IDIOPATHIC AVASCULAR NECROSIS OF THE SCAPHOID

T. J. HERBERT and M. LANCHETTA

St Luke's Hospital Hand Unit, Sydney, Australia

Idiopathic avascular necrosis of the scaphoid is a rare condition. A review of the literature shows a variety of conditions labelled as spontaneous avascular necrosis or Preiser's disease. In this paper we report on a study of eight patients with idiopathic avascular necrosis affecting only the proximal pole of the scaphoid. Seven of these patients had positive ulnar variance. The possible aetiology is discussed and the natural history has been studied. A staging system is proposed, as this helps to determine the prognosis and appropriate management. Two of our patients were managed conservatively; the others were treated successfully by partial silastic replacement of the scaphoid.

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Avascular necrosis of the carpal bones can affect the lunate (Kienböck, 1910), the pisiform (Olah, 1968), the capitate (Urman et al, 1977; Rahme, 1983) and the scaphoid. When the whole scaphoid becomes avascular, following a fracture or an injury, this condition has been called Preiser's disease (Bray and McCarroll, 1984), after Preiser's original article in 1910. Although avascular necrosis may occur without any known history of injury (Ekerot and Eiken, 1981), it may be difficult to exclude the possibility of mild or repetitive trauma (Allen, 1983).

Other known causes of avascular necrosis include steroid therapy (Milgram and Riley, 1976), chemotherapy (Harper et al, 1984) and connective tissue diseases such as systemic lupus erythematosus (Urman et al, 1977) or progressive systemic sclerosis (Kawai and al, 1983). In the latter, high doses of steroids are generally used, so that it is difficult to establish whether the primary cause of the scaphoid necrosis is the disease itself or the steroid therapy.

The literature contains reference to 14 cases of idiopathic avascular necrosis of the scaphoid, (Guelpa et al, 1980; Ekerot and Eiken, 1981; Allen, 1983; Ferlic and Morin, 1989; Alnot et al, 1990; Table 1).

We present our experience with eight cases of idiopathic avascular necrosis of the scaphoid. In all cases only the proximal pole of the bone was affected (Table 2).

The history and physical signs were remarkably similar in all cases. All patients complained of increasingly severe radial-sided wrist pain of spontaneous onset, without any significant history of trauma. Physical examination showed marked synovial swelling and tenderness around the dorso-radial aspect of the wrist.

Range of motion and function were severely impaired because of pain, a condition we recognize as an "irritable wrist". X-ray findings varied, depending on the duration of the disease. Since this is a progressive disorder, we have attempted to classify our patients according to the X-ray appearance at the time of examination. The following staging system was used:

— Stage 1. Normal X-rays
  Positive bone scan
— Stage 2. Increased density of proximal pole
  Generalized osteoporosis
— Stage 3. Fragmentation of proximal pole
  + pathological fracture
— Stage 4. Carpal collapse pattern, osteoarthritis.

CASE REPORTS

Case 1
A healthy 46-year-old male company director (LA) presented with a 14-month history of pain in his dominant right wrist. There was no history of injury. He had tenderness and swelling over the anatomical snuff box and the dorsum of the right wrist. The range of motion was reduced by approximately 50%.

X-ray films showed complete avascular necrosis of the proximal half of the scaphoid with a pathological fracture and significant carpal collapse deformity (stage 4; Fig 1). He had been treated previously with a single cortisone injection, with significant pain relief. His symptoms were not considered severe enough to warrant surgery, and he was fitted with a wrist support and advised to undergo regular review. Over a period of 29 months the situation has remained unchanged.
Table 2

<table>
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<th>Case no.</th>
<th>Name</th>
<th>Sex</th>
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<th>Side</th>
<th>Previous treatment</th>
<th>Initial X-rays</th>
<th>Stage</th>
<th>Ulnar variance</th>
<th>Treatment</th>
<th>Op. findings</th>
<th>Last X-rays</th>
<th>Follow-up (mths)</th>
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<td>Steroid injection</td>
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<td>+</td>
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<td>—</td>
<td>Unchanged</td>
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<td>M</td>
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<td>AVN PP</td>
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<td>+</td>
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<td>AVN PP Path #</td>
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D: dominant; ND: non-dominant; MUA: manipulation under anaesthetic; AVN PP: avascular necrosis of the proximal pole; Path #: pathological fracture; CCD: carpal collapse deformity; RC OA: Radio-carpal osteoarthritis.

Case 2

A fit 36-year-old female cook (JM) presented with a 5-month history of pain in her dominant left wrist. No history of trauma could be elicited. On examination, swelling was noted in the anatomical snuff box and the dorsum of the wrist at the scapho-lunate junction. The wrist joint was "irritable". Initially, X-ray films appeared normal, although a bone scan showed intense reaction adjacent to the scaphoid (stage 1; Figs 2a and b). Positive ulnar variance was noted. X-rays taken 2 months later showed the development of changes consistent with a diagnosis of avascular necrosis of the proximal pole of the scaphoid (stage 2; Figs 2c and 3a).

Arthroscopy showed marked synovitis on the radial side of the wrist, with an intact scapho-lunate ligament. The articular cartilage over the scaphoid was intact although soft to probing over the proximal pole.

Plans were made to admit the patient to hospital for a "revascularization" procedure, but these had to be cancelled for social reasons. Her wrist remained painful, and 6 months later X-rays showed increased collapse of the proximal pole of the scaphoid, with the development of a fracture line at the junction between healthy and unhealthy bone (stage 3; Fig 3b). By the time she was admitted to hospital for treatment (5 months later), X-rays showed a clear fracture line and complete avascular necrosis of the proximal pole (Fig 3c).

At operation, the articular cartilage over the proximal pole was soft and unhealthy, but there was no external sign of fracture or of scapho-lunate ligament damage. An osteotomy was carried out through the waist of the scaphoid, the proximal fragment was excised and replaced with a stabilized silicone prosthesis (Fig 3d). The patient's symptoms were relieved although she did require treatment for an associated de Quervain's syndrome before she was able to return to work. Histological examination of the excised bone confirmed a diagnosis of avascular necrosis with a pathological fracture through the trabeculae at the junction between healthy and unhealthy bone.

Case 3

A healthy 39-year-old right handed book-keeper (FS) presented with chronic right wrist pain, which was preventing her from performing simple daily activities,
including driving. The patient was unable to recall any history of injury. 9 years previously she had undergone surgical release of the first extensor compartment for a possible de Quervain's syndrome, without relief of the symptoms.

On examination she had a very irritable wrist, with pain and swelling around the scaphoid. There were signs of mild median nerve compression at the wrist, but no thenar muscle wasting. X-ray films showed complete avascular necrosis of the proximal pole of the scaphoid (stage 3; Fig 4). The proximal pole of the scaphoid was excised and a partial silicone replacement performed; intra-operatively there was no sign of fracture or scapholunate ligament tear. 32 months after operation the patient showed good function of the hand, with excellent range of motion and grip strength, and no pain. X-rays showed good function of the prosthesis, with no signs of silicone synovitis. However, early radio-carpal osteoarthritis was noted, suggesting some increase in the carpal collapse deformity.

Case 2

A healthy 39-year-old right-handed storeman (PS) presented with a 10-year history of right wrist pain. 9 years previously he had been diagnosed elsewhere as having idiopathic avascular necrosis of the proximal pole of the right scaphoid. This was treated surgically by a Russe type bone graft to the scaphoid, with reasonable symptomatic relief. However, there was no improvement in the
SCAPHOID AVASCULAR NECROSIS

Fig 3 (a–d) For legend please see page 178.
Case 5
A healthy 55-year-old housewife (GC) presented with a 7-month history of pain in her dominant right wrist. There was no specific history of trauma to the wrist, although she did recall having sustained a severe electric shock to the arm a few days before the onset of her symptoms. She had undergone manipulation of the wrist under anaesthesia with an intra-articular steroid injection with no relief before presenting to us.

On examination she had a very painful wrist suggesting synovitis of the radio-carpal joint; the range of motion was severely restricted by pain and her grip strength was markedly reduced. X-rays showed sclerosis and slight narrowing of the proximal pole of the scaphoid, consistent with avascular necrosis; there was no evidence of fracture (stage 2). Bone scan showed markedly increased uptake around the scaphoid. Positive ulnar variance was noted.

At operation, the avascular proximal scaphoid was excised and partial silicone replacement performed. The scapho-lunate ligament was oedematous and loose, and there appeared to be a small avulsion fracture at its insertion into the proximal pole of the scaphoid. 7 years after operation, the patient is happy with the result, although she still has mild discomfort and weakness.

Case 6
A healthy 60-year-old secretary (JR) presented with a history of chronic pain in her non-dominant left wrist. Although there was no history of acute trauma, she recalled that the onset of pain and swelling in the wrist followed an intense session of typing. On examination, she was found to have marked swelling over the tendon of abductor pollicis longus at the base of the thumb. Tenderness in the anatomical snuff box and over the dorsum of the wrist, which was irritable. X-ray films showed avascular necrosis of the proximal pole of the scaphoid (stage 2) and marked positive ulnar variance (Fig 5a). The patient was fitted with a wrist support and advised to rest the wrist. Her symptoms have remained controlled with conservative treatment. Follow-up X-ray films show the development of a pathological fracture through the proximal pole of the scaphoid (Stage 3; Fig 5b). She remains under review and shows no further progression of the disease.

Case 7
A fit 66-year-old doctor (IR) presented with a 6-month history of increasing pain and weakness in his dominant
right wrist, preventing him from lifting heavy objects or playing croquet. There was no history of significant injury.

On examination, he had swelling and tenderness over the anatomical snuff box and the dorsum of the scaphoid. X-ray films showed changes consistent with avascular necrosis of the proximal pole of the scaphoid (stage 2; Fig 6). The patient underwent a period of conservative treatment of splinting and rest, but this failed to relieve his symptoms. At surgery, the proximal pole of the scaphoid was found to be completely avascular and necrotic; a silicone prosthesis was used to replace it. The removed bone was sent for histological examination, which confirmed the diagnosis of avascular necrosis. He reports marked improvement following surgery and has been able to resume playing croquet.

Case 8

A 51-year-old shop assistant (BW) presented with a 4-month history of pain in her dominant right wrist following a bump on the radial side of the wrist. She had signs of a mild de Quervain's syndrome treated conservatively. 3 years earlier she had received two steroid injections for a painful sternoclavicular joint. Clinically, she had classical signs of avascular necrosis of the scaphoid with an "irritable wrist", and she had signs of mild carpal tunnel syndrome. X-rays showed avascular necrosis of the scaphoid with fragmentation of the proximal pole and signs of a pathological fracture (stage 3). Positive ulnar variance was noted. Surgical treatment was carried out, with replacement of the proximal pole with a silicone implant. 6 months post-operatively the patient's wrist was stable and she had regained virtually full range of motion. She reported excellent pain relief to the extent that she had been able to resume normal work and household duties.

DISCUSSION

The aetiology and pathogenesis of avascular necrosis of the carpal bones remains uncertain. A carpal bone, or a fragment thereof, may be completely devascularized (i.e. completely stripped of all its soft tissue attachments)
Fig 6  Case 7. Early avascular necrosis of the proximal pole (Stage 2).

...and survive if it is repositioned or if the fragment is reattached, yet avascular necrosis may occur following simple fractures of the scaphoid, lunate and capitate. Idiopathic avascular necrosis, by definition, occurs in the absence of major trauma or pre-existing fracture, although a patient may have a scaphoid fracture without being aware of it, and the fracture may not always be obvious on X-ray. Our review of the literature suggests that a number of cases labelled as idiopathic avascular necrosis of the scaphoid may well have been due to a pre-existing undiagnosed fracture. We have reviewed carefully all available X-ray films in the patients reported in this series, and are confident that in none of these was there a pre-existing fracture.

Gelberman and Menon (1980) have shown that the scaphoid receives its blood supply through the areas of soft tissue attachment. The dorsal vasculature enters through numerous small foramina along the spiral groove and dorsal ridge. These feeding vessels arise from the dorsal scaphoid branch of the radial artery and from the dorsal radial carpal arch and account for approximately 80% of the total blood supply to the scaphoid; the remaining 20% is supplied by palmar vessels entering the tubercle and distal pole. These authors failed to demonstrate any intraosseous connection between these two areas of blood supply and were unable to show any significant blood supply entering the proximal pole through the area of attachment of the scapho-lunate ligament.

However, we have seen numerous cases in which a small proximal pole fragment remains viable when the scapho-lunate ligament must be its main source of blood supply (Herbert, 1990). Similarly, we have seen patients in whom avascular necrosis of the proximal pole of the scaphoid has occurred after traumatic avulsion injuries of the scapho-lunate ligament (Herbert, 1990). It seems, therefore, that in a certain percentage of cases, the proximal pole must receive a blood supply through the attachments of the scapho-lunate ligament. If this is the case, any interference with this blood supply could lead to the development of segmental avascular necrosis of the proximal pole of the scaphoid. This situation is, in many ways, analogous to the hip joint, where damage to the ligamentum teres may lead to the development of segmental avascular necrosis of the femoral head in susceptible patients (Kamegaya et al, 1989).

Thus certain patients may be unusually susceptible to mild or repetitive trauma which may be sufficient to disrupt the blood supply entering the proximal pole by way of the scapho-lunate ligament. This theory is supported by the findings of Guelpa et al (1980) and Ferlic and Morin (1989), who each reported a case of bilateral idiopathic avascular necrosis of the scaphoid. Whilst we have yet to encounter this problem, we do have a number of patients with bilateral avascular necrosis associated with proximal pole fractures following relatively minor trauma, highly suggestive of an underlying predisposing factor. Case 5 in this series would further appear to support this hypothesis: at operation, damage to the scapho-lunate ligament was apparent, which could well have occurred as a result of the forces associated with an electric shock.

It is well-known that repetitive stress on the wrist can cause damage to the scapho-lunate ligament and this could be sufficient to interfere with the blood supply to the proximal pole of the scaphoid in susceptible patients. In seven of our eight patients, it was the dominant wrist that was affected. It is interesting to speculate whether manipulation and/or steroid injection of the wrist (Cases 1 and 5) could have affected the outcome in any way.

The reason for the preponderance of females in this series (75%) compared with scaphoid fracture (6%) is uncertain. In all except one of our cases, plain X-rays suggested a positive ulnar variance, although in none of these were specific views for ulnar variance taken, so that some allowance should be made for radiological error. Nevertheless in case 6 the positive variance was striking, and it is difficult not to conclude there may be some correlation between ulnar variance and differential loading in the radio-carpal joint. This certainly warrants further investigation.

Clinically, the onset of idiopathic avascular necrosis of the proximal pole of the scaphoid is heralded by increasing pain and stiffness of the wrist. Examination shows signs of an “irritable” wrist joint with tenderness...
and swelling over the dorsum of the wrist due to chronic synovitis. Histological examination of the synovium has shown chronic inflammatory changes only, with no diagnostic features.

It is hardly surprising that two of our patients also had signs of carpal tunnel syndrome, which resolved once the synovitis had settled. Cases 2 and 6 appear also to have had a related De Quervain's syndrome which settled following treatment.

In the early stages radiological examination may be normal, though bone scan is likely to show intense activity around the scaphoid (Stage 1). Before long, osteoporosis occurs, sparing the ischaemic proximal pole which thus appears relatively dense on X-ray (Stage 2). The dead bone is later replaced by scar tissue and the normal trabecular bone structure disappears, producing a "ground-glass" appearance. The bone tends to collapse and deform, and cystic changes and areas of sclerosis are commonly seen. In the later stages, pathological fracture may occur at the junction of healthy bone with the avascular fragment (cases 2, 6 and 8; Stage 3). As the scaphoid deforms, progressive carpal collapse occurs leading to the development of radio-carpal osteoarthritis (Stage 4). Thus, as with Kienböck's disease, idiopathic avascular necrosis of the scaphoid is a progressive condition so that treatment and prognosis depend on the stage of the disease.

In the early stages one should consider the possibility of reversing the pathological process. In our only Stage 1 patient, we had planned to carry out a vascularized bone graft; unfortunately, by the time she came to surgery she had progressed to Stage 3 and required a salvage procedure.

Similarly, in Stages 1 and 2 it is tempting to consider "unloading" the scaphoid in some way (e.g. radial osteotomy) in order to prevent progressive collapse deformity. If positive ulnar variance could be proved to be of significance, then presumably ulnar shortening would be indicated in selected cases. Once the bone has undergone complete avascular necrosis (Stage 3), these changes are almost certainly irreversible so that reconstruction is no longer indicated.

If the symptoms are not severe enough to justify a surgical procedure, the patient may benefit from conservative measures, as in cases 1 and 6. When surgery is indicated, our preferred treatment to date has been a partial silastic replacement of the scaphoid combined with a local synoveotomy. A percutaneous osteotomy is carried out through the waist of the scaphoid and the entire proximal fragment is removed in one piece; a Swanston silastic scaphoid prosthesis is then cut down in such a way that it has a square stem distally, which can be locked into a suitably prepared "peg hole" in the distal fragment. The implant is thus stabilized and provides an effective spacer which appears to prevent further carpal collapse. We always choose the smallest possible prosthesis (size 0 or 1) as this prevents overloading the implant and therefore reduces wear problems to the minimum. Using the technique we have described previously (Herbert, 1990), this procedure has provided lasting pain relief with improved range of motion and wrist function. To date, none of these patients has developed any signs of silicone synovitis.

The use of a silicone prosthesis does not preclude the possibility of carrying out partial or total fusion at a later date, should wear become a problem. Similarly, if scaphoid shortening results in symptomatic radio-carpal impingement, the symptoms may be relieved by carrying out a limited radial styloectomy.

In conclusion, we consider that idiopathic avascular necrosis of the scaphoid is a distinct entity, similar in many ways to Kienböck's disease of the lunate. It is incorrect to call it Preiser's disease since the condition that Preiser described occurred following definite trauma, and was almost certainly avascular necrosis due to fracture. Whilst most previous reports in the literature refer to idiopathic avascular necrosis affecting the whole of the scaphoid, we believe that the problem always starts in the proximal pole of the bone.

As the disease progresses, the scaphoid collapses to the extent that the distal pole becomes deformed and arthritic, mimicking the appearance of avascular necrosis. The prognosis would appear to depend on the stage of the disease, and treatment should be planned accordingly.

It appears that the aetiology may be related to interference in the extrinsic blood supply to the proximal pole of the scaphoid in susceptible patients, and it appears that positive ulnar variance may be significant. These hypotheses warrant further investigation.

References


