

Congenital Proximal Radioulnar Synostosis with an Unvisualized Radial Head: A Case Report and Review of Imaging Classification

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Abstract

Case Report

Congenital proximal radioulnar synostosis (PRUS) is a rare developmental anomaly characterized by abnormal proximal fusion of the radius and ulna, resulting in variable limitation of forearm rotation. We report the case of a 14-year-old female patient who underwent CT examination of the left forearm. CT demonstrated complete proximal osseous fusion of the radius and ulna extending over approximately 43 mm, associated with non-visualization of the radial head. Multiplanar and three-dimensional reconstructions clearly demonstrated the location and extent of the osseous fusion. The conventional Cleary–Omer classification could not be confidently applied because the radial head was not individually visualized. This case highlights the value of multiplanar and three-dimensional CT reconstructions in defining complex osseous anatomy and recognizing atypical morphological patterns of PRUS.

Keywords: Radioulnar synostosis, proximal radioulnar synostosis, Cleary–Omer classification, computed tomography, radial head, congenital anomaly.

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INTRODUCTION

Congenital proximal radioulnar synostosis (PRUS) is a rare malformation characterized by osseous or cartilaginous fusion of the proximal portions of radius and ulna [1]. Fewer than 700 cases of congenital radioulnar synostosis (CRUS) have been reported in the literature up to 2024 [2]. The main functional impairment is limitation of forearm pronation and supination. However, compensatory movements at the shoulder and wrist may allow patients to maintain relatively preserved function, with only a slight cosmetic deformity [1,3].

Although the exact etiology remains unclear, a genetic association has been reported in approximately 25% of cases [4]. Bilateral presentation of PRUS is reported in 60–80% of patients [5].

Several classification systems have been proposed. The Cleary–Omer classification remains one of the most frequently used systems and is based mainly on the type of synostosis and the morphology and position of the radial head [6]. However, atypical forms, particularly those in which the radial head cannot be identified, may not fit neatly into the four conventional categories [3].

We present a case of proximal osseous radioulnar synostosis associated with non-visualization of the radial head, emphasizing the CT findings and the classification challenge posed by this morphology.

CASE REPORT

A 14-year-old female patient presented with a longstanding limitation of her left forearm function. Clinical examination revealed marked restriction of pronation and supination. Limb innervation and perfusion were normal. There were no other concomitant malformations, no history of previous trauma, fracture, or surgery involving the elbow or forearm. She previously performed X-rays on her left forearm suspecting radioulnar synostosis. CT scan of the left forearm was requested by her physician.

CT confirmed complete proximal osseous fusion of the radius and ulna. The synostosis started at the level of 2.3 cm below the coronoid process of the ulna, extending over approximately 4.3 cm. The osseous bridge was clearly delineated on axial and multiplanar reformatted images, as well as on three-dimensional volume-rendered reconstructions (figure 1 and 2). At the proximal radioulnar region, the radial head was not separately visualized. The proximal radius appeared

fused to the ulna, with the proximal ulnar extremity remaining in articulation with the humerus (figure 2).

No additional osseous abnormality was identified. The distal radioulnar joint and wrist articulation were preserved, without evidence of associated osseous abnormality.

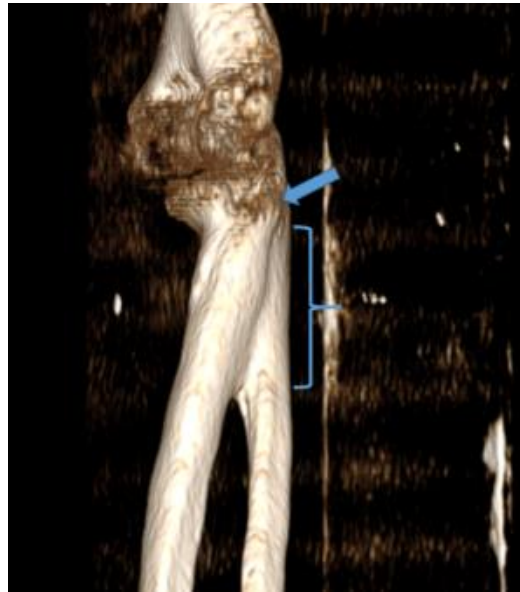


Figure 1: Three-dimensional volume-rendered CT reconstruction demonstrating an extensive proximal radioulnar synostosis measuring approximately 4,3 cm in length (bracket). The synostotic segment extends from the proximal radioulnar region distally along the forearm, with separate visualization of the radius and ulna distal to the fused segment. The radial head is not individually identifiable (arrow).

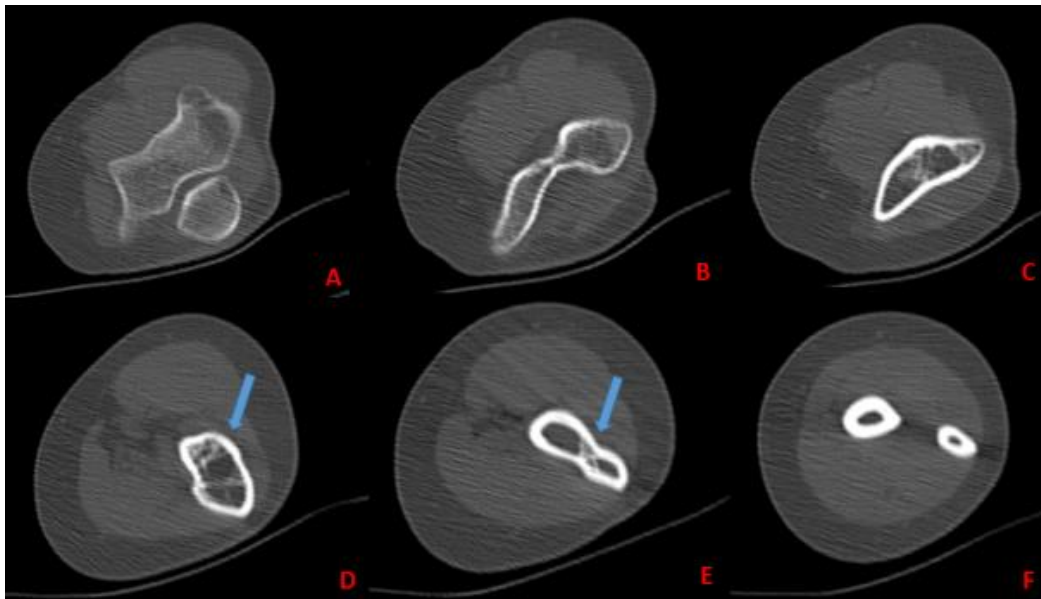


Figure 2: Axial CT images of the left elbow and forearm (A–F), obtained from proximal to distal, demonstrating complete proximal osseous radioulnar synostosis. The proximal radius is incorporated into the osseous bridge and the radial head cannot be individually identified (arrows, D–E). More distally, the radius and ulna become separately identifiable (F)

DISCUSSION

Congenital radioulnar synostosis was first described by Sandifort in 1793 [3]. PRUS results from an abnormality segmentation during early fetal development [7]. The condition is usually sporadic, although familial cases and genetic associations have been reported. PRUS may also occur as part of broader congenital or genetic syndromes, including

chromosomal abnormalities and associated upper-limb malformations [8,9].

The principal imaging feature of PRUS is abnormal osseous continuity between the proximal radius and ulna. Imaging assessment should characterize the extent of the synostosis, the morphology and position of the radial head, the relationship of the fused segment

to the elbow, and any associated osseous abnormalities [1,3]. Radiographs are generally sufficient for diagnosis, whereas CT, particularly with multiplanar and three-dimensional reconstructions, provides detailed assessment of the osseous anatomy and may be useful for preoperative planning [3,10,11].

Several classification systems have been proposed. Early radiographic classifications included the two-type system described by Wilkie, which distinguished proximal radioulnar fusion from cases associated with congenital radial head dislocation [10]. The Cleary–Omer classification describes four morphological patterns: type I, fibrous synostosis without definite osseous fusion; type II, osseous synostosis with a relatively normal radial head; type III, osseous synostosis with a hypoplastic and posteriorly dislocated radial head; and type IV, short osseous synostosis with an anteriorly dislocated, hypoplastic or mushroom-shaped radial head [6]. In the present case, the radial head was not visualized. Therefore, direct assignment to Cleary–Omer type II, III, or IV was not possible. This limitation is recognized in the literature. Tachdjian's classification includes a “headless” pattern, in which the radial head may be fused to the ulna or completely absent [12]. More recently, Li *et al.*, proposed an MRI and radiography-based anatomical classification in which type III is defined by an unobservable radial head associated with osseous synostosis with the ulna [3].

In our patient, CT demonstrated a complete proximal osseous synostosis with non-visualization of the radial head. The proximal ulna remained articulated with the humerus, while the radius and ulna were separately visualized distally, with preserved distal radioulnar and wrist articulations. These findings could not be confidently categorized within the conventional Cleary–Omer classification and highlight the value of multiplanar and three-dimensional CT reconstructions in defining the anatomy of atypical PRUS.

Although CT provides excellent assessment of the osseous abnormalities, PRUS may be associated with soft tissue abnormalities around the proximal radioulnar joint, including contracture of the pronator muscles and interosseous membrane and absence of the supinator muscle [10]. Dynamic magnetic resonance has recently been proposed as a complementary imaging technique for assessing the soft tissue and cartilaginous abnormalities associated with PRUS [10].

Beyond radiological classification, the main challenge in CRUS is determining the appropriate management, as no clear guidelines are currently available and the literature remains limited to level IV evidence [13]. Conservative management may be appropriate in patients with limited functional impairment, whereas surgical treatment, including

corrective derotation osteotomy, may be considered in patients with significant functional limitation [11].

CONCLUSION

Congenital proximal radioulnar synostosis is a rare malformation with variable morphological patterns. Our case illustrates an atypical form characterized by an extensive proximal osseous fusion and non-visualization of the radial head, which cannot be confidently classified using the conventional Cleary–Omer system alone. Multiplanar and three-dimensional CT reconstructions provide detailed assessment of the osseous anatomy, facilitating preoperative planning in such atypical presentations.

Declarations

Conflict of interest: None.

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