Kienböck's Disease

Synonyms: lunatomalacia, lunate malacia, idiopathic lunate necrosis, avascular lunate necrosis, lunate osteonecrosis

Kienböck's disease is poorly understood. It involves impaired vascularity of the lunate bone in the wrist which leads to osteonecrosis of the lunate and marked wrist degeneration. It is usually unilateral. It was first described by Robert Kienböck in 1910.

Aetiology

The cause of Kienböck's disease is not known, although various factors have been proposed. These include:

- Single or repetitive microfractures leading to vascular disruption of the fragments.
- Recurrent compression of the lunate between capitate and distal radius.
- Extreme wrist positions or repetitive compression loading of the wrist.
- Negative ulnar variance, which occurs when the ulna projects more proximally than the radius, leading to increased load transmission through the radio-lunate joint.
- Various differences in lunate morphology, such as size and inclination, that lead to altered trabeculation and therefore structural strength.
- Venous plexus abnormalities that may lead to obstructed venous drainage and increased intraosseous pressure.

It is traditionally regarded as an occupational disease caused by hand-arm vibration but a systematic review found that the evidence for this association is weak.

Use of agents such as corticosteroids or systemic disease such as osteoporosis could in theory predispose to Kienböck's disease but a study of bilateral disease did not find such risk factors. There appears to be an association between osteoporosis and Kienböck's disease when it occurs in the elderly, when there is also a higher prevalence among women.

Epidemiology

It occurs most often in adult males between 20 and 40 years of age. Its incidence and prevalence are unknown but it is considered a rare disease.

Presentation

History

- It is difficult to diagnose in the early stages when symptoms are similar to wrist sprain.
- It presents insidiously as a dull intermittent ache over the lunate, which is medial to the scaphoid and lateral to the triquetral in the proximal carpus. It articulates distally with the capitate and proximally with the radial head.
- The pain may radiate up the wrist and forearm.
- There is associated stiffness at the wrist.
- It may be aggravated by activity and relieved by rest.
- There may be a history of a single injury or repetitive injury/compression loading.
- It may be picked up as an incidental finding on X-rays and it does not always cause pain or interfere with activities of daily living.
Examination

- There may be no, or only very subtle, signs, such as swelling over the radio-carpus signifying a synovitis.
- Passive dorsiflexion of the middle finger produces the characteristic pain.
- There may be limitation of wrist flexion and extension when compared with the other side.
- There is often a weakened grip.

Symptoms are progressive and chronic as the lunate bone progressively collapses and degenerative changes occur.

Differential diagnosis

Wrist sprains and any cause of arthritis in the wrist joint may give similar symptoms.

Investigations

- Wrist X-ray:
  - Initially, this may be normal or show sclerosis of the lunate.
  - It may show negative ulnar variance.
  - The lunate shows progressive loss of height, and fragmentation.
  - The lunate collapse causes further degenerative joint changes (because of carpal instability) with bone cysts within the lunate.
  - Eventually, degenerative changes may involve the whole wrist.
  - Measurement of ulnar variance requires a zero rotation view on a PA radiograph of the wrist in neutral pronation/supination. Evidence of its importance in Kienböck's disease is conflicting.
  - X-rays may show no specific abnormality in the early stages and so MRI scans are essential if early Kienbock's disease is suspected.

Associated diseases

Osteoarthritis.

Staging

The modified Lichtman classification, based on radiological and MRI findings, is used to guide management of Kienböck's disease:

- Stage I: symptoms are similar to wrist sprain. Normal X-rays or a possible linear fracture. MRI may be useful to confirm diagnosis.
- Stage II: symptoms of recurrent pain and swelling. The lunate bone becomes hard/sclerotic and X-ray shows sclerosis (indicating the bone is infarcting). CT and MRI scanning may be useful to assess the condition of the lunate bone.
- Stage III A, B and C: increasing pain, weakened grip and limited wrist movement occur. The infarcted bone collapses and breaks up, causing a shift in position of the surrounding bones.
- Stage IV: degeneration of the surrounding bones causes arthritis of the wrist.

Other classification systems are described that take account of different imaging technologies, including gadolinium contrast MRI (Schmitt) and arthroscopic evaluation of articular cartilage (Bain).

Management

Treatment is controversial, partly because the natural history of the disease is poorly understood; symptoms and disability do not correlate with progression.

No treatment +/- splinting is an option at all stages, particularly in the young and the elderly, as they have the best prognosis. A systematic review concluded that no surgical treatment is superior to any other and that there are insufficient data to show any benefit of a particular intervention over placebo or the natural history of the disease.

Early disease

The aim is to reduce compressive loading of the lunate, encouraging revascularisation and preventing lunate collapse. Referral to an orthopaedic or hand surgeon is recommended. The aim of reducing lunate loading and encouraging revascularisation may require surgery.

- Synovectomy, scapho-trapezio-trapezoid (STT) joint pinning, neurectomy and radial forage (arthroscopic drillings) are surgical options that may be considered at any stage of disease.
- Establishing a negative ulnar variance (unloading the lunate fossa and distributing load to the scaphoid fossa) by:
  - Radial shortening.
  - Ulnar lengthening.
  - Fusion of the capitate and hamate.
  - STT joint fusion (up to stage III).
Establish neutral ulnar variance by radial wedge osteotomy.
Vascular bone graft.

**Late disease**
These procedures are for more advanced disease and used less often.

- Lunate reconstruction:
  - Vascularised bone graft.
  - Medial femoral condyle osteochondral reconstruction.
  - Lunate replacement.
  - Proximal row carpectomy.
  - Capitate lengthening.
  - Scapho-capitate fusion.
  - Pyrocarbon replacement.

- Wrist reconstruction:
  - Radio-scapho-lunate or scapho-capitate fusion.
  - Proximal row carpectomy with hemiarthroplasty.

- Wrist salvage:
  - Total wrist fusion or arthroplasty.

**Complications**
Stiffness and progressive loss of wrist function are well-described sequelae of the condition. Grip strength deteriorates by 40% between stages II and IV of the disease.

**Prognosis**[1]
If left untreated it may progress, passing through the various stages described by Lichtman. However, this is not inevitable, although it is not known why or how the disease ultimately subsides in some people[8]. Children younger than 12 appear to have least progression and normal lunate anatomy may be restored simply following immobilisation. Elderly patients appear to be most likely to progress to stage IV disease; however, they do well symptomatically without surgery, despite radiographic progression. The greatest difficulty in the management of Kienböck's disease lies in the inability to identify which patients will have progressive, symptomatic disease and which will eventually enjoy resolution of the disease, without any intervention.
Prevention

Awareness of this condition can prompt earlier diagnosis and corrective measures to prevent progression of the disease. However, it should be remembered that symptoms can be mild for many years with no treatment[6].

Further reading & references


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