# **Case Report**

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# Presier's disease: idiopathic avascular necrosis of scaphoid in a case presenting with wrist pain in young male

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## **ABSTRACT**

A 25-years-old male presented with complain of pain in right wrist and difficulty in gripping objects. No history of trauma to the right wrist joint. On local examination, tenderness was present in right anatomical snuff box. Tenderness was elicited by axial compression on right first metacarpal with decreased range of the motion at the right wrist compare to the left side. Routine blood investigation was within normal limits. X-ray of the right wrist joint showed minimal sclerotic in right scaphoid. On MRI right wrist joint, low intensity signal was seen involving the whole right scaphoid bone on T1 weighted, T2 weighted and STIR images with loss of normal marrow signal intensity. So according to the Herbert and Lanzetta it was stage 4 and Kalainov et al, type 1 avascular necrosis. Diagnosis of idiopathic avascular necrosis of the right scaphoid bone was postulated based on clinical and radiological findings. Patient was treated with vascularised pedicle bone graft from the right distal radius. The patient was gradually improved clinically with subsidence of pain and improvement in the grip strength over 1 year.

Keywords: AVN, MRI, Wrist joint

#### **INTRODUCTION**

Preiser's disease is an idiopathic avascular necrosis of scaphoid bone. It is a rare condition with female preponderance. The most common symptoms are pain in the wrist joint, decreased range of motion and loss of grip strength. The other possible etiology of avascular necrosis is prolonged use of corticosteroid therapy and trauma. The management of Preiser's disease is controversial because it is a rare disease. Treatment is provided according to the stage of the disease. Conservative management or vascularised bone grafts from iliac crest, distal radius and second metacarpal has been used to treat early disease. Options of treatment

available for advanced stage are the salvage procedure like wrist arthrodesis, proximal carpal row excision, wrist denervation, scaphoid excision and scaphoid replacement.

#### **CASE REPORT**

A 25-years-old male presented with complain of pain in right wrist and difficulty in gripping objects. No history of trauma to the right wrist joint. No complain of pain in the other joints of the body. No history of any drug intake or blood disorder. On physical examination of patient, his blood pressure of 110/80 mm Hg (normal), respiration rate of 16/min (normal), pulse rate of 84/min (normal).

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On local examination, tenderness was present in right anatomical snuff box. Tenderness was elicited by axial compression on right first metacarpal with decreased range of the motion at the right wrist compare to the left side. No evidence of any deformity of right wrist joint and hand.



Figure 1: Plain x-ray right wrist joint antero-posterior view shows fracture of right scaphoid bone with minimal sclerotic changes.

His haemoglobin (Hb) was 13.0 gm% (normal), total white blood cell 9700 cells/mm3 (normal), erythrocyte sedimentation rate was 12 mm at the end of 1 hour (normal). Test for the serum C-reactive protein and HIV (Human rheumatoid factor were negative. Immunodeficiency Virus) and HbSAg (Hepatitis B antigen) were negative. Then patient refereed to the department of the radiology for X-ray and MRI study. Xray of the right wrist joint showed minimal sclerotic in right scaphoid. (Figure 1) On MRI right wrist joint, low intensity signal was seen involving the whole right scaphoid bone on T1 weighted, T2 weighted and STIR images with loss of normal marrow signal intensity. So according to the Herbert and Lanzetta it was stage 4 and Kalainov et al. type 1 avascular necrosis.



Figure 2: Coronal T1 weighted MRI right wrist joint shows low signal intensity involving the whole right scaphoid bone with loss of normal marrow signal intensity.



Figure 3: Coronal T2 weighted MRI right wrist joint shows low signal intensity involving the whole right scaphoid bone with loss of normal marrow signal intensity with few high signal intensities cystic changes within.

Little small cystic signal intensity was seen in T1 weighted, T2 weighted and STIR images (Figure 2, 3, and 4) Diagnosis of idiopathic avascular necrosis of the right scaphoid bone was postulated based on clinical and radiological findings. Patient was treated with vascularised pedicle bone graft from the right distal radius. The patient was gradually improved clinically with subsidence of pain and improvement in the grip strength over 1 year.



Figure 4: Coronal STIR MRI right wrist joint shows low signal intensity involving the whole right scaphoid bone with loss of normal marrow signal intensity with few high signal intensity cystic changes within.

### **DISCUSSION**

Preiser's disease is an idiopathic avascular necrosis of scaphoid bone. It is a rare condition. Female are more affected than the male. The average age of presentation is 42 years. Dominant side is more affected twice than the non-dominant due to repeated microtrauma.

Proximal branch of the radial artery supply blood to the scaphoid bone primarily. Dorsal branches enter the distal aspect of the bone and run proximally. Volar and laterally based vessels have also been described and enter at the level of the scaphoid tubercle.<sup>2</sup>

The most common symptoms are pain in the wrist joint, decreased range of motion and loss of grip strength. The other possible etiology of avascular necrosis is prolonging use of corticosteroid therapy and trauma. Thumb hypoplasia and ulnar variance are still probably a risk factor plays a role in the development of Preiser's disease.<sup>3</sup> In study by Herbert and Lanzetta, seven out of eight of the patients had positive ulnar variance.<sup>4</sup> Staging of Preiser's disease (Herbert and Lanzetta)<sup>4</sup>-

- Stage 1: Normal radiographs, MRI changes, Positive bone scan
- Stage 2: Increased density of proximal pole and Generalised osteoporosis
- Stage 3: Fragmentation of proximal pole +/pathological fracture
- Stage 4: Carpal collapse, osteoarthritis.

So, our case was the stage 4 according to the Herbert and Lanzetta classification. According to the Kalainov et al, two categories of avascular necrosis by the location of necrosis, type 1 in which necrosis involved the entire scaphoid bone while type 2 involves only the proximal pole.<sup>5</sup> Type 1 patients had poorer functional outcome than type 2 patients. Our patient was belonging to the type 1 category. The management of Preiser's disease is controversial because it is a rare disease. Plain radiography and MRI is needed for the diagnosis of Preiser's disease according to the Kalainov et al, to define the type.<sup>5</sup>

In the absence of trauma, any areas of low-signal intensity on T1-weighted MR images should raise the suspicion of idiopathic AVN as the carpal bones do not show the same subchondral pattern of AVN as larger bones.<sup>6</sup> Treatment is provided according to the stage of the disease. Conservative management consist of immobilization in a plaster cast for 3 months and other modalities like arthroscopic debridement and radial osteotomy have been successful in early stage.<sup>7-9</sup>

According to the study of Harpf et al, vascularized bone grafts from iliac crest, distal radius and second metacarpal has been used to treat early disease. <sup>10</sup> Use of local pedicle grafts had less pain and greater movement of the wrist at a mean follow-up of three years according to the Moran et al the treatment of Preiser's disease is promising. <sup>11</sup> Options of treatment available for advanced stage are the salvage procedure like wrist arthrodesis, proximal carpal row excision, wrist denervation, scaphoid excision and scaphoid replacement.

#### **CONCLUSION**

Preiser's disease is an idiopathic avascular necrosis of scaphoid bone. It is a rare condition with female preponderance with average age of presentation is 42 years. Patent presented with pain in the wrist joint, decreased range of motion and loss of grip strength. Plain radiography and MRI is needed for the diagnosis of Preiser's disease. Treatment is provided according to the stage of the disease.

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