CASE REPORT OPEN ACCESS

The Diagnostic Dilemma of Heyde Syndrome: A Case Report

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Heyde syndrome is a multisystem disorder characterized by the triad of aortic stenosis (AS), gastrointestinal bleeding, and acquired von Willebrand syndrome. We present the case of an 80 years old male that presented to us with a recurrent gastrointestinal bleed and syncope. Upon investigation it was found that the patient has clinically severe AS. On physical examination he had a grade 4/6 systolic murmur at the second right intercostal space radiating toward the right carotid artery. An upper GI endoscopy was planned which reported proximal jejunal AVM consistent with a Dieulafoy lesion. The history of lower gastrointestinal bleeding (angiodysplasia) and the presence of aortic stenosis were consistent with Heyde syndrome. Given the patient's age and a high EuroSCORE, the patient was referred for transcatheter aortic valve implantation. The procedure was uneventful and the patient was discharged once stable. The patient did not experience gastrointestinal bleeding in the periprocedural period.

Keywords: Heyde; Dieulafoy Lesion; Cardiology; Gastroenterology

Citation: Sohail R, Yazdaan HM, Hoor A. The Diagnostic Dilemma of Heyde Syndrome: A Case Report. THE STETHO 2024;5(2):1-4

INTRODUCTION

This medical condition was first documented by a notable physician Dr Heyde. He rerported to the New England Journal of Medicine that he found a correlation between old aged patients with aortic stenosis and Atrioventricular Malformations (AVM). Hence, the triad of AV Malformations in the GI tract, Aortic Stenosis and Von Willebrand Disease is known as the Heyde Syndrome. The exact

pathophysiology is still under investigation. The first case of Heyde syndrome was reported in 1958 [1]. The disease has high prevalence in the old ages [2] and interestingly the GI bleed did actual resolves with the replacement of the valve [3]

CASE REPORT

We present the case of an 80 years old male that presented to us with a recurrent gastrointestinal bleed and syncope. Upon investigation it was found that the patient has clinically severe AS. On physical examination he had a grade 4/6 systolic murmur at the second right intercostal space radiating toward the right carotid artery. The findings of a rectal examination (including testing for fecal occult blood) were normal. Oxygen saturation was 98% on room air. Rest of the physical exam was also normal.

Laboratory tests showed a hemoglobin of 110 (normal 130–170) g/L, platelets 190 (normal 150–400) × 109/L. Troponins and other cardiac markers were within the normal range. Electrocardiography on admission was suggestive of severe aortic stenosis, a heavily calcified aortic valve with a mean aortic valve gradient of 33 mm Hg. An upper GI endoscopy was planned which reported proximal jejunal AVM consistent with a Dieulafoy lesion. The history of lower gastrointestinal bleeding (angiodysplasia) and the presence of aortic stenosis were consistent with Heyde syndrome.

Computed Tomography (CT) scan of the chest without contrast revealed that the heart as mildly enlarged. There is calcification seen in Left Anterior Descending (LAD) artery. A treatment for the replacement or repair of the aortic valve was recommended.

Given the patient's age and a high EuroSCORE, the patient was not considered a suitable candidate for surgical aortic valve replacement and was referred for transcatheter aortic valve implantation. A Transcutaneous Aortic Valve Replacement (TAVR) was planned. Successful right transfemoral transcatheter aortic valve replacement resulting in no residual gradient or perivalvular leak. Excellent hemodynamic result appreciated. The procedure was uneventful and the patient was

discharged once stable. The patient did not experience gastrointestinal bleeding in the periprocedural period.

A Six-month follow-up was unremarkable, with no further episodes of gastrointestinal bleeding reported. The patient's ventricular function had improved.

DISCUSSION

The association between chronic gastrointestinal bleeding due to angiodysplasia and calcific aortic stenosis was first described in 1958 by Edward Heyde and has since been termed Heyde syndrome. Not until 1992 did Warkentin and colleagues elucidate the role of acquired coagulopathy (depletion of high-molecular-weight multimers of von Willebrand factor) in the pathogenesis of Heyde syndrome. [4] However, despite several years of work, the Pathophysiology of Heyde Syndrome still remains elusive. Degenerative aortic stenosis is the most common valvular heart disease in older patients and is one of the main causes of morbidity and mortality in this age group. There is an increased risk for bleeding in few of the patients due to the acquired von willebrand disease type 2, which is due to the unfolding of vWF multimers as it passes through the stenosed valve and its subsequent exposure to the ADAMTS13 enzyme which cleaves it and makes the patient susceptible to bleeding. [5]

Angiodysplasia is another age-related degenerative process, characterized by dilated, thin-walled tortuous vessels in the mucosa and submucosa of the gastrointestinal tract, including arterioles, capillaries and venules. Angiodysplasia accounts for 1%–6% of hospital admissions for gastrointestinal bleeding. It may occur throughout the gastrointestinal tract, but most particularly in the right colon and cecum. [6]

Management of Heyde syndrome often requires a multidisciplinary approach, and treatment options include medical therapy, endoscopic interventions, colon surgery and aortic valve replacement. Treatment modalities used in von Willebrand disease (e.g., desmopressin, octreotide or supplementation of factor VIII or von Willebrand factor) are usually insufficient or ineffective for acquired type IIA von Willebrand syndrome. Administration of von Willebrand factor–factor

VIII concentrate immediately before surgery should be considered for patients who have transient improvement in von Willebrand factor activity with a test dose. [7-8]

More recently, transcatheter aortic valve implantation has emerged as a feasible option for patients with severe aortic stenosis whose risk is deemed too high for surgical aortic valve replacement. This was also the approach in our case and with the TAVR, patient's symptoms improved significantly.

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