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CONGENITAL PROXIMAL RADIOULNAR SYNOSTOSIS: RADIOGRAPHIC FEATURES IN TWO CASES WITH REVIEW OF THE LITERATURE

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ABSTRACT

Background: **Congenital radioulnar synostosis (CRUS) is an uncommon developmental defect caused by failure of segmentation between the radius and ulna during early embryonic life. Despite its rarity, it is the most prevalent congenital functional abnormality affecting the elbow. Clinical presentation varies widely, ranging from subtle cosmetic concerns to significant restriction of forearm rotation with consequent limitations in activities of daily living.**

Case 1: **A 4-year-old girl presented with inability to fully extend her left elbow since birth, accompanied by progressively worsening difficulty performing daily tasks involving the affected limb. There was also a history of absent left thumb noted at birth. Plain radiographs of the left forearm demonstrated symmetrical shortening of the radioulnar diaphysis, hypoplasia and posterior dislocation of the radial head, absence of the distal radial epiphysis, and osseous fusion of the proximal radius and ulna. Radiographs of both hands further revealed asymmetry of the left hand with aplasia of the first digit.**

Case 2: **A 13-year-old girl was evaluated for lifelong inability to extend both elbows, with increasing functional impairment of her forearms during routine activities. Plain radiographs of both forearms showed bilateral, symmetrical osseous fusion of the proximal radius and ulna with posteriorly dislocated radial heads. Additional findings included symmetrical bowing of the radial and ulnar shafts, penciling of the distal ulnar metaphysis, and positive ulnar variance.**

Conclusion: **These cases demonstrate the diverse clinical spectrum of CRUS and emphasize the critical role of radiographic assessment in accurate classification and detection of associated anomalies. Prompt diagnosis and comprehensive functional evaluation are vital for optimal management planning.**

Keywords: Congenital radioulnar synostosis, Musculoskeletal anomaly, Thumb aplasia, Children, Orthopaedics, Radiology

INTRODUCTION.

Congenital radioulnar synostosis (CRUS) is a rare congenital musculoskeletal abnormality[1] affecting the proximal radioulnar articulation, which may cause significant limitation of forearm rotation, particularly pronation and supination in affected individuals.[2] First described by Sandifort in 1793, the condition arises from failure of longitudinal segmentation of the cartilaginous anlage of the forearm during the fifth to seventh week of embryonic development, leading to persistence of a primitive osseous or fibrous bridge between the two bones.[3-6]

The precise aetiology of CRUS is unknown, although mutations in genes regulating bone morphogenic protein (BMP) signaling, such as SMAD6, have been implicated.[2] Congenital radioulnar synostosis may also occur as part of a few syndromic associations, including Tetrasomy X, Poland, Cornelia de Lange, Holt-Oram, Crouzon, and Apert syndromes.[5,7]

Fewer than 700 cases have been reported in the literature.[7] Despite its rarity, CRUS represents the most common congenital anomaly of the proximal radioulnar articulation.[8] There is no recognised sex predilection, and bilateral involvement occurs in approximately 60% of cases.[3,4]

Given the limited published data from sub-Saharan Africa, we report two cases of CRUS in Nigerian girls, illustrating unilateral and bilateral presentations and highlighting the characteristic radiographic features and associated skeletal anomalies.

CASE PRESENTATION

CASE 1

A 4-year-old girl presented due to limitation of daily activities involving the left upper limb, and inability to fully extend the left elbow since birth. There was also a history of an absent left thumb noted since birth. She was the second of three children in a monogamous family, with no known congenital abnormalities among her siblings.

The antenatal, perinatal, and postnatal periods were uneventful. Immunisations were up to date, and there was no history of trauma or previous surgical intervention involving the left upper limb. Her developmental milestones and academic performance were satisfactory.

Musculoskeletal examination revealed asymmetry of the forearms with shortening of the left forearm. There was limited extension at the elbow joint with restricted forearm rotation - pronation and supination. The first digit of the left hand was absent. No other abnormality was noted.

Plain radiographs of both forearms (Figure 1) revealed symmetric shortening of the left radius and ulna, hypoplasia of the left radial head with posterior dislocation, absence of the left distal radial epiphysis, and bony fusion of the left proximal radius and ulna. Radiographs of both hands (Figure 2) also revealed asymmetry of the hands with the left hand demonstrating visibly shorter phalanges and aplasia of the first digit. The right forearm and hand appeared normal.

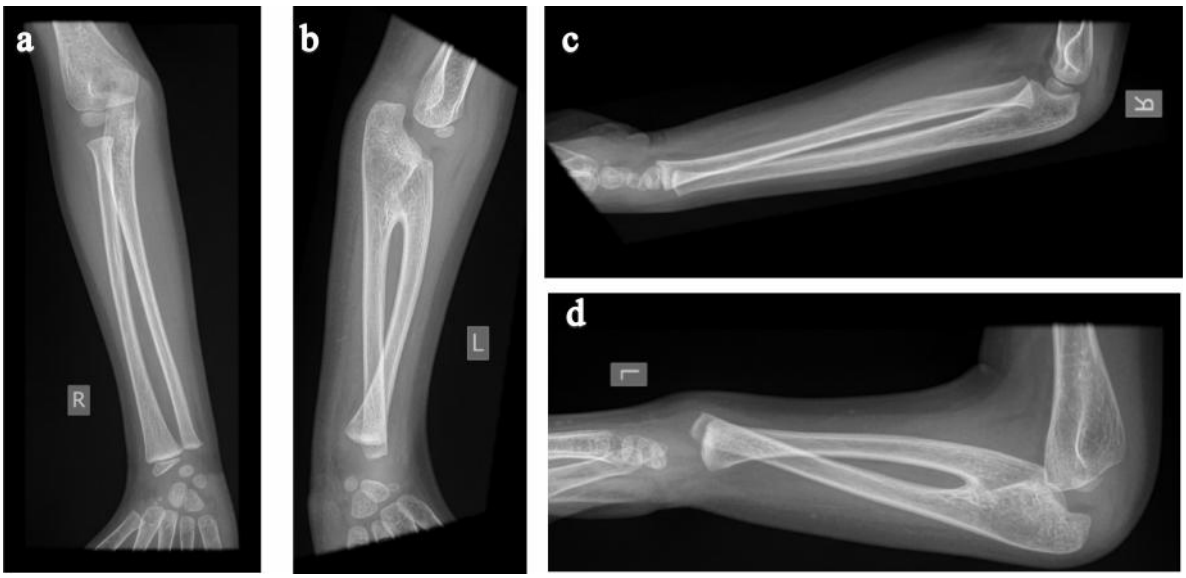

Figure 1: Plain radiographs of both forearms showing bony fusion of the left proximal radius and ulna, symmetric shortening of the left radius and ulna, and hypoplasia of the left radial head, which is also dislocated posteriorly (b & d). The left distal radial epiphysis is also absent (b).

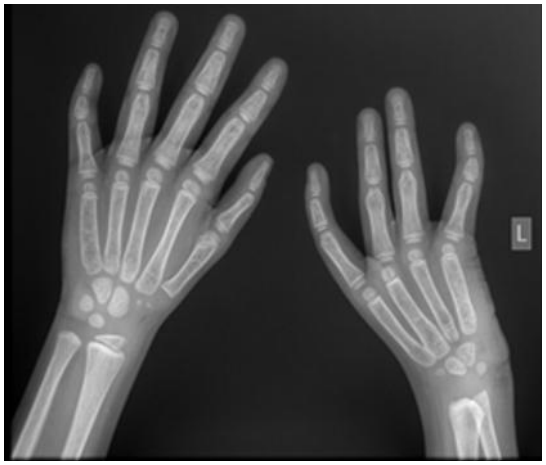

Figure 2: Plain radiograph of both hands showing asymmetry of the hands with aplasia of the first digit and abnormally deviated little finger

The patient was subsequently referred to the orthopaedic surgery team for further evaluation and possible corrective osteotomy.

CASE 2

A 13-year-old girl was referred on account of inability to fully extend both elbows since birth. This was associated with functional limitations in the use of her forearms during daily activities, including turning doorknobs and brushing her teeth. She was the first child in the family, with no known congenital abnormalities among her siblings.

No adverse events occurred during her pregnancy, birth, and postnatal period. No prior trauma or surgical intervention on both upper limbs.

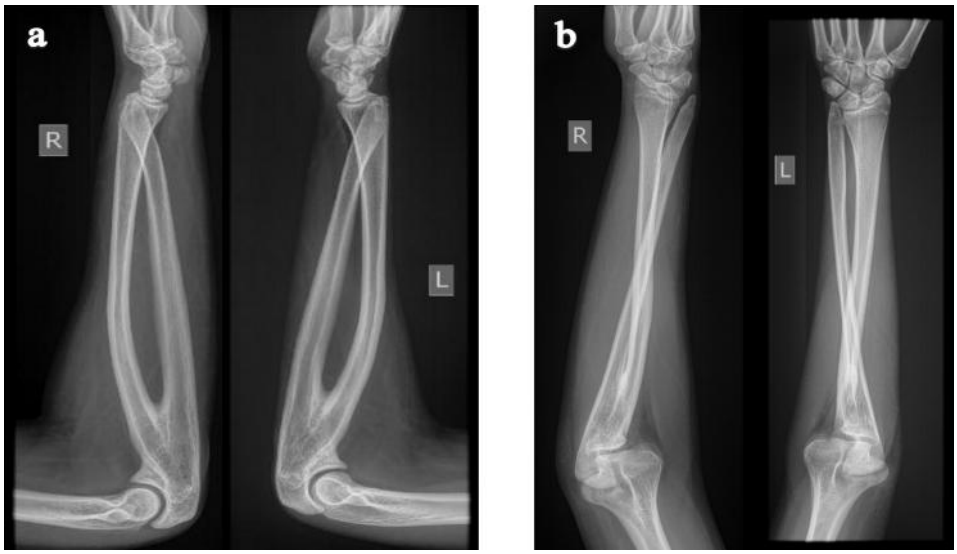

Figure 3: Anteroposterior (a) and lateral (b) radiographs of both forearms showing symmetric bony fusion of the proximal radius and ulna with posterior dislocation of the radial heads, bilateral symmetric bowing of the radius and ulnar shafts, and bilateral pencilling of the distal ulnar metaphysis with positive ulnar variance.

Musculoskeletal examination revealed restricted extension at the elbow joints with limited forearm range in pronation and supination.

Plain radiographs of both forearms (Figure 3) showed bilateral symmetrical bony fusion of the proximal radius and ulna with posterior dislocation of the radial heads, symmetric bowing of the radial and ulnar shafts, and bilateral pencilling of the distal ulnar metaphysis with positive ulnar variance.

She was subsequently referred to the orthopaedic surgery team for further evaluation and consideration of corrective osteotomy.

Discussion

Congenital radioulnar synostosis (CRUS) is a congenital musculoskeletal anomaly resulting from disturbed segmentation of radius and ulna in early foetal life.[2,8,9] It is a rare anomaly that has no sex predilection and usually presents in children aged between 2 and 6 years of age.[3,8] Clinical presentation varies widely, ranging from mild cosmetic concerns with minimal functional limitation—often contributing to delayed diagnosis—to more severe pronation deformities that significantly impair daily activities[2], as demonstrated in the two cases reported here. Patients with unilateral CRUS who have a fixed pronation angle of 30^0 can perform daily activities well, while those with bilateral CRUS or a fixed pronation angle $>60^0$ may experience more severe limitations in daily activities.[4] This pattern aligns with the two cases presented here, in which bilaterality was demonstrated in the second patient with pronounced functional limitation.

Cleary and Omer in 1985[10] proposed four types of congenital synostosis, which is the most widely used classification system for CRUS.[2] In type I the synostosis does not involve the bone and is associated with a reduced, normal-appearing, radial head; there is a visible osseous synostosis in type II with no other associated findings; type III is osseous synostosis with a hypoplastic and posteriorly dislocated radial head, and type IV is represented by a short osseous synostosis with an anteriorly dislocated radial head often with a 'mushroom shaped' morphology.[5,7] Based on this classification, both patients in this report correspond to type III CRUS, which is the most common morphologic type.[2]

Additional anomalies were demonstrated in the first case - including ipsilateral symmetric shortening of the radial and ulnar diaphyses, absent distal radial epiphysis, disorganised carpal bone ossification, and aplasia of the thumb – all of which may suggest a syndromic association. CRUS has been reported in association with various limb-patterning syndromes and skeletal dysostoses, including thumb aplasia, syndactyly, polydactyly, and cervico-vertebral schisis.[9,11,12] For example, Faust *et.al.*[7] described a 29-month-old male with concurrent CRUS, ipsilateral thumb aplasia, and Chiari malformation, and suggested a possible syndromic genetic basis. We did not conduct genetic investigations in the index cases due to lack of parental consent

Confident diagnosis and phenotyping of congenital radioulnar synostosis is typically established with plain radiographs alone.[5,10] Computed tomography with

three-dimensional reconstruction may be required for detailed preoperative evaluation of the synostosis.[4,13]

Early identification of CRUS is essential to optimise functional outcomes and guide appropriate management. It also allows timely stratification into conservative versus surgical pathways and facilitates physiotherapy and adaptive strategies where indicated. Conservative treatment is usually employed in asymptomatic cases with minimal functional deficits[6,13], whereas operative management is required in disabling CRUS.[1] Surgical techniques may involve mobilisation surgery, which excises the bar, and the created space is filled by a tissue e.g., fat or fascia. A rotational osteotomy, which places the forearm in a more functional position, is another surgical alternative.[2,8,13]

CONCLUSION

Congenital radioulnar synostosis is an uncommon musculoskeletal anomaly with variable functional impact. Radiography remains crucial for diagnosis, classification, and identification of associated skeletal abnormalities. The presented cases highlight both unilateral and bilateral presentations, as well as the importance of recognising additional anomalies that may suggest underlying syndromic associations. Early identification and appropriate functional assessment are essential to guide optimal management for favourable long-term outcomes.

Author conflicts of interest

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Consent statement

Informed consent for publication of this manuscript was obtained from the parents of the patients.

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