ORIGINAL ARTICLE

Idiopathic avascular necrosis of the scaphoid: 10 new cases and a review of the literature. Indications for Preiser’s disease

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Summary

Introduction: Avascular necrosis of the scaphoid (Preiser’s disease) is a rare entity for which there are no treatment guidelines to date. The aim of this study was to delineate the optimal treatment at different stages of this disease, based on an analysis of the cases treated in our department.

Materials and methods: Ten wrists (nine patients) were retrospectively reviewed after a mean 92 months of follow-up. The initial diagnosis was stage II avascular necrosis in four cases, stage III in four cases and stage IV in two cases. These patients were treated by conservative treatment (non surgical) in three cases, vascularized bone graft in two and palliative treatments (proximal row carpectomy or partial fusion) in five. Radiological examination, sometimes completed by MRI, and functional assessment of range of motion, grip strength and Mayo Wrist Score were performed.

Results: Completely different results were obtained in the two stage II cases treated by conservative treatment, while functional results improved in the two cases treated by vascularized bone graft with regression of necrosis on MRI. Pain improved following palliative treatment in 4/5 stage III and IV wrists. Functional results were satisfactory with conservative treatment in one stage IV case for 13 years but worsened at the final follow-up assessment.

Discussion: A review of the literature, including 126 cases in 29 articles clarified the role of conservative treatment, vascularized bone grafts and proximal row carpectomy in the treatment of avascular necrosis of the scaphoid. Conservative treatment is ineffective in the early stages and newly always results in disease progression. In contrast vascularized bone grafts can stop or even reverse damage at stage II. Palliative treatment is indicated when facing irreversible lesions.

Level of evidence: Level IV — Retrospective study.

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Introduction

Idiopathic avascular necrosis of the scaphoid was first described in 1910 by Preiser [1]. This disease develops insidiously beginning with pain and sometimes swelling around the anatomical snuffbox, which may be associated with loss of strength and reduced range of motion in the wrist. The natural history of this disease frequently involves progression to carpal collapse and osteoarthritis after fragmentation or pathological fracture of the scaphoid [2]. Thus certain authors prefer to treat this entity surgically in the early stages to stop disease progression. Based on the many indications for conservative treatment, others, who hope for spontaneous recovery of the associated vascular anomalies, consider surgery to be too invasive if imaging results remain satisfactory. In later stages of the disease, surgical treatment is a more unanimously accepted option because of the development of what are considered to be irreversible lesions. At present, there is no consensus on a treatment strategy based on the generally accepted classification of Herbert and Lanzetta [2] and it is difficult to identify reliable therapeutic indications based on disease progression. We reviewed 10 wrists with avascular necrosis of the scaphoid and compared our results with a review of the literature to identify treatment strategies that stabilized disease progression in the early stages (stages I and II) or provided optimal functional benefit in the later stages of the disease (stages III or IV).

Materials and methods

Inclusion and exclusion criteria

Patients presenting with pain in the radial side of the wrist associated with avascular necrosis of the scaphoid were included in the study. The diagnosis of avascular necrosis of the scaphoid was usually based on hypoperfusion of all or part of the scaphoid after gadolinium enhanced nuclear magnetic resonance (NMR) imaging. In three patients who presented with an indication for palliative treatment (proximal row carpectomy or partial fusion with excision of the scaphoid), the suspected diagnosis on X-ray was later confirmed by histological examination of the necrotic scaphoid.

Patients presenting with avascular necrosis of the scaphoid secondary to fracture were excluded.

Characteristics of the study population

Nine patients, or 10 wrists with avascular necrosis of the scaphoid were followed in our unit. They were retrospectively reviewed and evaluated by an observer who was independent from our surgeons.

The nine patients included six men and three women. The mean age at the first consultation was 45 years old (24–68). The reason for consulting was pain in the radial side of the wrist in all patients associated with swelling in two cases. Pain was secondary to trauma in two cases (fall from a motorcycle and while playing rugby). The other etiological factors included long-term cortisosteroid treatment in three patients. One of these patients had also received chemotherapy for a breast tumor. This patient presented with avascular necrosis in both scaphoids (patient 6).

Except for this one case of bilateral necrosis, six patients were affected on the dominant side and two on the non-dominant side.

Imaging tests included initial and follow-up X-ray associated with NMR imaging in certain cases. Six cases of necrosis were found in the proximal pole, and four cases of diffuse necrosis of the scaphoid were identified. These imaging results also made it possible to classify cases into the four stages of the Herbert and Lanzetta classification system [2]. There were no cases of stage I necrosis in our series (normal X-ray and abnormal signal of the scaphoid on another imaging test). On the other hand, there were four stage II wrists (increased scaphoid density), four stage III (fragmentation and/or pathological fracture of the scaphoid) and two stage IV (collapse with or without osteoarthritis) wrists. Finally AP X-rays showed six wrists with negative ulnar variance, two with positive ulnar variance and two with neutral variance.

Treatment strategies

Three of the 10 cases received conservative treatment, which included occasionally wearing a flexible brace, local (cortisone) injections or oral analgesics without surgery. Two cases with stage II lesions, and in one case with stage IV lesions received conservative treatment.

Two vascularized grafts harvested from the volar aspect of the distal radius were performed for stage II disease. This was a vascularized bone graft on a pronator quadratus pedicle in the first case [3] and a vascularized bone graft pedicled on the volar carpal artery according to the Kuhlmann technique for the second [4,5]. The graft was attached with two pins after curettage of the necrotic area in both cases. Treatment was completed by immobilization with a volar splint that stabilized the base of the thumb for 5 weeks.

Proximal row carpectomies were performed in three cases of stage III disease.

Capitolunate arthrodesis and four-corner fusion both associated with a scaphoidectomy were performed in one case of stage III and stage IV disease, respectively.

Histological examination confirmed the diagnosis of avascular necrosis in all cases that underwent surgery and required scaphoid excision.

Study method

Following initial treatment, functional assessment included an evaluation of pain on a scale from 0–10 and calculation of the Mayo Wrist Score. Results were considered excellent if the score was 90 points or above, good between 80–89 points, average between 65–79 points and poor if it was less than 65 points. Patients evaluated their level of satisfaction as not satisfied, somewhat satisfied, satisfied and very satisfied.

Clinically the three axes of wrist range of motion were evaluated by goniometry, and grip strength by Jamar dynamometry. This was then compared to the value in the contralateral side and expressed as a percentage of the latter.
When the scaphoid was preserved a radiographic assessment was performed to determine disease progression. In the other cases the quality of palliative treatment was assessed.

**Results**

The final follow-up was performed after a mean 92 months (6–156 months).

Results are summarized in Table 1.

**Stage II necrosis**

Four wrists with stage II necrosis were treated, two by conservative treatment and two with a vascularized bone graft. At 6 years of follow-up one of the patients who received conservative treatment presented with a flexion-extension arc of 145° and a grip strength of 81% with no pain. Stage II lesions had progressed to stage IV after 34 months of follow-up in another patient who presented with stage IV lesions in the contralateral wrist (patient 6). The flexion-extension arc was limited to 35° and associated with significant pain. Range of motion was normal in the two vascularized bone grafts after 11 years of follow-up and grip strength was comparable to the contralateral side. Only the first patient still had exercise-induced pain. Results of a follow-up MRI confirmed revascularization of the scaphoid.

**Stage III necroses**

Three proximal row carpectomies were performed when scaphoid fracture or fragmentation developed. All patients reported reduced or resolved pain, despite reduced range of motion and grip strength. Only one patient was dissatisfied with the results because of a septic complication requiring surgical lavage. At this same stage, capitulunate arthrodesis in a fourth patient resulted in a flexion-extension arc of 90° and a grip strength of 80% of the contralateral side, without pain.

**Stage IV necrosis**

Three wrists with Stage IV necrosis were treated in two patients. One patient (patient 6) reported occasional but mild symptoms for 13 years in her right wrist, although initial X-rays showed scaphoid collapse. Pain increased at the final consultation (Visual Analog Scale [VAS] = 4/10) and this patient requested surgical treatment. After 7 years of follow-up in this wrist, the development of pain in the contralateral left side was diagnosed as stage II Preiser’s disease. Proximal row carpectomy was performed when the disease progressed to stage IV after 34 months of follow-up and was associated with severe pain. Three years later the patient’s flexion-extension arc had increased from 35 to 95° with no pain and the Mayo Wrist Score was excellent. Finally, the third wrist presented with stage IV lesions and was treated with four-corner fusion. At 14 months of follow-up, the results were poor in this patient with a pain score of 5/10.
Review of the literature and discussion

How much hindsight do we have on avascular necrosis of the scaphoid?

The very low incidence of Preiser’s disease makes it impossible to perform a robust prospective study. For the moment, existing epidemiological, therapeutic and prognostic notions are based on retrospective series with a limited number of patients or case reports. Thus, a meta-analysis would probably help validate certain data. Nevertheless, missing data and differences in the presentation of results make it impossible to harmonize the results of these different articles.

Thus, we performed an in-depth review of the literature to determine if the results in our series could be extrapolated to a larger number of patients.

We found 29 articles published between 1980 and 2011 reporting patients with Preiser’s disease, including 21 case reports [2,6–25] and eight original studies [26–32]. Only five series included 10 cases or more. A total of 126 cases were reviewed in addition to the 10 cases presented in this study.

Epidemiological factors

The gender of the patient was identified in 132/136 diseased wrists and included 77 women and 55 men. The mean age of 113 patients was 42 (9–76). There does not seem to be a peak age for the disease, except that it was rare before the age of 20 (four patients) and after 70 (four patients).

When dominance and the side of the diseased wrist were reported, the dominant side was affected two times more frequently (n = 39) than the non-dominant side (n = 20). This could be due either to a microtraumatic origin to the disease or more severe functional difficulty when the dominant limb was affected, causing the patient to consult.

Symptoms seemed to develop slowly because the delay between the first symptoms and the first consultation, which was mentioned in 60 patients, was 31 months (0–276). The reason for consulting, except in one patient who consulted for an atypical carpal tunnel syndrome, was always pain (67 cases) sometimes associated with swelling (26 cases), a loss of range of motion (23 cases) or grip strength (12 cases).

Long-term corticosteroid treatment (reported in 20 cases) and a history of trauma (18 cases) seemed to play a role in the pathogenesis of avascular necrosis of the scaphoid. The mechanism and intensity of the initial trauma was rarely mentioned in the latter cases. Moreover, considering the very high incidence of benign wrist traumas, the reality of a cause and effect relationship between trauma and avascular necrosis remains difficult to establish.

A microtraumatic origin is still highly probable as suggested in the study by Buttermann which showed that vascularization of the scaphoid was interrupted when the wrist is in a position of 60° of palmar flexion and 15° of ulnar deviation after tightening of the extensor carpi radialis brevis muscle [33]. Although thumb hypoplasia was only found in six cases [20,23,31,32], it is still probably a risk factor because this disease is so rare in the general population. The possibility that ulnar variance plays a role in the development of Preiser’s disease has been discussed by certain authors, as in Kienböck’s disease [34]. Seven out of eight of the patients in the study by Herbert and Lanzetta [2] had positive ulnar variance. Analysis of our 10 cases shows that most cases presented with negative ulnar variance (six cases). However, no tendency was identified in the literature because our analysis of all of the articles shows 19 cases with negative ulnar variance, 17 cases with positive ulnar variance and 10 cases with neutral variance. This suggests that there is no potential indication for osteotomy in Preiser’s disease.

The other etiological factors suggested were tobacco (three cases), alcohol (three cases), chemotherapy (three cases), kidney transplantation (three cases), diabetes (two cases), scleroderma (two cases), myelodysplastic syndromes (one case), vasculitis (one case), hypercholesterolemia (one case), scapholunate advanced collapse (one case). None of these can be identified as direct causes because of their low frequency.
A 25-year-old man who developed wrist pain after a fall from a motorcycle: a: heterogenous appearance of the scaphoid on X-ray, hyperfixation of tracer on scintigraphy and necrosis of the entire scaphoid on MRI; b: following a vascularized bone graft: revascularization of the scaphoid; less pain, recovery of joint range of motion (flexion/extension arc 140°) and grip strength (grip 96% of the contralateral side).

Radiological features

The stage of the disease was reported in 85 cases and included: 12 cases of stage I disease, 36 stage II, 26 stage III and 11 stage IV.

Kalainov et al. identified two categories of avascular necrosis according to the location of necrosis, type 1 in which necrosis involved the entire scaphoid and type 2 in which the necrosis was only found in the proximal pole [28]. Type 1 patients presented with general etiologies and had poorer functional results than type 2 patients, whose pathogenesis seems to be associated with local factors. Although our series does not confirm this tendency (both of our cases with a traumatic etiology involved total necroses and two thirds of the cases of long-term corticosteroid treatment involved local necrosis) our analysis of the 76 cases in the literature that reported the area of necrosis confirms Kalainov's observations. Thus, the 37 type 1 and 38 type 2 cases observed were associated with a general etiology in 13 and five cases and a local etiology (traumatic or hypoplasia)
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Figure 3  A 53-year-old patient consulting for pain and loss of range of motion in the right wrist secondary to a breast tumor treated by corticosteroid therapy and chemotherapy. a: Heterogenous appearance of the scaphoid on the initial X-ray (Herbert and Lanzetta stage II); b: X-ray of the same wrist 3 years later showing collapse of the scaphoid and early osteoarthritis of the radial side of the scaphoid (stage IV); c: proximal row carpectomy. At 36 months of follow-up, pain has completely disappeared, flexion/extension arc 95°.

Figure 4  A 29-year-old patient referred for right wrist pain. No etiological factor was identified: a: scaphoid collapse and necrosis of the two thirds of the proximal pole identified on MRI; b: four-corner fusion with denervation of the posterior interosseous nerve: persistent pain 14 months after surgery with flexion/extension arc of 90°.

in seven and 11 cases, respectively. The functional results in type 1 patients were indeed poorer following all the different treatments, with a mean flexion-extension arc of 74.5° (n=24), grip strength of 70% of the contralateral side (n=26) and a Mayo Wrist Score of 62 points (n=21). The values obtained for these same parameters in type 2 patients were respectively 101° (n=12), 85% (n=10) and 81 points (n=6).

These observations support the hypothesis that a continuum may exist between type 2 avascular necrosis and necroses of the proximal pole observed secondary to fractures. Damage to the dorsal vascular network, which is the only blood supply for the proximal pole of the scaphoid [35], could explain these localized types of Preiser’s disease [13]. Moreover, this notion is not new, since Allen had already suggested the possibility of vascular damage in a report of 2
cases of avascular necrosis of the scaphoid in 1983 [6]. Moreover, it is possible that these necroses were secondary to unidentified fractures [13]. The report by Filan and Herbert in 1995 of four cases presenting with necrosis after union of a scaphoid fracture, is a good example of this possibility [36].

**Therapeutic strategies**

To our knowledge, none of the published series objectively compares one type of treatment strategy to another, and none of the treatments has been shown to be more effective than the others to date. Moreover the numerous treatment options in the literature make it difficult for the surgeon to choose the indication (Table 2).

In our series, functional results in the two vascularized bone grafts performed for early stage necrosis were very encouraging with significant improvement in necrosis on imaging. Results of non-surgical treatment were mixed for this early stage of the disease. Finally, although palliative treatment did not restore strength or physiological range of motion, it helped reduce pain in later stages of the disease.

These observations confirm results in the literature:

- 83% of stage I and 42% of stage II cases were managed with conservative treatment. Numerous authors find this non-invasive approach to be the most reasonable option. Based on a study of nine patients, five of whom received non-surgical treatment, Vidal et al. [32] recommended non-surgical treatment in most patients. Nevertheless, the results of conservative treatment in early stage disease vary and are often mediocre. Only four out of 10 patients with stage I disease treated non-surgically had little or no pain and good long-term results (strength and range of motion more than 80% of the contralateral side and/or a Mayo Wrist Score above 80 points). Moreover, radiological follow-up of eight of these patients showed that initial lesions had worsened in all adults (Fig. 1). A cure was only obtained in one patient, a 12-year-old child [14] and the disease did not progress in only one patient, a 9-year-old child [25]. Follow-up results were similar in 15 other patients who initially presented with stage II disease and were treated conservatively. In this group only four patients presented with a range of motion greater than 120° (or 80% of the contralateral side) and grip strength of more than 80% at final follow-up. Only two of them had no pain. Radiological signs of disease progression in these 15 patients, which was reported in 10 cases, showed progression towards fragmentation of the scaphoid or osteoarthritis in 90% of the cases (Fig. 1). Moreover six patients required additional surgery. No predictive factor could be identified in the few patients with little progression or who were asymptomatic. Moreover, there is no way to predict whether these results will remain stable over time. Therefore, for us, conservative treatment is only indicated in pediatric patients in whom cases of spontaneous revascularization have been observed [14,18] or in late stage forms with mild symptoms, before performing palliative surgery;
- the indication for a vascularized bone graft should be reserved for stage II avascular necroses (Fig. 2). Functional results of this treatment option, which was performed in 15 stage II cases, were encouraging. Clinical results reported in seven cases showed a mean range of motion of 102°, a grip strength of 82% of the contralateral side and a Mayo Wrist Score of 75 points. Moreover this is the only treatment that stopped radiological disease progression, since in the 10 cases that were followed-up, initial lesions only worsened in 40% of wrists (Fig. 1). Moreover, no additional surgery was required in any of these cases. Despite these good results, there were no reports of vascularized bone grafts in any stage I wrists. Although it may seem difficult to prescribe this treatment in cases with no radiographic evidence of lesions, these results suggest that this option could at least prevent the natural disease progression. On the other hand, the indication for this treatment in stage III disease in which the architecture of scaphoid is already compromised is less clear. Nevertheless, Sokolow et al. [37] recommend vascular bone graft to preserve the morphology of the wrist as much as possible. We only found seven cases of vascularized bone graft at this stage: all with good results. Moreover, five of these were reported by Moran et al. [29] who only recommends this treatment for stages I and II;
- the few number of traditional grafts makes it impossible to determine if this treatment should be an option for Preiser’s disease. Although certain authors report results that are comparable to vascularized bone grafts [37], the physiopathology of this disease does not support using this procedure;
- the role of proximal row carpectomy (Fig. 3) is difficult to define because the stage of the wrists that were treated with this option was only reported in seven out of 19 cases. Nevertheless, this treatment option seems to be effective for pain. Thus in 14 patients the mean VAS was 1 [0—5]. By providing effective pain relief, this option may therefore be interesting in degenerative forms of the disease. For Alnot et al. [7] and De Smet [27], it is the reference treatment whatever the stage;
- partial fusion (Fig. 4) is rarely used (Table 2). We did not find any explanation for the choice of proximal row carpectomy by the different authors in the literature. Our divergent results in two patients did not further clarify this choice.

**Disclosure of interest**

The authors declare that they have no conflicts of interest concerning this article.

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