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Is osteonecrosis of the lunate bone an underestimated feature of systemic sclerosis? A case series of nine patients and review of literature

Marc Frerix, MD[a,*,1], Kai Kröger[a,b,1], Gabor Szalay, MD[c], Ulf Müller-Ladner, MD[a], Ingo Helmut Tarner, MD[a]

[a] Department of Internal Medicine and Rheumatology, Justus-Liebig-University Giessen, Kerckhoff-Clinic Bad Nauheim, Benekestr. 2-8, Bad Nauheim 61231, Germany
[b] Department of Radiotherapy, University Hospital Münster, Münster, Germany
[c] Department of Trauma Surgery, University of Giessen and Marburg, Giessen, Germany

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ABSTRACT

Osteonecrosis of the lunate bone, also known as Kienböck's disease, is a very rare disease of unknown cause. Until today, only six cases of osteonecrosis of the lunate bone in patients with systemic sclerosis (SSc) have been reported in the literature. It is unknown whether these few cases reflect only a coincidence of two rare diseases or whether osteonecrosis of the lunate bone is a potential currently underestimated disease-associated feature of SSc. In this study, we report the clinical course of nine SSc patients with magnetic resonance imaging proven osteonecrosis of the lunate bone and discuss associated disease characteristics and potential underlying pathophysiological mechanisms. Overall, our observations suggest that osteonecrosis of the lunate bone is a frequent and so far under-recognized manifestation of SSc which might be linked to SSc-related vasculopathy. It is important to distinguish osteonecrosis of the lunate bone from wrist arthritis in SSc patients because the clinical treatment is different. In general, the clinical progression of osteonecrosis of the lunate bone seems to be slow in SSc patients. As most of the patients have only minor complaints, watchful waiting in combination with analgesic therapy seems to be a feasible treatment approach in most patients whether an operative intervention might be necessary in rapid progressive cases.

Introduction

Osteonecrosis of the lunate bone, first systematically described in 1910 known as Kienböck's disease or lunate malacia [1], is a very rare disease of unknown cause without reliable data on its pathophysiology and prevalence in the general population. Moreover, there is still little information about the natural history of the disease [2] and despite several surgical treatment approaches, the evidence to favor a particular surgical treatment is limited [3].

Systemic sclerosis (SSc) is an orphan disease with an estimated prevalence between 150 and 300 cases per million in most populations worldwide [4]. Approximately one out of four SSc patients is affected by synovitis, which is predominantly localized at the wrist and sometimes clinically inapparent [5].

Until today, only six cases of osteonecrosis of the lunate bone in SSc patients have been described in the literature, with bilateral manifestation in three patients [6–9]. However, the question, whether lunate changes are a specific disease-associated manifestation of SSc organ involvement and how to treat the affected patients has not yet been resolved. Recently, our study group analyzed 38 hands of 26 SSc patients in a low-field magnetic resonance imaging (MRI) study. Besides inflammatory arthritis in 79% of the examined hands, we observed an unexpected high frequency of erosions in 45% and pathologic bone marrow edema in 18% of lunate bones [10]. These lesions seem to be different from arthritis and resemble early stages of lunate osteonecrosis [11]. The coincidence of two rare diseases raises the question, whether osteonecrosis of the lunate bone is a manifestation secondary to SSc, possibly as a consequence of SSc vasculopathy.

Case report series

In this case series, we describe the clinical course of nine SSc patients with MRI-proven osteonecrosis of the lunate bone. Five patients have been classified as limited cutaneous (lcSSc) and four

* Corresponding author.
E-mail address: m.frerix@kerckhoff-klinik.de (M. Frerix).
[1] Contributed equally.

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patients as diffuse cutaneous (dcSSc) disease subtype in accordance with the criteria by LeRoy et al. [12] all patients fulfilled the recently published new ACR/EULAR criteria for classification of systemic sclerosis [13]. Seven patients were female (77.8%), the mean age was 61.3 years (range: 48–71), and the mean disease duration 10.7 years (range 4–20) since the first non-Raynaud symptom. All patients suffered from severe Raynaud's phenomenon, seven (77.8%) patients had digital ulcers. In the following, we provide a concise report of each case with focus on the osteonecrosis of the lunate bone, The lunate bone changes have been classified as suggested by Lichtman et al. [14].

- *Stage I*: Overall unremarkable radiographs except possible linear fracture, but diffuse T1 signal alteration of the lunate bone.
- *Stage II*: Sclerosis resulting in density changes of the lunate but no collapse present.
- *Stage IIIA*: Partial collapse of the lunate bone without alteration of the carpal joint alignment.
- *Stage IIIB*: Collapse with proximal migration of lunate bone and fixed flexion of the scaphoid.
- *Stage IV*: Resembles changes of stage III with additional carpal arthritis.

An overview of the most important disease characteristics and comedication is presented in the Table.

Patient 1: 68-Year-old male dcSSc patient with arthritis and unilateral destructive lesions of the right lunate bone

A 68-year-old man suffered from dcSSc including severe Raynaud's phenomenon (RP), rapid progressive dermal fibrosis, lung fibrosis, MRI-proven inflammatory myocarditis, gastrointestinal and musculoskeletal involvement.

When he developed bilateral wrist pain for the first time, a low-field MRI of the right hand revealed severe carpal arthritis. Additionally, besides a palmar radial cyst there was a severe proximal lesion of the lunate bone and an impingement of the triquetral bone associated with a positive ulnar variance (Fig. 1A). He previously received methotrexat and cyclophosphamide for treatment of skin and cardiac involvement. Because of ongoing bilateral polyarthritis of the wrists and feet refractory to conventional treatment, the therapy was changed to biannual 2 × 1000 mg rituximab infusions, with good clinical response regarding wrist and muscular pain and increasing pain at the end of treatment cycles. At time of follow-up 3 years later, the patient did not report any wrist pain. Clinical and ultrasound examination showed no signs of synovitis, thus the arthritis was in remission according to clinical parameters. However, because of the previously described large lesion of the lunate bone suggestive of osteonecrosis, a low-field MRI of the right hand was repeated. The known cyst had increased in size and an effusion in the scaphotrapezotrapezoidal joint had occurred. The lunate bone still showed the previously described pathologic signal alterations but no significant worsening. The shape of the lesion remained intact but with considerable bone-marrow edema (Fig. 1B). In contrast, X-ray studies of the wrist revealed only minor changes of the lunate bone and early osteoarthritis of the first metacarpophalangeal joint (Fig. 1C). This corresponds to stage 2 in the Lichtman classification [11].

To evaluate potential vasculopathy associated with osteonecrosis, a nailfold videocapillaroscopy (NVC) and duplex sonography of the patient's distal upper extremity arteries were performed. NVC showed a typical late SSc pattern with loss of capillaries and neovascularization (but without giant capillaries or hemorrhages). Duplex sonography revealed a bilateral total occlusion of the distal ulnar arteries (beginning approximately 10 cm proximal of both wrists) without compensatory increased blood flow in both radial arteries.

Besides systemic sclerosis vasculopathy itself, low-dose corticosteroid therapy could have served as a potential risk factor for developing osteonecrosis. Other potential risk factors for osteonecrosis of the lunate bone such as negative ulnar variance, repetitive trauma to the wrist, or coagulation disorders were not present in this patient.

Patient 2: 71-Year-old male lcSSc patient with suspected osteonecrosis of the right lunate bone

A 71-year-old man with lcSSc predominantly suffering from sclerodactyly, severe RP with recurring finger-tip ulcerations, gastrointestinal and interstitial lung involvement underwent a low-field MRI of both hands even without clinical symptoms for the purpose of a clinical study on joint involvement in SSc [10]. It revealed an early clinical asymptomatic stage of lunate osteonecrosis in his right hand with bone marrow edema in the T2-weighted images and diffuse signal alterations in the T1-weighted images after application of contrast agent corresponding to a Lichtman stage between 1 and 2 as plain radiographs were normal but MRI revealed multiple fracture lines (Fig. 2A) [11]. The lunate bone of his left hand did not show any signs of osteonecrosis, only a small cyst. As the patient's only symptoms regarding his hands were RP, sclerodactyly with joint contractures, and digital ulcers secondary to calcinosis cutis, no treatment regarding the changes in the lunate bone was initiated.

At 3 years later, upon physical examination the wrist's range of motion was found to be limited on both sides. An MRI of the right hand showed a considerable progress of signal alterations of the lunate bone which had started to collapse and the cartilage had thinned (Fig. 2B). A radiograph was obtained which showed osteoarthritis of both wrists and a slightly indented right lunate bone with increased density but only minimal changes of the left lunate bone structure (Fig. 2C). Thus, the process in the right lunate bone had progressed to stage 3A of the Lichtman classification. Of note, the patient denied wrist pain at any time.

NVC showed a typical late SSc-pattern with avascular areas, neovascularisation, bushy capillaries, and only a few microhemorrhages. Both ulnar arteries were occluded at the wrist confirmed by duplex sonography. With respect to additional risk factors for osteonecrosis, the patient worked as a stonemason for 15 years and was taking low-dose prednisolone until his first hospital admission 8 years ago.

Patient 3: 50-Year-old female lcSSc patient with unilateral MRI changes of the complete right lunate bone

This woman with lcSSc had a history of severe RP and finger-tip ulcers for 5 years until she complained of wrist pain. A low-field MRI was performed and showed carpal arthritis in both wrists. Therefore, therapy with methotrexate was started. At 3 months later, the carpal arthritis improved clinically and on MRI. At 2 years later, prednisolone was introduced for treatment of pericardial effusion even a cardiac MRI did not show any signs of permyocarditis. At time of this visit, a review of the previous low-field MRI studies of both the wrists 2 years ago showed an already present large area of hypointensity in the T1-weighted sequence in the proximal part of the right lunate bone. Almost the entire bone was edematous in the STIR-sequence and showed abnormalities in the uptake of contrast agent. In contrast, the left lunate did not show any signs of osteonecrosis, just several small cysts. Therefore, MRI of the right wrist was repeated and confirmed the previous findings with diffuse signal alterations, but no significant progression of the disease (Fig. 3A and B). At the same time,