A Case of Heyde Syndrome: Management and Anti-Coagulation

Muhammad Siddiqi, M.D., PGY-1, Hamza Naveed, M.D., PGY-2, Ashley Charales, M.D., PGY-2, Marloe Prince Jr., M.D.

Background

- Heyde syndrome is a rare clinical diagnosis whose clinical manifestations include Aortic Stenosis (AS), gastrointestinal bleed, angiodysplasia, and anemia.
- In patients diagnosed with severe AS, **the incidence of Heyde syndrome is 3%.**
- The pathophysiology of the disease involves **an acquired von Willebrand Factor (vWF) deficiency** secondary to AS in elderly patients **with intestinal dysplasia** which is thought to result in GI bleeding.
- In these cases, it is important to recognize the syndrome as there has been correlation between aortic valve replacement and improvement and/or resolution of Heyde syndrome.

Case

- A 71-year-old male with a history of diabetes and hyperlipidemia presents with acute blood loss anemia due to GI bleed.
- On arrival the patient was transferred to ICU due **to Hgb of 5.5 and hemodynamic instability.** After stabilization of Hgb after transfusion **of 5 units of blood**, GI proceeded with **Upper GI endoscopy revealing multiple bleeding arteriovenous malformations (AVMs) in the stomach**. Fulguration was done to stop the bleeding by argon plasma.
- After stabilization of the GI bleed, cardiology was brought on board due to **elevated troponin levels, peaking at 9.2, and a severe crescendo-decrescendo systolic murmur heard in the 2nd intercostal space.** Due to the patient's repeated hospitalizations due to GI bleeds as well as some for shortness of breath, cardiology decided to evaluate for severe aortic stenosis.
- Transthoracic echocardiogram was done and revealed **severe aortic stenosis with valve area of 0.6 cm2, mean gradient of 78 mmHg, and peak velocity of 5.52 m/s**, with no wall motion abnormalities and an ejection fraction of 55-60%.
- Aortic valve replacement was pursued due to severe stenosis with symptoms as well as likely cause of AVMs → Heyde Syndrome
- Due to age and risk factors, **patient was scheduled for TAVR 5 weeks after discharge from hospitalization**
- After TAVR, discussion on **anti-coagulation was made between cardiology, gastroenterology, and patient. Patient was started on aspirin monotherapy due to high risk GI bleeds** even though usually DAPT for valve replacement

Images

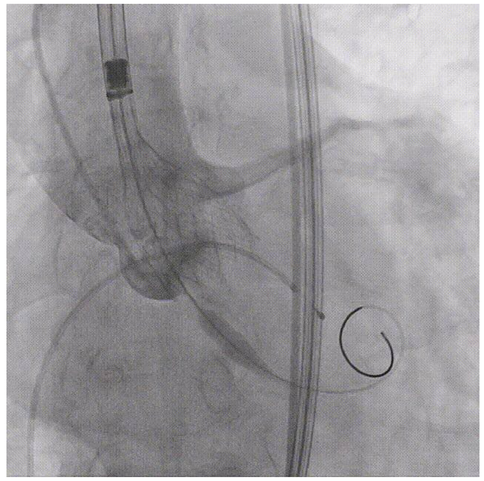

Figure 1: Deployment of Evolut 29mm Aortic Valve during TAVR procedure
医心. (n.d.). 阜外精彩案例集 | TAVR创新交流论坛:阜外医院 Evolut Pro植入1例纯局麻、清醒TAVR. 知乎专栏. https://zhuanlan.zhihu.com/p/549516224?just_published=1

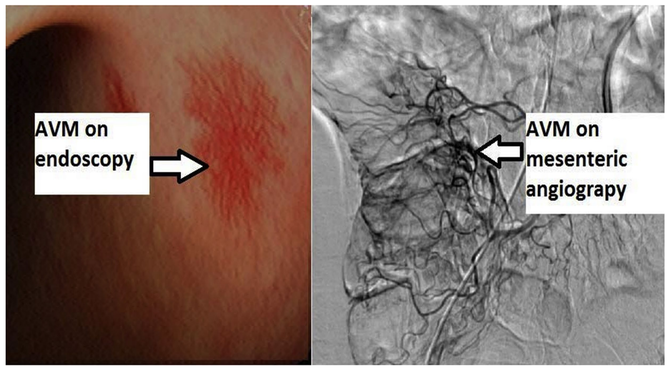

Figure 2: Endoscopy and CTA findings of bleeding AVM in mesentery. Vuddanda, V., Jazayeri, M., Turagam, M. K., Lavu, M., Parikh, V., Atkins, D., Bommana, S., Yeruva, M. R., Di Biase, L., Cheng, J., Swarup, V., Gopinathannair, R., Olyaee, M., Ivaturi, V., Natale, A., & Lakkireddy, D. (2017). Systemic octreotide therapy in prevention of gastrointestinal bleeds related to arteriovenous malformations and obscure etiology in atrial fibrillation. JACC: Clinical Electrophysiology, 3(12), 1390–1399. https://doi.org/10.1016/j.jacep.2017.04.022

Discussion

- Heyde syndrome patients can present **with life threatening GI bleeds** which will require urgent intervention but will also need to evaluate manage severe and/or symptomatic cases of aortic stenosis
- Many current studies identify **the AVMs and GI bleeds in these patients as acquired von Willebrand Factor deficiencies** due to wall shear stress due to severe aortic stenosis
- Some preliminary studies have shown that GI bleeding due to angiodysplasia have **been curable through aortic valve replacement**
- Many possibilities for adjusting protocols or guidelines for patients with aortic stenosis or new onset GI bleeds could improve patient outcomes while also making the use of healthcare resources more efficient
- Should patients with moderate to severe aortic stenosis who have not pursued valve replacement have **different screening criteria for colonoscopies?**
- Should patients with lower GI bleeds **have echocardiograms done to evaluate for aortic stenosis?**

Conclusion

- Heyde syndrome is rare, with little research done regarding it
- Many preliminary cases have shown improvement with valve replacement, should there be changes in how we pursue valve replacement in patients high risk for GI bleed
- Guidelines on screening for high risk patients could be adjusted to improve patient outcomes and introduce primary prophylaxis
- Anti-coagulation decisions on patients need to be further evaluated due to balancing bleeding risk with valve replacement

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