Hemophilia Pseudotumor

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A 38 year old hemophilic male, presented with enlarging right thigh swelling from 6 years with a fractured femur (Figure 1). He received multiple blood transfusions and Factor VIII. His brother and nephews also have the disease. Examination confirmed a markedly enlarged thigh with 2 oozing masses, larger one of 33x18 inches. His factor VIII level was <2%.

X-ray right hip and femur (Figure 2) showed osteopenia, expansile osteolytic lesions, cortical thinning, and extensive soft tissue mass, scalloped extrinsic erosion of shaft with radiating trabeculae, suggestive of Hemophilic Pseudotumour which were confirmed by MRI (Figures 3, 4).

Hemophilic pseudotumor or “blood cysts”, described by Starker¹ are a rare complication of Hemophilia, observed in only 1-2% of patients with severe disease,² consisting of extravasated clotted blood surrounded by a fibrous capsule. It affects bones, lungs, and abdomen and stomach wall. Morbidity occurs by compression on surrounding structures including bone destruction, muscle and skin necrosis. Complications include perforation, abscess, fistula, pathologic fractures, compartment syndromes, joint contractures, vascular erosions and distortion of natural anatomy.

CT or MRI is indicated as an imaging modality. There is little consensus regarding management strategies. A conservative approach includes factor replacement and immobilization. Cyst fluid aspiration is contraindicated. Surgery is recommended early; however due to risk of uncontrollable hemorrhage radiotherapy is a viable alternative which induces injury to blood vessels and fibrosis of cellular component.

References


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