CASE REPORT

Single-stage management of a neglected radial club hand deformity in an adult

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Accepted 14 January 2015

SUMMARY

Radial club hand is an intercalary congenital deformity involving the forearm, wrist and hand. A congenital absence of radius (partial or complete) and ulnar bowing are classical radiographic abnormalities seen in this condition. This deformity is usually treated surgically in infants and young children but the management of this problem in an adult is complex and challenging. We present a neglected case of an adult with severe and rigid deformity that was successfully treated by one-stage correction involving ulnar osteotomy and wrist arthrodesis, simultaneously.

CASE PRESENTATION

A 23-year-old man presented with deformity of the right forearm and wrist ([figure 1](#)) since birth. The patient was born through normal vaginal delivery in a rural setting where prenatal screening for musculoskeletal congenital anomalies was not possible. Even after birth, proper assessment and management of this anomaly was not carried out due to lack of adequate healthcare infrastructure. The patient's wrist was stiff and he could only move his fingers, which deviated away from his body; the flexion of the fingers was impaired. He was unable to grasp. He was working as an assistant in a physician's clinic. His elbow and shoulder functions were totally preserved. Over the years he had adopted the use of his left hand for routine activities, however, with very limited function. But he could not perform meaningful activities with his affected right hand. He wanted to have cosmetic correction of his wrist and forearm deformity with improved function of his right hand.

There was severe radial deviation of the right wrist with bowing of the ulna ([figure 1](#)). The patient's left forearm was 25 cm long whereas it was only 15 cm on the right side; there was marked atrophy of the right forearm. There was only a single forearm bone on palpation. Sensation and perfusion of the hand were intact. The right humerus was normal with range of motion -10° to 130° at the elbow joint. A general examination was unremarkable. There were no other associated congenital anomalies.

Plain radiograph of the affected limb revealed complete absence of radius (Heikel's grade IV) with almost 45° lateral bowing of the ulna ([figure 2](#)). Soft tissue release of contracted structures on the radial side was performed along with ulnar closed wedge osteotomy, centring at the apex of the deformity ([figure 3](#)); it was fixed with a 3.5 mm low contact dynamic compression locking plate. Proximal carpectomy was undertaken to bring the ulna to the centre of the wrist. Wrist arthrodesis was performed ([figure 4](#)) after denuding the distal and remaining carpal bones, and the wrist was stabilised by fixation using a 3.5 mm wrist arthrodesis plate (Synthes); bone grafting was carried out using the bone retrieved from the ulnar osteotomy fragment. Proximal row carpectomy and ulnar osteotomy were two crucial steps that led to correction of the deformity, and maintenance of the correction. The wrist and forearm were kept in a splint for 3 weeks. The postoperative period was uneventful.

The patient was able to flex and extend his fingers appropriately and he developed reasonable grasp.

Figure 1 Radial club hand with severe radial deviation of the wrist and finger deformity.

Figure 2 Anteroposterior radiograph showing complete absence of the radius and severe bowing of the ulna.

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To cite: Vaishya R, Agarwal AK, Vijay V, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-208682

Figure 3 Oblique radiograph confirming severe bowing of the ulna and deviation of the wrist.

There was satisfactory correction of the deformity of wrist and, to some extent, of the fingers as well (figures 5 and 6).

At a follow-up of 6 months, the patient was able to flex and extend his fingers appropriately and he developed a reasonable grasp. Range of motion at the metacarpophalangeal joint was full and he could now brush his hair and feed himself using the right hand. His osteotomy and arthrodesis had also healed.

GLOBAL HEALTH PROBLEM LIST

Musculoskeletal disorders constitute an important global health problem. Failure to diagnose and properly manage musculoskeletal deformities in childhood can lead to significant disabilities later in adulthood. In developing countries, poverty, illiteracy, ignorance and inadequate medical facilities are all associated with a higher incidence of neglected musculoskeletal disorders. Screening programmes during pregnancy and early childhood can lead to early diagnosis and proper management.

GLOBAL HEALTH PROBLEM ANALYSIS

The occurrence of radial club hand is usually sporadic, but it may be associated with any of several other defects or

Figure 4 Anteroposterior and lateral radiographs showing corrective ulnar osteotomy and fixation of the ulna and wrist arthrodesis with a plate.

Figure 5 Dorsal view (with forearm in pronation) of the wrist and forearm showing good postoperative correction of the deformity.

syndromes. More than half of those born with radial dysplasia have the deformity in both limbs.¹ Radial club hand occurs due to longitudinal failure of formation of parts along the preaxial border of the upper limb. It occurs more commonly in boys than in girls, and more on the right than the left side. The cause of this congenital deformity is unknown but it appears to be more of a sporadic mutation than an inherited one.² It results from a defect occurring in embryo development during the fourth to seventh week in utero. Malformations occurring at this stage may occur from failure of formation or differentiation of the mesenchymal anlage. The significance of this event is that other organs forming at this stage may also be affected and hence the acronym VACTERL (vertebral, anorectal, cardiac, tracheal oesophageal, renal) anomalies.³

Heikel,⁴ in 1988, classified congenital club hand based on the quantity and quality of the existing rudimentary radius.

Type 1: Short distal radius

Type 2: Hypoplastic radius

Type 3: Partial absence of radius

Type 4: Total absence of radius

Global classification involves the regional components of the deformities. Most but not all tissues involved in the pathology are taken into account. The states of the thumb, carpus, distal and proximal radius are accounted for in this classification.⁵

The age of presentation of our patient was late, at 23 years. Walia *et al*,² in 2006, also reported a case that presented

Figure 6 Palmar view (with forearm in supination) of the wrist and forearm showing good postoperative correction of the deformity.

relatively late at the age of 10 years. Progression of the deformity and cosmetic issues prompted the patient to present at this stage. The issue of stigmatisation of criminality related to extremity loss prevalent in certain countries is also a reason for the patient demanding correction and not amputation in this particular case. All these issues justified the need for surgical intervention. Conservative and surgical interventions with gradual corrections have both been used with various levels of success in the past for the treatment of radial club hand.⁶ Conservative management involves passive stretching of the tight radial structures and the elbow, which must begin shortly after birth. Splinting and casting must be started as soon as the forearm matures enough to accommodate the splint.⁷ However, there appears to be no role of conservative management in a person with radial club hand presenting at the age of 23 years.

Surgical intervention revolves around centralisation of the hand on the wrist. This is advocated at 1 year in order to provide a foundation for the development of the hand motor function, improvement in length and realisation of the developing immature brain.⁸ Osteotomies are indicated if the ulna exhibits more than 30° of angulation. This corrects angulation, provides early alignment and healing, reduces soft tissue tensioning, redirects force and improves the mechanical advantage of the hand. As an adjunct to this, various tendon transfers have been advocated.

Some innovative German procedures include reconstruction of the distal radius with non-vascularised fibular and tibial grafts, and microvascular free transfer to support the radial side of the wrist; these have been described in the literature.⁹ Various forms of uniplanar external fixators and multiplanar ring external fixators have also been used in children aged less than 10 years with success.¹⁰ The principles of treatment outlined include realignment of deviated wrist, suspending of the wrist on the forearm, ensuring finger motion, promoting growth of the forearm, prevention of pain and reduction of progression of the deformity. The late presentation of this index patient precluded strict adherence to these principles. To a patient coming from another country, gradual correction using the multiplanar external fixator, which involves longer duration of treatment and multiple follow-up, was not acceptable. The patient was willing to undergo a one-stage procedure to correct his deformities. Single-stage correction of radial club hand in an adult patient has not being described in the literature.¹¹ The degree of stiffness was such that a proximal row carpectomy had to be performed in order to free the tethers and to centralise the hand on the wrist. The one-stage procedure using corrective osteotomy of ulna and wrist arthrodesis helped the patient to gain good functional outcome soon after the procedure.

Patient's perspective

I was delighted to have significant cosmetic correction of my wrist and hope I will get a better match for my marriage. I could now also use my hand much effectively and can do many more activities of daily living independently. I never thought that the correction of my deformity was possible, as all the other doctors in my country refused to operate and give me any assurance about the outcome of the treatment.

Learning points

- ▶ Radial club hand deformity is a congenital condition where the radius is partially or totally absent; it is associated with bowing of the ulna.
- ▶ Most of these patients are treated by surgical correction in childhood.
- ▶ Untreated radial club hand deformity in an adult is a rare and challenging problem to treat.
- ▶ Single-stage correction in severe and rigid deformity in an adult is a complex corrective surgery but can be achieved by combined ulnar osteotomy and wrist arthrodesis.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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