



UNIVERSITY of
ROCHESTER
MEDICAL CENTER

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DEPARTMENT OF IMAGING SCIENCES

Imaging Sciences Interesting Cases

CASE 139

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CLINICAL PRESENTATION: Patient is a 42-year-old African-American male with a life long history of severe hemophilia A, now presenting with recurrent elbow and shoulder pains accompanied by restricted range of motion.

IMAGING FINDINGS: Large subchondral cyst formation, widened trochlear and radial notch, and overgrowth with flattening of the radial head (best seen on the lateral view). There is also loss of the joint space.





Figure 1A&B.

DIAGNOSIS: Hemophilia Arthropathy

DISCUSSION: Hemophilia is a disorder of blood clotting that is inherited as a gender linked recessive trait. It is estimated to affect approximately 1 in 10,000 males. Hemophilia A is caused by the absence, deficiency or defective functioning of plasma coagulation factor VIII. Hemophilia B (Christmas disease) is caused by an abnormality of factor IX. The function of factor VIII is to accelerate the rate of cleavage of factor X by activated factor IX in the multienzymatic reaction leading to coagulation. Minor trauma or seemingly spontaneous bleeding can cause a hemarthrosis in patients with hemophilia and recurrent hemarthrosis can result in early arthropathy. These changes may mimic rheumatoid arthritis. The elbow is the predominant site of arthropathic involvement in the upper limb of patients with hemophilia. The elbow is involved in 25% to 87% of cases, but rarely is it the only joint affected. Chronic hemophilic synovitis of the elbow joint often leads to enlargement of the radial head. The margins of this hypertrophic radial head become rough and irregular, impinging against the proximal ulnar facet. By virtue of its increased size and irregular shape, the radial head acts as a mechanical block to forearm rotation, generating synovial impingement,

hemorrhage, and pain. Limited forearm rotation can result in considerable impairment of activities of daily living especially if the involved extremity is on the dominant side.

REFERENCES:

1. Brower AC. Arthritis in Black and White. Philadelphia, PA: W.B. Saunders Company, 1988:336-337.
2. Greenspan A. Orthopedic Radiology: A Practical Approach. 3rd ed., New York: Lippincott Williams & Wilkins, 2000:497.