Dorsal radiocarpal dislocation in a patient with Goldenhar syndrome: case report

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Abstract

Background Fracture–dislocations of the carpus are rare, generally occurring after high-energy trauma. Goldenhar syndrome is among a group of genetic abnormalities associated with radial limb defects. We present a case of a dorsal radiocarpal dislocation in a patient with Goldenhar syndrome after a low-energy fall. To our knowledge, there has been no previous report of radiocarpal dislocation in the setting of Goldenhar syndrome.

Methods This patient with Goldenhar syndrome had a dorsal radiocarpal dislocation in the setting of an absent scaphoid and dysplastic distal radius. A computed tomography scan, recognized as a useful modality to evaluate the wrist and scaphoid, was used to rule out any other osseous trauma or avulsion fractures.

Results Closed reduction and 6 weeks of immobilization resulted in a successful treatment.

Conclusions The incidence of radiocarpal dislocations in patients with Goldenhar syndrome and the appropriate long-term treatment for patients with Goldenhar syndrome with radiocarpal dislocations require further investigation.

Keywords Dorsal radiocarpal dislocation · Dysplastic distal radius · Goldenhar · Scaphoid aplasia

Introduction

Fracture–dislocations of the radiocarpal joint are rare. They account for only 0.2 % of wrist injuries [4]. These dislocations occur with or without associated fracture of the distal radius and/or ulna after high-energy trauma [3, 6, 7]. The oculoauriculovertbral spectrum, including Goldenhar syndrome, is a group of various genetic abnormalities that result in craniofacial, cardiac, pulmonary, renal, and skeletal abnormalities, including scoliosis and (rarely) radial defects [1, 9, 10]. We report a case of a radiocarpal dislocation in a patient with Goldenhar syndrome after a low-energy fall onto an outstretched hand while playing basketball. Radiographs showed thumb hypoplasia, scaphoid aplasia, and a dysplastic distal radius, consistent with previous reports of a rare subcategory of patients with oculoauriculovertbral spectrum.

Case Report

A 22-year-old right-hand-dominant male with a history of Goldenhar syndrome presented to the emergency department with right wrist pain after a fall while playing basketball. His medical history consisted of congenital cervicothoracic scoliosis, horseshoe kidney, recurrent eye infections, and a history of a chronically subluxated right hypoplastic thumb metacarpophalangeal joint for which he underwent arthrodesis 15 years before with no complications. On examination, he had notable right wrist deformity with pain and swelling, as well as paresthesias in a median nerve distribution. Radiographs showed a dorsal radiocarpal joint dislocation, without evidence of fracture, and an abnormal-appearing distal radius and proximal carpal row (Fig. 1a, b).

After intravenous analgesia, the wrist was reduced with manual longitudinal traction and volarly directed pressure on the carpal bones. Once the carpus was reduced, the

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Fig. 1 Radiographs of the right wrist after injury. **a** Posteroanterior. **b** Lateral

patient had symmetric range of motion from 70 ° of flexion to 45 ° of extension. He also was noted to have improved sensation in the median nerve distribution. Postreduction radiographs showed an acceptable reduction, scaphoid aplasia, and a dysplastic distal radius (Fig. 2a, b). A computed tomography scan was obtained to rule out fracture to the carpal bones that may have occurred during dislocation (Fig. 3a–c). The patient was placed in a volar resting splint and treated with 6 weeks of immobilization.

Discussion

Oculoauriculovertebral spectrum, including Goldenhar syndrome, is a constellation of clinical findings ascribed to dysplastic development of the first and second branchial arches as well as the first pharyngeal pouch and branchial cleft. It occurs at a population frequency of 0.03 % and is suspected to arise secondary to a defect in blastogenesis, although there is no consensus on its cause [10]. The asymmetric underdevelopment of the external ear, middle ear, mandibular rami, and other facial structures results in a classic hemi-facial microsomia in these patients [1]. These findings may occur in isolation or in conjunction with other

systemic findings. Other organ systems involved in this syndrome include cardiac, pulmonary, renal, central nervous, and musculoskeletal systems [1, 10].

Orthopaedic findings often include kyphosis, scoliosis, hemivertebra, fusion of single vertebrae levels throughout the spine, and congenital dislocation of the hip [1, 9]. One series found orthopaedic abnormalities as high as 61 % of patients reviewed [1]. However, other series report orthopaedic abnormalities between 20 and 60 % [1, 9].

Various authors have reported on the association of radial defects and the oculoauriculovertebral spectrum. In one series of patients with Goldenhar syndrome, the most common abnormalities were hypoplasia of the thumb and hypoplasia and/or agenesis of the radius [10]. Less common are thumb agenesis, preaxial polydactyly, and triphalangeal thumb [10]. These radial anomalies occurred unilaterally or bilaterally, with most involving the right side [10].

In the normal wrist, the proximal surface of the carpus is an oblong condyle that articulates with the biconcave and triangular distal radius and the triangular fibrocartilage [6, 8]. Combined with the intrinsic and extrinsic ligaments of the wrist, the radiocarpal joint allows mobility and flexibility while appropriately offloading and dissipating pressure throughout the carpal bones and ligaments [8]. Various

Fig. 2 Radiographs of the right wrist after reduction. **a** Posteroanterior. **b** Lateral

Fig. 3 Computed tomography scans of the right wrist carpal bones after reduction. **a** Axial. **b** Sagittal. **c** Coronal

models have been described to explain wrist motion, including a Slider-crank mechanical model for wrist motion and a three-column system [8]. In both descriptions, the scaphoid serves as the main source of stability, with the extrinsic radiocarpal ligaments, capsule, and scaphoid and lunate fossa of the distal radius all contributing to stability [8].

Radiocarpal dislocations have been described extensively in the literature, with many case reports describing mostly dorsal fracture–dislocations arising after high-energy trauma [5]. Although few series exist, two classification systems have been developed to describe these injuries. In a small series, Moneim et al. [7] grouped these injuries into radiocarpal fracture–dislocation without associated intercarpal dissociation (type I) or radiocarpal fracture–dislocation with an associated intercarpal dissociation (type II). Type II radiocarpal dislocations in this classification scheme represent a more complex pattern of injury, with concomitant scapholunate or lunotriquetral intercarpal ligament injuries [7]. A second classification, proposed by Dumontier et al. [3], also described two patterns of injuries: group 1, radiocarpal dislocations that are purely ligamentous or with a small cortical avulsion fracture; and group 2, radiocarpal fracture–dislocations associated with a large radial styloid fracture fragment [3].

The management of radiocarpal dislocations varies. Non-operative management has been successful and is described in several series [5, 7]. Ilyas et al. [6] proposed a treatment algorithm based on radial, intermediate, and ulnar columns and stability, with open reduction and internal fixation indicated for the presence of radial styloid fracture, lunate facet fracture, or ulnar styloid fracture or suture anchor repair recommended in the presence of ligamentous avulsion (repair of radioscapohocapitate ligament, short radiolunate

ligament, or ulnocarpal ligament, respectively). All patients were immobilized for a period of six weeks, with patients having unstable fractures requiring additional fixation with a radiolunate pin or external fixation [6].

To our knowledge, there has been no previous report of radiocarpal dislocation in the setting of Goldenhar syndrome. Other genetic abnormalities have been associated with chronic bilateral wrist dislocation, including trisomy 21 (characterized by ligamentous laxity). Patients with Goldenhar syndrome have a reported increased incidence of congenital dislocation of the hip and radial limb defects [1]. In the current patient with Goldenhar syndrome and associated radial dysplasia, with hypoplastic thumb, scaphoid aplasia, and dysplastic distal radius, we postulate the normal mechanics of the wrist had been altered enough that a low-energy fall was sufficient to create a dorsal radiocarpal dislocation. Given that his dislocation occurred after a low-energy fall, it is likely that the preexisting radial anomaly and Goldenhar Syndrome put this patient at greater risk for this injury. A computed tomography scan, recognized as a useful modality for evaluating the wrist and scaphoid, was used to rule out any other osseous trauma or avulsion fractures [2]. Additional study is needed to determine the long-term outcomes of nonoperative and operative management in this group of patients.

This patient with Goldenhar syndrome had a dorsal radiocarpal dislocation in the setting of an absent scaphoid and dysplastic distal radius. Management consisted of closed reduction with appropriate intravenous analgesia and splinting. The incidence of radiocarpal dislocations in patients with Goldenhar syndrome and the appropriate long-term treatment for patients with Goldenhar syndrome with radiocarpal dislocations require further investigation.

Conflict of interest statement The authors declare that they have no conflict of interest.

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