International Journal of Innovative Research in Medical Science (IJIRMS)
Volume 01 Issue 10 December 2016, ISSN No. – 2455-8737
Available online at - www.ijirms.in

Open Access Journal | Case Report | ISSN - 2455-8737

Haemophilic Arthropathy and Pseudotumour in an Elderly Male

Kunal Mahajan MD[*1], Sanjay Rathour MD[2], Ashish Dhiman MD[3], Gunjan Gupta MD[4]

Indira Gandhi Medical College Shimla

Case description *- A 62 years old male presented with h/o recurrent episodes of pain and swelling in both knee joints leading to flexion deformity on left side.(Figure1A) He also had a soft tissue swelling in right thigh for last 30 years, which was nontraumatic and gradually progressive in size. On examination, a well-defined swelling of 15 x 10 cm was present over right thigh, which was compressible, nonreducible, and non-tender. (Figure2A) History of recurrent episodes of epistaxis and spontaneous gum bleeds since early childhood was also present. Laboratory investigations revealed increased clotting time with increased activated partial thromboplastin time (aPTT). Platelet count, bleeding time and prothrombin time were normal. Factor VIII levels were low (2%) suggestive of moderate haemophilia. X-ray right thigh and bilateral knees confirmed it to be case of haemophilic arthropathy with pseudotumour. (Figure1B and 2B) Hemophilic pseudotumor is a rare complication occurring in 1–2% of patients with haemophilia. [1]Repetitive bleeding into the bones and soft tissues results in pseudotumor, whereas repetitive bleeding into the joints leads to arthropathy.[2] Pseudotumours are most common in thigh, while haemophilic arthropathy classically involves knee joint. Soft-tissue pseudotumors manifest as nonspecific masses at radiography. They may demonstrate internal calcifications or ossifications. Most of the morbidity from pseudotumors is due to their compressive effect on surrounding structures including bone destruction. Haemophilic arthropathy of knee tends to be asymmetric in involvement.Changes include Juxta-articular osteoporosis, irregular appearance of the subchondral surface,multiple subchondral cysts and widening of the intercondylar notch.[3] This report describes two important musculoskeletal complications of haemophilia. Long term factor replacement is essential for treatment and surgery is required in pseudotumours which are progressively increasing and for end stage haemophilic arthropathy.*

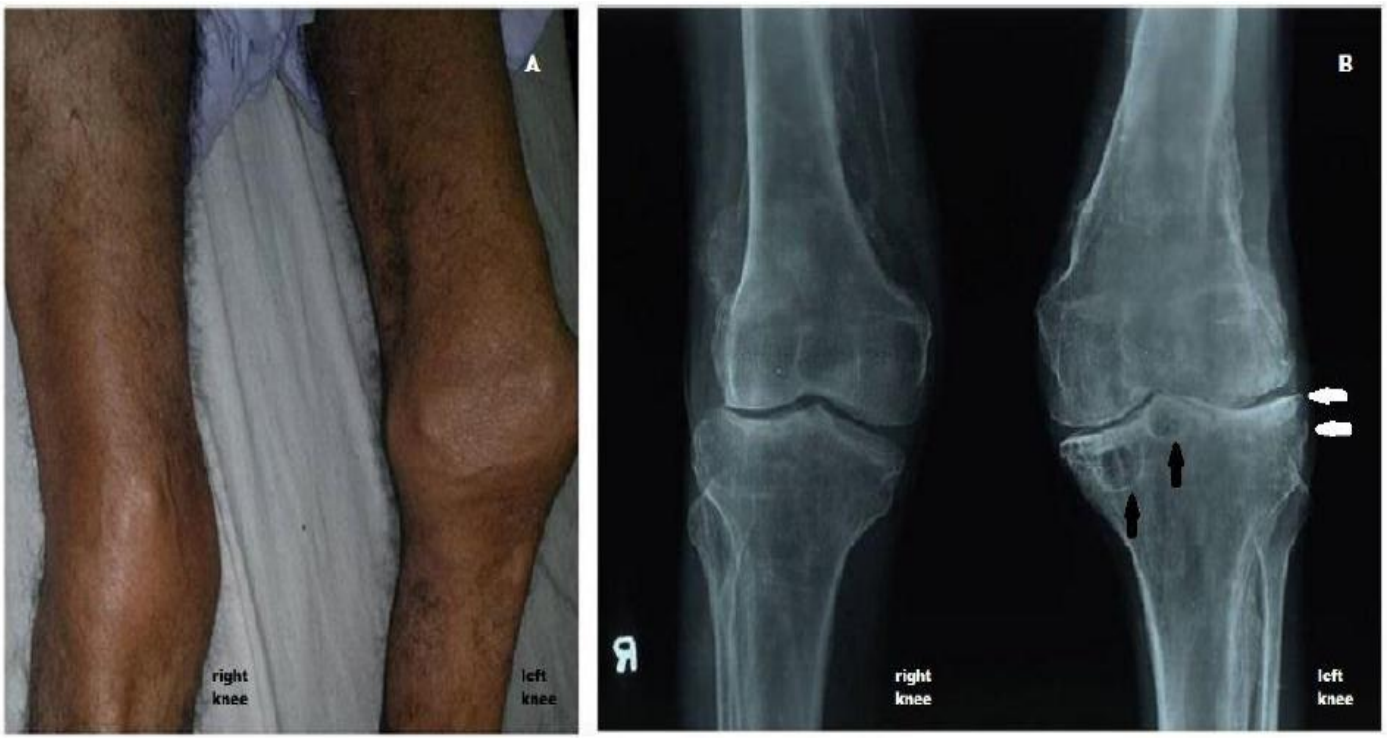

Figure 1. (A) Bilateral swollen knee joints with marked deformity of left knee joint. (B) Anteroposterior radiograph of bilateral knee joints which shows degenerative changes with decreased joint space, juxta-articular osteoporosis, subchondral sclerosis and broadening of the intercondylar notch. Note the osteophytes (white arrows) and multiple subchondral bone cysts(black arrows) in left knee joint.

Corresponding Author - Kunal Mahajan MD

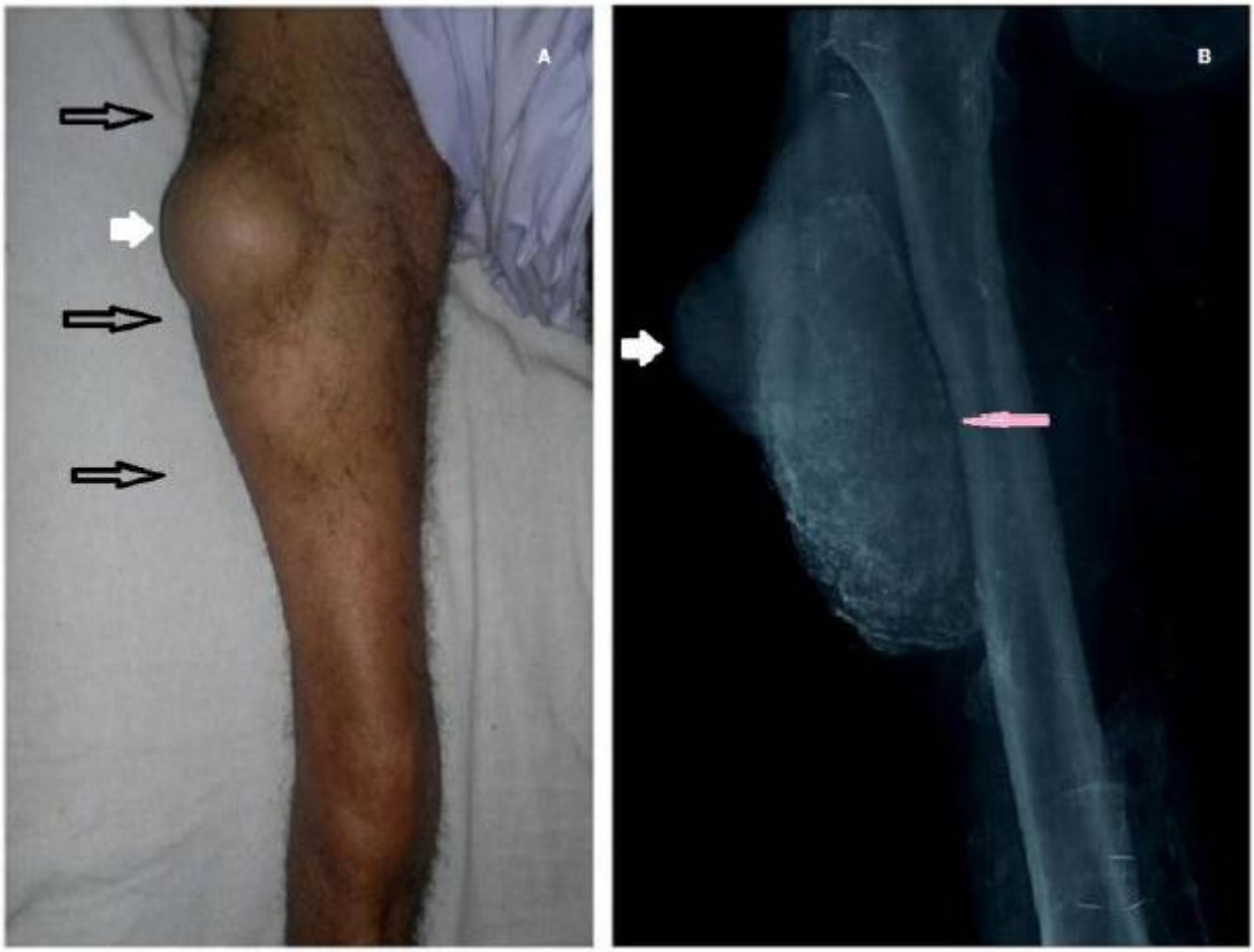

Figure 2. (A) Image of right thigh showing the extent of pseudotumour (black arrows). Note the rounded protuberant lump present in its centre (white arrow). (B) Anteroposterior radiograph showing the pseudotumour which demonstrates internal calcifications. The pseudotumour is compressing the shaft of femur (pink arrow). White arrow represents the protuberant lump which is not calcified.

Implications/learning points-

1. Most of the patients with moderate to severe haemophilia experience chronic degenerative changes (hemophilic arthropathy) by the second or third decade of life.
2. Knee is the classical joint involved, with asymmetrical involvement in majority of cases.
3. Another problem in hemophilia is the spontaneous development of muscle hematomas. When a hematoma is not totally resolved, rebleeding will tend to occur, eventually resulting in a hemophilic pseudotumor.
4. Long term factor replacement is necessary in such patients.
5. Surgery is advocated for resistant cases, end stage joint disease and for progressively increasing pseudotumours.

References –

[1] Brant EE, Jordan HH. Radiologic aspects of hemophilic pseudotumor in bone. American Journal of Roentgenology 1972; 115:525–39.
[2] J. S. Park and K. N. Ryu. Hemophilic Pseudotumor Involving the Musculoskeletal System: Spectrum of Radiologic Findings. American Journal of Roentgenology 2004; 183:55-61.
[3] Arnold WD, Hilgartner MW. Hemophilic arthropathy. Current concepts of pathogenesis and management. J Bone Joint Surg Am 1977; 59(3):287-305.

Corresponding Author - Kunal Mahajan MD