Avascular necrosis of scaphoid-Preiser’s syndrome.

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ABSTRACT: Preiser’s syndrome is a rare avascular necrosis affecting the carpal scaphoid. Osteoarthritic changes of the articular cartilage, local synovitis, and loose fragments are the most common findings associated with this syndrome. We report a case of patient with Preiser’s syndrome, without a traumatic history, presenting with pain, swelling and functional impairment of the wrist. Radiography wasn’t sufficient for the diagnosis, so MRI and scan were done to clearly establish type and extension of the lesion. Differential diagnosis may be sometimes difficult and the therapeutic approach mainly depends on the stage of disease. Idiopathic avascular necrosis of the scaphoid must be included in the differential diagnosis of persistent pain on the radial side of the wrist with no history of injury, although rare cases have been reported in the literature.

Key words: Preiser’s syndrome, scaphoid, wrist

INTRODUCTION
Idiopathic avascular necrosis of the scaphoid was first described in 1910 by Preiser [1,2]. That 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 wrist motion. Avascular necrosis of the carpal bones can affect the lunate, the pisiform, capitate and the scaphoid.

Although avascular necrosis may occur without any known history of injury [3], it may be difficult to exclude the possibility of mild or repetitive trauma. Other known causes of avascular necrosis include steroid therapy [4], chemotherapy and connective tissue diseases such as systemic lupus erythematosus [5] or progressive systemic sclerosis[6].

The natural history of this disease frequently involves progression to carpal collapse and mid carpal osteoarthritis after fragmentation or pathological fracture of the scaphoid. [7].

According with Herbert and Lanzetta classification there are four stages. Stage I refers to normal X-rays and positive bone scan, stage II is characterized by increased density of proximal pole and generalized osteoporosis, stage III is related with fragmentation of proximal pole and pathological fracture, while advanced osteoarthritic changes and carpal collapse are the features of the stage IV.

Also, Kalainov et al identified two categories of avascular necrosis regarding the location of necrosis, type 1, necrosis involved the entire scaphoid bone and type 2 where the necrosis is only found across the proximal pole. Type 1 patients usually have systemic diseases and exhibit poorer functional results than type 2 patients, whose pathogenesis seems to be associated with local factors.[8]

CASE REPORT
A 55-year-old woman presented with 18-month history of pain on her dominant right wrist. No history of trauma could be elicited. On examination, swelling was noted along anatomical snuffbox and tenderness at the dorsum of the wrist was presented. The patient had previously undergone a decompression of the
first dorsal compartment of the wrist while De Quervain tenosynovitis had been misdiagnosed as well as local corticosteroid injections and a long-term immobilization of the wrist without any improvement of symptoms. The grip strength was measured at about 20% of the contralateral one. No causative factors for ischaemic necrosis were reported. X-ray findings demonstrated necrosis of scaphoid bone (dense opacity involving entire the bone) and fragmentation of the proximal pole. MRI and scan findings were consistent with avascular necrosis of the scaphoid bone with a pathological fracture (collapse) of the proximal half of the scaphoid without evidence of periscaphoid arthritis (Fig. 1). Based on these findings that case was classified as Herbert and Lanzetta type III and type I according to Kalainov classification. Proximal row carpectomy has been proposed as the method of choice to manage that type of disease. Under regional block a dorsal longitudinal approach centered over the third dorsal compartment was used. Degenerative changes regarding the lunate fossa and capitate were not observed. Proximal row carpal bones were removed using small rongeurs avoiding injury to palmar wrist ligaments. Postoperatively, a wrist splint was worn for one month and a rehabilitation protocol to gain motion and grip strength was applied. After four months, VAS (Visual Analogue Score) was improved to 2 from 8, grip strength reached 50% of the contralateral one and the range of motion approached 40% of the normal. At one year, VAS score improved to 1, grip strength was measured as 65% and motion at the 60% of the normal ones.

DISCUSSION

The aetiology and pathogenesis of avascular necrosis of the carpal bones remains uncertain. A carpal bone may be completely devascularized (i.e. completely stripped of all its soft tissue attachments) as consequence from various fracture patterns. In contrast, idiopathic avascular necrosis, occurs in the absence of major trauma or pre-existing fracture. 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. We have reviewed carefully all available X-ray films of our patient without any evidence of a pre-existing fracture.

Clinically, the onset of idiopathic avascular necrosis of the scaphoid is presented with 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.

Figure 1. MR image consistent with ischaemic necrosis of scaphoid bone.
It is hardly surprising that patients also may have signs of carpal tunnel syndrome and De Quervain’s syndrome, which resolved once the synovitis has settled.

As with Kienbock’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. Once the bone has undergone structural deformities due to the necrosis (stages III and IV) these changes are almost certainly irreversible so that reconstruction is no longer indicated. In stage I the patients received conservative treatment, which included occasionally bracing, local cortisone injections and oral analgesics. Vascularized grafts harvested from the volar aspect of the distal radius were performed for stage II disease. Proximal row carpectomy has been proposed for stage III disease. Also, a partial silastic replacement of the scaphoid bone combined with a local synovectomy has been reported in the management of selected type III cases. Additionally, if scaphoid shortening results in symptomatic radio-carpal impingement, the symptoms may be relieved by a limited radial styloidectomy.

A capitotolunate arthrodesis and four-corner fusion both associated with a scaphoidectomy are recommended for stage IV disease. Especially, regarding the proximal carpectomy it is considered a reliable method to manage the pain and grip strength deficit. Based on 2 separate cohorts of patients, each with a minimum follow-up period of 10 years, one can conclude that patients having a proximal row carpectomy will achieve over 80% of normal grip strength and 60% of normal motion [9, 10], results comparable with those regarding our case.

Idiopathic avascular necrosis of the scaphoid must be included in the differential diagnosis of persistent pain on the radial side of the wrist with no history of injury, although rare cases have been reported in the literature.

Ισχαιμική νέκρωση του σκαφοειδούς οστού-Σύνδρομο Preiser

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ΠΕΡΙΛΗΨΗ: Το σύνδρομο Preiser είναι μια αστεία άσηπτη νέκρωση που αφορά στο σκαφοειδές οστάρι του καρπού. Οστεοαρθρίτιδες, η επιχώρια υμενίτιδα, και συχνά παθολογικά κατάγματα του οσταρίου είναι τα πιο κοινά ευρήματα που σχετίζονται με αυτό το σύνδρομο. Παρουσιάζουμε την περίπτωση μιας ασθενούς με σύνδρομο Preiser, χωρίς ιστορικό κάκωσης, με άλγος, οίδημα και λειτουργική δυσλειτουργία του καρπού. Η απλή ακτινογραφία δεν ήταν επαρκής για τη διάγνωση, έτσι ο απεικονιστικός έλεγχος με MRI και σπινθηρογράφημα οστών ήταν αναγκαίος για τη διάγνωση. Η διαφορική διέγνωση συχνά είναι δύσκολη και η θεραπευτική προσέγγιση βασίζεται κυρίως στο στάδιο της διαταραχής.

Λέξεις-κλειδιά: Σύνδρομο Preiser, σκαφοειδές, καρπός
REFERENCES


