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Case report: Congenital pseudoarthrosis ulna with radial head extrusion

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Abstract
Isolated congenital pseudoarthrosis of the ulna bone is not a common entity. We present such a case of a 13year teenager, she visited us with a deformity of her right forearm (since birth), and extruded forearm bone from the lateral aspect of the elbow joint. The examination and investigation confirmed the diagnosis of congenital pseudoarthrosis of the ulna bone with extrusion of the radial head and we treated her condition by the creation of a single bone forearm as a salvage procedure.

Keywords: Congenital, neurofibromatosis, one bone forearm, pseudoarthrosis, ulna

Introduction
Isolated congenital pseudoarthrosis of the ulna bone is not a common entity. The occurrence of bony malformation like tibiofibular and radioulnar pseudoarthrosis in patients with generalized neurofibromatosis (type 1) is a well-known fact even literature supports the association. These malformations can easily be diagnosed clinicoradiologically. Very few cases have been reported in the literature so far and treatment protocol is not yet well defined for this condition (1-6). If a child of this deformity remains uncorrected for a long period, secondary deformities will develop. The main aim of treatment is to prevent the bony deformity from progressing further, to achieve a good union of bone ends and an essentially stable hand with a near-normal range of movement.

Case report
A 13years old teenager visited us with a gross deformity of the right upper limb. The right forearm was curved and the elbow joint was unstable by birth. Excessive medial bowing of the ulna was present with extrusion radial head from the lateral aspect of the elbow joint. The range of movement of the elbow joint was restricted, with a flexion range from 20-90 degrees and forearm supination and pronation were restricted to 20degree. A 3cm shortening was also noted in the affected forearm. The neurovascular structure of the right forearm was normal and the girl had a near-normal hand functions (fig 1.a). We performed a radiological examination in the form of plain x-rays of the right upper limb, which revealed a pseudoarthrosis of the ulna bone at the middle and distal third junction of the ulnar diaphysis with atrophy of distal third, the radius bone had a normal contour. Still, the radial head was dislocated anterolaterally and extruding out with a wound over the lateral aspect of the elbow (fig1.b). Anticipating a high chance of surgical site infection for a direct corrective surgery, an elbow spanning JESS was applied on the lateral aspect of the elbow and gradual distraction was carried out with regular wound dressing till the extruded radial head was well within a healed skin coverage over it (fig 1.c). We planned and performed a salvage procedure of reconstruction of double forearm bone to a single forearm bone using modified Henry's approach. The proximal part of the radius and distal part of the ulna was resected. The proximal stump of ulna and distal part of radius brought into alignment with square nail passed from tip of olecranon process under c-arm guidance (fig 2. a). The elbow and forearm were immobilized with an above elbow cast with the elbow flexed to 90 degree with the forearm in the supine position. At 2 months follow up, plain x-rays performed and bony ends had consolidated well (fig 2. b), the above-elbow cast was removed at the end of 2 months and elbow joint was mobilized.

The patient was told to avoid lifting heavy weights for another 3months till the remodelling of the bone occurred well. At 13months follow-up, the bone ends had remodelled completely (fig 2. c). The child had an elbow flexion of 110degree and the forearm had a normal range of supination (fig 2. d), but the pronation was restricted to 30 degree and the hand functions were near normal.

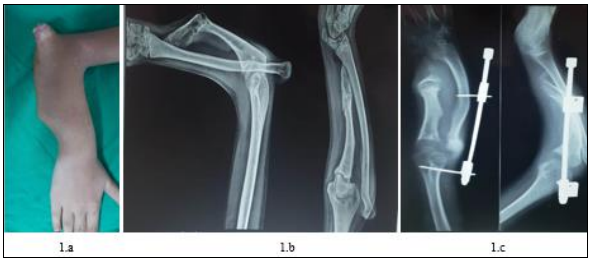

Fig 1: a. Girl had a near-normal hand functions**, b.** A wound over the lateral aspect of the elbow**, c.** An elbow spanning JESS was applied on the lateral aspect of the elbow

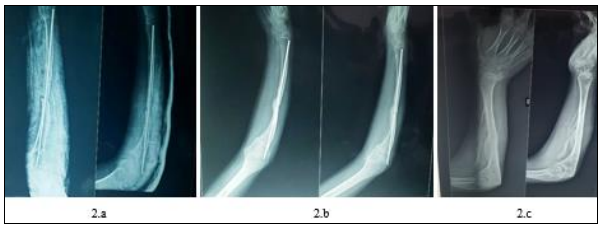

Fig 2: a. The proximal stump of ulna and distal part of radius brought into alignment with square nail passed from tip of olecranon process under c-arm guidance. **b.** bony ends had consolidated well. **c.** the bone ends had remodelled completely.

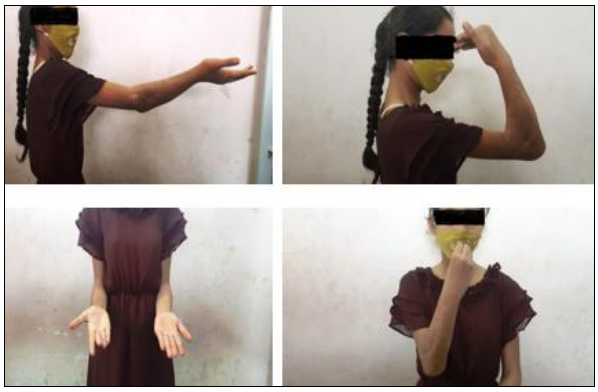

Fig 2d: The child had an elbow flexion of 110degree and forearm had a normal range of supination

Discussion

Deformity and malformation of the forearm bones occur in association with various syndromes. Congenital pseudoarthrosis of the forearm bones is not so common entity. Among the two forearm bones, the ulnar bone is more commonly deformed than the radius. The most common area affected is the junction of the distal and middle third shaft of the forearm bones [1]. The condition can be easily diagnosed and many patients usually report this deformity very soon after birth, but in some cases parents neglect and present only when the secondary deformities occur and affects the childs daily activities and with the cosmetic issue. Good quality plain x-rays of the affected area is sufficient to diagnose the forearm bone deformity. The typical radiological appearance in case of congenital pseudoarthrosis are, obvious gap, affected bone ends are sclerozed and tapering and medullary canal is usually seen obliterated. The malformed bone causes a theatering affect on the growth of unaffected bone resulting in the bowing of the later, the degree of bowing directly proportional to the duration of pseudoarthrosis. In our case, the girl had pseudoarthrosis from birth for 11years, but the radius was not proportionately bowed this may be because of early anterolateral dislocation of the radial head. A wide range of surgeries have been mentioned in the literature and the most popular ones are pseudoarthrosis excision and filling the gap with vascularized fibular graft and salvage procedure of reconstruction of dual bone forearm into a single-bone forearm. For an effective vascularized free fibula grafting and a good union a very important prerequisite is that radial head should be in a normal position, other disadvantages of vascularized free fibula graft includes resorption of graft, morbidity of donor site and steep learning curve and expertise needed for the procedure [3]. The single bone forearm procedure described by Lloyd Roberts [4] is a not technically demanding, shorter and easier procedure with a very good union rate [4]. In our case we had a dislocated radial head so we had to go with a salvage single bone reconstruction procedure. The principle behind creation of single bone forearm is that it maintains the stability of elbow and wrist joint and length of the forearm in turn maintaining the effective length of the long flexors of the forearm.

In this case we did an end to end fusion to avoid bone grafting and to have a good chance of bone union due to the compressive forces achieved at the 2 bone ends. At 10 weeks follow-up, x-rays showed a good bone union. Elbow and wrist joint range of movements were near normal with good hand functions, a 2cm shortening of the forearm was noted but did not cause any functional disability.

Conclusion

With our experience of this case and also after reviewing the literature, we conclude that congenital pseudoarthrosis of the ulna is a rare entity, which needs early surgical intervention. Single bone forearm procedure is a viable option in cases where radial head is not in normal position. Though a salvage procedure, it achieves a stable elbow and wrist joint with a near normal hand functions.

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