Severe Hemophilic Arthropathy of the Elbow and Knee

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The patient, a 60-year-old Afro-Caribbean man with moderate untreated hemophilia A (factor VIII deficiency) presented with chronic pain, deformity, and contractures of the right knee and the right elbow, having been lost to medical followup for several decades.

Plain radiographs of both joints revealed the presence of severe destructive changes, most notably incongruity of joint surfaces, cartilage irregularity, joint space narrowing, osteophyte formation, and subarticular bone cysts. The increased radiodensity of the thickened synovium is thought to result from deposition of hemosiderin[1,2].

The striking new bone formation, most evident in the right elbow (Figure 1a), as well as the extensive disorganization of both joints, is reminiscent of Charcot joints, a rare and very late manifestation of hemophilic arthropathy. The widened epiphysis of the right knee results from hyperemia and inflammation during childhood (Figure 1b).

Recurrent hemarthroses occurring in this sex-linked inherited coagulation disorder result in hemosiderin deposits in the synovium, which then stimulate synovial hypertrophy, the release of cytokines, and the influx of chronic inflammatory cells. Both the hypertrophic synovium and the inflammatory milieu result in cartilage damage and finally the growth of fibrotic tissue in the joint[3].

REFERENCES

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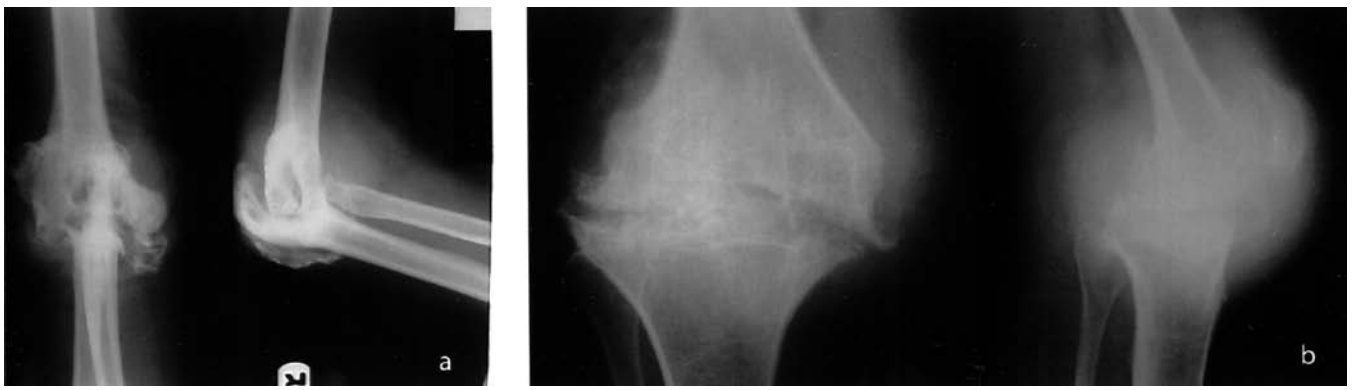

Figure 1. Anteroposterior and lateral views of the right elbow (a) and the right knee (b) in a patient with hemophilia A.