A Second Mitral Valve Replacement in a Patient With Hereditary Hemorrhagic Telangiectasia (Osler's Disease)

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SUMMARY

A 62-year-old female with Osler’s disease was admitted to our hospital because of fever and cardiac failure. The patient had undergone a mitral valve replacement (MVR) using a Carpentier-Edwards prosthetic valve 14 years earlier. A bacterial examination of arterial blood identified Streptococcus mitis. No arteriovenous malformations were detected in visceral organs. The patient underwent MVR using the same prosthetic xenograft after conservative treatment and management of repetitive epistaxis and decayed teeth. Intra- and postoperative bleeding were typical of a mitral valve reoperation. This is the first reported experience, to the best of our knowledge, of a second MVR in a patient with Osler's disease. (Jpn Heart J 2004; 45: 885-888)

Key words: Mitral valve replacement, Hereditary hemorrhagic telangiectasia, Osler’s disease, Redo surgery

OPERATIVE reports of cardiac surgery in patients with hereditary hemorrhagic telangiectasia (Osler's disease) are rare1) probably due to coexisting arteriovenous malformations (AVMs) and perioperative bleeding tendency concerning extracorporeal circulation (ECC). We performed the first successful mitral valve replacement (MVR) in 1987 in a patient with Osler's disease.2) The patient returned to our hospital because of infective endocarditis and cardiac failure due to primary tissue failure of a Carpentier-Edwards prosthetic valve. After the management of repetitive epistaxis and decayed teeth and close exploration of a combined arterial-venous fistula, the mitral valve was replaced again with the same prosthetic xenograft. Intra- and postoperative bleeding were typical for cases of a reoperation for mitral valve. We report this rare experience focusing on the perioperative management and surgery.

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CASE REPORT

A 62-year-old female was admitted to our hospital on an urgent basis because of fever and cardiac failure in January 2003. The patient had undergone a MVR 14 years earlier due to degenerative mitral valve regurgitation. In the previous operation a Carpentier-Edwards prosthetic valve, 27 mm in diameter, was selected because of Osler's disease associated with repetitive epistaxis. Telangiectasias were found in the skin and buccal and nasal mucosa. Her elder sister had also experienced a repetitive epistaxis. Hematological examination revealed anemia (Hb 8.0 g/dL) and a slight decrease in the platelet count (14.4 × 10⁴/mm³). Bleeding time was 150 seconds preoperatively. No defect in the coagulation mechanism was found and platelet function was normal. A bacterial examination of arterial blood on admission revealed the presence of Streptococcus mitis. Transesophageal echocardiograms revealed primary tissue failure of the mitral xenograft associated with moderate regurgitation and possible small vegetation.

Following a diagnosis of infective endocarditis and cardiac failure due to primary tissue failure of the prosthetic valve, the patient initially received conservative treatment using antibiotics and diuretics. Visceral AVMs combined with Osler's disease were also explored by brain magnetic resonance imaging (MRI), thoracic and abdominal computed tomography (CT), gastrointestinal endoscopy, and other techniques. No AVMs were detected in the brain, lung, liver, gastrointestinal tract, or urinary region. After the management of repeated nasal bleeding and decayed teeth, a second MVR was performed on the 32nd day after admission (Figure 1). The mitral valve was approached via the right-sided left

![Figure 1](chart.png)

Figure 1. Hospital course after admission. The second MVR was performed after the management of systemic infection. PIPC = piperacillin sodium; CEZ = cefazolin sodium; BE = bacterial examination of arterial blood.
atrium under the assistance of normothermic extracorporeal circulation (ECC) and blood cardioplegia. The valve was replaced with a Carpentier-Edwards prosthetic valve (27 mm in diameter). Intra- and postoperative bleeding was almost typical of a mitral valve reoperation. Bleeding around the sternum was slightly excessive, however, it was successfully managed using an argon-beam laser coagulator. Just before closing the chest, 20 units of platelets were infused to reinforce hemostasis. The total volume of intraoperative hemorrhage was 593 mL. The duration of ECC and the aortic cross clamping time were 195 and 122 minutes, respectively. The Carpentier-Edwards prosthetic valve removed was thick and calcified at the edge of the leaflets. There was no tearing of the leaflets or apparent vegetation (Figure 2). Bacterial examination of the valve was negative. The postoperative course was uneventful without any bleeding and the patient was discharged three weeks after surgery.

**DISCUSSION**

Osler's disease affects multiple organ systems including telangiectases and AVMs of the skin, mucosa, and viscera. With respect to the mucosa, epistaxis is the most common and a wide variety of treatments have been used. In our case, repetitive epistaxis was observed and telangiectasias were identified in the skin.
and buccal and nasal mucosa. Visceral involvement includes pulmonary, gastrointestinal, and cerebral AVMs, which have been reported predominantly in adults. Visceral AVMs and these complications are sometimes life threatening, and lung and liver transplantations have recently been performed in patients with Osler’s disease. Before cardiac surgery using ECC, careful preoperative screening of systemic AVMs is indispensable. Fortunately, this case had no visceral AVMs and the patient could undergo a second MVR following the management of systemic infection, repetitive epistaxis, and decayed teeth. Concomitant operations, including cardiac surgery, seem to be inappropriate considering the usage of ECC, except for small and peripheral pulmonary lesions. If postoperative bleeding may occur in spite of close preoperative evaluation, angiography of the brain or visceral organs or endoscopy of the respiratory or gastrointestinal tracts should be urgently performed.

In the field of cardiac surgery, the number of reported cases with Osler’s disease still remains small. We reported this same case previously as being the first experience of open-heart surgery and this patient also became the first case of a second MVR. Ruggieri, et al reported an operative case with pulmonary valve stenosis. A tissue valve should be selected for patients with hemorrhagic disease. We used the same type of Carpentier-Edwards prosthetic valve as in the first replacement operation, taking into consideration the repetitive epistaxis. Young, et al also reported an aortic valve replacement operation with a tissue valve for a patient with von Willebrand’s disease. In this case, intra- and postoperative bleeding was typical of a redo mitral valve operation under normothermic ECC. In conclusion, open-heart surgery itself may be performed safely even in a reoperative case with Osler’s disease.

**REFERENCES**