of smoke”, thus, the Japanese term “Moyamoya” (3). Children with MD may present with various symptoms such as aphasia, dysarthria, hemiparesis, and seizures. Less common presentations such as syncope, visual changes, and chorea can occur (4). Although the first sign was seizure in our patient, MD was not diagnosed because of the nonspecific findings on cranial MRI. After experiencing syncope twice while exercising, he was diagnosed with pulmonary hypertension based on echocardiography findings, and the secondary causes were excluded. Since there was co-occurrence of pulmonary hypertension and seizure, the cranial MRI was re-evaluated, and MD was suspected due to collateral vessels seen in both hemispheres. This is a very rare presentation of MD in a child. Therefore, it was emphasized that MD must be kept in mind in all patients diagnosed with IPAH, particularly those who have had concomitant seizures.

The co-occurrence of pulmonary hypertension and MD has been previously reported in very few cases (5-7). Among them, RNF213 homozygosity was found in two patients. Therefore, this gene may cause a novel entity involving the brain and lung together. We also plan to investigate this gene mutation in our patient.

There is no consensus about therapy in children with MD and severe pulmonary hypertension because of the rarity of this entity. In one study, two cases with MD and pulmonary hypertension were reported, and both died after vasoreconstructive surgery for MD (8). Thus, surgery in patients with MD may have poor prognosis, particularly in those with severe pulmonary hypertension, and it should be discussed very carefully with the parents. The risk of surgery was expected to be high due to pulmonary hypertension in our patient, and the parents refused the surgery.

There was one report that showed bosentan therapy improved the blood flow in the cerebral hemispheres in a child with MD (9). Although the role of endothelin pathways in the pathogenesis of MD has not been studied, this case suggested the use of endothelin receptor antagonists to improve the cerebral circulation. However, bosentan and tadalafil combination therapy did not improve the pulmonary artery pressures in our patient; we do not know about the blood flow to both hemispheres yet. Additional studies are needed to determine whether the patients may benefit with bosentan or other specific pulmonary artery hypertension treatments to improve the circulation of cerebral hemispheres and pulmonary artery pressures.

Conclusion

In conclusion, MD should be considered in children with seizures accompanied with pulmonary hypertension, and the risk of shunt surgery is very high in these patients. It is not known whether bosentan or other specific pulmonary artery hypertension treatment influence the mortality or morbidity in this rare entity.

References


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Unexpected complication during transcatheter aortic valve replacement: Balloon that cannot inflate!

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Introduction

In the treatment of severe aortic stenosis, transcatheter aortic valve replacement (TAVR) procedure is increasingly being used. In this report, we present a case of balloon rupture during
The valve that was pulled back with the other system was checked; no structural or functional problem was found (Fig. 1). Then the same valve was loaded into a new delivery system from the same femoral artery but at another puncture place. Fortunately, the valve system was successfully implanted (Video 2). Paravalvular insufficiency was not observed during the control aortography (Video 3).

Discussion

To the best of our knowledge, this is the first study to report such a complication; during the process, the balloon burst without being inflated. The rupture may have been caused by the fact that the valve–balloon axis was different during the loading of the valve into the balloon.

In literature, the number of cases reporting balloon rupture during TAVR operation is limited (1-3). Cases of balloon rupture associated with aortic valve have been reported mostly during aortic balloon valvuloplasty in the preoperative period. In these reports, balloon rupture development has been associated with the presence of a bicuspid aortic valve, sudden high-pressure inflation of the balloon, or intense calcification in the ascending aorta (4, 5). In literature, we could not find any case in which the balloon was ruptured without being inflated during the TAVR operation, similar to our case.

The preoperative preparations are extremely important to foresee the complications that may occur during the TAVR operation. At this stage, evaluation of the calcification and tortuosity of the ascending and descending aorta as well as the detailed evaluation of the valve with TEE and CT is extremely important. In the presence of advanced aortic tortuosity, a harder wire may be helpful in solving the problem. Alternatively, subclavian or aortic pathway may be preferred.

There are some prosthetic valves that can be retrievable or not during the TAVR operation. The Edwards Sapien S3 valve is not repositionable or retrievable, making precise deployment critical. However, it can be reused when it is withdrawn before inflation. In our patient, the balloon never inflated, thus the valve never opened.

Conclusion

Although TAVR opens new horizons in the treatment of patients with severe aortic stenosis, it has brought new complications too. However, in these cases, successful implantation of the same valve is possible with timely detection and correct management of complications.

References

increasingly used since they are shown to shorten the duration of intensive care and hospitalization and can be applied in patients with other system problems, can reduce blood loss and transfusion needs, and have lower rates of mortality and morbidity (3, 4).

Certain special endovascular systems have been developed for treating complex aneurysms of the major side branches arising from or nearby the aneurysm sac. Fenestrated stent grafts (5), chimney technique and multilayer flow modulator (MFM) are among the best known endovascular systems (6, 7).

Case Report

A 64-year-old male patient presented to our clinic with chest and back pain. His medical history revealed previous treatment for hypertension and diabetes mellitus for the last 10 years. Critical stenosis in the left and right coronary arteries was detected in coronary angiography; contrast computed tomography showed a 7-cm diameter aortic aneurysm starting from the distal of the left subclavian artery and extending to proximal of the celiac trunk (Fig. 1). Two MFMs (Cardiatis, CTMS 40150) were placed in the aneurysm region for endovascular intervention. However, the targeted region could not be reached due to the insufficient flexibility of the transmission system of the stent and/or anatomy of the aortic arch, an observation that was evident with the presence of the proximal end of the stent angled to the aortic axis (Fig. 2). Consequently, the procedure was terminated; the patient underwent open heart surgery 3 days later, and arterial cannulation was performed from the right subclavian artery after sternotomy. Subsequently, aortotomy was performed in the ascending aorta after total circulatory arrest under antegrade cerebral perfusion. The angulation (Fig. 3) of the MFM stent was corrected by manual guidance. Afterward, an MFM stent was im-