Idiopathic avascular necrosis of the scaphoid and lunate

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Abstract

Idiopathic avascular necrosis (AVN) of the carpal bones of the wrist is rare. The lunate and the scaphoid are the two most commonly affected, the lunate being the more common of the two. AVN of a fractured proximal pole of the scaphoid is a well-known problem following trauma; however idiopathic AVN of the scaphoid not related to acute trauma is a rare event with few cases described in literature.

There is no consensus on the optimal surgical management of these conditions, although treatment is largely based on the stage at presentation and the expertise and repertoire of the surgeon. In this paper, we present a review of the symptoms, classifications, risk factors and management of these rare conditions.

Keywords avascular necrosis wrist; idiopathic avascular necrosis; Kienböck's disease; osteonecrosis; Preiser's disease

Avascular necrosis of scaphoid

The scaphoid is the most frequently fractured carpal bone and, due to its precarious blood supply, avascular necrosis (AVN) of the proximal pole is not uncommon after such fractures. Spontaneous or idiopathic AVN of the scaphoid, also known as Preiser's disease, is however very rare.[1]

The original cases of AVN of the scaphoid presented by Preiser in 1910 were related to fractures.[2] The modern understanding of the disease, however, is idiopathic AVN of the scaphoid in the absence of any fracture. The aetiology remains unknown but several risk factors have been identified in the literature. These include trauma, steroid use, systemic vascular disorders and hypoplasia of the scaphoid.[3,4] A number of case reports have also described an association with chemotherapy in the absence of concomitant steroid therapy.[5,6]

Preiser's original paper compared the disease with Kienböck's and he believed the aetiologies were similar. Some authors have suggested an association with positive ulna variance in the disease's aetiology.[7] Others, however, have described a similar incidence of the disease in patients with positive, negative and neutral ulna variance,[8,9] which suggests that the role of ulnar variance is less than certain. Most patients present with insidious pain and little or no history of trauma. There is no known preponderance for gender, handedness or chronological age, although most cases reported in the literature are described in adults aged 20–70 years[8] and only few reports of the condition presenting in children.[10,11] Clinical investigations, as with other conditions in orthopaedics, start with history, examination and imaging. Radiographs and MRI scans form the basis of diagnosis. Herbert and Lanzetta[7] proposed a staging system based on radiological and MRI findings to help determine prognosis and management (Table 1). The disease is also classified based on the location of the AVN. The two types described based on MRI findings are type 1, in which the signal change of necrosis affects the entire scaphoid (100%) and type 2, where there is only partial (range 33–66%) involvement of the scaphoid, found predominantly in the proximal pole. Patients who present with type 1 changes are often more refractory to treatment, whilst type 2 patients tend to have a more favourable outcome.[8] The natural progression of the disease is from scaphoid fragmentation and collapse onwards to local or pancarpal arthritis (Table 1).

Due to the rarity of the condition and a lack of prospective studies there is no consensus on the ideal treatment for idiopathic AVN of the scaphoid. Conservative treatment is often limited to symptomatic treatment. Temporary immobilization of the wrist has some success, particularly in type 2 AVN of the scaphoid. Most authors recommend exhausting conservative measures at first, as the results of surgery are variable and sometimes unpredictable.

Surgical treatment can be broadly categorized into scaphoid-sparing procedures and scaphoid-sacrificing procedures.

Surgical treatment

Scaphoid-sparing procedures

A number of procedures have been described in the quest to manage patient's symptoms and preserve the scaphoid. These include partial debridement of the necrosed scaphoid, closing wedge radial osteotomy and revascularization procedures.

Partial debridement: arthroscopic partial debridement of the scaphoid has been used in a limited number of cases with reports of good symptomatic relief in the short to medium term. These are often, however, single patient reports.[12] Some authors have combined partial debridement with partial replacement of the resected, necrosed bone with pyrocarbon implants with short-term improvement in pain but persistent stiffness.[13,14] The success of debridement alone, or indeed any scaphoid-preserving technique, may be limited by chronic degenerative changes in the radioscaphoid joint.

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Staging of Preiser's disease

Stage	Radiological findings
1	Normal X-ray. Abnormal findings on MRI. Positive bone scan
2	Proximal pole sclerosis of the scaphoid. Generalized osteoporosis
3	Fragmentation of the proximal scaphoid pole with/without pathological fracture
4	Periscaphoid collapse, fragmentation and osteoarthritis

Table 1

Closing wedge procedures: this is not commonly performed for Presier's disease. Nine cases were reported in the literature by the same author over a 5-year period. Two of the cases had concomitant Kienböck's disease. A closing wedge osteotomy of the distal radius was performed to decompress the radioscaphoid joint in an attempt at offloading the radius. Results are variable, with moderate improvement of pain in the short term.[15,16]

Revascularization: this is the most commonly performed procedure for idiopathic AVN of the scaphoid.[8] The results are variable and in some cases revascularization has been incomplete. Better outcomes are reported for early stage (stage 2) of the disease but the disease may still progress following revascularization surgery.[17] There have, however, been cases reported where radiological progression of the disease has been halted.[8] There is no standard technique described for revascularization procedures for Preiser's disease and most techniques are based on experience from the treatment of fracture-related AVN of the scaphoid. Options include vascularized bone graft from the distal radius,[18] free vascularized iliac crest bone graft[19] and pronator quadratus pedicle bone graft.[20] Some authors have combined revascularization procedures with joint distraction of the radiocarpal joint in order to offload the scaphoid, with inconclusive results.[18] Although commonly performed, revascularization procedures may be technically challenging and in some cases impossible due to fragmentation or to finding poor cortical bone on which to incorporate the graft. Therefore the patient should also be consented for salvage procedures such as excision and arthrodesis.

Scaphoid-sacrificing procedures

These include partial excision with replacement, total replacement, complete excision with arthrodesis and proximal row carpectomy. These procedures are more commonly performed in moderate to severe disease stages.

Total replacement: the use of silicone implants to replace the scaphoid has had a varying degree of success. Historically silicone has been shown to produce an inflammatory response coupled with the generation of particulate debris when used in arthroplasty. The limited number of cases available makes it difficult to determine whether it remains a viable treatment option. One case report with a follow-up of 8 years reported persistent symptomatic relief at final follow-up,[21] whilst a case series of total replacement in three patients reported a poor outcome with complications including subluxation of the implant. The authors concluded by suggesting a conservative approach based on their experience[22]

A potential alternative is a titanium scaphoid implant developed by Swanson in 1989. Its use for AVN related to fractures has been described[23] but there are currently no papers evaluating its use for Preiser's disease.

Excision and arthrodesis: excision of the scaphoid combined with capitolunate arthrodesis or four-corner fusion is performed for the later stages of Preiser's disease (stages 3 or 4). The results of four-corner fusion for scaphoid non-union and scapholunate advanced collapse have been described with favourable outcomes in the literature. But there have been fewer cases reported for Preiser's disease, although both patient groups present with similar symptoms of pain and/or deformity of the wrist. Parlini et al. described a technique of pancarpal arthrodesis in 15 patients, one of whom had stage 4 Preiser's disease. All patients were reported to have union of their arthrodesis and significant improvement of symptoms.[24] Several techniques for wrist arthrodesis have been described, including those with or without the use of plates.[24,25] When combined with autologous graft, the fusion rates are high.[25]

Proximal row carpectomy: the use of proximal row carpectomy has been well documented in the literature for several wrist conditions[26] but is generally avoided in younger patients who carry out manual or heavy work. It has been shown to produce a marked reduction in pain in Preiser's disease. For some authors this is the optimal form of treatment.[9]

Kienböck disease

Although the disease is associated with Robert Kienböck, a radiologist, who described the characteristic X-ray changes, AVN of the lunate was first described by Peste, who in 1843 noticed collapse of the lunate in some cadavers. Patients may present with pain, stiffness or weakness. The exact aetiology is poorly understood but several contributing factors have been described including mechanical and vascular factors.

Mechanical factors

Negative ulna variance is associated with increased loads and elevated intra-osseous pressure in the lunate. This increase in pressure is believed to be a contributing factor to developing Kienböck's disease. As the disease progresses there is marked negative ulnar variance, which is thought to contribute to further collapse of the lunate.[27] The role of negative ulnar variance has, however, been described as inconclusive by some authors. Van Leewen et al. compared the ulnar variance of 166 patients with Kienbock's disease with a similarly matched control group to evaluate the diagnostic value of ulnar variance in both groups. They found a high incidence of negative ulnar variance in both groups and concluded that the prevalence of the disease in those with neutral or positive variance makes the role of negative variance inconclusive.[28] It has also been noted that if ulnar variance played a predominant role in the diseases aetiology, it would be far more common, particularly in patients who undergo

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