A Rare Case of Bleeding Disorder: Acquired Hemophilia

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I. BACKGROUND

Acquired hemophilia A, a rare bleeding disorder caused by neutralizing autoantibodies against coagulation factor VIII, occurs in both men and women without a previous history of bleeding. Patients typically present with an isolated prolonged activated partial thromboplastin time due to FVIII deficiency. Neutralizing antibodies are detected using the Nijmegen-modified Bethesda assay.

II. CASEPRESENTATION

A 33 year old female P1L1A2 presented with history of multiple joint pain and myalgia for past 8 months and compartment syndrome of left forearm for which fasciotomy was done and developed bleeding from fasciotomy site. Blood panel

revealed Low HB, Elevated APTT, ANA profile negative, APLA profile negative. In view of isolated APTT, an APTT mixing study was done which revealed prolonged Aptt- 85.3 -Inference is an INHIBITOR is present. Inhibitor Bethesda assay was done 108.8 BU (positive if > 0.6BU). Factor VIII assay DONE <1% (very low). Findings suggestive of acquired haemophilia. Patient was started on IV steroids, inj FEIBA, Inj Factor VII, IV antibiotic, PRBC. Bleeding decreased and aPTT reduced patient. Discharged with Oral steroids tapered, Oral antibiotics, and Review with inhibitor screening and factor VIII assay after 1 week.

III. DISCUSSION

The hallmark features of Acquired haemophilia are isolated elevation of APTT, bleeding is first noticed during surgery, there will be extensive ecchymosis and subcutaneous hematomas, not involving joints

as compared to congenital hemophilias. Whenever there is an isolated apt elevation do a mixing study followed by BETHESDA assay. Due to the complexity of diagnosis and treatment, immediate consultation with a hemophilia centre initiated whenever AHA is suspected.