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An Interesting Case of Upper GI Bleeding

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Abstract

Introduction

Von Willebrand disease is the most common inherited bleeding disorder. It is of three types type 1, type 2, type 3 with type 2 having 4 subtypes 2A, 2B, 2M, 2N. Type 1 is the most common with 80% of cases. There is decrease in VWF protein, VWF levels and Factor VIII levels in type 1.

Acquired VWD is rare and occurs in patients with underlying lympho - proliferative disease including MGUS, multiple myeloma and waldenstormmacroglobulinemia.

Case Report

A 60 - year old male presented to medicine opd with complaints of easy fatigability, black colored stools ,giddiness since 8 days associated with breathlessness on exertion . Yellowish discoloration of skin, eyes, urine since 4 days. Patient had history of recurrent epistaxis and bleeding tendencies which could not be controlled and had received multiple blood transfusions in the past 20-25 yrs. Patient had decreased sleep and appetite and was chronic Tadi drinker for 30 years and consumes 4-5 bottles per day. Patient father

had history of consanguineous marriage. Patient elder brothers had similar complaints of bleeding tendencies. On examination he was a febrile, pulse of 110bpm,blood pressure of 100/70mmhg. On physical examination pallor, icterus and grade 1 clubbing was present. Systemic examination was normal.

On routine examination: Hb was 3.8 g/dl, TLC was 5600, Platelet was 3,86,000, MCV 89.1,RDW 20.70. Retic count 4.00%, Peripheral blood smear showed nucleated rbc's, moderate anisocytosis, mild poikilocytosis, few tear drop cells. Blood group B positive, LFTs, RFTs, SE , FLP ,urine r/m were normal. VIT B12 was 169 (reduced), iron studies : ferritin 10.36 and iron 21 (reduced), TIBC 309 (normal), Transferrin saturation 6.8% (reduced) , Stool occult blood positive, prothrombin time 16.30 sec, INR 1.38,APTT 50.30sec, bleeding time 1min 16 sec, clotting time 4min 40 sec, viral markers negative.

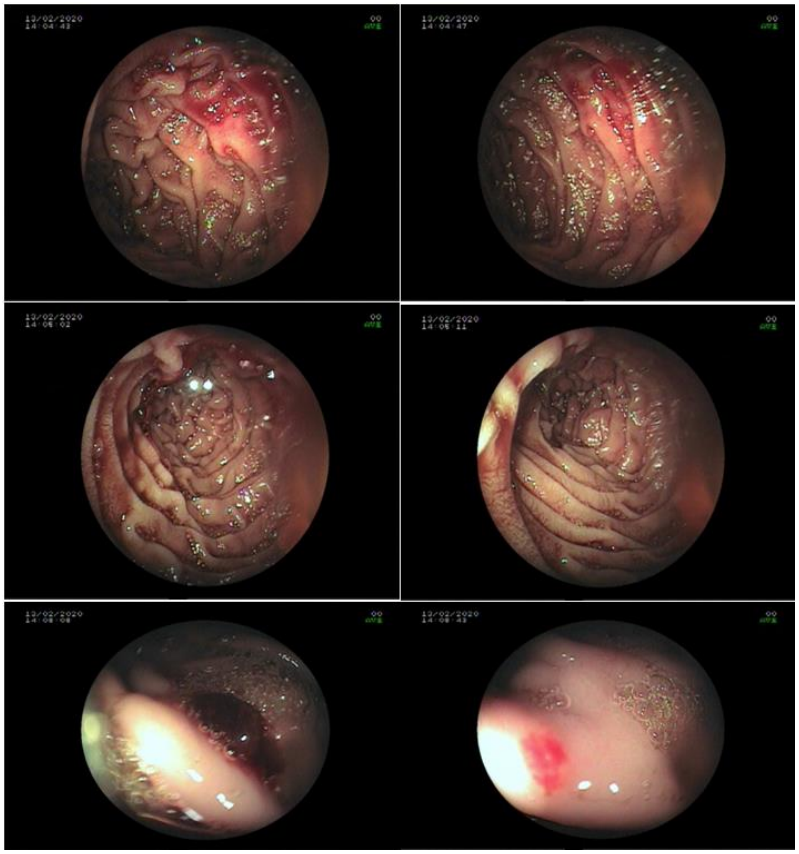
Bone marrow aspiration and biopsy report showed erythroid hyperplasia with dimorphic maturation. Von willebrand factor antigen was 3% (normal : 48-241),coombs test negative, Factor VIII activity 5.20% (normal 50-150%).

Ultrasoundabdo - pelvis was showed heterogenousechotexture of liver .Liver elastography suggestive of chronic liver parenchymal disease F4 Stage and 2d echo had mild concentric left ventricular hyperytrophy.

Gastroscopy showed few erosions with adhesive clots seen in stomach and few erosions with active ooze seen in D2 part of duodenum. (Figure 1)

Patient was diagnosed as Von willibrand disease with Dimorphic anemia . Patient was treated with with blood transfusions, cryoprecipitate and haematinic supplements given.

Figure 1



Discussion

VWD is the most commonly inherited bleeding disorder. It was first described in Aland Islands by Erik von Willebrand. The disease prevalence is about only 1%. More often it is detected in women. The disease may be severe in people with 'O' blood group[9].

It occurs as a result of decrease in plasma levels or defect in von Willebrand factor which is a large multimeric glycoprotein. Monomers of this glycoprotein undergo N-glycosylation to form dimers which get arranged to give multimers. Binding with plasma proteins (especially factor VIII) is the main function of von Willebrand factor[1].

The disease is of two types: Inherited and acquired . Inherited are of three major types. They are type 1, type 2, and type 3; in which type 2 is sub-divided into 2A, 2B, 2M, 2N. Type 1 is more prevalent than all other types. Mucocutaneous bleeding is mild in type 1 and mild to moderate in types 2A, 2B, and 2M. Type 2N is similar to haemophilia.

Type 1 is seen in 60%-80% of the cases[7], Type 2 in 20-30%, Type 3 in less than 5% of all the cases. Acquired VWD occurs most often in individuals of 40 years with no prior bleeding history.

The pathophysiology of each type is dependent on the qualitative or quantitative defects in von Willebrand factor. The diagnosis is based on von Willebrand factor antigen, von Willebrand factor activity assay, FVIII coagulant activity and some other additional tests[1].

Desmopressin is a synthetic derivative of vasopressin. It is chemically 1-desamino-8-D-arginine vasopressin. Through V2 receptors, it stimulates the release of VWF from endothelial cells. DDAVP increases the plasma concentration of VWF through cyclic AMP-mediated release of VWF from endothelial cell Weibel-Palade bodies[12] . The standard dose of DDAVP is 0.3 mg/kg intravenously in 30-50 ml of normal saline over 30 min [13]. Nasal dose contains 0.1 ml of a 1.5 mg/ml solution. Dose is one puff for those whose body weight is <50 kg and two puff for those who are >50 kg weight[14]. It is not indicated for type 2B VWD as there was fall in the platelet count after its use[14]. It is not clinical use in type 3. Side effects include hyper or hypotension, headache or gastrointestinal upset, facial flushing[13]

Humate-P® and Alphanate SD/HT® are the plasma-derived concentrates to replace VWF[14]

References

1. Von Willebrand Disease: An OverviewK. PavaniBharati* and U. Ram Prashanth Author information Article notes Copyright and License information Disclaimer Received 2010 Jul 13; Revised 2011 Jan 10; Accepted 2011 Jan 15. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3224412/#ref71
2. National heart, lung, and blood institute diseases and conditions index. vonWillebrand disease. [Last accessed on 2010 May 14]. Available from: http://www.nhlbi.nih.gov/health/dci/Diseases/vWD/vWD_All.html .
3. Sadler JE, Mannucci PM, Berntorp E, Bochkov N, Boulyjenkov V, Ginsburg D, et al. Impact, diagnosis and treatment of von Willebrand disease. ThrombHaemost. 2000;84:160–74. [PubMed] [Google Scholar]
4. Sadler JE, Gralnick HR. Commentary: A new classification for von Willebrand disease. Blood. 1994;84:676–9. [PubMed] [Google Scholar]
5. Sadler JE. A revised classification of von Willebranddisease.For the subcommittee on von

Willebrand Factor of the scientific and standardization committee of the International society on thrombosis and haemostasis. ThrombHaemost. 1994;71:520–5 [PubMed] [Google Scholar]

6. Favaloro EJ, Soltani S, McDonald J. Potential Laboratory Misdiagnosis of Haemophilia and von Willebrand disorder owing to cold activation of blood samples for testing. Am J ClinPathol. 2004;122:686–92. [PubMed] [Google Scholar]
7. von Willebrand EA. Hereditary pseudohaemophilia [English translation] Haemophilia. 1999;5:223–31. [PubMed] [Google Scholar]
8. Sadler JE. Biochemistry and genetics of von Willebrand factor. Annu Rev Biochem. 1998;67:395–424. [PubMed] [Google Scholar]
9. Levy GG, Motto DG, Ginsburg D. ADAMTS 13 turns 3. Blood. 2005;106:11–7. [PubMed] [Google Scholar]
10. Anonymous molecular basis of von Willebrand disease and its clinical implications. Haematologica. 2004;89:1036. [PubMed] [Google Scholar]
11. Goodeve AC, James P. Von Willebrand Disease. [Last accessed on 2010 June 14]. Available from: http://www.ncbi.nlm.nih.gov/bookshelf/br.fcgi?book=geneandpart=vonwillebrand .
12. Kaufmann JE, Oksche A, Wollheim CB, Gunther G, Rosenthal W, Vischer UM. Vasopressin-induced von Willebrand factor secretion from endothelial cells involves V2 receptors and cAMP. J Clin Invest. 2000;106:107–16. [PMC free article] [PubMed] [Google Scholar].
13. Mannucci PM, Ruggeri ZM, Pareti FI, Capitanio A. 1-Deamino-8-d-arginine vasopressin: A new pharmacological approach to the management of haemophilia and von Willebrand's disease. Lancet. 1977;1:869–72. [PubMed] [Google Scholar]
14. National Heart, Lung, and Blood Institute, U.S. Department of Health and Human services. The diagnosis, evaluation, and management of von Willebrand disease. [Last accessed on 2010 July 5]. Availablefrom: http://www.nhlbi.nih.gov/guidelines/vwd/4_managementofvwd.htm