as an anomaly of longitudinal segmentation. It is presumed that some factor inhibits the interzonal mesenchyme between the cartilage anlage of the developing radius and ulna from undergoing dissolution during the seventh week of intrauterine life. Thus, the interzonal mesenchyme persists and undergoes chondrification, ossification, and eventually synostosis. The proximal one-third of the forearm is the most common site of involvement. About 40 per cent of the cases are unilateral and 60 percent bilateral. Males are affected more commonly than females. Approximately one-third of patients with radio-ulnar synostosis have associated anomalies involving the cardiovascular, genitourinary, gastrointestinal, central nervous and musculoskeletal systems. Our patient has none.

Wilkie in 1914 had described two types of synostosis based on radiographic appearance.[1] Our patient has bilateral Type-II deformity.

Cleary and Omer in 1985[7] observed four distinct patterns of radio-ulnar synostosis radiologically. Our patient had bilateral Type IV radio-ulnar synostosis as per Cleary's classification.

Treatment options for CRS are limited to surgery, with the most dependable option being rotational osteotomy through the synostosis site[8] .Because most affected individuals have adapted through increased mobility in the wrist and shoulders, the indications for surgery have not been clearly established. Consideration must be given to the functional and cosmetic effects of the synostosis.

Surgical intervention is rarely indicated for unilateral CRS because of compensation through the contralateral limb. However, most clinicians agree that any forearm fixated beyond 70 degrees of pronation is an indication for surgery.[9] But in our case patient has extensive synostosis but no functional deformity and so has denied any surgical intervention. Although patient in this study demonstrated no pathological findings in the wrist or shoulder the presence of proximal or distal joint disease would be a relative indication to improve the position of a forearm that cannot be actively compensated for by motion of an adjacent joint.

CONCLUSION:

Congenital radioulnar synostosis is a rare deformity, frequently bilateral, and more commonly seen in male patients, with multifactorial etiology including both sporadic, mutation and undefined genetic patterns. It is because of the failure of the longitudinal segmentation and the persistence of the cartilaginous anlage between the radius and ulna during the seventh week of development. The position of the forearm or severity of synostosis was not found to be related to subjective functional limitations. The operative treatment for synostosis is rarely indicated, less emphasis should be placed on the single factor of the position of the forearm, and the functional limitation to the day to day activities must be taken into the consideration for the operation. Our patient had denied surgical correction because he has no objective or functional limitations of movements inspite of having sever degree of synostosis. No study has objectively compared the preoperative functional limitation of the patients with their postoperative functional improvement in order to justify surgical intervention. So main indication of the operative treatment is functional impairment because of the synostosis.

REFERENCES:

1. Wilkie DPD. Congenital Radio Ulnar Synostosis. Br J Surg 1914;1:366-375
2. Lewis WH. The development of the arm in man. Am J Anat., 1: 145-183,1901
3. LOVELL W.W. and WINTER R B (editor): Pediatrics Orthopaedics. Philadelphia, J.B. Lippincott, 1978.
4. MITAL M.A.: Congenital Radioulnar Synostosis and Congenital Dislocation of the Radial Head.Orthop.Clin.North.America. 7: 375-383,1976,
5. SIMMONS B.P.: SOUTHMAYD,W.W.;and RISEBOROUGH,E.J.: Congenital Radioulnar Synostosis. J.Hand Surg., 8: 829-838,1983
6. Online Mendelian Inheritance in Man, OMIM(TM). Johns Hopkins University, Baltimore, MD. Radioulnar synostosis: MIM Number:179300.http://www.ncbi.nik.gov/entrez/dispomim.cgi,id-179300
7. Cleary JE, Omer GE Jr. Congenital proximal radioulnar synostosis. Natural history and functional assessment. J Bone Joint Surg. 67!: 539-545, 1985.
8. Green WT, Mital MA. Congenital radio-ulnar synostosis: Surgical treatment. J Bone Joint Surg 61A:738-743,1979
9. Ogino T. Hikino K: Congenital radio-ulnar synostosis: compensatory rotation around the wrist and rotation osteotomy. J Hand Surg (Br).12:173-178,198