

# **Hemophilic Pseudotumor: Case Report**

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### **ABSTRACT**

Hemophilia B is characterized by deficiency in Factor IX, which results in prolonged oozing after injuries, tooth extractions, or surgery. In severe hemophilia B, spontaneous joint bleeding is the most frequent complication . Hemophilic pseudotumor is a rare complication in hemophiliacs with an incidence of 1-2% . A hemophilic pseudotumor is an encapsulated, chronic, slowly expanding hematoma . Pseudotumors that occur in muscles with broad tendon insertions oftenly progress to cause severe pressure erosion of adjacent bones.

We describe a case of a 52 years old male (Jehovah's Witness) with history of Hemophilia B, legal blindness in both eyes and left thigh swelling for many years, presented in ED with oozing of

blood from left thigh after minimal trauma to his left leg .  
Patient refused any blood transfusion .

He denied any allergies, smoking, alcohol and use of any illicit substances. His home medications includes folic acid and multivitamins.

Physical examination was remarkable for blindness in both eyes, left thigh swelling greater than 24 inches in diameter.

He has severely limited range of motion on left hip and knee joint with motor strength of left leg 1/5. Right leg had normal motor and sensory functions.

Admission labs were significant for Hb of 7.8 gm/dl and Hct of 24.4%. PT 13.3 seconds; PTT 90 seconds; Factor IX assay less than 3%. Patient was transfused with Factor IX concentrate and started on Aranesp, Ferrous Sulphate and Multivitamins.

Next day, patient was found to have profuse bleeding from left thigh swelling . A Pressure dressing was applied and patient was transfused with more Factor IX concentrates.

CT scan of left thigh showed complete resorption of left femur with 24x25x34 cm enormous hematoma [fig 1, 2]. He refused any surgical intervention initially but afterwards agreed and undergoes total disarticulation of left hip joint.

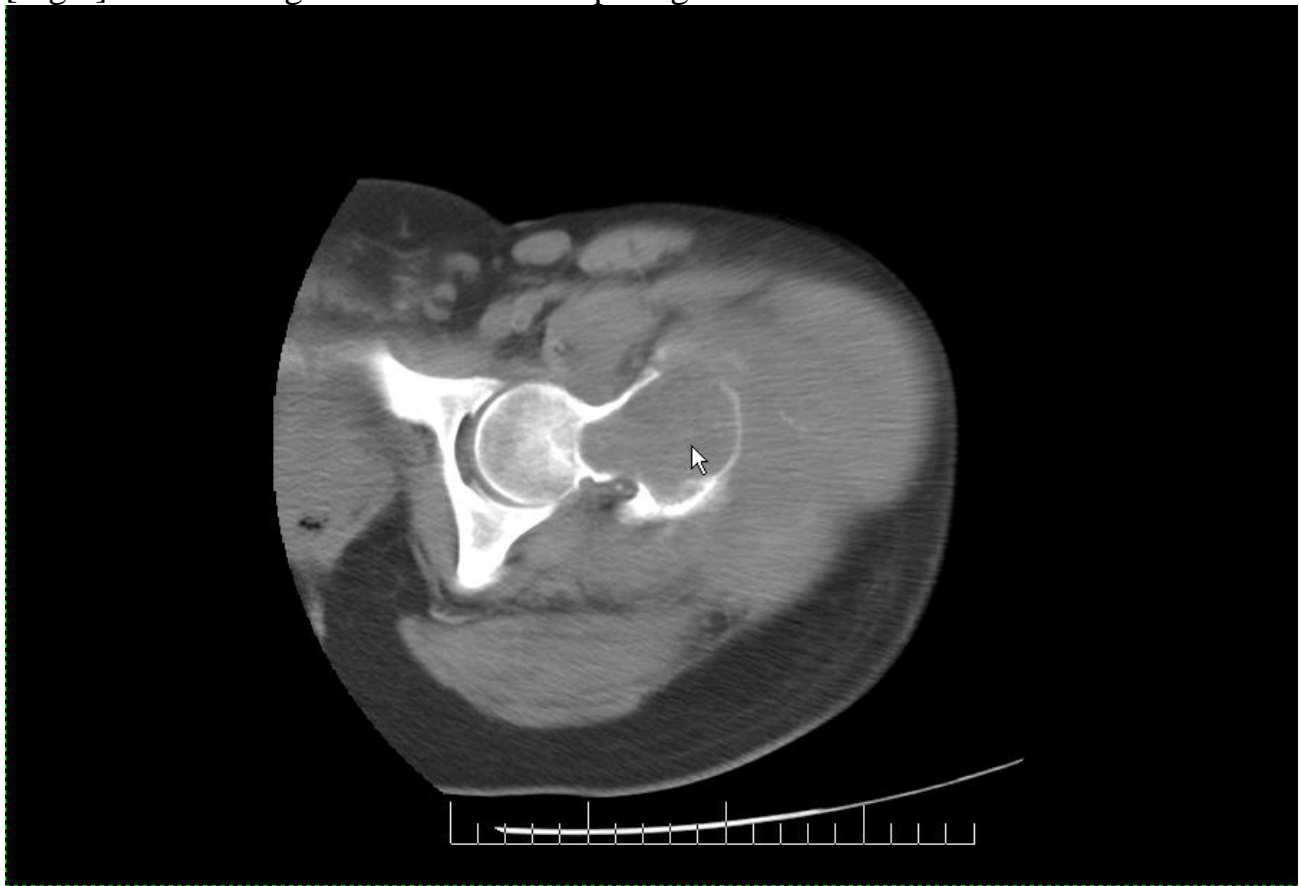


(Fig 2 ) CT scout film showing complete destruction of the left femur with a 24x25 hematoma.

He received 8 units of autologous PRBCs during surgery.

He tolerated the procedure well and after two weeks,  
discharged home with plans for physical rehabilitation.

[Fig 1] shows huge hematoma with sparing of femoral head



Hemophilic pseudotumor was first described in 1918 by Starker.

They are often asymptomatic or stable for long periods of time. CT and MR imaging are useful for determining the extent of the pseudotumor in bones and soft tissues, respectively.

The radiological diagnosis of pseudotumor can be made confidently when characteristic imaging findings are seen in a patient with a severe coagulation disorder.

Therefore, it is vital that the radiologist make the diagnosis of pseudotumor based on patient history and imaging findings.

Percutaneous drainage or biopsy of a pseudotumor is contraindicated due to the high incidence of complications, including life-threatening bleeding, fistula formation, and infection. Therapy for hemophilic pseudotumor aims at preserving function; however, no standard therapy exists. It includes conservative therapy with immobilization and clotting factor replacement, whereas surgical management yields the best results for pseudotumors that have been present for years and/or for those cases refractory to conservative measures .

In summary, therapy for hemophilic pseudotumor should be selected on an individual basis.