Annals of Cardiovascular Surgery

Case Report
Published: 08 Aug, 2022

A Case of Successful Aortic Valve Replacement for Heyde's Syndrome with Perioperative Evaluation of Multimer of Von Willebrand Factor

Shun Hiraga, Yoshihiro Hayata, Tomoaki Hirose, Ryohei Fukuba, Junichi Takemura, Rei Tonomura, Sayaka Tamada, Kazuhiro Mitani, Masaya Hanalawa and Shinya Yokoyama*

Department of Thoracic and Cardiovascular Surgery, Nara Medical University, Japan

Abstract

Heyde's syndrome is defined as a combination of aortic stenosis and recurrent gastrointestinal bleeding from angiodysplasia; it is recently known as von Willebrand disease type 2A, with loss of a large multimer of von Willebrand factor. We report a successfully treated case of Heyde's syndrome with intestinal bleeding and angiodysplasia. An 87-year-old man presented with recurrent gastrointestinal bleeding and advanced anemia. Angiodysplasia was observed in the intestine on capsule endoscopy; the diagnosis was Heyde's syndrome. Gel electrophoresis revealed the loss of large multimer of von Willebrand factor. We performed aortic valve replacement. After the operation, the large multimer of von Willebrand factor decreased in the early phase, then increased, and finally normalized. We report a case of successful aortic valve replacement for Heyde's syndrome with evaluation of the multimer of von Willebrand factor in the perioperative period.

Introduction

Heyde's syndrome was first reported in 1958 by EC Heyde, a general practitioner from Vancouver, Washington [1]. He reported on 10 patients who presented with aortic valve stenosis and severe gastrointestinal bleeding. Recently Heyde's syndrome has been defined as aortic valve stenosis with recurrent Gastrointestinal (GI) bleeding from angiodysplasia and one of the acquired von Willebrand disease subtypes. High-molecular-weight multimers of von Willebrand Factor (vWF) is deformed and decreased by shear stress due to transvalvular aortic gradients in patients' having Heyde's syndrome. Thus, GI bleeding occurs due to intestinal angiodysplasia, due to which, advanced anemia can easily occur. We report the case of an 87-year-old man with Heyde's syndrome. He developed recurrent gastrointestinal bleeding and severe anemia before the surgery. Capsule endoscopy revealed angiodysplasia in the intestine and he was diagnosed with Heyde's syndrome. Preoperative gel electrophoresis showed the loss of large multimer of vWF. We performed aortic valve replacement and postoperative GI bleeding ceased gradually. Preoperative evaluation of the multimer of vWF revealed a temporary increase in large multimer of vWF in the early postoperative phase followed by normalization. We report a case of successful treatment of Heyde's syndrome.

Case Presentation

An 87-year-old man had advanced anemia due to recurrent GI bleeding and needed repeated transfusions. He was referred to our hospital and small intestinal capsule endoscopy revealed multiple angiodysplasia in the small intestine (Figure 1). Angiodysplasia was not seen in any other lesions. He was treated with soft coagulation and clipping; however, recurrent GI bleeding was observed. A transthoracic echocardiogram showed severe aortic valve stenosis, with a peak aortic pressure gradient of 73 mmHg. Aortic valvular surface was 0.8 cm^2 and left ventricular ejection fraction was 77%. Heyde's syndrome was suspected; therefore, multimer of vWF was examined. Gel electrophoresis revealed loss of the large multimer of vWF, while laboratory findings showed anemia (hemoglobin, 8.7 g/dl), normal platelet count, and coagulation function. Factor VIII was not decreased in the preoperative evaluation. He was diagnosed with Heyde's syndrome and we performed aortic valve replacement. He was transfused with 22 units of red cell contents and 2 units of fresh frozen plasma. Under standard cardiopulmonary bypass conditions with mild hypothermia, the ascending aorta was cross-clamped. The aortic valve had 3 cusps; all were heavily calcified. The valve and calcified regions were excised, and 21 mm Magna-Ease valve prosthesis (Edwards Lifesciences, Irvine, California, USA) was implanted. Aortic cross-clamped time was 72 min and

OPEN ACCESS

Correspondence:
Shun Hiraga, Department of Thoracic and Cardiovascular Surgery, Nara Medical University, 840, Shijyouchou, Kashihara city, Nara, 634-8522, Japan, Tel:+81-744-22-3051; Fax:+81-744-24-8040;
E-mail: s.hiraga.0730@gmail.com

Received Date: *15 Jul 2022*
Accepted Date: *02 Aug 2022*
Published Date: *08 AUg 2022*

Citation:
Hiraga S, Hayata Y, Hirose T, Fukuba R, Takemura J, Tonomura R, et al. A Case of Successful Aortic Valve Replacement for Heyde's Syndrome with Perioperative Evaluation of Multimer of Von Willebrand Factor. Ann Cardiovasc Surg. 2022; 5(1): 1031.

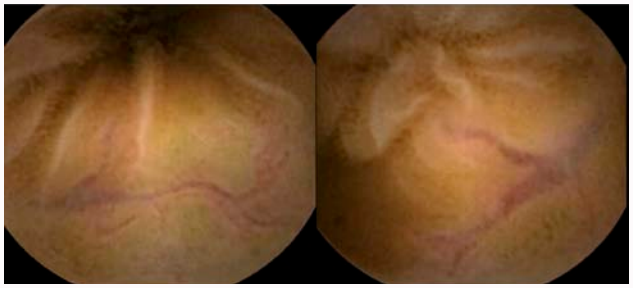

Figure 1: No bleeding point was confirmed on small intestinal capsule endoscopy, however multiple angiodysplasia were observed in the small intestine.

CPB time was 138 min. Tarry stool was seen on Post Operative Day (POD) 7 and laboratory findings showed anemia (hemoglobin, 7.6 g/dl). He was transfused with 12 units of red cell contents during the postoperative period and tarry stool gradually ceased. Postoperative gel electrophoresis revealed that the loss of large multimer of vWF improved to a normal level on POD 2 (Figure 2). The patient's postoperative course was uneventful and he was discharged on POD 50.

Discussion

Heyde initially reported 10 cases of aortic valve stenosis with GI bleeding in 1958 [1]. He described the association of aortic valve stenosis with iron deficiency anemia due to arteriovenous malformation of the gastrointestinal duct. In 1992, Olearchyk described the combination of aortic stenosis and recurrent GI bleeding from intestinal angiodysplasia as Heyde's syndrome [2].

Angiodysplasia refers to enlarged and fragile angioplasty vessels, often found in the mucosa and submucosa of the colon, stomach, small intestine, and nose with recurrent episodes of bleeding [3]. Coexistence of angiodysplasia and aortic valve stenosis has been reported in 7% to 29% of patients with gastrointestinal angiodysplasia, and GI bleeding may occur in 3% of patients with severe stenosis [4]. Batur et al. [5] reported that the prevalence of aortic valve stenosis of any severity in patients with angiodysplasia was 2 to 3 fold greater than in the general population. Furthermore, moderate to severe aortic valve stenosis is reported to be 4.1 times more common in those with angiodysplasia [5]. In our report, the source of GI bleeding was not identified initially with upper and lower GI endoscopy; however, on capsule endoscopy, intestinal angiodysplasia was observed and bleeding from there was noted.

Recently, Vincentelli et al. [6] reported that Heyde's syndrome is caused by acquired von Willebrand disease type 2A. VWF is a plasma glycoprotein involved in subendothelial platelet adhesion and subsequent platelet-platelet interaction. Collagen and other subendothelial tissues are exposed in the damaged vessel wall, and vWF following in the plasma binds to them. Platelets then bind to the A1 domain of collagen-bound vWF through glycoprotein Ib/IX. ADAM TS-13 cleaves the Tyr1605-Met1606 peptide bond in the vWF A2 domain to reduce vWF molecular size. The larger multimers of vWF are more likely to bind to platelets and collagen [7]. In a previous report on Heyde' syndrome, the multimers of vWF are subjected to high fluid shear stress as they pass through the stenotic aortic valve, making it susceptible to cleavage with ADAM TS-13. Reduction of larger multimers of vWF inhibits primary hemostasis, resulting in a bleeding tendency [6]. The multimers of vWF can be estimated using gel electrophoresis, and we continued to estimate the multimers of vWF in the perioperative period. We observed that the larger multimers of vWF decreased in preoperative term.

King et al. [8] reported that among 16 patients with unexplained GI bleeding and concomitant aortic valve stenosis who underwent aortic valve stenosis, only 1 patient continued bleeding postoperatively. Moreover, they reported that in 37 patients who underwent abdominal exploration or bowel resection, 35 had persistent bleeding. Recently, Thompson et al. [9] reported that aortic valve replacement decreases the risk of GI bleeding in patients with

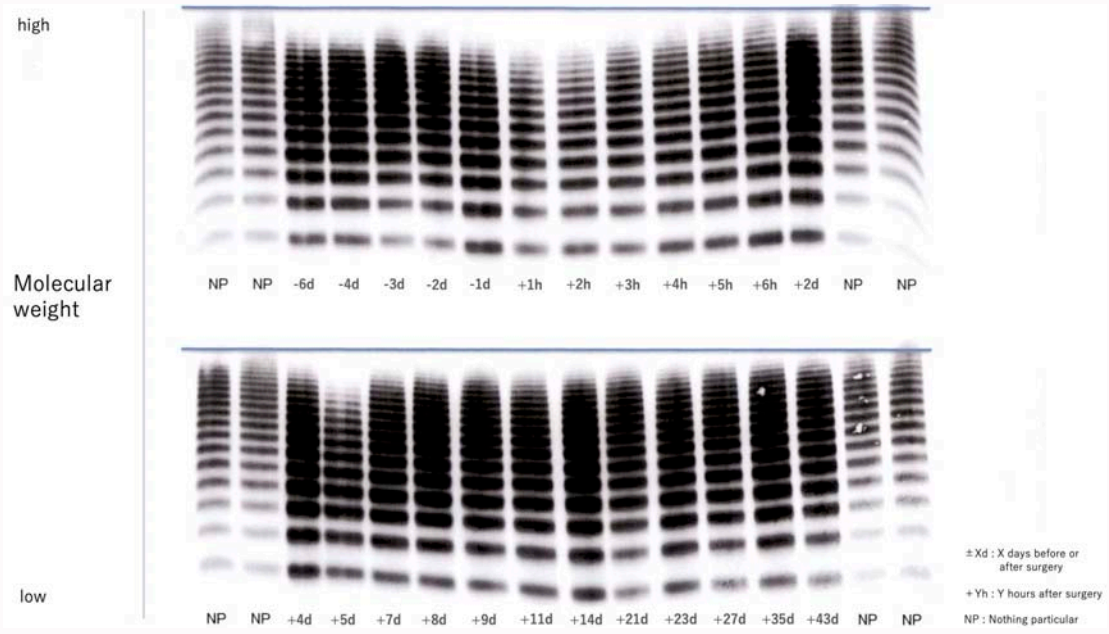

Figure 2: vWF multimer analysis revealed that the high molecular vWF gradually decreased from the 6th day before the operation and its level was the lowest 1h to 2 h after the operation. Subsequently, it gradually recovered and decreased once on the 5th day after the operation, but it improved after the 7th day and became almost normal on the 43rd day after the operation.

Heyde's syndrome in approximately 80%. Aortic valve replacement is an effective treatment for Heyde's syndrome. Thus, in asymptomatic patients of aortic valve stenosis with angiodysplasia and secondary blood loss, aortic valve replacement is indicated.

Conclusion

We encountered a case of Heyde's syndrome. We performed aortic valve replacement following which GI bleeding gradually improved, postoperatively. Evaluation of the multimers of vWF with gel electrophoresis revealed a decrease in the large multimer of vWF in the early postoperative phase.

References

1. Heyde EC. Gastrointestinal bleeding in aortic stenosis. N Engl J Med. 1958;259:196.
2. Olearchyk AS. Heyde's syndrome. J Thorac Cardiovasc Surg. 1992;103(4):823-4.
3. Gola W, Lelonek M. Clinical implication of gastrointestinal bleeding in degenerative aortic stenosis: An update. Cardiol J. 2010;17(4):330-4.
4. Mishira N, Moda P, Khanna A. Heyde's syndrome; an unusual cause of lower gastrointestinal bleeding. Ann Nigerian Med. 2012;6:54-5.
5. Batur P, Stewart WJ, Isaacson JH. Increased prevalence of aortic stenosis in patients with arteriovenous malformations of the gastrointestinal tract in Heyde syndrome. Arch Intern Med. 2003;163(15):1821-4.
6. Vincentelli A, Susen S, Le Tourneau T, Six I, Fabre O, Juthier F, et al. Acquired von Willebrand syndrome in aortic stenosis. N Engl J Med. 2003;349(4):343-9.
7. Sadler JE. New concepts in von Willebrand disease. Annu Rev Med. 2005;56:173-91.
8. King RM, Pluth JR, Giuliani ER. The association of unexplained gastrointestinal bleeding with calcific aortic stenosis. Ann Thorac Surg. 1987;44(5):514-6.
9. Thompson JL, Schaff HV, Dearani JA, Park SJ, Sundt TM, Suri RM, et al. Risk of recurrent gastrointestinal bleeding after aortic valve replacement in patients with Heyde syndrome. J Thorac Cardiovasc Surg. 2012;144(1):112-6.