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Case Report

A Case of Gastrointestinal Bleeding Associated to Aortic Valve Stenosis

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ABSTRACT

The relationship between aortic valve stenosis and gastrointestinal bleeding caused by arteriovenous malformations, known as Heyde's syndrome (HS), is poorly understood. Recently, acquired type 2A von Willebrand syndrome was shown to be the most likely etiological mechanism of anaemia. We describe a 73-year-old female admitted with weakness of lower extremities. Her history was hypertension, type 2 diabetes and lumbar spine osteoarthritis. Echocardiography showed severe aortic valve stenosis. iron deficiency anaemia with hemoglobin was 7.4 g/dL. The patient was recovery and anemia resolved after surgical aortic valve replacement.

Keywords: Aortic stenosis; Anaemia, Heyde's syndrome; Gastrointestinal bleeding

Introduction

In 1958, Dr. Edward C. Heyde first described an association between calcific aortic stenosis and gastrointestinal bleeds without a clear unifying mechanism in a letter to the editor in the New England Journal of Medicine^{1,2}. In the 1970s and 1980s, associations were made between Heyde's findings, gastrointestinal angiomyolipoma, and decreased high molecular weight multimers. Reports where interventions were aimed specifically at treating the gastrointestinal bleeding had bleeding recurrences.

However, when the aortic valve was replaced, bleeding was alleviated in the majority of cases. Interestingly, replacing the aortic valve alleviated the bleeding in >90% of cases. In the early 2000s, the syndrome was further described to include severe aortic stenosis inducing an acquired von Willebrand coagulopathy due to shear stress and gastrointestinal arteriovenous malformations (AVMs) causing moderate to significant bleeding³. An estimated 7-24% of patients with unknown origin of gastrointestinal bleeding were later found to have aortic stenosis⁴. Of patients found to have aortic stenosis with gastrointestinal bleeding, acquired von Willebrand syndrome is seen in up to 67%⁵.

Severe aortic stenosis is defined as an aortic valve area $\leq 1.0 \text{ cm}^2$, with an aortic velocity $\geq 4.0 \text{ m/s}$, and/or a mean transvalvular gradient $\geq 40 \text{ mmHg}$ ⁶. Generally a disease of the elderly, the prevalence of aortic stenosis varies from 0.2% at ages 50-59 years, to 1.3% at ages 60-69, 2.9% at ages 70-79, and 9.8% at ages 80-89⁷. The classic clinical manifestations are heart failure, syncope, and angina. These manifestations are described in the context of cardiac symptoms. Here, we discuss a case of severe aortic stenosis with gastrointestinal bleeding.

Case Presentation

This case involves a 73-year-old woman who initially presented with weakness of lower extremities and dyspnea for two weeks duration. Her history diseases were hypertension, type 2 diabetes and lumbar spine osteoarthritis. She had no significant cardiac family history with no history of sudden cardiac death. Physical examination was remarkable for a 3/6 crescendo systolic murmur heard best at the right upper sternal border. Electrocardiography showed sinus rhythm 95 bpm, left ventricular hypertrophy (Figure 1). Chest Xrays showed a large cardiac shadow (Figure 2). Transthoracic echocardiography revealed a severe aortic valve stenosis, peak velocity of 4.10

m/s across the valve and peak gradient 67 mmHg (**Figure 3**). The patient's native aortic valve area was 0.6 cm^2 ; moderate aortic valve regurgitation. Laboratory tests including routine full blood count showed moderate hypochromic microcytic anemia with Hb: 7.4g/dL, renal and liver profiles were unremarkable. Abdominal ultrasound was normal.

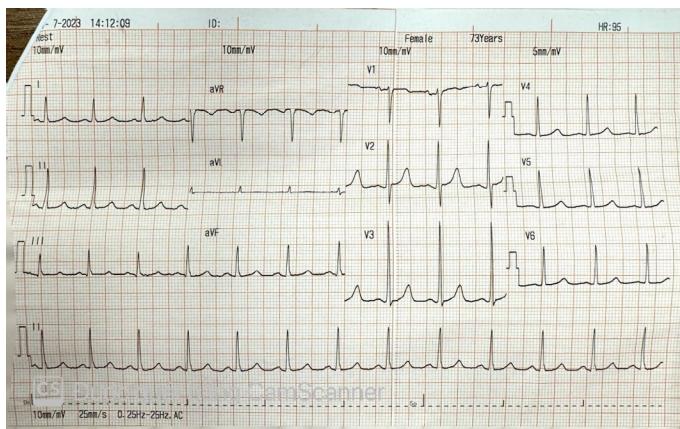


Figure 1: Electrocardiography showed sinus rhythm 95 bpm, left ventricular hypertrophy.

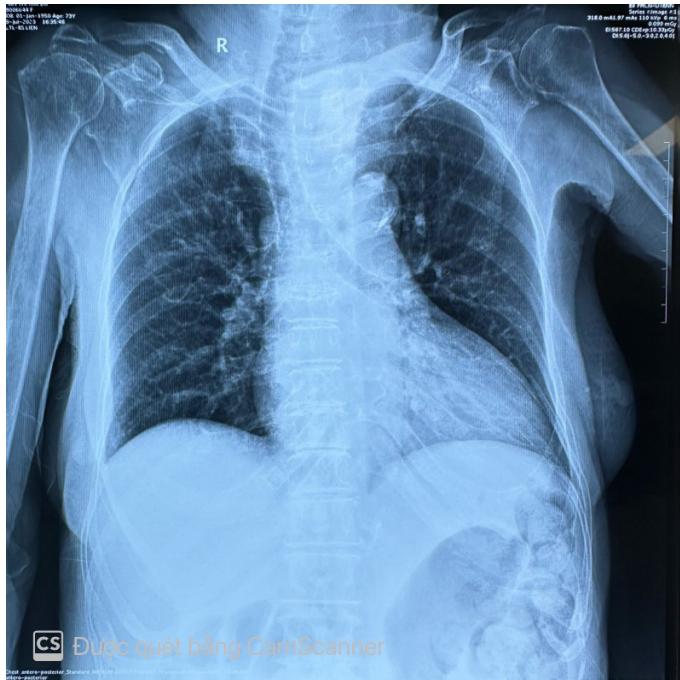


Figure 2: Chest Xrays showed a large cardiac shadow.

She electively underwent aortic valve replacement with a bioprosthetic valve. In the perioperative period, the patient had no recurrent anemia. She was discharged with no episode of recurrent gastrointestinal bleeding.

Discussion

Heyde's syndrome is named after Dr E C Heyde who in 1958 described ten cases of aortic stenosis and massive gastrointestinal bleeding for which he could discover no cause². The prevalence of the condition is unknown. In one retrospective analysis, 2.6% of patients with aortic stenosis, compared with 0.025% of a control group, had idiopathic gastrointestinal bleeding⁸. Subsequent reports have implicated gastrointestinal angiodyplasia as a possible source of the bleeding but the precise relation between aortic stenosis and angiodyplasia is unclear. A causal relation is possible, but another possibility is that they coexist as age-related phenomena.

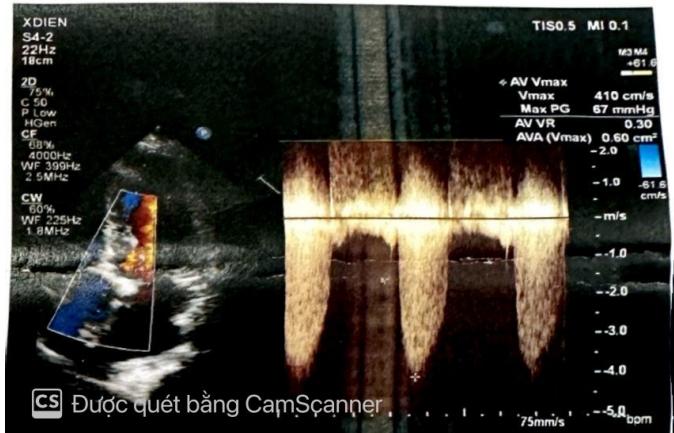


Figure 3: Continuous wave Doppler Echography revealed Vmax 4.10 m/s, maximum pressure gradient 67 mmHg, AVA 0.6 cm².

The blood vessel dilatation in gastrointestinal angiodyplasia is thought to arise from long-term obstruction within the submucosal mesenteric venous system, leading to loss of precapillary sphincters and formation of arteriovenous communications. A possible mechanism for the bleeding in Heyde's syndrome is an acquired form of von Willebrand's disease (type IIa) due to the aortic stenosis. High shear rates across the stenotic valve lead to increased consumption of high-molecular-weight multimers of von Willebrand factor (vWF). The subsequent functional deficit of vWF predisposes to bleeding. In a recent study, 21% of patients with severe aortic stenosis experienced skin or mucosal bleeding. Most of the patients had vWF abnormalities, the severity of which related to the degree of stenosis⁹. Gastrointestinal angiodyplasia may be diagnosed by upper gastrointestinal endoscopy, enteroscopy, colonoscopy or capsule endoscopy. The lesions are found most commonly in the small bowel.

For bleeding angiodyplasia, one treatment option is combined oestrogen and progesterone therapy, which can reduce transfusion requirements by up to half¹⁰. However, the possible benefits of hormone therapy should be set against cardiovascular risks. An alternative is to treat bleeding angiodyplasia at endoscopy with argon beam diathermy, but it is unknown whether this affects the natural history of the disease and risk of subsequent bleeding. Surgical options are bowel resection or aortic valve replacement. There have been several case reports of cessation of bleeding following aortic valve replacement¹¹. In a retrospective study of 91 patients with aortic stenosis and unexplained gastrointestinal bleeding King *et al.* found that bleeding ceased in 93% of patients treated by valve replacement compared with 5% of those managed by laparotomy with or without bowel resection¹².

Conclusion

Doctors need to be thoughtful and consider Heyde syndrome in patients with severe aortic stenosis and gastrointestinal bleeding secondary to angiodyplasia. Doctors should also be vigilant in patients with Heyde syndrome presenting with gastrointestinal bleeding after undergoing aortic valve replacement, as gastrointestinal bleeding might take time to resolve completely in these patients, assuming they had a successful aortic valve replacement.

Conflict of Interest

None declared.

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