A bloody painful knee: delayed presentation of haemophilic arthropathy

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DESCRIPTION

A 52-year-old South Asian man (recently arrived in the UK) with moderate haemophilia B was referred to orthopaedics with a painful right knee. Over the past 30 years he had experienced multiple flares of pain and swelling, each advancing the degree of stiffness and pain. He struggled to walk, with a fixed flexion deformity of 15° and active flexion restricted to 90°. He had no history of trauma, had not sought prior treatment, and other joints were unaffected (figure 1A, B).

Haemophilic arthropathy is usually multiarticular, but can be monoarticular, with the same joint involved on multiple occasions.[1] Ankles are commonly affected in children; knees, elbows and ankles in adolescents and adults. Haemarthrosis can be acute or subacute. Chronic toxic effects of blood and an inflamed synovium result in a mass eroding into cartilage and subchondral bone, causing subarticular cyst formation. Severe haemophilic arthropathy—permanent joint disease secondary to repeated haemarthrosis—is seen in 50% of cases. Pettersson and Arnold-Hilgartner are two of many scoring systems; however, X-ray can underestimate the degree of joint destruction.[2]

In acute bleeds, factors should be administered to achieve levels of 40–50%. Early prophylactic

Learning points

- Haemophilia A (factor VIII deficiency) and haemophilia B (factor IX deficiency) are X linked recessive diseases (1 in 5000 live male births) associated with excessive, prolonged bleeding. Eighty per cent of bleeding occurs in the joints, typically first occurring in childhood.
- Haemophilia B is associated with less severe arthropathy and better surgical outcome. Severe haemophilia (factor <0.01 IU/mL or <1%) results in a high risk of spontaneous bleeds (haemarthrosis in 75–90%). Moderate (0.02–0.05 IU/mL or 2–5%) or mild (0.06–0.40 IU/mL or 6–40%) disease results in bleeds following minor trauma or invasive interventions.
- Plain X-ray is first-line imaging, supplemented by ultrasound to identify effusions and synovial proliferation. MRI and CT may aid preoperative planning. Joint aspiration for Gram stain, microscopy and culture (after correcting coagulation abnormalities) can exclude infection or crystal arthropathy.

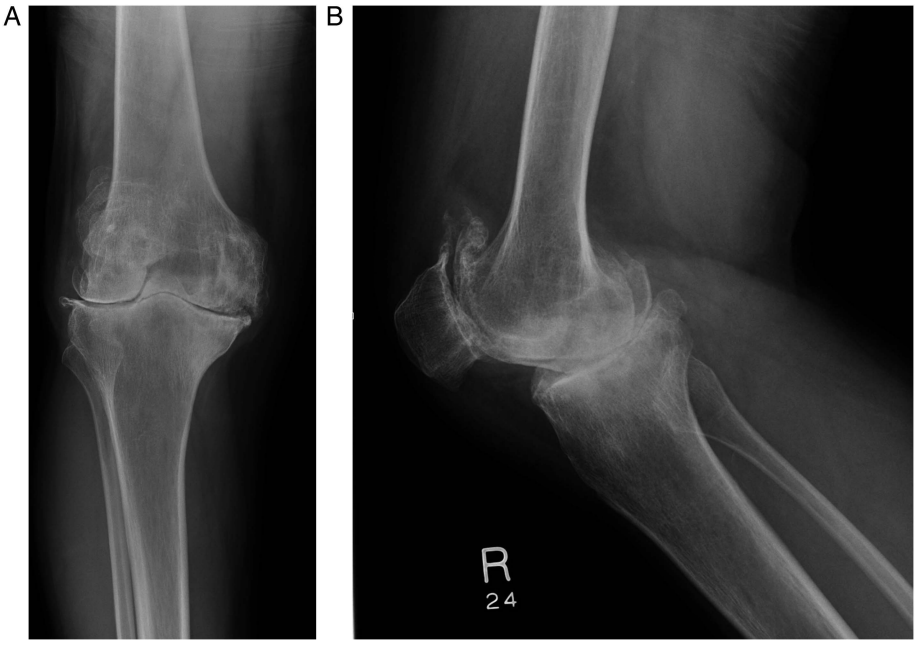

Figure 1 (A) Anteroposterior (AP) and (B) lateral radiographs of a patient's right knee. These show severe tricompartmental osteoarthritis, broadening of the intercondylar notch, squaring of the inferior patellar margin, bulbous femoral condyles and flattened condylar surfaces.

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factor, viscosupplementation, avoidance of trauma and careful follow-up can prevent or delay arthropathy.[3] Advanced joint disease, as seen here, is not uncommon in patients presenting from developing nations without early factor treatment. Arthroscopic synovectomy may be helpful in early disease. Surgical arthrodesis or arthroplasty can improve joint function in end-stage disease—as seen in our patient. Older implants show higher risk of prosthesis failure due to higher wear and aseptic loosening, although recent series have demonstrated component survival of >85% at 10–15 years.[4]

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