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Obscure cause of gastrointestinal bleeding hidden in heart: Heyde syndrome

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Abstract
Heyde syndrome is a triad of gastrointestinal bleeding, aortic stenosis, and angiodysplasia of the colon. It is a rare cause of gastrointestinal bleeding. Patients with Heyde syndrome are assumed to be at increased risk for gastrointestinal (GI) bleeding via arteriovenous malformation (AVM) because of acquired deficiency of Von Willebrand Factor (VWF) resulting from mechanical destruction of Von Willebrand Factor multimers as they pass turbulently through the narrowed aortic valve. This is most commonly seen in the elderly age group in which recurrent bleeding can be fatal if not treated aggressively and can deteriorate the quality of life due to severe anemia, recurrent hospitalization, and multiple blood transfusions without absolute cure due to lack of a definite diagnosis. Our patient was admitted to the hospital multiple times and received multiple blood transfusions but the diagnosis was missed. Once the diagnosis was established (aortic stenosis as a cause of gastrointestinal bleeding), the patient was cured with no further bleeding episodes. Hence clinicians need to have a high degree of suspicion of Heyde syndrome while treating gastrointestinal bleeding, especially if no cause of bleeding is found. Till date only a few cases have been reported, hence reporting of this case may enlighten the clinicians about this rare diagnosis and its management.

Keywords: arteriovenous malformation; gastro-intestinal bleeding; heyde syndrome; aortic stenosis; von willebr and factor

Introduction
Heyde syndrome is the association between gastrointestinal (GI) bleeding from arteriovenous malformation (AVM) and aortic stenosis. Von Willebrand factor (VWF) is assumed to be critical in the pathway for adequate (physiologic) suppression of angiogenesis. Patients with Heyde syndrome are assumed to be at increased risk for GI bleeding via AVM formation because of acquired von Willebrand syndrome resulting from mechanical destruction of von Willebrand multimers as they pass turbulently through the narrowed aortic valve [1]. It is a rare but potentially fatal condition if not managed appropriately. Thus, it is prudent that physicians are more aware about Heyde syndrome so that it can be identified in a timely manner and managed appropriately [2].

Case report
A 71 years old female presented to the emergency department with the complaint of passing black-coloured stool for the last one week associated with generalized weakness and easy fatigability. She was a known case of hypertension (HTN), hypothyroidism, and diabetes mellitus (DM) and was on regular medication. Her prior medical history revealed multiple hospital admissions due to upper gastrointestinal bleeding managed with blood transfusions and supportive medication. Attained menopause 20 years ago. There was no history of smoking, alcohol intake, or illicit drug use. On examination, the patient appeared lethargic, with a blood pressure of 101/70 mm of Hg, a pulse of 110 beats/minute, a respiratory rate of 20 breaths/minute, and oxygen saturation of 92% at room air. She had conjunctival pallor. Cardiovascular examination revealed a systolic ejection murmur at right upper sternal border and a systolic murmur at apex. Laboratory tests showed a WBC count of 5410 /cumm, haemoglobin-5.10 g/dL, and a platelet count of 2,12,000/cumm, HbA1c-9.3, serum iron 13 mcg/dL, peripheral blood smear revealed normocytic hypochromic RBCs. Kidney, thyroid, and liver function tests were within the normal range. Echocardiography revealed severe aortic valve stenosis with thickened and calcified aortic valve with an aortic valve area of 0.6 cm^2 , mean gradient across aortic valve of 53 mm Hg and LVEF of 60% (Figure 1). Stool occult blood was positive. Upper GI endoscopy showed normal findings. Ultrasonography (USG) of abdomen was suggestive of mild fatty liver. Colonoscopy was suggestive of colonic angiodysplasia in the transverse colon (Figure 2) and descending colon (Figure 3).
The patient was treated with intravenous administration of the proton pump inhibitor, oral iron supplement as well as four units of blood transfusion along with other supportive medications. Based on her history of chronic GI bleeding, anemia, and aortic stenosis and colonic angiodysplasia she was diagnosed with Heyde syndrome.

Surgical management for aortic valve replacement was advised. The patient underwent a minimally invasive aortic valve replacement. At present, nine months after the procedure, the patient has been asymptomatic with no further episodes of bleeding.

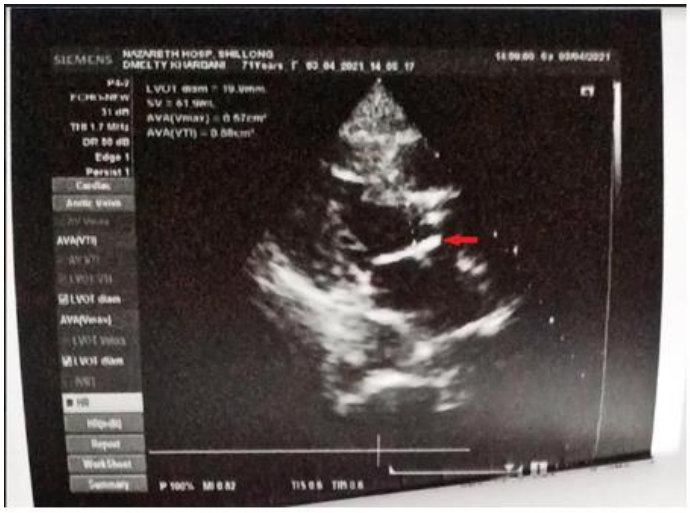

Fig 1: Parasternal long-axis view of a two-dimensional echocardiogram, showing a calcified and stenotic aortic valve (red arrow)

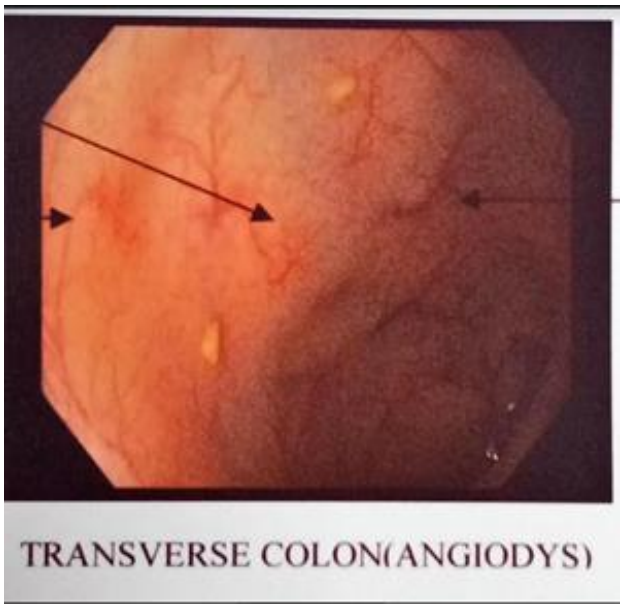

Fig 2: Angiodysplasia in the transverse colon (black arrows)

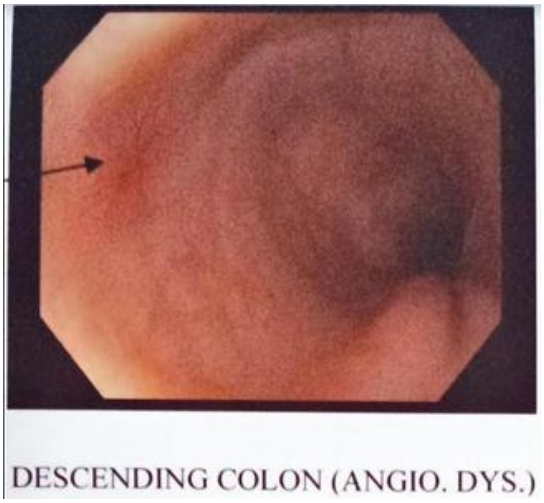

Fig 3: Angiodysplasia in the descending colon (black arrow)

Discussion

Earlier, Heyde EC *et al* [3]. and then Schwartz BM *et al*. were the first to report a possible association between AS and bleeding from angiodysplasia in 1958. This was called 'Heyde's Syndrome' after the name of author Edward C. Heyde.

The recognition of Heyde syndrome is important for patient management, to screen for, and detect an early stage aortic valve stenosis in patients with GI bleeding or occult GI bleeding. In patients with aortic valve stenosis (which may help the early diagnosis of potentially harmful conditions) [3]. Most importantly, it has been reported that aortic valve replacement may have a dual treatment effect in these patients, that is, resolution of the altered left ventricular outflow hemodynamics and of the intestinal bleeding [4].

The proposed pathophysiologic explanation involves a high shear stress around the stenotic aortic valve which increases the breakdown of high-molecular-weight multimers of von Willebrand factor, thus causing a coagulopathy (type IIA von Willebrand syndrome) which facilitates bleeding in predisposed areas, such as the vascular malformations of angiodysplasia or other bleeding-prone lesions [4].

In this case with GI bleeding of many years was found to have severe aortic stenosis and angiodysplastic lesions in the colon, diagnosed as a case of Heyde syndrome. The case was managed with aortic valve replacement leading to no further episodes of GI bleed. This is a rare diagnosis, which exists more frequently than diagnosed. This is a curable cause of GI bleeding, which can be cured and can reduce morbidity, mortality, hospital admissions, and the need for blood transfusions if diagnosed early.

The evidence that AS is the root cause of coagulopathy in HS is compelling, with 5% to 20% of these patients manifesting recurrent bleeding with severity depending on the gradients across a stenotic AV. VWS-2A arises from degradation of VWF multimers by shear stress across the AV, resulting in coagulopathy. These multimers are required to maintain hemostasis in high-flow conditions, such as in intestinal arteriovenous malformations in patients with AS. The pathogenesis of angiodysplasia could be due to low-grade chronic hypoxia that stimulates sympathetic vasodilation reflex progressing to fixed smooth muscle relaxation and true ectasia. These could also be due to mucosal hypoxia caused by cholesterol emboli or by the altered pulse waveform secondary to AS [5].

Conclusion

Gastro-intestinal bleeding of unknown cause is a common diagnosis in the hospital setting. Heyde syndrome should be kept in the differential diagnosis in such cases for early diagnosis and management. This is most commonly seen in the elderly age group and recurrent bleeding can be fatal if not treated aggressively and can deteriorate the quality of life due to severe anemia, recurrent hospitalization, and multiple blood transfusions without absolute cure in absence of a diagnosis. This patient was admitted to the hospital multiple times and received multiple blood transfusions but the diagnosis was missed. Once the diagnosis was established, patient was cured with no further bleeding episodes. Hence clinicians need to have a high degree of suspicion of Heyde syndrome while treating GI bleed, especially if no cause of bleeding is found.

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