5-2020

The Role of Transcatheter Aortic Valve Replacement in Heyde Syndrome

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Abstract

Introduction: This is a case of a pleasant 81 year-old male who presented to the ED complaining of chest pain with activity for the past 3-4 weeks. He reports of chest pain with minimal exertion of walking across the parking lot to the car. This has been a substantial change from his baseline, he currently works as a mason. He was referred to the ED by his primary care physician after having abnormal lab result showing low hemoglobin of 7.6. During the admission he underwent work up of his chest pain and symptomatic anemia. The patient's anemia was attributed to intestinal arteriovenous malformations seen in the cecum and ascending colon during colonoscopy. He had a 2-D ECHO showing severe aortic valve stenosis. The patient was diagnosed with Heyde syndrome which is the association acquired von Willebrand deficiency with aortic stenosis and gastrointestinal angiodysplasia. Here we discuss the pathophysiology of Heyde syndrome and the role of TAVR in treatment of our patient.

Presentation

Patient is a 81 year old male with a past medical history of hypertension, hyperlipidemia, chronic kidney disease stage 2, skin cancer (diagnosed in 2014) and BPH who presented to the hospital with chief complaint of chest pain on exertion. He states that he has been having chest pain with normal daily activity for the past 3-4 weeks. The chest pain is described as localized pressure to the chest and is nonradiating in nature. He denies any associated shortness of breath with this chest pain. On presentation the pain was rated a 6/10. He notes that the chest pressure occurs after taking about 70 steps which has significantly reduced his work as a mason. The pain is relieved after several minutes of rest. More recently his chest pain required longer rest time to resolve which prompted him to visit his primary care physician. several days prior to presentation. Lab work was collected at the PCP office and he was given referral to follow with a cardiologist. Upon visit to the cardiologist's office he was scheduled for a pharmacologic stress test in 1 month and was prescribed Imdur 30mg daily. He had taken the Imdur as prescribed without significant relief. The morning of presentation he received a call from his PCP of an abnormal lab result, hemoglobin of 7.6. He denied any other complaints of fevers, chills, sweats, nausea, vomiting, headache, lightheadedness, dizziness, visual changes, palpitations, paroxysmal nocturnal dyspnea, orthopnea, shortness of breath, hemoptysis, abdominal pain, changes in bowel habits, melena, hematochezia, leg pain/cramping.

Hospital Course

During hospitalization he was noted to be anemic and have hemoglobin of 7.5 on presentation. He was transfused 2 units packed red blood cell. He underwent colonoscopy which revealed multiple AVMs in the right colon. There was no plan for capsule endoscopy. ECHO revealed severe calcific tricuspid aortic valve stenosis. He did undergo outpatient iron supplementation for iron deficiency anemia. He underwent outpatient cardiology evaluation for aortic valve replacement and later a transcatheter aortic valve replacement.

Discussion

Heyde syndrome is a multisystemic disorder characterized by the triad of aortic stenosis (AS), gastrointestinal bleeding and acquired von willebrand syndrome.

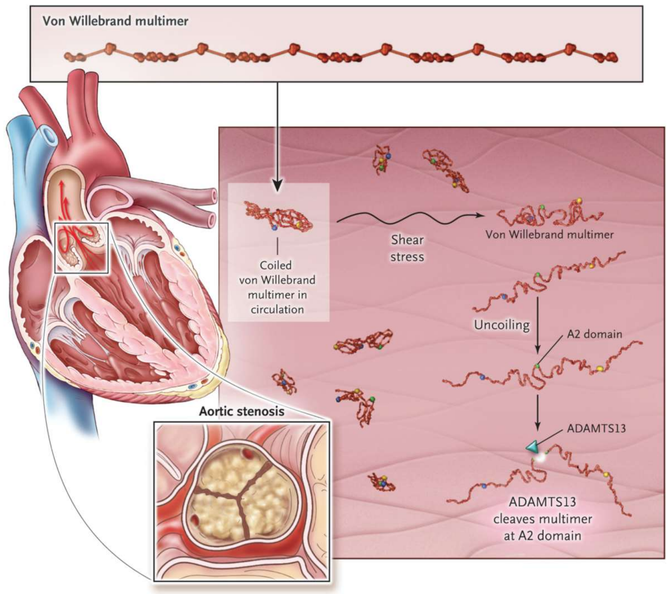
It is suspected that Heyde syndrome occurs as a result of the increased force through the stenotic aortic valve that results in cleavage of large von Willebrand multimers. The increased shear force is thought to cause transformation of the von Willebrand factor into an uncoiled form in plasma. This uncoiled von Willebrand factor is recognized by a plasma protease, ADAMTS13 which binds to the A2 domain resulting in cleavage of the von Willebrand factor. As a result, the von Willebrand multimers are reduced in size and become less able to maintain hemostasis. (11)

Angiodysplasia is thought to be common in vascular aging. It is suspected that the combination of vascular aging and the acquired von Willebrand factor deficiency from the shear force of stenotic aortic valve results in bleeding from the angiodysplastic lesions. In a study of patients diagnosed with angiodysplasia of the GI tract over 10 years, ECHO significantly correlated with aortic stenosis. (9).

The role of transcatheter aortic valve replacement (TAVR) in Heyde syndrome is the correction of the underlying AS. Of the triad of Heyde's syndrome the valvular disorder remains the component that maybe the easiest to address at this time. TAVR is a minimally invasive procedure that involves placement of a new aortic valve over the existing stenotic valve with the ultimate goal of reduction in peak gradient through the valve. A retrospective study of 91 patients with AS and chronic unexplained GI bleeding showed that bleeding ceased in 93% of patients treated with valve replacement. (10) Replacement of this valve can result in removal of one of the causes of increased shear forces on the large von Willebrand factor multimers.

Post-TAVR our patient has maintained his hemoglobin levels and has not had any further hospitalizations for symptomatic anemia.

Photos



Conformational Change and Proteolytic Cleavage of von Willebrand Factor in Aortic-Valve Stenosis (5)

Pertinent Testing

- Upon presentation EKG showed Normal sinus rhythm without any ST changes.
- He was initially started on 1 unit PRBC transfusion.
- Iron Studies: Iron 10 ug/dL, Ferritin 8 ng/mL, Iron Saturation 2%
- Stool guaiac positive
- He underwent EGD and colonoscopy. EGD showed no inflammation, ulceration, mass lesions or scalloping of the folds. Colonoscopy showed significant number of arteriovenous malformations in the right colon, unclear of AVMs in the small bowel.
- ECHO which revealed severe aortic stenosis with peak aortic velocity of 4.48 m/s and peak/mean gradient 80.1 mmHg/ 49.8 mmHg. Calculated aortic valve area 0.81 cm^2.
- TEE: LVEF 58%, severe calcific tricuspid aortic valve stenosi.
- Left heart Cardiac Catheterization: Mild nonobstructive coronary artery disease with severe aortic valve stenosis and mild pulmonary hypertension.
- von Willebrand Factor antigen pre TAVR: 187%
- von Willebrand Factor antigen post TAVR : 140%
- Post TAVR ECHO: LVEF 58%, Peak gradient 29.6 mmHg, mean gradient 14.3 mmHg

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