



A person with severe Factor IX (FIX) deficiency and a swollen knee: an unexpected finding



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Objective: To describe the clinical approach and differential diagnosis of a patient with severe F IX deficiency and a swollen, painful knee.

Methods: Case description of 45 year old HIV+ male with severe F IX deficiency and advanced bilateral knee arthropathy who developed sudden onset of severe right knee pain, swelling, and loss of range of motion with no history of trauma.

Results: The symptoms did not respond to immediate factor IX infusions and he went to a local hospital. Factor IX level on admission was 77%, he was afebrile with a grossly swollen, warm, tender right knee. He continued to receive factor IX and intravenous pain medications. The knee remained painful and swollen, despite therapeutic factor IX levels. Radiographs showed severe narrowing of the tibiofemoral and patellofemoral joints with joint effusion. No acute fracture was identified. On the second hospital day an arthrocentesis was performed which yielded 180 ml of pink-tinged fluid. Analysis of the joint fluid revealed intra and extracellular calcium pyrophosphate crystal deposition (CPPD), diagnostic of pseudo-gout. The patient obtained immediate pain relief after the aspiration and was discharged on day 4 on therapeutic doses of factor IX. The subsequent outpatient treatment consisted of naproxen, short course of oral steroids supplemented with intra-articular steroid injections, and colchicine. He did not have a recurrence of the pseudogout and 10 weeks later underwent successful bilateral total knee arthroplasties. Five months post hospitalization he is doing well, without colchicine therapy, and with no recurrence of pseudogout.

Conclusion: Inhibitor formation and joint infection are the primary considerations in the differential diagnosis of a swollen joint in hemophilia not responsive to factor replacement. Gout and pseudogout are generally not considered. However, if not diagnosed, these conditions may result in significant morbidity.

References:

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