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Case Report: A Rare Case of Haematuria

Dr. Deepika Gnanasekaran

Senior House Officer, Internal Medicine, Global Medical Center, Salem, India

Abstract: ***A 63 year old man with a past medical history of diabetes with gross haematuria for one month. He had no prior history of haematuria or mucosal bleeds and denied having any trauma. His activated partial thromboplastin time was prolonged and factor 8 levels were low. The patient was subsequently managed with steroids and prothrombin complex concentrate which has a recombinant factor VIII inhibitor bypass activity. This is a very rare cause of haematuria.***

Keywords: Haematuria, Acquired factor VIII deficiency, Autoantibody, Factor vill inhibitor bypass activity, Acquired Hemophilia.

1. Introduction

Acquired hemophilia is a rare disease that results from development of auto antibodies against factor VIII. These auto antibodies are called inhibitors and can lead to bleeding which may be severe. Acquired haemophilia is different from the congenital type as it has no genetic predisposition and hemarthrosis are generally absent. Bleeding can be superficial like mucosal or subcutaneous or can be deep like intra-articular or intra-abdominal. It usually occurs in elderly patients with comorbidities and can be associated with underlying conditions such as diabetes, infections, malignancies or autoimmune diseases; however, in about half of the cases, it can be idiopathic.

The primary goal of treatment includes hemostasis followed by eradication of the inhibitors. Management can be difficult and mortality risk remains high due to underlying comorbidities, bleeding, and complications associated with the treatment. The disease affects 1 to 1.5 per one million people annually and is likely under diagnosed or misdiagnosed [1, 2]. The diagnosisis based on isolated prolongation of APTT not corrected by PTT correction and confirmation by Bethesda assay. We report the case of an elderly male with AHA presenting a shaematuria.

2. Case Presentation

A 63 year old man presented with a one month history of painless gross haematuria without clots. He had no history of loss of weight or appetite. No history of trauma or travel to foreign countries. He was a known diabetic and hypertensive patient on regular treatment. He was not on any anticoagulants. He complained of epistaxis and bruising following insulin injection.

Clinical Findings: On examination, he was hemodynamically stable. Abdominal exam was unremarkable. Full blood count showed haemoglobin to be 9.8 g/dL. Urinalysis revealed significant haematuria. Bleeding time, clotting time, liver function tests, urine culture and urine cytology were unremarkable. Ultrasound and contrast KUB showed no calculus or mass lesions. Cystoscopy with retrograde pyelogram revealed frank haematuria from right ureteric orifice. Ureteroscopy did not reveal any pathology such as urothelial tumors, calculi except for frank haematuria.

The patient continued to have haematuria and received six units of packed RBC and six units of fresh frozen plasma. He underwent renal angiogram which was found to be normal.

His activated partial thromboplastin time (APTT) was raised but D-dimer, fibrinogen and fibrin degradation product were normal. APTT correction study revealed antibodies against factor VIII. The patient was subsequently managed with prednisolone and Factor VIII inhibitor bypass activity after which haematuria settled.

3. Discussion

Acquired hemophiliais idiopathic condition Acquired haemophilia usually presents in pregnancy and also in elderly population and is associated with malignancy [3], skin diseases, and infections. Incidence is about one case per year. It can present asepist axis, bruising throughout the body and rarely haematuria. In general, a patient with hemophilia is missing or has a low supply of; two common factors that affect blood clotting are factor VIII and factor IX. Among three main forms, there are three types of hemophilia and the patient is presented with type A.

Therapeutic Intervention: Diagnosisis based on

- Isolated prolonged APTT not corrected by PTT correction study
- Bethesda assay showing factor VIII inhibitor activity
- Reduced factor eight levels.
- Treatment based on the level of factor Ill inhibitor activity. Inhibitor assays <5-DDAVP or human porcine factor VIII infusion. Inhibits assay >5-activated prothrombin complete concentrate (FEIBA) or recombinant factor VII is needed [4]. Corticosteroids, cytotoxic drugs such as cyclophosphamide and high dose intravenous immunoglobulin can be used alone or in combination to eradicate the autoantibodies [5].

4. Conclusion

If elderly man presents with haematuriaand there is finding of isolated APTT elevation one should have a high index of suspicion for acquired haemophilia especially when all other urological investigations were normal.

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