Case Report

Bentall Operation for Prosthetic Valve Endocarditis with Hereditary Hemorrhagic Telangiectasia

Hideki Morita, MD, Naoyuki Kimura, MD, PhD, Koichi Yuri, MD, Koichi Adachi, MD, Atsushi Yamaguchi, MD, PhD, and Hideo Adachi, MD, PhD

Hereditary hemorrhagic telangiectasia (HHT; Osler–Weber–Rendu syndrome) is an uncommon disease characterized by abnormal telangiectasias and arteriovenous malformations that cause recurrent bleeding. Here, we present the case of a patient with HHT, who had a history of pulmonary and hepatic arteriovenous malformations and endocarditis of a prosthetic aortic valve that was caused by methicillin-resistant Staphylococcus aureus. The patient underwent the Bentall operation after coil embolization for pulmonary arteriovenous malformations. The postoperative course was uneventful.

Keywords: Osler–Weber–Rendu syndrome, hereditary hemorrhagic telangiectasia, Bentall operation, prosthetic valve endocarditis, methicillin-resistant staphylococcus aureus

Introduction

Hereditary hemorrhagic telangiectasia (HHT; Osler–Weber–Rendu syndrome) is characterized by an abnormality of the peripheral blood vessels that often causes recurrent episodes of bleeding. Here, we report the case of a patient with HHT who underwent the Bentall operation for the treatment of prosthetic valve endocarditis caused by methicillin-resistant Staphylococcus aureus (MRSA) and present the patient’s 20-month follow-up. The postoperative course was uneventful. To our knowledge, this is the first published case describing the use of the Bentall operation in a patient with HHT.

Case Report

The patient was a 61-year-old female who had been diagnosed with HHT 30 years previously when she presented with nasal telangiectasias as well as pulmonary and hepatic arteriovenous malformations (AVMs). Excision of a nasal hemangioma responsible for recurrent bleeding episodes had been performed 2 years previously.

The patient arrived at our institution in April 2010 with cardiogenic shock due to acute endocarditis of the aortic valve caused by MRSA infection. An emergency aortic valve replacement was performed using a St. Jude Medical® Regent 21-mm prosthetic mechanical valve (St. Jude Medical Inc., St. Paul, MN, USA). Vegetations and perforation of the left coronary cusp were evident, but the aortic annulus was intact. After the 8-week antibiotic treatment following surgery, the patient was discharged and was able to lead a normal life. However, the patient returned to our institution in March 2011 with sepsis caused by MRSA infection. An emergency aortic valve replacement was performed using a St. Jude Medical® Regent 21-mm prosthetic mechanical valve (St. Jude Medical Inc., St. Paul, MN, USA). Vegetations and perforation of the left coronary cusp were evident, but the aortic annulus was intact. After the 8-week antibiotic treatment following surgery, the patient was discharged and was able to lead a normal life. However, the patient returned to our institution in March 2011 with sepsis caused by MRSA infection. Although antibiotic therapy was initiated, a paravalvular leakage occurred 2 weeks after admission, and prosthetic valve endocarditis was diagnosed. Coil embolization for a pulmonary AVM was performed in May 2011 (Fig. 1). Bleeding from a gastric mucosal telangiectasia occurred in May 2011, but active bleeding was not evident during gastric endoscopy.
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A 6-month follow-up revealed that the patient had not experienced any new bleeding complications except epistaxis. After 3 months of warfarin medication, the incidence of epistaxis decreased significantly. A secondary coil embolization for a pulmonary AVM was performed in November 2011.

Discussion

HHT is an inherited autosomal dominant trait, with a prevalence of 1 in 5000–8000. It is caused by mutations of the endoglin gene or of Activin receptor-like kinase type 1, both of which are essential for angiogenesis and
normal vascular maturation. These mutations result in the formation of telangiectasias and AVMs, which tend to bleed. They are usually localized in the skin and mucosa, causing epistaxis and gastrointestinal bleeding, but AVMs also occur in the pulmonary and hepatic circulation in 30% of patients and in the cerebral circulation in 10%–20%. In this case, we observed repetitive epistaxis, gastric bleeding, and three pulmonary and hepatic AVMs. The AVMs were highly susceptible to infection, and the pulmonary AVMs were treated with coil embolization twice.

In the field of cardiac surgery, the number of reported cases of HHT remains small. To our knowledge, our patient is the first with HHT to undergo the Bentall operation following aortic valve replacement. Although a mechanical valve was selected in the first replacement operation, we used a tissue valve in the second, taking into consideration repetitive epistaxis and gastric bleeding. Ishikawa, et al. also reported the replacement of mitral valve with a tissue valve in a patient with HHT. Benzadón, et al. recommend maintaining the activated clotting time >400s, which prevents bleeding by decreasing the activity of the coagulation system during surgery.

The postoperative course of our patient was uneventful. Recurrent infection was not evident at a 6-month follow-up, but this patient will require careful follow-up in future.

Disclosure Statement

The authors have no conflict to disclosure.

References