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Case Report

Preiser's disease in teenage female: A rare case report ☆

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ABSTRACT

Preiser's disease or idiopathic avascular necrosis of the scaphoid is a rare condition where ischemia and necrosis of the scaphoid bone occurs without previous fracture. It is thought to be caused by repetitive micro trauma or side effects of drugs (e.g., steroids or chemotherapy) in conjunction with existing defective vascular supply to the proximal pole of the scaphoid. Wrist radiography or CT coupled with MRI is the imaging modality of choice in the diagnosis of this rare entity.

Here, we report a case of Preiser's disease of the left wrist in a 17-year-old female patient who presented with left wrist pain of 2 years duration in the absence of trauma history or causative drug use. The diagnosis was made by wrist X-ray and MRI. She was managed by Physiotherapy and wrist immobilization using wrist and forearm support as well as NSAIDS (Meloxicam).

Introduction

Preiser's disease is a rare condition where ischemia and necrosis of the scaphoid bone occurs without previous fracture and Idiopathic avascular necrosis of the scaphoid is usually referred to as Preiser's disease, although other potential causes have also been taken into consideration. It is uncommon, and the detail of its natural history is not well known [1]. Although the pathophysiology is debatable, patients frequently report wrist pain and restricted wrist motion [2].

Diagnosis can be made with progressive radiographic and clinical sequence of osteosclerosis and subsequent scaphoid fragmentation, in the clinical setting of no previous fracture or trauma [3]. Treatment is NSAIDs and observation in minimally symptomatic patients. A variety of operative procedures are available depending on the severity of the disease and the patient's symptoms [1].

Abbreviations: NSAIDs, non-steroidal anti-inflammatory agents; MRI, magnetic resonance imaging; CT, Computed Tomography.

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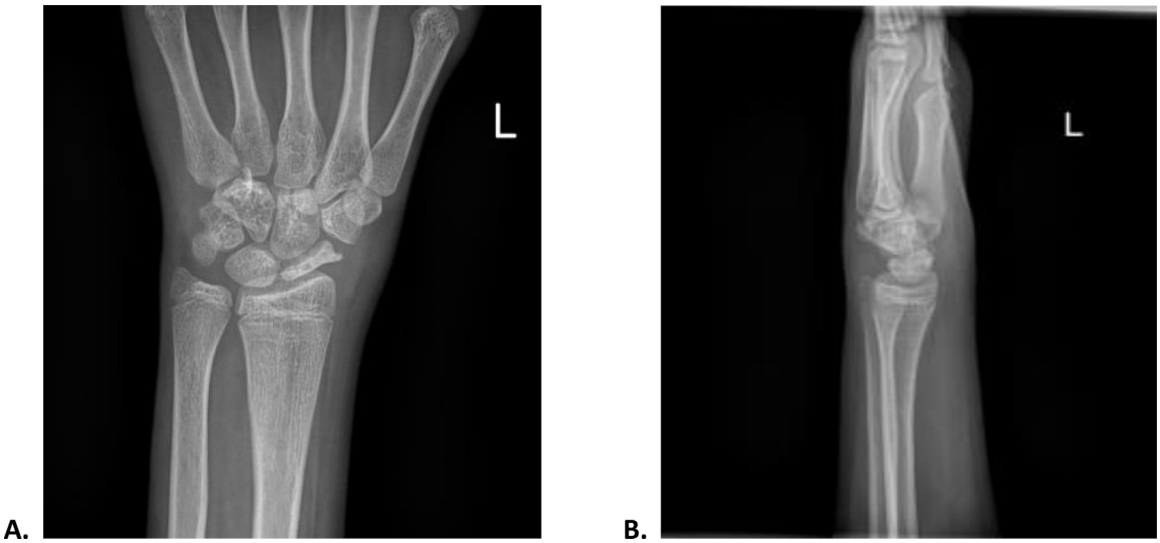

Fig. 1 – AP and lateral of the left wrist radiography showing volume loss/ collapse and sclerosis of the scaphoid bone.

Here, we report a case of Preiser's disease in a 17-year-old female patient diagnosed by imaging findings and clinical data.

A case history

A 17-year-old female patient presented with left wrist pain for more than 2 years duration. The patient described the pain as dull aching which worsens during wrist movements and when picking up objects using her left hand. Although the patient denies the use of steroids, she admits to using over-the-counter NSAIDs for pain relief. Otherwise, she had no chronic medical illness, no medication use, or trauma history.

On physical examination, she had left wrist swelling with the fullness of the anatomic snuff box. She also had limitation of active wrist range of motion (especially during radial deviation) due to pain, with full passive range of motion. Upon examination, Finkelstein's maneuver was negative. The Physical exam of the contralateral wrist was normal.

She was investigated for the above complaint with left wrist radiograph and MRI. The wrist radiograph showed collapsed and sclerosed left scaphoid bone with no evidence of scaphoid fracture (Fig. 1). The joint spaces were maintained and the rest of the osseous structures were normal. There was no soft tissue swelling. MRI showed a scaphoid bone with T1 and T2 low marrow signal intensity and marked height loss (Fig. 2). Based on the radiograph, MRI findings, and supporting clinical data the diagnosis of Preiser's disease was made.

After settling the diagnosis the orthopedic surgeon discussed the choice of management with the patient and her parent's. Surgical and nonsurgical options were offered and parents opted for nonoperative management with physiotherapy and wrist immobilization with wrist and forearm supports as well as NSAIDs (Meloxicam) were given.

Discussion

Preiser's disease, a rare disorder, causes ischemia and necrosis of the scaphoid bone when there hasn't previously been a fracture or nonunion of the scaphoid bone [3] and also in our case, the patient had no history of trauma or previously diagnosed scaphoid fracture. There are few reported cases of Preiser's disease in the literature review [4]. A recent systematic review of literature review identifies only 170 cases [1].

Preiser first characterized idiopathic avascular necrosis of the scaphoid in 1910 [5]. There is still significant debate about its pathogenesis, development, and management. The vascularity of the proximal two-thirds of the scaphoid appears to be in danger due to the reduction of dorsal blood flow, leading to osteonecrosis [4,5]. Osteonecrosis is also linked to long-term corticosteroid usage, chemotherapy, trauma, collagen disorders, and alcoholism. Women, between the ages of 20 and 70, are more likely to develop the condition, more in their dominant hand [6]. There is only 1 case described in children under the age of 10 [7]. Localized pain on the lateral side of the wrist (anatomical snuff box) is the main feature of the clinical picture. On palpation, the scaphoid has a hypersensitive area, and there may occasionally be slight swelling and a loss of strength. Only in severe disease does the amplitude of motion decrease [8]. Similarly, our patient also had pain in the lateral side of the wrist which worsens during wrist movements and had mild swelling and fullness in the anatomic snuff-box with decreased active wrist range of motion.

Diagnosis is usually made by radiography or coupled with CT and/or MRI. Radiographic Imaging evidences of scaphoid sclerosis, fragmentation, and collapse of the scaphoid and MRI imaging evidences more specifically showing partial or diffuse involvement of scaphoid bone are the imaging clues of Preiser's disease, in the absence of history of trauma . [9,10] The diagnosis of Preiser disease is made in patients with imaging evidence of scaphoid osteonecrosis in the setting of no history of trauma or scaphoid fracture. In our case, the pa-

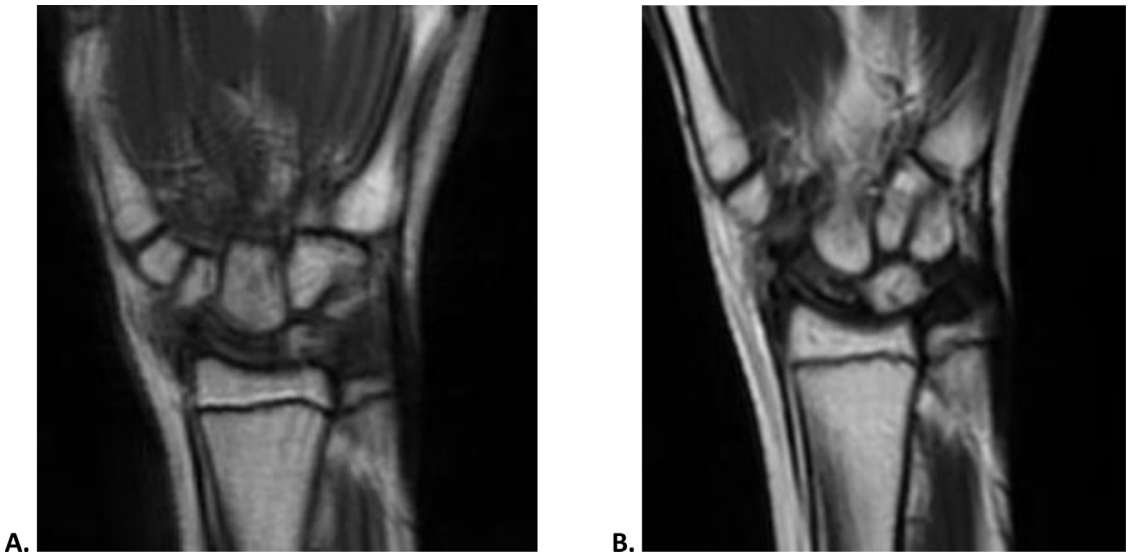

Fig. 2 – T1 and T2 coronal MRI showing T1 and T2 low marrow signal intensity with height loss of the scaphoid bone.

tient had no history of trauma, which makes the diagnosis of Preiser's disease likely with the typical imaging findings.

Herbert and Lanzetta proposed a radiographic staging system of scaphoid avascular necrosis, based on the progression of the disease:

Type I—normal radiographs and CT scans showing abnormal findings.
Type II—increased density of the proximal pole with generalized osteopenia.
Type III—fragmentation and collapse with or without pathologic fracture.
Type IV—fragmentation and collapse with osteoarthritis [10].

Our patient's radiograph showed collapse and sclerosis of the scaphoid which is Type III according to the Herbert and Lanzetta staging system.

Magnetic resonance imaging (MRI) helps in disease staging. Kalainov and his colleagues identified two types of avascular necrosis of the scaphoid as seen in MRI:

Type I—necrosis or diffuse ischemia.
Type II—partial necrosis [9].

So our patient MRI showed diffuse T1 and T2 low signal intensity and based on Kalainov staging our case is -Type-1.

Treatment includes both non-surgical and/or surgical management. Given, the rarity of Preiser's disease,the management option is not yet well defined .The treatment options depends on the progress of the disease which in turn is assessed by imaging [4]. The nonsurgical management includes use of nonsteroidal anti-inflammatory drugs, immobilization with casts and rest [11]. surgical management options include prosthesis replacement, carpectomy, carpal arthrodesis, radial osteotomy, and vascularized bone graft [8]. De Smet was the first to describe the scaphoid graft for revascularization, in 2000 [12]. Using a distal radius vascularized graft of dorsal retrograde flow, Moran and his colleagues reviewed and treated 8 cases of Preiser's disease and also sokolo and his colleagues treated 2 patients and it showed satisfactory results, with improved pain and range of motion of every patient in both studies [4,13].

So for our patient, both surgical and nonsurgical options were offered and parents opted for non-operative management with Physiotherapy and wrist immobilization using wrist and forearm support as well as NSAIDS (Meloxicam).

Conclusion

Preiser's disease is a rare condition and the natural history is not fully understood. Even though rare, it should be considered in the differential diagnosis of patients who present with localized wrist pain, in the appropriate clinical setup. The diagnosis involves both clinical and imaging studies. There is no consensus for the single best effective management but various surgical techniques and conservative management options are being tried.

Availability of data and materials

The data supporting the findings of the case are available upon request to the corresponding author.

Authors' contributions

All authors contributed to the conduct of this research and read and approved the final version of the manuscript.

Ethics approval

This study was approved by the standing research ethics committee of the Department of radiology, school of Medicine, Addis Ababa University.

Patient consent

Written informed consent was obtained from the patient's parents for publication and any accompanying images. A copy of written informed consent is available for review by the Editor–in–chief of this journal on request.

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