CASE REPORT

Kienbock’s disease and juvenile idiopathic arthritis

Nicholas M. Desy*, Mitchell Bernstein, Edward J. Harvey, Elizabeth Hazel

ABSTRACT: Kienbock’s disease or osteonecrosis of the lunate is an uncommon cause of wrist pain. Though there have been several reports of cases in patients with various rheumatologic diseases, the precise etiology has currently not been established. We report a case of Kienbock’s disease that occurred in a patient with juvenile idiopathic arthritis. To our knowledge, this is the first case report with an association between these two conditions.

Keywords: Kienbock’s disease, osteonecrosis, juvenile idiopathic arthritis, lunatomalacia, avascular necrosis

INTRODUCTION

The etiology of Kienbock’s disease, also known as osteonecrosis of the lunate, remains controversial. It commonly occurs in patients twenty to forty years of age and presents with pain and stiffness in the dorsomedial aspect of the wrist. Several risk factors have been established to help explain its etiology: acute or repetitive trauma, variation in blood supply to the lunate, differences in the anatomy and shape of the lunate bone, and venous congestion (1, 2). Abnormal biomechanics at the radiocarpal joint between the distal radius and ulna has also been implicated in Kienbock’s disease (3, 4). Positive ulnar variance describes the length relationship between the articular surfaces of the radius and ulna at the radiocarpal joint. Positive ulnar variance indicates that the ulna is longer than the radius, while negative ulnar variance indicates that the ulna is shorter at the wrist joint. In neutral ulnar variance 80% of the axial load at the wrist is transmitted through the distal radius. As ulnar variance decreases to more negative values, the load transmission across the distal radiocarpal joint increases, subsequently exposing the lunate to abnormal higher pressures and potentially increasing the risk of Kienbock’s disease (3, 4).

Kienbock’s disease is also associated with systemic lupus erythematosus (SLE) (5-8), antiphospholipid antibody syndrome (9), sickle cell anemia (10), and Crohn’s enteritis (11). Multiple hereditary osteochondromata (12), carpal coalition (13, 14) and congenital shortening of the radius and ulna in Langer-Giedion syndrome (15), are other anatomic abnormalities that have been reported with Kienbock’s disease. Rheumatic diseases, including scleroderma (16-18), rheumatoid arthritis (19), gout (20, 21) and dermatomyositis (22) have been published in association with Kienbock’s, but there have been no identifiable cases in patients with juvenile idiopathic arthritis (JIA). A literature review is done to illustrate the proposed etiologies of Kienbock’s disease and its association with other rheumatologic conditions.

CASE REPORT

A 20-year-old right-handed female with known rheumatoid factor negative polyarticular JIA presented to the clinic because of pain and limited range of motion in the left wrist. She was diagnosed with JIA at the age of nine after a two-month history of pain and swelling in both hands and knees. She reported difficulty with recreational activities. During the course of her illness several other joints progressively became involved. During the first year of treatment, she was prescribed nonsteroidal anti-inflammatory medication and low-dose prednisolone. To help control her symptoms she required disease-modifying antirheumatic drugs. Her medications included methotrexate 20 mg weekly, folinic acid 2.5 mg weekly, and etanercept 25 mg twice a week.

Three weeks prior to presentation, the patient experienced a severe flare of her arthritis due to non-compliance with her medication. This lead to persistent left wrist pain and limited range-of-motion. On examination she demonstrated synovial thickening of her left wrist with no palpable effusion. Magnetic resonance imaging of her left wrist showed mild synovial thickening and erosive arthropathy throughout the carpus, radiocarpal, and carpometacarpal joints. In addition, there is possible sclerosis and edema, since she had collapse of the lunate with mixed signal intensity. Plain radiographs revealed negative ulnar variance, sclerosis, and loss of lunate height (Fig. 1). The imaging was compatible with Stage 4 osteonecrosis of the lunate (23). The patient was managed non-operatively and at two-year follow-up was asymptomatic with no concomitant worsening of lunate osteonecrosis on radiograph.

DISCUSSION

The etiology of Kienbock’s disease remains unclear (Table 1). The current literature indicates that most cases of Kienbock’s disease develop without a history of trauma and posit that extrascleal and intrascleal blood supply to the lunate have a role in the disease process. The extrascleal blood supply is formed by a series of dorsal and volar vascular arches (1, 24). The intrascleal blood supply is made up of branches entering the volar and dorsal poles however the composite of this vasculature is variable (1, 25-27); branches demonstrate different anastomosis patterns inside the lunate, or the lunate may be supplied by only one dorsal or volar branch or By both a dorsal and volar arterial supply the lunate without any anastomosis (26). When an anastomosis does exist, it can be characterized as a you, x, or y pattern, depending on the amount of vessels supplying each pole (24, 27). Depending on the intrascleal vascular anatomy

Figure 1: A. Coronal T-weighted fast-spin-echo magnetic resonance image showing collapse of the lunate with mixed signal intensity suggesting sclerosis and edema consistent with Kienbock’s disease (arrow). Erosive arthropathy throughout the carpus and negative ulnar variance are also noted. B. Antero-posterior plain X-ray of the left wrist demonstrates sclerosis and slight loss of height involving the lunate compatible with Kienbock’s disease (arrow). Negative ulnar variance and degenerative changes are also seen.
of the lunate, certain lunate bones are predisposed to Kienbock’s disease. This concept was highlighted in a case report of Kienbock’s disease associated with sickle cell anemia (10). The osteonecrosis was thought to have developed from an at-risk lunate - single volar arterial supply - along with significant vascular sickling and stasis. Venous congestion has also been attributed to the pathogenesis of Kienbock’s disease (2). During surgery, Jensen measured increased pressure inside the lunate compared with the radial styloid and capitale. He concluded that the higher pressure was caused by venous congestion leading to osteonecrosis of the lunate.

Negative ulnar variance is also implicated in the pathogenesis of Kienbock’s disease (3, 4, 28). The altered relationship between the ulna and radius at the distal radioulnar joint modifies the biomechanics at the radiolunate joint and increases strains on the lunate. This postulation is still controversial because several studies, including a meta-analysis, have shown that negative ulnar variance is not a risk factor for developing Kienbock’s disease, (29, 30). On the contrary, Ledoux et al. performed a finite-element analysis on cadaveric lunate bones and found that the progression of a fracture of the lunate was present with negative ulnar variance, a high lunate uncovering index, which is the amount of lunate outside the lunate fossa of the radius compared to the amount of lunate articulating with the lunate fossa, and angulated trabeaculae (31). This suggests that given the circumstance, the lunate can be at risk for developing osteonecrosis due to abnormal stresses.

Further cases have also reported patients with conditions that may have caused altered stresses to the lunate, which in turn led to Kienbock’s disease. Schuind et al. reported a case of Kienbock’s disease associated with congenital shortening of the ulna as seen in Langer-Giedion syndrome (15). It was suggested that Kienbock’s disease developed from microfractures sustained by an abnormal stress distribution (15). Multiple hereditary osteochondromata in the forearm was also found in association with Kienbock’s disease and was attributed to an excess load on the lunate by negative ulnar variance, but with no carpal slip (12).

Systemic lupus erythematosus has been associated with avascular necrosis of bone. In 1977, Urman presented several cases of patients with SLE and osteonecrosis of the carpal bones, including a case report of a patient with SLE and Kienbock’s disease (8). The patient also had a history of Raynaud’s phenomenon and was taking high-dose corticosteroids. In SLE patients treated with corticosteroids and who developed osteonecrosis, there was one patient who developed Kienbock’s osteonecrosis of the lunate. This patient was treated with low-dose corticosteroids compared to those who did not develop Kienbock’s disease. Mok et al. reported a case of bilateral Kienbock’s disease in a patient with SLE (5). They believed that low-dose corticosteroids would not cause the Kienbock’s disease occurred in this patient due to an increase in intra-articular pressure within the wrist compartment, causing impingement of venous return, with or without ankylosis to the lunate, and subsequent osteonecrosis (19).

In addition to the various risk factors mentioned above, Kienbock’s disease b is prevalent in specific patient populations. Rooker et al. found an increased prevalence of Kienbock’s disease in a group of patients with cerebral palsy (34). Several changes occur at the wrist, including narrowing of the intercarpal spaces, premature ossification of the carpal bones, and early fusion of the ulnar epiphysis leading to a shorter ulna (negative ulnar variance) (35). The wrist ultimately becomes displaced ulnarily and volarly leading to a dislocation of the wrist and bayonet deformity. Therefore, it is possible that the abnormalities in the wrist associated with JIA could lead to abnormal stresses and or pressures in the wrist that led to Kienbock’s disease. Furthermore, the erosive changes in the joint may lead to a change on the normal force patterns in the patient’s carpus. Seven years prior to the onset of Kienbock’s disease, our patient was also treated with low-dose corticosteroids which could have also disordered circulation and led to the development of lunate osteonecrosis.

The precise etiology of Kienbock’s disease remains elusive. Several theories attempt to explain its pathogenesis, which suggests that it may be multifactorial. Many risk factors have also been identified; steroid treatment, a predisposing rheumatologic disease, a variation in lunate blood supply, and possibly negative ulnar variance. Our case demonstrates the possible role of long-term corticosteroid therapy and Raynaud’s phenomenon in Kienbock’s disease and JIA. It also suggests that Kienbock’s disease could be a possible cause of wrist pain and stiffness in patients with JIA.

REFERENCES