Rare cause of paediatric wrist pain unmasked by minor trauma

Fahim Patel, Arani V Sridhar

DESCRIPTION

A 10-year-old girl was referred to paediatric rheumatology with a 6-month history of a painful, swollen left wrist associated with functional limitation and disturbed sleep. She initially had a minor fall, and X-rays at the time showed no bony abnormalities (figure 1). Blood tests were normal, including inflammatory markers and autoimmune screen. Wrist MRI showed significant synovial thickening and avascular necrosis of the left lunate (figure 2). She received non-steroidal anti-inflammatories and physiotherapy, but a year later, continues to have chronic regional pain with allodynia and hyperalgesia.

Kienböck disease, an eponym for avascular necrosis of the lunate bone, is of unknown aetiology and incidence. The proposed trigger is trauma in patients with a skeletal and vascular susceptibility. It is the most common cause of adult aseptic osteonecrosis of the upper extremity, usually in dominant hands of men aged 20–40 years. Paediatric Kienböck is rare, presenting as pain, stiffness, swelling and reduced power, often after an innocuous fall. Diagnostically, this is challenging because the mechanism suggests

Figure 1 A plain radiograph of the left wrist taken shortly after the injury shows no visible alterations.

Figure 2 Coronal T1-weighted (A) and coronal T2-weighted fat-suppressed (B) MRI of the left wrist. The lunate bone has low T1 signal (white arrow) and high T2 signal (blue arrow) due to bone marrow oedema and bony changes associated with avascular necrosis. The synovium exhibits the same pattern of signal change due to thickening and reactive inflammatory changes. Negative ulnar variance is also present: there is a significant association with Kienböck disease.

Patient’s perspective

Our patient received non-steroidal anti-inflammatories and physiotherapy, but a year later, continued to have chronic regional pain with allodynia and hyperalgesia. Despite this, she remains upbeat and continues to live a normal childhood and has coped well with the recent increase in computer usage and typing associated with remote schooling as a consequence of school closures during the COVID-19 pandemic.

Learning points

► Kienböck disease is an eponym for avascular necrosis of the lunate bone. It is of unknown aetiology and incidence, but the proposed trigger is trauma in patients with a skeletal and vascular susceptibility. In the majority of cases (approximately 80%), there is a significant association with negative ulnar variance.
► Kienböck disease is a rare paediatric diagnosis, but the most common cause of adult aseptic osteonecrosis of the upper extremity, usually affecting the dominant hands of men aged 20–40 years.
► Kienböck disease can develop insidiously and should, therefore, be considered in both adults and paediatric patients presenting with a chronic non-resolving or partially-resolving wrist trauma.
Images in…

a soft tissue injury whereas the chronicity mimics juvenile idiopathic arthritis.

Radiographical severity is defined by Lichtman classification and used to guide non-curative surgical or conservative management.4 This aims to relieve pressure on the lunate bone and restore perfusion. Anti-inflammatory medications are offered prior to surgical joint levelling to reduce pain, swelling and deformity.4

We emphasise that clinicians consider this rare, destructive pathology in their differential diagnosis for paediatric chronic wrist pain and swelling, especially in those presenting weeks after a seemingly innocuous hand trauma.

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