CASE REPORT OPEN ACCESS

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Heyde Syndrome: A Case Report and Review of Literature

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The triad of AV Malformations in the GI tract, Aortic Stenosis and Von Willebrand Disease is known to us as the Heyde Syndrome. We present the case of a 72 years old male patient who presented with Heyde Syndrome Secondary to Aortic Stenosis (Mean gradient of 36) with Proximal Jejunal Arteriovenous malformation-Dieulafoy lesion, seen on Push enteroscopy and capsule endoscopy. The patient had a porcelain Aorta visible on CT and MRI. A Transcutaneous Aortic Valve Replacement-TAVR was planned to replace the Aortic Valve. Edwardes S3 Prosthetic Size 26 Valve was used. A follow up Echocardiography 3 months after the surgery shows that there is no Aortic Valve stenosis. The patient also underwent a follow up Anterograde Double Balloon Enteroscopy which shows that the AVMs had diminished and are currently non bleeding. The high risk patient with Heyde Syndrome and a Porcelain Aorta recovered with the use of TAVR.

Keywords: TAVR; Heyde Syndrome; Dieulafoy Lesion; Porcelain Aorta

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INTRODUCTION

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

A 72 years old male with a Past Medical History of hyperlipidemia presented with fourth episode of GI Bleed and Syncopal episodes. A moderately severe Aortic Valve stenosis was found Echo. Patient had no Cardiac high risk factors, No DM, no CKD, no CV, no cardiac symptoms and a functional capacity ≥4 METS.

On Physical examination he had systolic crescendo decrescendo murmurs (gallops or Rubs). JVP was not raised. Rest of the physical exam was normal.

GI Endoscopy showed Proximal Jejunal Arteriovenous malformation consistent with a Dieulafoy lesion. Echocardiography showed moderately severe Aortic Valve Stenosis with mild Aortic Valve Regurgitation. There is severe Calcification. AV area is 1.25 cm square by continuity, VTI. The dimensionless index is 0.36. The peak gradient is 59mm Hg and the mean gradient is 31mm Hg. The Left Ventricular function was normal. Echocardiography Shows there is moderately severe Aortic Valve Stenosis with mild Aortic Valve Regurgitation. There is severe Calcification.

CT Chest without contrast-The heart is mildly enlarged with coarsely trileaflet aortic valve. There is calcification seen in LAD and circumflex distributions. Mild aneurysmal dilation of Ascending Thoracic Aorta.

A Cardiac Cath was advised for the indication Preoperative assessment before valvular surgery: AUC Score = 7. The result was that there are calcified coronary arteries with no obstructive disease. Treatment for the replacement or repair of Valve was recommended.

He had a full work up by Cardiology including Echo, Cardiac Cath and other relevant investigations. Due to the Porcelain Aorta a Transcutaneous Aortic Valve Replacement-TAVR (sometimes referred to as TAVI) was planned. Successful right transfemoral transcatheter aortic valve replacement using a Edwardes S3 Prosthetic Size 26 Valve resulting in no residual gradient or perivalvular leak. Excellent hemodynamic result appreciated. The surgery was uneventful and the patient was discharged after he got stable.

A follow up Echocardiography 3 months after the surgery shows that there is no Aortic Valve stenosis. The peak gradient is 25 mm Hg and the Mean gradient is 13 mm Hg. Follow up Echocardiography with no aortic valve stenosis Post TAVR. The patient also underwent a follow up Anterograde Double Balloon Enteroscopy which shows that the AVMs had significantly reduced and bleeding has resolved. The patient has fully recovered and the use of TAVR results in resolution of AVMs. GI Endoscopy Post TAVR (2 months follow up) shows significant reduction and resolution of the GI Bleed and AVMs.

DISCUSSION

Despite several years of work, the Pathophysiology of Heyde Syndrome still remains elusive. 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. [4] There has been a debate as to treat the vW disease (now called von willebrand Syndrome) or proceed with surgery. A high percentage (92%) of patients with aortic stenosis had an altered vWF function [5], further supporting the involvement of vWF targeted therapies but a very and multidimensional approach needs to be considered by the Physician in managing patients who present with such signs & symptoms. The investigations need to be thoroughly evaluated and a decision should be made. Along with the classical symptoms those who present with Risk that they care high risk for Open Heart Surgery a decision to operate should be very carefully made, this may lead to the use of TAVR as a suitable treatment option.

In this case, treatment of Aortic Stenosis with TAVR significantly diminished AV malformations and resolution of GI Bleed from the AVMs. One can hypothesize that the etiology may lie in the calcified stenosed valve and the physical alteration of a biomarker may be the cause.

Although this is a single case report, it is demonstrated that possible utility of TAVR in patients with Heyde syndrome who have poor high risk for surgical aortic valve replacement, TAVR provides a viable alternative to surgical AVR in patients who are a prohibitive risk for systemic heparinization (cardiopulmonary bypass) and may improve the GI Bleed as well.

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