International Journal of Advanced Research and Review

www.ijarr.in

HEYDE'S SYNDROME: A CASE REPORT

R.Rada[1], S.Tazi[1], H. Yahyaoui [1 2], M. Ait Ameur [1 2], M. Chakour [1 2]

[1]Laboratory of Hematology of Avicenne military hospital, Marrakesh
[2]Faculty of medicine and pharmacy, Cadi Ayyad University, Marrakesh, Morocco

ABSTRACT

Heyde syndrome is a multisystem disorder characterized by the classical triad of aortic stenosis, gastrointestinal (GI) angiodysplasias, and acquired von Willebrand syndrome. GI angiodysplasias, common in older patients, are tortuous, thin walled blood vessels seen in the mucosa or submucosa of the GI tract and are highly prone to rupture resulting in GI bleeds.

We report a case of a 60-year-old woman with recurrent gastrointestinal bleeding and aortic valve stenosis presented with bleeding from intestinal angiodysplasia, the exploration of von willebrand factor revealed willebrand disease 2A, the patient was diagnosed with Heyde's syndrome.

We report throw a case of Heyde syndrome the role of hematology laboratory in the diagnostic of this rare pathology.

Keywords: Heyde's syndrome, von Willebrand factor, diagnosis.

INTRODUCTION

First described by Edward Heyde in 1958, Heyde syndrome (HS) is a multisystem disorder characterized by the classical triad of aortic stenosis (AS), angiodysplasia of the gastrointestinal (GI) tract, and the presence of an acquired von Willebrand syndrome (von Willebrand syndrome type 2A, abbreviated here vWS-2A).[1,2]

The exact incidence and prevalence of the disease is unknown, but it is commonly seen in older individuals [3].

Management of Heyde syndrome involves replacement of the stenotic aortic valve, but valve replacement requires antiplatelet and/or anticoagulant therapy, which may be contraindicated in the setting of severe bleeding. [4]

We report throw a case of Heyde syndrome the role of hematology laboratory in the diagnostic of this rare pathology.

CASE REPORT

The patient was male, 60 years old, with a medical history of chronic anemia and valvulopathy on dilated heart disease and hypertension under treatment, the patient was a chronic smoker who had weaned himself off smoking 1 year ago.

No family history of a bleeding syndrome. The patient was admitted for an anemic syndrome with rectal bleeding. The patient received a hemostatic treatment with a fibroscopy which showed active rectal bleeding with a gastric tumor.

The bleeding time was prolonged, the willebrand factor immunoassay (vWFAg) was 302% and the measurement of ristocetin cofactor activity (WF-RCo) was 13% with a ratio of vWFCo to vWFAg = 4.2% (normal value >70%) which is in favor of a type 2A willebrand disease (by default of multimerization of willebrand factor).

DISCUSSION

Heyd4s syndrome is characterized by the presence of a classical triad that includes as, acquired coagulopathy, or AVWS, and the presence of angiodysplasia in the GI tract.[5] The pathogenesis of AVWS is believed to be secondary to the increased sheer stress forces on the blood as it flows through a stenotic valve. These sheerforces induce significant conformational change in the high molecular-weight (HMW) von Willebrand factors (vWF) subsequently exposing the A2 domain of vWF, which can be cleaved by ADAMST13, a well-known plasma protease.[6]

As a result, the HMW vWF multimers are significantly smaller in size and less competent for hemostasis when compared with the larger vWF multimers.[7] Due to increased interactions between the vWF multimers and the platelets, there is increased degradation and clearance of these vWF.[8]In our case the clinical triad was complete.

It is important to consider the possible presence of Heyde syndrome if metallic AVR is being considered, as there is a need for lifelong anticoagulation subsequently. In vWS-2A, routine screening tests for vWS are usually normal. The gold standard is gel electrophoresis of vWF [9]. vWS-2A is characterised by absence of large vWF multimers seen on SDS-agarose electrophoresis [10]. The sensitivity of various tests for vWS-2A has been ranked as follows: gel electrophoresis (most sensitive), PFA-100 closure time, skin bleeding time, vWFristocetin cofactor activity and vWFantigen level (least sensitive) [2].

In our study, the diagnostic approach to Willebrand disease is based on the immunoassay of willebrand factor (vWFAg) and the measurement of ristocetin cofactor activity (WF-RCo) with the measurement of the vWFCo/ vWFAg ratio;

Although aortic valve replacement is the most preferred therapy for patients with HS, providers must be cautious and due consideration should be excised for the prescription of antiplatelet and oral anticoagulant agents as they may increase the risk of GI bleeding.[2] Many elderly patientsmay be unfit for AVR or may refusesurgery. Conservative management includes oral iron supplements but regular blood transfusions may be necessary. Combined oestrogen and progesterone have been used to reduce bleeding from angiodysplasia, although the mechanism of action is not understood [2]. In serious cases this could halve transfusion requirements [10]. In patients with severe recurrent bleeding, endoscopy with laser therapy may be an option. In these circumstances treatment with octreotide may be considered [11].

CONCLUSION

Heyde syndrome is a complex disorder, resulting from interactions between aortic stenosis, intestinal angiodysplasia and acquired vWS-2A.
In vWS-2A, routine screening tests for vWS are usually normal. The gold standard is gel electrophoresis of vWF.
Early diagnosis and appropriate treatment of Heyde syndrome is essential but requires teamwork and liaison between different specialities.

REFERENCES

1. Theis SR, Turner SD. Heyde Syndrome. StatPearls Publishing; 2020.

2. Hudzik B, Wilczek K, Gasior M. Heyde syndrome: gastrointestinal bleeding and aortic stenosis. CMAJ. 2016;188:135-138
3. Dushyant Singh Dahiya et al.Heyde Syndrome: An Unusual Cause ofGastrointestinal BleedingJournal of Investigative Medicine HighImpact Case ReportsVolume 9: 1–4https://doi.org/10.1177/2324709621997279
4. Andrew B. Song et al. Systemic bevacizumab for refractory bleeding and transfusion-dependent anemia in Heyde syndrome. blood advances , 12 OCTOBER 2021 • VOLUME 5, NUMBER 19
3. Heyde E. Gastrointestinal bleeding in aortic stenosis. N Eng J Med. 1958;259:196.
5. Loscalzo J. From clinical observation to mechanism—Heyde'ssyndrome. N Eng J Med. 2012;367:1954-1956.
6. Warkentin TE, Moore JC, Anand SS, et al. Gastrointestinal bleeding, angiodysplasia, cardiovascular disease, and acquired von Willebrand syndrome. Transfus Med Rev. 2003;17:272-286.
7. Pate GE, ChandavimolM, Naiman SC et al. Heyde's syndrome: a review. J Heart Valve Dis 2004; 13: 701–12.
8. Gill JC, Wilson AD, Endres-Brooks J et al. Loss of the largest von Willebrand factor multimers from the plasma of patients with congenital cardiac defects. Blood 1986; 67: 758–61.
9. Warkentin TE, Moore JC, Anand SS et al. Gastrointestinal bleeding, angiodysplasia, cardiovascular disease, and acquired von Willebrand syndrome. Transfus Med Rev 2003; 17: 272–86.
10.Henne S, Denzer U, Seitz U et al. Recurrent gastrointestinal bleeding and aortic valve stenosis (Heyde syndrome): need for valve replacement? Z Gastroenterol 2007; 45: 245–9.
11. Morishima A, Marui A, Shimamoto T et al. Successful aortic valve replacement for Heyde syndrome with confirmed hematologic recovery. Ann ThoracSurg 2007; 83: 287–8.