

Obojestranska Kienböckova bolezen z obojestransko negativno ulnarno varianto: poročilo o primeru

Bilateral Kienböck's disease with bilateral negative ulnar variance: a case report

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Izvleček

Namen: Kienböckova bolezen se navadno javlja enostransko, in sicer na dominantni roki. Obojestranska avaskularna nekroza lunatuma je izjemno redko opisano stanje, še manj pogosta pa je sočasna pojavnost z negativno ulnarno varianto – ulno minus.

Poročilo o primeru: Članek opisuje bolnika, pri katerem smo z magnetno resonanco ugotovili obojestransko avaskularno nekrozo lunatuma. Prav tako je na magnetni resonanci potrjena ulna minus, ki je bila ugotovljena z RTG-slikanjem.

Zaključek: Čeprav je povezava Kienböckove bolezni in negativne ulnarne variante še vedno nejasna, bi ta obojestranska sočasnost pri obravnavanem bolniku lahko prispevala k nadaljnjemu raziskovanju etiologije avaskularne nekroze lunatuma.

Abstract

Purpose: Kienböck's disease usually occurs unilaterally on dominant hands. As such, bilateral avascular necrosis of the lunate, in concurrence with bilateral negative ulnar variance (NUV), is a very rarely described condition.

Case report: The case of a patient diagnosed with bilateral avascular necrosis of the lunate is presented. Plain film and magnetic resonance imaging confirmed bilateral NUV was also present.

Conclusions: Although the relationship between Kienböck's disease and NUV remains unclear, the bilateral concurrent appearance of both might instigate a discussion of the possible etiology of avascular lunate necrosis.

INTRODUCTION

Kienböck's disease or lunatomalacia is a condition characterized by avascular necrosis (AVN) of the lunate bone, which most commonly occurs unilaterally. Although mechanical, anatomic, and systemic mechanisms have all been implicated in the development of the Kienböck's disease process, no specific etiologic mechanism has been identified; as such, the complete pathogenesis remains unclear (1–5).

Different stages of negative ulnar variance (NUV) have been demonstrated after collapse of the lunate, indicating that it plays a role in the development and progression of Kienböck's disease (3, 6).

Magnetic resonance imaging (MRI) is important in the diagnosis and staging of Kienböck's disease, especially in the early stages (1). This report describes a patient with concurrent bilateral Kienböck's disease and NUV in both wrists.

CASE PRESENTATION

A 28-year-old male presented with moderate wrist pain of one-year duration in both arms. The pain started gradually, after an apartment renovation, first appearing on the non-dominant left wrist; half a year later, the right wrist became painful as well. No specific injury was reported, and no carpal tunnel syndrome symptoms were present. The patient had no history of systemic disease.

Clinical examination showed symmetrical wrists without signs of inflammation or swelling, symmetrical grip strength, painful flexion and extension in both wrists, and preserved pronation and supination. Watson's test on the left wrist was painful, with the characteristic "clunk". The right wrist was pain-free during the test. Watson's test also revealed bilateral effusion over the scapholunate area.

Plain films (Figure 1) showed a fissure of the lunate on the left wrist, and bilateral NUV, which was -3 mm on the right arm, and -2.5 mm on the left arm according to the Gelbermann method (7). There were



Figure 1. Plain film showing the fissure of the left lunate (thin arrow) and the partial collapse (thick arrow) of the right lunate. Note also the bilateral NUV.

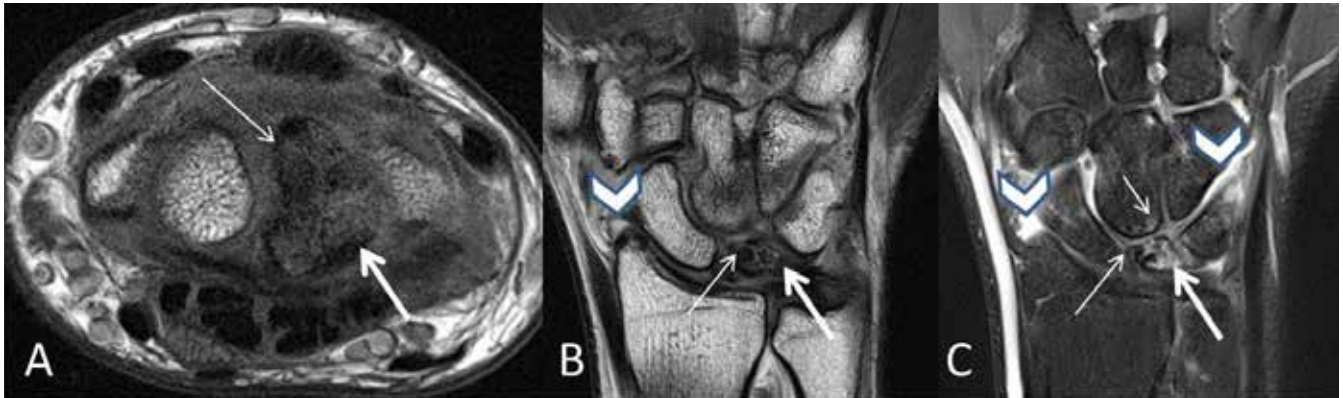


Figure 2. Transverse T1-weighted (A) and coronal non-fat-suppressed (B) and fat-suppressed (C) proton density (PD)-weighted MRI images of the right wrist. There is a collapse and fragmentation of the lunate. At the navicular side, there is predominantly a hypointense signal of the fragments indicating sclerosis (thin arrows), whereas in the larger part of the bone (thick arrows) there is a moderately T1 hypointense and PD hyperintense signal, indicating bone marrow edema (BME), which is much more easily observed on the fat-suppressed image. Note the small area of the BME at the proximal part of the capitulum, indicating mild impaction with the collapsed lunatum (short arrow). Note also the effusion surrounding the radial half of the scaphoid bone as well as in the triquetrohamate joint, indicating possible reactive synovitis (chevrons). The NUV is clearly visible even without detailed measurements.

no signs of osteoarthritis. The radiolunate coverage area was not significantly decreased (8).

An MRI confirmed fissure of the left lunate, and additionally showed a fragmented and irregular appearance of the right lunate on PD (proton-density) and PD FAT SAT (proton-density with fat suppression) sequences, with effusions in the ulnocarpal and intercarpal joints (Figures 2, 3).

These findings are consistent with a diagnosis of bilateral avascular lunate necrosis, stage 3A, according to Lichtmann et al. (9).

The patient is scheduled for bilateral radial shortening.

DISCUSSION

Bilateral Kienböck's disease is a rare condition, present in only 4% of all patients with AVN of the lunate bone (4). Concurrent bilateral NUV makes this case even rarer. Determining the etiology of AVN of the lunate bone is important for finding the most efficacious treatment.

The etiology of osteonecrosis in general remains controversial (4). The fact that the lunate is almost the only bone of the carpus that may undergo complete necrosis suggests that the process is somehow related to its vascularity, biomechanics, osseous anatomy, and/or to the morphological features of the distal forearm and the surrounding carpal bones (2). NUV is also associated with changes in the triangular fibrocartilage (TFC). The thickness of the TFC is proportional to the NUV; the shorter the ulna, the thicker the TFC (10). These changes modify normal support for the ulnar half of the lunate bone, reinforcing the effects of repeated trauma, leading to an increased risk of microfracture, and eventual necrosis (11).

The increased loading between the capitate and ulnar parts of the radius, which damages the radial part of the lunate, was named the "nutcracker effect" (8, 10). This was well observed on the left wrist of our patient, where he suffered a fracture of the lunate bone.

An MRI is useful in the diagnosis of AVN, and is especially important in its early stages, when Kienböck's

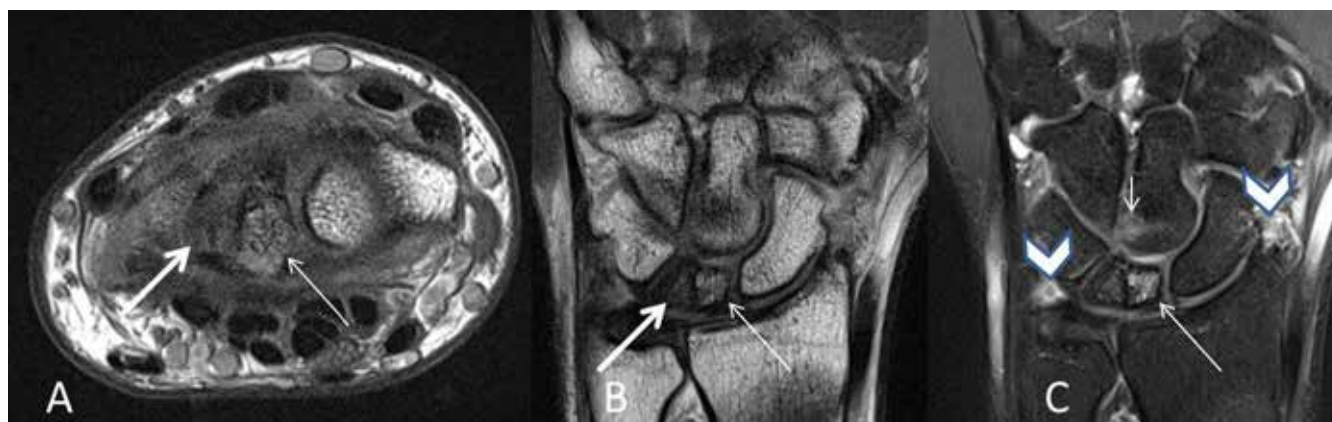


Figure 3. Transverse T1-weighted (A), and coronal PD without (B) and with (C) fat suppression MRI of the left wrist. A fissure of the lunate separates the radial (thin arrow) fragment with the T1 mildly hypointense, and PD hyperintense signal, indicating the BME and the ulnar (thick arrow) fragment with T1/PD moderately hypointense, and fat-suppressed PD mildly hyperintense bone marrow signal, indicating mixed sclerotic, and oedematous structure of the bone. Note the preserved shape of the lunate, and a small focus of BME (short arrow) in the proximal capitate, indicating possible impaction. Note also a thickened synovium at the tips of ulnar, and radial styloids (chevrons). Again, note the shorter ulna.

disease is suspected. Normal bone marrow has a high signal intensity on T1-weighted imaging (T1) because of adipose and hematopoietic cells (12).

Since the presented patient is not a manual worker nor a professional sportsman, it is quite unusual that he developed bilateral AVN of the lunate directly after a brief exertion, during the apartment renovation. We suggest that the bilateral Kienböck's disease in the presented case originates from patient-related pathological tendencies and morphological predispositions, such as NUV, rather than from mechanical overload on the joints alone.

NUV seems to be the crucial predisposition in the presented case, leading to some kind of vascular obstruction, which caused AVN of the lunate bone. NUV probably resulted in the transmission of a mechanical force to the radial side of the lunate bone, and NUV is correlated with a weaker radioscapholunate ligament or a thicker TFC; these factors together probably contributed to the development of vascular pathology of the lunate bone.

The relationship between lunate AVN and NUV remains unclear, therefore we consider the presented case as a contribution to the debate and valuable information for future investigations.

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