CASE REPORT

New onset Heyde’s syndrome presenting after total aortic valve replacement

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Abstract

Heyde’s syndrome is a well-documented entity in the medical literature that can result in life-threatening gastrointestinal (GI) bleeding. It is a syndrome that is characterized by the combination of GI angioectasias and aortic stenosis. In most cases, the GI angioectasias resolve entirely with correction of the stenotic valve by way of total aortic valve replacement (TAVR). Rarely will cases recur after TAVR. Our case consists of an 84-year-old woman who presented with three new gastric angioectasias several months after undergoing TAVR. This is an exceedingly rare presentation of Heyde’s syndrome. To our knowledge this is the first such case reported in the medical literature.

INTRODUCTION

First described in 1958 by Dr Edward C Heyde, Heyde’s syndrome is a rare cause of life-threatening gastrointestinal (GI) hemorrhage associated with aortic stenosis [1]. The mechanism of hemorrhage in this entity occurs secondary to GI angioectasias, which are also known as angiodysplasias. Current data suggests that up to 80% of cases occur in the cecum and ascending colon. The descending colon follows as the third most common location [1]. This makes the rarity of our case twofold in which Heyde’s syndrome presented with the formation of new GI angioectasias after TAVR in an unusual location (gastric body).

CASE REPORT

The patient is an 84-year-old woman with a medical history significant for aortic stenosis, hypertension, hyperlipidemia and asthma who presented to our institution’s emergency department after noticing dark stools for 3 days associated with light-headedness. Of note the patient had undergone total aortic valve replacement (TAVR) 3 months prior to presentation at another institution. Records were obtained and showed that her hospital course was complicated by deep venous thrombosis and an ischemic stroke. Also during that admission an esophagogastroduodenoscopy (EGD) and colonoscopy were done showing erosive gastropathy and diverticulosis respectively. She was...
later discharged on dual anti-platelet therapy (Aspirin and Plavix) to complete 6 months. On presentation to our institution she reported only being compliant with Plavix. Notable lab work during her admission included hemoglobin of 5.1 gm/dL, hematocrit of 16.2%, platelets of 152 (10^3/μL), PT of 11.1, PTT of 22.3, INR of 1.1, creatinine of 1.2 mg/dL, and BUN of 62 mg/dL. An echocardiogram was also done which revealed an elevated mean aortic valve gradient of 19 mmHg consistent with mild aortic stenosis. While in the emergency department the patient was transfused 3 units of packed red blood cells and was taken for emergent EGD. The procedure was promptly done and showed three mucosal points of brisk bleeding in the gastric body (Fig. 1). Hemostasis was achieved after injection of 12 mL 1:10 000 epinephrine and 4 hemostatic clips (Fig. 2). After the procedure and transfusions, the patient’s hemoglobin remained stable at 7.4 g/dL. During her hospital course, the patients cardiologist was contacted who recommended resuming aspirin given her history of CVA and recent aortic valve replacement. The patient remained vitally stable, had no more episodes of dark stools and was safely discharged home with a follow up appointment.

**DISCUSSION**

Calculating the true prevalence of Heyde’s syndrome has been an arduous task. This is mainly attributed to the fact that both aortic stenosis and GI angioectasias are commonly found in the elderly, and can occur independently of one another, oftentimes asymptotically. In spite of this, there is an abundance of data linking these conditions to one another. One study in particular, found that patients with calcific aortic stenosis had an ~100-fold increased risk to develop GI bleeding when compared to the general population [2].

Present day the pathogenesis of Heyde’s syndrome is still a point of contention. Recent studies have postulated the destruction of von willebrand factor as the root cause. When passing through a stenosed aortic valve, vWF multimers are exposed to shear stress, which makes them more susceptible to proteolysis by the metalloproteinase ADAMTS13 [3]. This in effect, results in a form of von willebrand disease (vWD) type 2a, which in most cases manifests with mild to moderate mucocutaneous bleeding [4]. This theory however does not

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**Figure 1:** Esophagogastroduodenoscopy (EGD) showing three mucosal points of brisk bleeding in the gastric body.

**Figure 2:** Hemostasis achieved with injection of 12 mL 1:10 000 epinephrine and insertion of four hemostatic clips.
explain how aortic stenosis is able to specifically afflict the GI tract. Another theory that may clarify this association postulates that chronic hypoxia from aortic stenosis leads to sympathetic induced vasodilation and smooth muscle relaxation, which in turn leads to formation of angioectasias and subsequent bleeding [5]. Considering our patient presented with persistent GI angioectasias after TAVR, we propose that a combination of the aforementioned theories contributed to its pathogenesis. The prior resulted in an underlying coagulopathy, and the later lead to the residual delayed formation of GI angioectasias. Alternatively, there could exist another unknown pathologic process that could explain the delayed presentation of GI bleeding in our case.

Regarding diagnosis, multiple modalities can be employed to identify Heyde’s syndrome. The first step would be to confirm the presence of aortic stenosis, either through echocardiography, or cardiac catheterization. These studies would show the stenotic valve and can accurately calculate cardiac pressure parameters. This is clinically relevant, as studies have shown that higher pressure gradients in aortic stenosis are directly proportional to von willebrand factor abnormalities [6]. The second step in diagnosing Heyde’s syndrome consists of confirming the presence of GI angioectasias. This can be best accomplished through EGD or colonoscopy. In cases of severe bleeding where endoscopic visualization may be obscured, selective mesenteric angiography may be employed [7]. In our case, colonoscopy was sufficient in identifying the angioectasias despite the severity of the hemorrhage.

Given the varied organ systems involved in its development, treatment of Heyde’s syndrome can oftentimes be a challenge to the clinician. This is especially true if the underlying disease process is not initially elucidated. From a hematologic standpoint, the clinician. This is especially true if the underlying disease process is not initially elucidated. From a hematologic, standpoint, the clinician. This is especially true if the underlying disease process is not initially elucidated. From a hematologic, standpoint, the clinician. This is especially true if the underlying disease process is not initially elucidated. This is especially true if the underlying disease process is not initially elucidated. This is especially true if the underlying disease process is not initially elucidated. From a hematologic standpoint, this is especially true if the underlying disease process is not initially elucidated. From a hematologic standpoint, this is especially true if the underlying disease process is not initially elucidated. This is especially true if the underlying disease process is not initially elucidated. This is especially true if the underlying disease process is not initially elucidated. This is especially true if the underlying disease process is not initially elucidated. From a hematologic standpoint, this is especially true if the underlying disease process is not initially elucidated. From a hematologic standpoint, this is especially true if the underlying disease process is not initially elucidated. This is especially true if the underlying disease process is not initially elucidated. This is especially true if the underlying disease process is not initially elucidated. Given the varied organ systems involved in its development, treatment of Heyde’s syndrome can oftentimes be a challenge to the clinician. This is especially true if the underlying disease process is not initially elucidated. From a hematologic standpoint, the clinician.

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Given the varied organ systems involved in its development, treatment of Heyde’s syndrome can oftentimes be a challenge to the clinician. This is especially true if the underlying disease process is not initially elucidated. From a hematologic standpoint, treatment options include heat probe and bipolar probe coagulation, neodymium-doped yttrium aluminum garnet laser, and argon plasma coagulation [9]. Hemosmoclip clips have also shown success and were indeed useful in achieving hemostasis in our patient. Despite the effectiveness of these measures, they have not shown long-term success as many cases have the tendency to re-bleed. Invariably, the only treatment option with proven, lasting success is TAVR. A recent retrospective chart review gathered 57 patients diagnosed with Heyde’s syndrome that subsequently underwent TAVR. Of those patients, 45 had no recurrence of bleeding spanning up to 15 years after undergoing TAVR [10]. Notwithstanding this strong evidence, our patient presented with new angioectasias after undergoing TAVR. This underscores the importance of endoscopic interventions to palliate bleeding, prevent transfusion dependence and reduce complications of severe anemia.

In conclusion, Heyde’s syndrome is a diverse pathology that encompasses various organ systems. With potentially life-threatening GI bleeding, early diagnosis and treatment with TAVR becomes essential. Although rare, the development of new onset angioectasias after undergoing TAVR is an important possibility to consider. Because of this, we recommend continued surveillance of Heyde’s syndrome patients for at least 6 months after TAVR. If present, endoscopic measures should be employed to palliate GI bleeding.

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CONFLICT OF INTEREST STATEMENT

None of the authors have any financial conflicts of interest.

ETHICAL APPROVAL

This case report was approved by the institute’s Institutional Review Board as per its policy.

CONSENT

Consent for participation was obtained from this patient.

GUARANTOR

Eric Omar Then, MD.

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