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Congenital Bilateral Proximal Radio-Ulnar Synostosis in a Nigerian Child

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Abstract

Congenital radio-ulnar synostosis (CRUS) is a rare skeletal malformation of the upper limb and the most common congenital functional disorder of the elbow joint, causing limitation in forearm rotational movements, which may lead to difficulties with some activities of daily living. We reported a 4-year-old girl with congenital bilateral proximal radio-ulnar synostosis who presented with functional discomfort due to limitation of the prono-supination movements of the forearms and abnormal gestures when handling objects. She has clinical and radiological features of congenital radio-ulnar synostosis (CRUS). However, the parents declined corrective surgery because of wrong perception of the condition to be normal. This case highlighted the poor health seeking behavior and wrong illness perception prevalent in the developing countries.

Keywords

Congenital Radio-Ulnar Synostosis, Upper Limb Congenital Anomalies, Prono-Supination

1. Introduction

Radio-ulnar synostosis (RUS) is the osseous union between the radius and ulna; and can occur in two general forms namely: congenital and post-traumatic radioulnar synostosis [1]. Congenital radioulnar synostosis (CRUS) is a rare condition in which there is an abnormal connection of the radius and ulna at birth leading to limited rotational movements of the forearm which may produce

some difficulties in daily living activities [1] [2] [3]. This condition is caused by a failure of segmentation between the radius and the ulna [4]. Sandifort, in 1793 originally described the congenital radioulnar synostosis [3]. More than 400 cases of CRUS have been reported in the world literature [5] [6]. The average age at diagnosis of CRUS is 6 years with a range from 6 months to 22 years [7] despite the fact that the malformation is present at birth.

The functional deficits associated with CRUS depend on the severity of the malformation and whether or not it is bilateral; and the management may be conservative or surgical [8].

The frequency of CRUS among all congenital upper limb deformities ranges between 1.12% and 9.35%. Among other musculoskeletal deformities, the frequency of CRUS ranges between 0.11% and 0.61% [8]. Although its exact aetiology is not clear, usually cases of CRUS are sporadic, but in a number of instances, inherited synostosis has been reported [6] [8]. A number of authors have reported a male preponderance of the disease while some reports have indicated that the disease has no sex predilection [1].

Currently, the attitude towards surgical intervention in patients with CRUS is controversial among orthopaedic surgeons. Indications for surgical treatment are related to bilaterality, the degree of deformity and the severity of the limitation of functions at the elbow joint(s) [1] [8]. Therefore, indications for surgery are based on individual functional limitations than on absolute forearm position [1] [8].

This case is reported to highlight the poor health-seeking behavior and illness perception of the biological parents which prevented compliance for comprehensive evaluation of the patient and the necessary follow-up to monitor the progression of the disease which is the prerequisite for appropriate management and counseling of the child for vocational career.

2. Case Report

A 4-year-old girl presented at the out-patient clinic of the department of Paediatrics and Child Health, Ladoke Akintola University of Technology (LAUTECH) Teaching Hospital, Ogbomoso with complaints of persistently and progressively having difficulty in lifting objects, turning both arms laterally, to supinate properly especially when feeding, accepting objects into an open palm or when trying to extend her hands outwardly noticed about 3 years prior to presentation. However, she had no difficulty in holding small objects and no significant difficulty in writing. No associated hyper-mobility of the wrists. There was no history of trauma, pain, or swelling of the arms nor previous fracture or surgical intervention. There was no history suggestive of bony abnormalities in other parts of the body. Past medical history and medication were of no significance. However, because the limitation in the forearms movement was painless, it was perceived not to be a serious problem and more so with very limited family income; her biological parents felt therefore, that it was unnecessary to seek medical help but she was later brought to our hospital by the maternal aunt.

The pregnancy including obstetric ultrasound was reported normal. The child's

growth and developmental milestones had been normal since birth.

The patient belong to low socioeconomic family and the total average monthly income of both parents was eighty thousand Naira (N80,000.00), about 78 U.S. dollars to care for the family of 2 children. Patient is the first of the two siblings in a monogamous family setting. Both parents possess National Certificate of Education (NCE) and were both teachers in elementary schools. No other family member had decreased forearm mobility or obvious skeletal malformation.

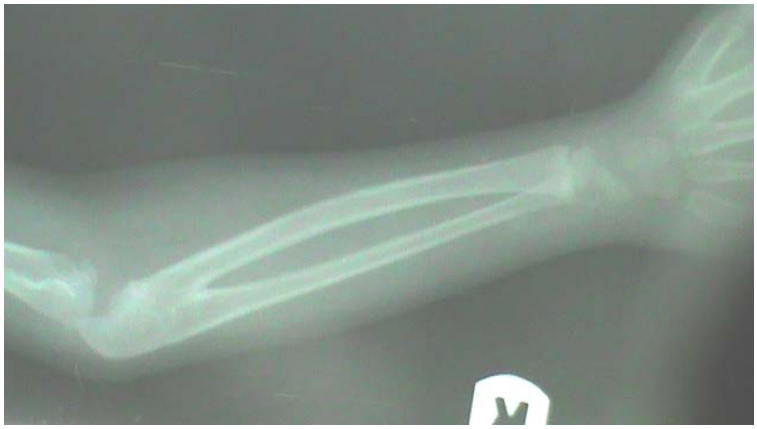
On physical examination, the child appeared well but had limited extension at the elbow joints, reduced forearm range of movement in pronation and supination bilaterally with supination worse than pronation. There was bilateral medial angular deviation of the elbows as well as exaggerated abnormal "carrying angle" of the elbows. There was reduction in the bulk of brachioradialis muscle bilaterally. No area of tenderness across the entire upper limbs and no loss of sensation. There was full range of movements across the shoulder, wrist and the small joints of the hands. The muscle power was grade 5/5 in all the limbs. There was no abnormality detected along the spine.

The radiographs of both upper limbs revealed a bony fusion of the radial head with the olecranon process of the ulna bilaterally. The medulla continued into the adjacent bone. There was associated posterior subluxation of the radial head bilaterally (**Figure 1(a)** & **Figure 1(b)**). The distal portion of the radius and ulna with the intervening shafts appeared normal. Both humeral outlines and shoulder joints, the wrists and the demonstrated metacarpals were preserved. There was no fracture line or other abnormality noted. The radiological skeletal survey also revealed no additional bony abnormality nor associated systemic disorders.

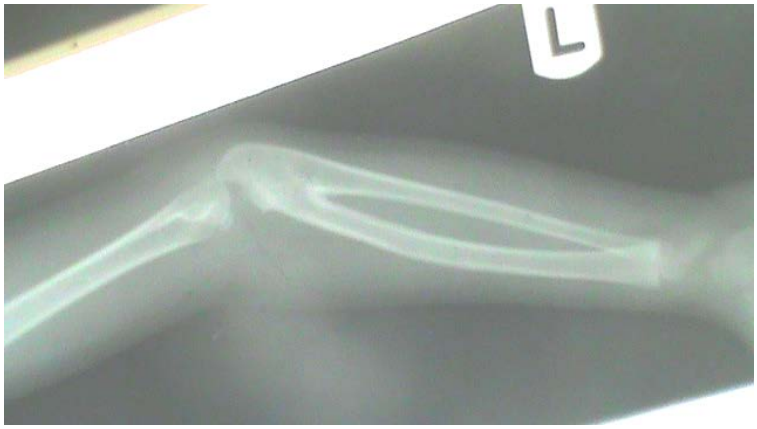
Total serum calcium was marginally low (1.9 mmol/L); but the serum phosphate (0.65 mmol/L) was within normal range. The haematological index was within normal range. Serum Electrolytes, Urea and Creatinine were within normal limit. The results of the laboratory investigations are shown in **Table 1**.

Table 1. Investigation results.

S/N	Investigation	Value	Reference rang	Remark
	Electrolyte, urea, creatinine, phosphate, alkaline phosphatase			
1	Sodium	135 mmol/L	120 - 140 mmol/L	Normal
	Chloride	97 mmol/L	90 - 110 mmol/L	Normal
	Bicarbonate	24 mmol/L	20 - 30 mmol/L	Normal
	Calcium	1.9 mmol/L	2.2 - 2.6 mmol/L	Low
	Phosphate	0.65 mmol/L	1.12 - 1.45 mmol/L	Low
	Alkaline Phosphatase	146 IU/L	44 - 47 IU/L	Normal
	Haematological Profile			
2	WBC total	6500/mm^3	6000 - 17,000	
	Neutrophil	40%	40 - 75	Normal
	Lymphocyte	60%	50 - 60	Normal
	Plateletes	400,000/mm^3	150,000 - 450,000	Normal



(a)



(b)

Figure 1. (a) Lateral view of the radioulnar synostosis; (b) Lateral view of the radioulnar synostosis.

The parents however, still perceived the illness (malformation) not to be a serious one and declined further evaluation of the child including referral to orthopaedic surgeon or follow-up at the Physiotherapy Unit despite extensive counselling. Patient was thereafter lost to follow up.

3. Discussion

The present case is a congenital bilateral proximal radio-ulnar synostosis. CRUS is a rare skeletal malformation caused by failure of segmentation between the radius and ulna [5]. However, it is the most common congenital functional disorder of the elbow joint [2]. Our patient presented with bilateral malformation. CRUS is bilateral in 60% - 80% of cases [3] [6] [7] [9]. The aetiology is not known [9]. It is a malformation which develops in the early foetal life between 34th and 37th days of the intrauterine age. Any influence of adverse factors in this period leads to a disturbed segmentation, which in turn causes malformation. The duration and severity of the insult determines the degree of subsequent synostosis. Endochondral ossification then proceeds and the cartilaginous synostosis ossifies either partially or completely, in the longitudinal or transverse

plane [1] [5] [10].

Our patient presented at age 4 year when functional limitation got worse although the mother had noticed the disease shortly after the age of 1 year. This suggests that the age at clinical diagnosis is determined by the severity of the limitation of movement [10]. The average age at diagnosis of CRUS is 6 years, with a range from 6 months to 22 years [1] [7].

In the low- and middle-income countries, where health Insurance is lacking, and the mode of payment for healthcare services is out-of-pocket, individual family's choice to seek healthcare for any member of the family has been shown to be influenced by factors such as beliefs, illness perception, proximity to healthcare facilities and illness severity. Additionally, socioeconomic class such as the household earning or income, perceived cost of treatment, gender of the decision maker, and perception of treatment efficacy will determine how early a family will seek healthcare for a sick member of their household [11] [12] [13] especially the child. The biological parents of our patient belong to the socio-economic class III. Their belief, illness perception and the household earnings were the main reasons identified for the delay in seeking medical care, refusal to comply with all the necessary investigations and to allow the child continue follow-up in the orthopaedic clinic. Although our patient was not having severe deformity and limitation of movement at the elbow joints at the age of 4 years when she presented to our hospital, some cases of CRUS, especially Cleary Type 1V [7] may have progressive decrease in elbow flexion which develops over time, mandating that such children be followed up in orthopaedic clinics until they achieve ssskeletal maturation [14] [15] [16]. Since our patient is still actively growing, her skeletal deformity and functional limitation may get worse, requiring orthopaedic specialist care in later life. Some authors consider it reasonable to start CRUS corrective surgery at the age of 3 years [8]. The radiological findings in our patient are in keeping with Cleary and Omer Type III bilaterally.

Various researchers have described and classified CRUS [7] [11] [12]. The original classification by Cleary and Omer [7] described four types radiologically: Type 1 is fibrous synostosis that does not involve bones and is characterized by normally appearing radial head; in type II there is bony synostosis, and the remaining bony structures do not reveal any other changes. Type III is osseous synostosis associated with a hypoplastic and posteriorly dislocated radial head. In type IV, there is a short bony synostosis with an anterior subluxation of the radial head.

CRUS commonly occurs between the proximal ends of the bones [1] as seen in the index case; while distal radioulnar synostosis is extremely rare [5].

It being an in-utero insult is expected to be associated with other abnormalities. Previous researchers have reported the association of CRUS with many orthopaedic and somatic diseases such as general skeletal abnormalities (hip dislocation, knee anomalies, clubfoot, polydactyly, syndactyly joint laxity) [1] [3] [8], chromosomal abnormalities (Klinefelter syndrome, Apert's syndrome, William's and Carpenter syndromes) [12] [14], haematological (acute lymphoblastic leu-

kaemia) [15], gastrointestinal, renal, neurological disorders [16], however, sporadic cases are common [8]. The disease (CRUS) was an isolated abnormality in our patient.

4. Conclusion

Congenital radio-ulnar synostosis remains a rare congenital bone abnormality with varying degrees of functional dysmobility and elbow deformity. It is commonly bilateral and appears clinically as a mild deformity in early life but with a delayed presentation may worsen. Thus, children with mild or subtle deformity or abnormal movement of the upper limb are encouraged to present early in life and should undergo a minimum of a plain radiograph of the affected limb(s) to rule out any skeletal deformity and to determine the modality of treatment. Early treatment may help the child to achieve more active use of the hands as well as for physical and mental growth.

Informed Consent

Informed consent was obtained from the parents of the patient for the manuscript.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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