CASE REPORT

Bilateral idiopathic avascular necrosis of the scaphoid

A RARE CASE OF PREISER’S DISEASE

Whereas avascular necrosis of the scaphoid after a fracture is well-documented, idiopathic avascular necrosis of the scaphoid (Preiser’s disease) is rare. Little is known of the aetiology of the condition and even less about the best course of management. We describe a rare case of bilateral Preiser’s disease. Possible aetiological factors and a summary of the current concepts of management are discussed.

Avascular necrosis (AVN) of the scaphoid after fracture is well recognised, but idiopathic AVN (Preiser’s disease) is rare. We describe a case of bilateral AVN of the scaphoid.

Case report
A 52-year-old right-handed Caucasian woman presented in April 2008 with an eight-month history of pain in the right wrist.

There was no history of trauma. She had developed bilateral wrist pain after moving a number of suitcases. The pain, which was localised to the radial aspect of the wrists, worsened over the following months until she was unable to drive, prompting her to see a physiotherapist in January 2008. By March, the pain in her left wrist had resolved, but persisted in the right.

Her medical history revealed a recent diagnosis of scleroderma with no complications, and chronic asthma. She had no drug allergies, was a non-smoker and took only salbutamol and used fluticasone and salmeterol inhalers.

On examination there was generalised tenderness around the right wrist which, although allowing a full range of movement, was uncomfortable. Examination of the left wrist was unremarkable. Plain radiographs of the right wrist showed no abnormality, but MRI with gadolinium enhancement revealed a diffuse abnormality of the marrow suggestive of AVN (Fig. 1). There was no evidence of a previous fracture.

She was managed conservatively with physiotherapy and, over the next eight months, the pain in the right wrist improved, but failed to resolve. The symptoms in her left wrist also recurred spontaneously. Plain radiographs of the left wrist showed sclerosis and fragmentation of the proximal half of the scaphoid (Fig. 2). A repeat MR scan of the right wrist and a new scan of the left wrist suggested gradual revascularisation of the right scaphoid (Fig. 3) with AVN of the proximal pole, and evidence of revascularisation and mild collapse on the left (Fig. 4).

She currently has good function in both wrists with only mild discomfort. She continues to be managed conservatively.

Discussion
The blood supply to the scaphoid arises primarily from branches of the radial artery. Dorsal branches enter the distal aspect of the bone and run proximally. Volar and laterally-based vessels have also been described and

![Fig. 1](MR scan of the right wrist (coronal T1 sequence). There is low-signal intensity of the whole scaphoid.)
enter at the level of the scaphoid tubercle.\textsuperscript{1} Idiopathic AVN of the scaphoid (Preiser’s disease)\textsuperscript{2} is an uncommon condition resulting from a disruption of this blood supply.\textsuperscript{3} Interestingly, the five patients originally described by Preiser all had a history of fracture, yet the eponym persists for idiopathic cases.

Vidal et al\textsuperscript{3} proposed a classification which included factors such as trauma, systemic disease and congenital mechanisms as possible causes. Our patient fitted this classification because vasculopathy is a characteristic feature of scleroderma\textsuperscript{4} and unilateral AVN of the scaphoid in conjunction with scleroderma has been reported.\textsuperscript{5}

Risk factors for AVN of the scaphoid include the use of corticosteroids, chemotherapy, excessive alcohol intake and smoking.\textsuperscript{6,7} Also, although the importance of ulnar variance has been investigated with conflicting conclusions, it is generally believed that negative ulnar variance is not a risk factor for Preiser’s disease.\textsuperscript{7,8}

\begin{table}[h]
\centering
\caption{Classification of Preiser’s disease (Herbert and Lanzetta)\textsuperscript{9}}
\begin{tabular}{|c|l|}
\hline
Stage & Description                                                                 \\
\hline
1     & Normal radiographs               \\
      & MRI changes                      \\
      & Positive bone scan               \\
\hline
2     & Increased density of proximal pole \\
      & Generalised osteoporosis          \\
\hline
3     & Fragmentation of proximal pole +/- pathological fracture                   \\
\hline
4     & Carpal collapse, osteoarthritis                                           \\
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Fig. 2
Plain radiograph of the left wrist. There is fragmentation and sclerosis of the proximal pole of the scaphoid.

Fig. 3
MR scan of the right wrist (coronal T1 sequence). Low-signal intensity is limited to the proximal pole of the scaphoid.

Fig. 4
MR scan of the left wrist (coronal T1 sequence). There is low-signal intensity at the proximal pole of the scaphoid.
A radiological classification of Preiser's disease has been proposed by Herbert and Lanzetta\(^9\) (Table I). Our patient had radiological findings suggestive of stage-3 disease (Fig. 2). Recently, however, two forms of Preiser's disease have been proposed, based on MRI findings.\(^7\) Type 1 is a diffuse avascular necrosis with widespread MRI changes: type 2 is more localised and involves less than half of the scaphoid.\(^10\) It has been suggested that type 1 has a poorer prognosis because of collapse of the scaphoid and subsequent degenerative change.\(^3,7\)

The management of Preiser's disease remains problematic. There are no guidelines and, because of its rarity, there are few data available. Kalainov et al.,\(^7\) in a study of 19 patients, recommended that all cases should be investigated by plain radiography and MRI, in order to define the type. Since the carpal bones do not show the same subchondral pattern of AVN as larger bones, it is suggested that, in the absence of trauma, any areas of low-signal intensity on T1-weighted images should raise the suspicion of idiopathic AVN.\(^10\)

A variety of methods have been used to treat the condition but in insufficient numbers to recommend an optimal solution. Treatment is directed towards the causative factors and tailored to the stage of the disease. Conservative management with immobilisation,\(^3\) arthroscopic debridement\(^11\) and radial osteotomy\(^12\) have each been tried, with good effect. More recently, the use of vascularised bone grafts has been used to treat early disease.\(^5\) Harpf et al\(^13\) reported a series of 60 patients in whom free iliac bone grafts were used successfully. Similar results have been reported in the treatment of idiopathic AVN of the lunate when progression of the disease was halted in 89% of patients.\(^14\) It seems possible that such treatment for Preiser's disease would also be successful.

The use of local pedicled grafts for Preiser's disease has been studied by Moran et al.,\(^6\) who reviewed eight cases. They found that patients had less pain and greater movement of the wrist at a mean follow-up of three years. The future for this treatment of Preiser's disease may also, therefore, be promising.

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References