Case Report

Concomitant Avascular Necrosis of the Lunate and Proximal Pole of the Scaphoid in a Thalassemia Minor Patient

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Abstract

Avascular necrosis (AVN) of multiple carpal bones is a very rare condition. This case report presents a patient with concomitant AVN of the lunate and proximal pole of the scaphoid that had led to severe joint space narrowing and degenerative arthritis in the radiocarpal and distal radioulnar joints (DRUJs). She was known to have thalassemia minor hemoglobinopathy. Wrist arthrodesis and distal ulna hemiresection-interposition arthroplasty (Bowers technique) for a DRUJ were offered and performed. The patient was satisfied with the clinical outcomes.

Keywords: Arthrodesis, avascular necrosis, Bowers technique, Kienbock’s disease, lunate bone, scaphoid bones, thalassemia, wrist

Introduction

Avascular necrosis (AVN) of the lunate is an uncommon disease and AVN of the scaphoid is a rare disorder. Concomitant AVN of the lunate and scaphoid is a very rare disorder. AVN of multiple carpal bones has been mentioned with a history of trauma, metabolic disorder, corticosteroid consumption, rheumatoid disorders, and systemic lupus erythematosus (SLE).[1-4] This case report presents a patient with concomitant AVN of the lunate and proximal pole of the scaphoid.

Case Report

The case was a 39-year-old female with pain and restricted range of motion (ROM) of her right wrist. She did not remember the exact beginning of her wrist pain, but she stated that it was started more than 20 years ago. She had carpal tunnel surgery for her right wrist pain 7 years before her visit; however, her pain continued and intensified from 1 year before. Her right wrist visual analog scale (VAS) for pain was 8 (range: 0–10). She did not mention any history of rheumatologic disorders, corticosteroid intake, and metabolic disorder; however, she was known to have thalassemia minor hemoglobinopathy. She had anemia and decreased red blood cell indices including mean corpuscular volume, mean corpuscular hemoglobin (MCH), and MCH concentration. Hemoglobulin electrophoresis confirmed thalassemia hemoglobinopathy.

The right wrist ROM was 0° flexion–10° dorsiflexion. The right forearm supination and pronation were painful at the distal radioulnar joint (DRUJ). The power grip of the right hand was 16 kg, while the left was 30 kg. Rheumatologic laboratory tests included normal ranges of rheumatoid factor, C-reactive protein, antinuclear antibody, anti-cyclic citrullinated peptide, and anti-DNA.

Plain radiographs of the right wrist demonstrated sclerosis, cyst formation, fragmentation, and severe collapse of the lunate and proximal pole of the scaphoid indicating AVN. Ulnar variance was neutral. There was severe degenerative arthritis at radiocarpal and DRUJs, and the joint spaces were lost [Figure 1]. A T1-weighted magnetic resonance imaging (MRI) demonstrated the low signal intensity of the...
lunate and proximal pole of the scaphoid [Figure 2], while a T2-weighted MRI demonstrated increased signal intensity in the lunate and proximal pole of the scaphoid and joint effusion in DRUJ [Figure 3].

Regarding the MRI findings, concomitant AVN of the lunate and proximal pole of the scaphoid was considered. Wrist arthrodesis and distal ulna hemiresection-interposition arthroplasty (Bowers technique) for DRUJ because of severe degenerative changes were offered and performed. Wrist arthrodesis was performed along the distal radius and third metacarpal bone with a ten-hole 3.5 dynamic compression plate and local bone graft.

Surgery sample from the wrist synovium and bony specimens from the lunate bone and proximal pole of the scaphoid were collected for histopathologic study. Histopathologic studies of the bone specimens from the lunate bone and proximal pole of the scaphoid confirmed AVN of the bones. Synovial biopsy demonstrated fibrofatty vascular tissue with chronic inflammation, fibrinoid deposition, and synovial hyperplasia with papillary appearance. There was no sign of infection and granuloma formation.

At 3 months postoperative, she had no pain and her VAS score became zero. The supination/pronation arc was 110°. Plain radiographs demonstrated satisfactory alignment and position of the wrist [Figure 4]. Power grip improves to 20 kg. She was satisfied with the clinical outcomes. The form of informed consent has been completed by the patient.

**Discussion**

Because of the rarity of AVN of multiple carpal bones and the insufficient number of reported cases, discussions over its treatment are contrasting. Involvement of more than a carpal bone is very rare. Based on existing hypotheses, various factors...
are involved in causing nontraumatic AVN in carpal bones such that thrombosis, fat embolus, or vasculitis in areas of bone with minimal collateral blood supply.[5] Furthermore, many different risk factors have a role in the pathogenesis of multiple AVN in carpal bones including corticosteroid use, smoking, gout, alcoholism, infections, hyperbaric events, storage disorders, marrow-infiltrating diseases, coagulation defects, hemoglobinopathies (e.g., sickle cell anemia), fat emboli, liver disease, hyperlipidemia Types II and III (but not Type I), hemophilia, rheumatoid arthritis, SLE, and pancreatitis.[4,5] Budoff reported simultaneous Kienböck’s disease and Preiser’s disease in a 50-year-old female with a remote history of steroid use but without a history of trauma or systemic illness.[3]

Factors that impair blood flow to the carpal bones appear to be involved in the simultaneous involvement of several bones. In another report by Bhardwaj et al., concomitant AVN of the scaphoid and lunate has been detected in patients who did not know the risk factors of taking some herbal medications of unknown content.[6] In these patients, biomechanical factors do not affect AVN of carpal bones so that most cases had a negative ulnar variance.[6] Treatment of cases of nontraumatic vascular necrosis with the involvement of more than one bone is highly challenging. Due to the progression of the disease, it was difficult to use common therapies based on the stages of disease.

Manohara et al. described a case of AVN of the proximal carpal row with a history of ligamentous perilunate dislocation. The patients declined further surgeries and chose nonsurgical treatments.[1] Didden et al. have reported a case of osteonecrosis of the entire proximal carpal row of the wrist, started briefly after intravenous bisphosphonate administration. Eventually, the patient was treated with a proximal row carpectomy.[3] Bayley and Simmons have reported a case of AVN of the proximal carpal row with a history of corticosteroid consumption, hyperlipidemia, and moderate intake of alcohol. The patient was treated with a radial-proximal carpal row arthrodesis.[2] Park et al. described a case of coexisting AVN of the scaphoid and lunate. They treated the patient with radioscapholunate arthrodesis.[4] In the report by Pulikkottil et al., a patient with simultaneous AVN of the lunate and scaphoid was treated with proximal row carpectomy which induced excellent pain relief and indicated increased wrist ROM.[7]

The association of AVN of the lunate and proximal pole of the scaphoid and thalassemia minor in this patient may be coincidental; however, the association of aseptic necrosis and thalassemia minor, mostly in the femoral head, has been described before.[8-11] Although the association of AVN of the lunate with sickle cell anemia[12] and spherocytosis[13] has been reported, to the best of our knowledge, concurrent AVN of the lunate and proximal pole of the scaphoid and thalassemia minor have not been previously reported. Long-standing hematological disorders, medullary hyperplasia, vascular events with possible bone marrow fat emboli, and an increase in intraosseous pressure might be the contributing factors.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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