



Case Report

A Rare Condition in the Differential Diagnosis of Arthritis, Preiser's Disease: A Case Report and Review of the Literature

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Abstract

Chronic wrist pain is a common clinical condition that negatively affects daily activities. Many diseases are involved in the etiology of chronic wrist pain. Identification of specific causes of pain increases treatment success and prevents disability. Detailed evaluation and necessary imaging methods for differential diagnosis of multiple clinical entities with similar history and examination findings are essential for accurate diagnosis and treatment success. Avascular necrosis is a progressive disorder resulting in partial or complete death of bone tissue cells due to impaired blood circulation of the bone, usually caused by nontraumatic or idiopathic etiology. Preiser's disease, avascular necrosis of the carpal scaphoid, is a rare disorder generally occurring on the dominant side and in women. This case report presents an uncommon disease that should be considered in the differential diagnosis while searching for the etiologic cause of chronic wrist pain.

Keywords: Preiser's disease, arthritis, differential diagnosis

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1. Introduction

Chronic wrist pain is a common clinical condition that negatively affects daily activities. The wrist joint's anatomical complexity and biomechanical properties can pose challenges for accurate diagnosis and appropriate treatment planning. The etiology of chronic wrist pain includes many diseases. Identifying specific causes of pain increases treatment success and prevents disability. Detailed evaluation and necessary imaging methods for differential diagnosis of multiple clinical entities with similar clinical findings are essential for accurate diagnosis and treatment success. The causes of chronic wrist pain include inflammatory diseases, crystalline arthropathies, osteoarthritis, De Quervain tenosynovitis, ganglion cyst, and avascular necrosis (AVN) of the carpal bones [1, 2].

AVN occurs when there's a disruption in the blood supply to a bone, leading to the death of bone tissue. This lack of blood flow can cause the bone to weaken and collapse over time. Although the condition can affect any bone, it is most commonly seen in the hip, knee, and shoulder. Preiser's disease was defined by Georg Preiser in 1910 as AVN of the scaphoid bone of unknown cause. Since its first description, 14 cases were reported by Herbert *et al.* and 126 cases by Lenoir *et al.* Bergman *et al.* documented 170 cases of AVN in the literature up to 2021 [3–5].

Preiser's disease is rare, and its natural history remains poorly understood. This condition, characterized by AVN of the scaphoid bone in the wrist, often lacks a well-defined progression pattern, making it challenging for researchers and clinicians to fully grasp how it develops and progresses over time. Although there is no definitive evidence, the underlying pathology is thought to result

from impaired blood supply to the scaphoid bone. Patients typically present with tenderness in the scaphoid region, radial-sided wrist pain and decreased wrist motion [6]. The etiology is unclear, however, it is thought that recurrent trauma, steroid use, chemotherapy, connective tissue diseases, biomechanical or anatomical variations may be risk factors. It has been reported to be more common in the female gender in case series in the literature, but it can be seen in different age groups and at any age. The incidence is higher on the dominant side. After a thorough history and clinical examination, the diagnosis of Preiser's disease can be guided by imaging techniques such as computed tomography (CT), magnetic resonance imaging (MRI), and arthroscopy. These tools help visualize the extent of bone damage and assess the condition of the scaphoid bone, aiding in accurate diagnosis and treatment planning. Since direct radiography makes early diagnosis difficult, MRI should be performed in the presence of clinical suspicion [3, 7]. Case series large enough to reveal the etiology and pathogenesis are rare in the literature. Therefore, treatment has not yet been standardized and treatment options depend on the progression of the disease [8].

This case report aims to present Preiser's disease. This rare disease should not be forgotten in the differential diagnosis while searching for the etiologic cause of chronic wrist pain, and to review the literature.

2. Case Presentation

A 74-year-old woman presented to the physical medicine and rehabilitation outpatient clinic with complaints of pain and swelling in the right shoulder and right wrist. In her history, the patient stated that right shoulder and right wrist pain started about four months ago and

there was no history of trauma. She was unresponsive to analgesics and myorelaxants and had night pain. Rheumatologic examination was unremarkable except for intermittent joint pain. Her medical history included hypertension, diabetes mellitus, and hypothyroidism. The patient was not using corticosteroids. Her family history and system examination were unremarkable. Physical examination revealed tenderness on palpation in the right shoulder and right wrist, swelling and increased temperature in the right wrist, and painful and limited range of motion. Other locomotor system examination was normal. Laboratory findings included C-reactive protein 4.37 (0–0.5mg/L), sedimentation 56 (0–25 mm/h), brucella negative, rheumatoid factor level 18.3 IU/ml (0–20 IU/ml), anticyclic citrulline peptide (<7U/ml) negative. There was no pathology in biochemical parameters. Doppler and superficial ultrasonography of the wrist and forearm were normal. There were no findings on hand-wrist radiography. MRI of the right hand and wrist, initially conducted with a suspected diagnosis of arthritis, revealed AVN of the scaphoid bone (Figure 1). When clinical and imaging findings were evaluated, nontraumatic AVN was considered. Immobilization was provided with a hand and wrist splint due to AVN. Nonsteroidal anti-inflammatory drugs (NSAIDs) were started for conservative treatment, and physiotherapy with transcutaneous electrical nerve stimulation, cold pack, and intermittent ultrasound was initiated. The patient was followed up clinically.

3. Discussion

Osteonecrosis occurs when there is insufficient blood supply to the bone due to impaired blood circulation. Osteonecrosis, which is often an irreversible event, develops in four stages. The initial stage of AVN is the “avascular” stage,

which commences following a bone infarction due to impaired blood supply. During this phase, ischemia also induces cartilage hypertrophy as part of the repair process, leading to endochondral ossification to address the bone defect. Radiologic changes are unclear at this stage. Osteopenia can result from the inflammatory response and activation of osteoclasts. The next phase is the “revascularization” stage, characterized by heightened osteoblastic activity due to the combined actions of osteoclasts and osteoblasts. In this stage, radiographic images typically display areas of increased intensity and enhanced sclerosis. Additionally, subchondral bone necrosis may lead to further bone destruction. The repair phase starts with revascularization, and while partial bone healing may sometimes be seen, it is pretty rare for the scaphoid bone. The degree of repair largely depends on the initial extent of damage and the amount of bone loss. Sometimes, the final repair may not fully compensate for the bone loss. Over time, the affected bone develops osteoarthritic changes and the joint space narrows. Cysts, osteosclerotic and osteoporotic areas occur. Deformity and instability may occur [8, 9]. The etiology of AVN of the scaphoid bone is still not fully understood. It may occur after fracture, repetitive trauma, systemic disease, steroid use, or idiopathically. Joint swelling and underdeveloped vascular networks that cause intraosseous and extraosseous disconnection and mechanical stresses such as repetitive strain and overload can cause impaired blood flow. Pain is the most critical symptom in AVN. On physical examination, it is usually seen that the movements in the affected joint are limited and accompanied by local tenderness. Osteoarthritic changes, joint space narrowing, areas of osteosclerosis and osteoporosis, and subchondral cysts are commonly observed. Following a thorough history and clinical examination, CT, MRI, and

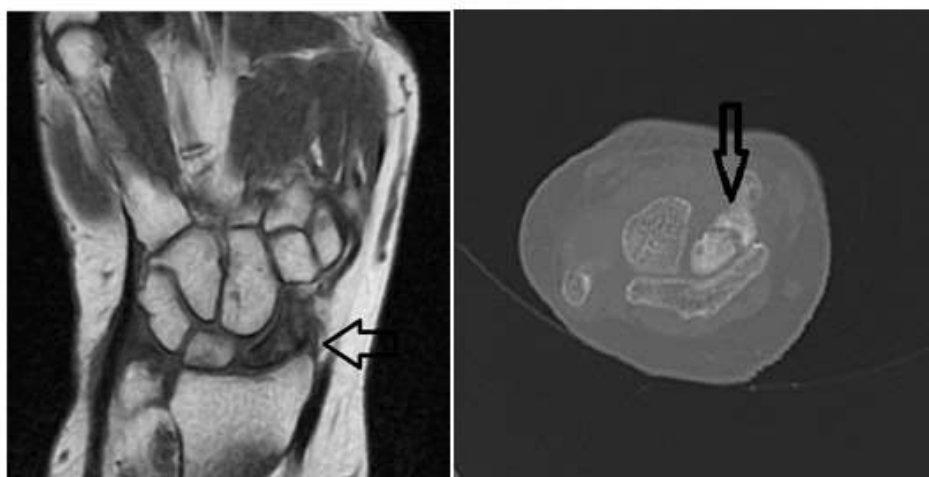


Figure 1: Magnetic resonance imaging of the right hand and wrist, and avascular necrosis of the scaphoid bone.

arthroscopy are crucial in assessing the scaphoid bone, wrist, and carpal instability [7]. MRI is the most sensitive method in early diagnosis and is a guide for the differential diagnosis of other diseases involved in the etiology of chronic wrist pain. With contrast-enhanced MRI, necrotic tissue (proximal), hypervascular repair tissue (middle), and viable bone (distal) can be visualized as three regions.

The most widely used radiologic classification was established by Herbert and Lanzetta in 1994. This classification includes four stages: Stage 1 is characterized by a normal X-ray image with a positive bone scan. Stage 2 shows increased density at the proximal end of the scaphoid and diffuse osteoporosis. Stage 3 is marked by fragmentation and/or pathologic fractures at the proximal end. Finally, Stage 4 is associated with carpal collapse and the development of osteoarthritis [3]. According to MRI records, Kalainov *et al.* determined that two types of disease state were possible. Type I is characterized by diffuse necrosis involving the entire scaphoid, and the prognosis is worse. On average, type II is characterized by necrosis located more proximally at 42% of the scaphoid, and prognosis is much better [8]. Due to its rarity and the fact that different treatment options have

been reported in the literature related to the case series, information regarding the indications for treatment varies. Various case series have been reported in the literature; however, treatment strategies for Preiser's disease have not yet been standardized. In the case series, patient factors and the degree of AVN were reported to be important in the treatment plan. Although conservative treatment was recommended for the early stage of Preiser's disease in a few cases, it was later reported that immobilization was not indicated in adult Preiser's disease. Surgical debridement, denervation, localized cortisone injection, radial wedge osteotomy, and vascularized or nonvascularized bone grafts have also been reported as treatment options. Vascularized bone grafting has been recommended for patients with stage 2 or 3 disease. In more severe cases, partial fusion procedures like proximal row carpectomy or four-corner fusion have been documented [10–12].

In this case, which had any known risk factors, we thought that repeated minor forceful traumas caused scaphoid AVN and followed our patient with nonsurgical treatments. Since cases at different stages of the disease are reported in the literature, treatment options differ according to the stage of the disease. However, there is

no consensus on a definite treatment protocol. According to MRI records of the patients, type II Preiser disease was discovered and immobilization was provided. NSAIDs and physiotherapy were used for conservative treatment. The surgeon did not recommend surgical treatments. Close monitoring was proposed, and a control MRI was planned after doing nonsurgical treatment. However, the patient did not come for follow-up, and the control MRI result could not be obtained.

4. Conclusion

Scaphoid bone AVN can occur due to nontraumatic causes and cause functional losses. In this case report, we presented an uncommon disease that should be considered in the differential diagnosis of arthritis while searching for the etiologic cause of chronic wrist pain.

Declarations

Acknowledgments

None.

Ethical Considerations

The authors declare that the study was conducted following the Declaration of Helsinki.

The patient completed a written consent form.

Competing Interests

The authors report no conflicts of interest related to the publication of this article.

Availability of Data and Material

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Abbreviations and Symbols

CT: Computed tomography

MRI: Magnetic resonance imaging

NSAIDs: Nonsteroidal anti-inflammatory drugs

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