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Proximal Translation of the Radius Following Arthroplasty of the Distal Radioulnar Joint in Hajdu-Cheney Syndrome

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INTRODUCTION

Several surgical procedures for the derangement of the distal radioulnar joint (DRUJ) have been developed such as the Darrach's, the Sauvé-Kapandji's, and the Bowers' procedures.[1,3] After arthroplasty of the DRUJ, limitation in the range of motion and wrist pain have been well documented, however, few reports related to problems of the elbow after these surgical procedures have been reported. In the present paper, we report asymptomatic marked proximal translation of the radius at the elbow joint after treatment of dorsal dislocation of the DRUJ using the Sauvé-Kapandji's procedure (S-K procedure) in a patient with Hajdu-Cheney syndrome, an idiopathic skeletal dysplasia with osteoporosis characterized by short stature, joint laxity, short clubbed fingers and toes, premature loss of teeth, and defective vision and hearing.[5,9,13]

CASE REPORT

The patient was a 46-year-old woman. The patient was the first-born and the product of a full-term normal delivery. There was no family history of bone disease and development of her intelligence was normal. In the patient's later childhood the patient's was diagnosed as having the Hajdu-Cheney syndrome from short stature, osseous dysplasia with osteoporosis, short clubbed fingers and toes, premature loss of teeth, defective hearing, acro-osteolysis, wide cranial sutures, and multiple wormian bones. The patient was right handed and had experienced mild pain in the right wrist and inability to extend the little finger 6 months before the patient visited us. Although the wrist pain did not increased, the patient experienced inability to extend the middle and ring fingers and visited us.

Physical examination revealed dorsal displacement and tenderness of the distal end of the ulna. Full range of motion in extension and flexion of the wrist and elbow, or in pronation of the forearm was noted. However supination of the forearm was limited to 60° due to wrist pain. Active motion of the thumb and the index finger was not limited. The patient could fully flex all joints of the middle, ring and little fingers, and extend their distal and proximal interphalangeal joints. However, extension of their metacarpophalangeal joints was restricted to 50° (Figure 1). Passive range of motion of these fingers was fully maintained. Relief and tenodesis effects of the extensor tendons of the fingers were not detected clearly. Grip strength in the right hand was 12 kg and in the unaffected left hand, 19kg. General joint laxity was detected. Anteroposterior view of the radiograph of the hand showed osteolysis of the distal phalanges, positive variance and deformity of the ulna and dissociation of the DRUJ (Figure 2). The radiograph of the forearm showed slight bowing deformity of the ulna and incongruity of the humeroradial joint, however, discrepancy of the length of these paired bones was not evident (Figure 3).

The patient was treated surgically. A curved dorsal incision was made on the dorsal aspect of the wrist and the extensor retinaculum was incised over the extensor digitorum communis (EDC). Very mild tenosynovitis and rupture of the extensor tendons, EDC (III, IV, V) and extensor digiti minimi, was observed. The ruptured extensor tendons were reconstructed with a tendon graft.[3] The graft of the palmaris longus was connected to the distal tendon stumps by an intercalary weave and then attached to the proximal tendon with an interlace weave technique in slightly greater extension than the normal cascade. Dorsal

dislocation of the DRUJ was managed by the S-K procedure.[1] A bone segment of the ulna was excised in order to create a space for the pseudoarthrosis site and articular surfaces of the radius and the ulna were excised until cancellous bone. The head of the ulna was opposed to the radius and pinned to it with two Kirschner wires.

The patient started active finger exercises at 3 weeks postoperative. By 6 months after surgery, there was no pain or limitation in the range of motion of the wrist or any of the fingers. Two years after surgery, the patient had no complaint concerning the patient's wrist, fingers, and elbow with full functional recovery. Radiographs of the wrist showed complete arthrodesis of the DRUJ and the pseudoarthrosis of the ulna (Figure 4). However, radiographs of the elbow showed marked anterior dislocation of the radial head and proximal translation of the radius (Figure 5).

DISCUSSION

Although Hajdu-Cheney syndrome is a congenital disorder, the correct diagnosis is rarely made until later childhood when characteristic symptoms occur. Clinical features included distinctive facial appearance, short stature, generalized osteoporosis, joint laxity, premature loss of teeth, retarded puberty, and defective vision and hearing.[5,9,13] On the radiographs, osteolysis of the distal phalanges of the hands and feet, wide cranial sutures, and multiple wormian bones were observed. Mostly all of the clinical features were detected in the patient described in the present report.

Rupture of the extensor tendons and disorder of the wrist joint has not been

reported in this syndrome. Rupture of the extensor tendons in rheumatoid arthritis is caused by the mechanical force associated with deformity of the wrist and tenosynovitis.[3] In the case presented, tenosynovitis of the extensor tendons was not severe and rupture occurred in the dominant hand. Mechanism of extensor tendon rupture was mainly considered to the mechanical force by the dorsal dislocated ulna and it was successfully treated with an intercalary tendon graft using the palmaris longus.

In Hajdu-Cheney syndrome, bowing deformities and cortical defects of the paired long bones, for example the tibia, fibula, radius, and ulna causes incongruity and arthrosis of the knee and ankle joint at both ends of these paired bones.[5,9] In the case presented, cortical defects of the radius and the ulna were not observed, however, bowing deformity was observed in the ulna.

Proximal translation of the radius was caused by resection of the radial head due to lack of bony stabilization at the humeroradial joint.[4,7,10] Fracture of the radial head and injury of the interosseous membrane caused translation of the radius with limitation in the range of motion of the wrist, forearm, and elbow.[2,6] A case report of osteoarthritis of the humeroradial joint due to proximal migration of the radius 25 years after an extensive Darrach's procedure at the age of the 15 suggested that the interosseous membrane plays an important role to the proximal translation of the radius.[11] The patients who had the posttraumatic disorder of the wrist with simultaneous involvement of the elbow was treated by the S-K procedure associated with insertion of a silicone implant of the radial head and they obtained a good functional recover of the wrist and elbow joints.[12] Biomechanical analysis of pressure distribution in the humeroradial joint using

fresh frozen human cadavers revealed that the force transmitted to the capitellum increased after the S-K procedure, however, that there was no significant differences between the forces before and after the S-K procedure.[8] By contrast the force transmitted to the capitellum after the S-K procedure with incision of the interosseous membrane significantly increased compared with the forces before the opertation.

In the patient reported here, anterior subluxation and proximal translation of the radial head had been promoted severely after surgical treatment of the DRUJ by the S-K procedure. It was speculated that dorsal dislocation of the ulna at the DRUJ and subluxation of the radial head was caused by incongruity of the radioulnar joint due to deformity of the forearm bones, joint laxity, and insufficiency of the interosseous membrane. Instability of the wrist due to the pseudoarthrosis site at the distal ulna created by the S-K procedure promoted proximal translation of the radius and instability of the forearm. The patient has no complaint of the elbow at present. However, we should monitor closely translation of the radius after arthroplasty of the wrist in the patient with general joint laxity and insufficiency of the interosseous membrane such as observed in Hajdu-Cheney syndrome because there is the possibility of pain or instability of the elbow occurring. We think that the wrist arthroplasty with resection of the ulna such as the S-K procedure should be performed very carefully in the patient with general joint laxity and insufficiency of the interosseous membrane.

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LEGENDS

Figure 1: The clinical appearance of the right hand.

Extension of the MP joints of the middle, ring, and little fingers was limited.

Figure 2: Radiographs of the hand (R indicates the right hand and L the left).

The anteroposterior view of the hand showed osteolysis of the distal phalanges, positive variance and deformity of the ulna, and dissociation of the DRUJ.

Figure 3. The radiographs of the forearm. Anterolateral (A) and lateral (B) view.

Bowing deformity of the ulna and incongruity of the humeroradial joint were observed, however, there was no discrepancy of length of these paired bones.

Figure 4: Radiograph of the wrist after surgical treatment. Anterorposterior (A) and lateral (B) view.

Complete arthrodesis of the DRUJ and the pseudoarthrosis of the ulna was shown.

Figure 5: Radiograph of the elbow after surgical treatment of the wrist. Anterorposterior (A) and lateral (B) view.

Marked proximal translation and anterior dislocation of the radial head were shown.