

CASE REPORT

Mitral valve plasty in a patient with multiple cerebral cavernous malformations: A case report

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Abstract

Cerebral cavernous malformations (CCMs) are blood vessel malformations, often untreated if asymptomatic. However, upon cardiac surgery with cardiopulmonary bypass, cerebral edema/hemorrhage may occur. We successfully performed mitral valve plasty for a case of severe mitral regurgitation with multiple CCMs. Preoperative head magnetic resonance imaging and strict perioperative management are important.

KEYWORDS

cardiac surgery, cardiopulmonary bypass, multiple cerebral cavernous malformations, perioperative neurological complications

1 | INTRODUCTION

Few reports have described cardiac surgery using cardiopulmonary bypass (CPB) in patients with brain tumors. When CPB is performed in patients with brain tumors, there may be an increased risk of cerebral hemorrhage, infarction, and edema.¹⁻⁸ In the present case report, we performed mitral valve plasty (MVP) in a patient with mitral regurgitation (MR) complicated by multiple cerebral cavernous malformations (CCMs). With strict perioperative management, the patient had good outcomes.

2 | CASE REPORT

A 54-year-old woman presented to our hospital with shortness of breath upon exertion. Her New York Heart Association (NYHA) classification was class 3. She had a history of multiple CCMs and had undergone surgical resection of CCM on the left cerebellum at 16 years of

age due to intratumoral hemorrhage. Physical examination at the time of her initial visit revealed no neurological abnormalities. Transthoracic echocardiography (TTE) revealed severe MR due to the posterior leaflet prolapse (Figure 1) (color Doppler area ratio, 0.41; vena contracta widths, 7.0 mm; regurgitant volume, 57.9 ml; regurgitant fraction, 0.63; and effective regurgitant orifice area of MR, 0.46). Left ventricular (LV) contractility was normal (LV diastolic diameter, 56 mm; LV systolic diameter, 33 mm; and LV ejection fraction, 0.71). Coronary angiography confirmed the absence of stenotic lesions in the coronary arteries. Intracardiac pressure measurements revealed a mean pulmonary artery pressure of 28 mmHg, pulmonary artery wedge pressure (PAWP) of 19 mmHg, v wave in PAWP of 31 mmHg, cardiac index of 2.28 L/min m², and LV end-diastolic pressure of 25 mmHg. Magnetic resonance imaging (MRI) of the head revealed multiple CCMs and microbleeding lesions (Figure 2). Since our patient had symptomatic severe primary MR, the American College of Cardiology/American Heart Association (ACC/

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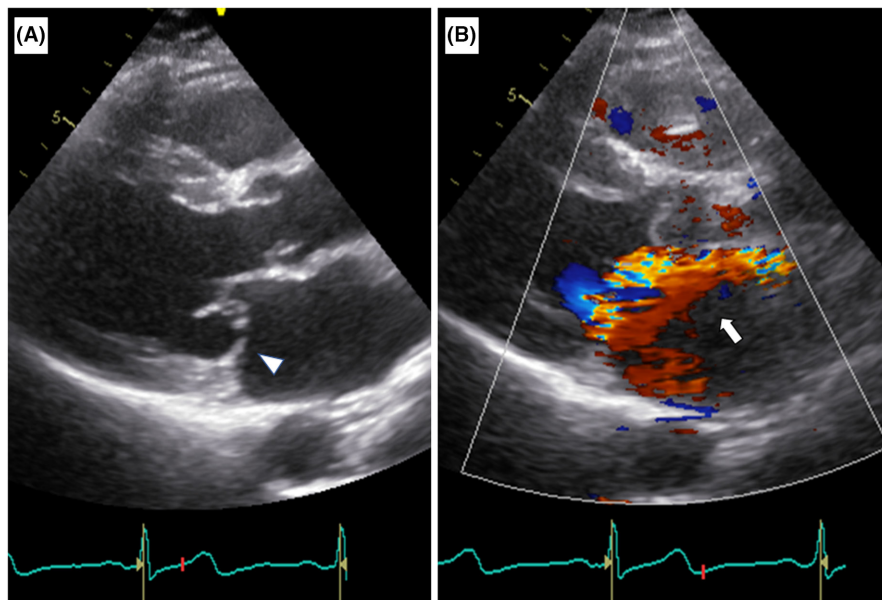


FIGURE 1 Transthoracic echocardiography (TTE). Parasternal long-axis view: (A). Prolapse at the posterior leaflet (P2) of the mitral valve (white arrowhead). (B). Severe mitral regurgitation with the jet was predominantly located on the anterior wall of the left atrium (white arrow).

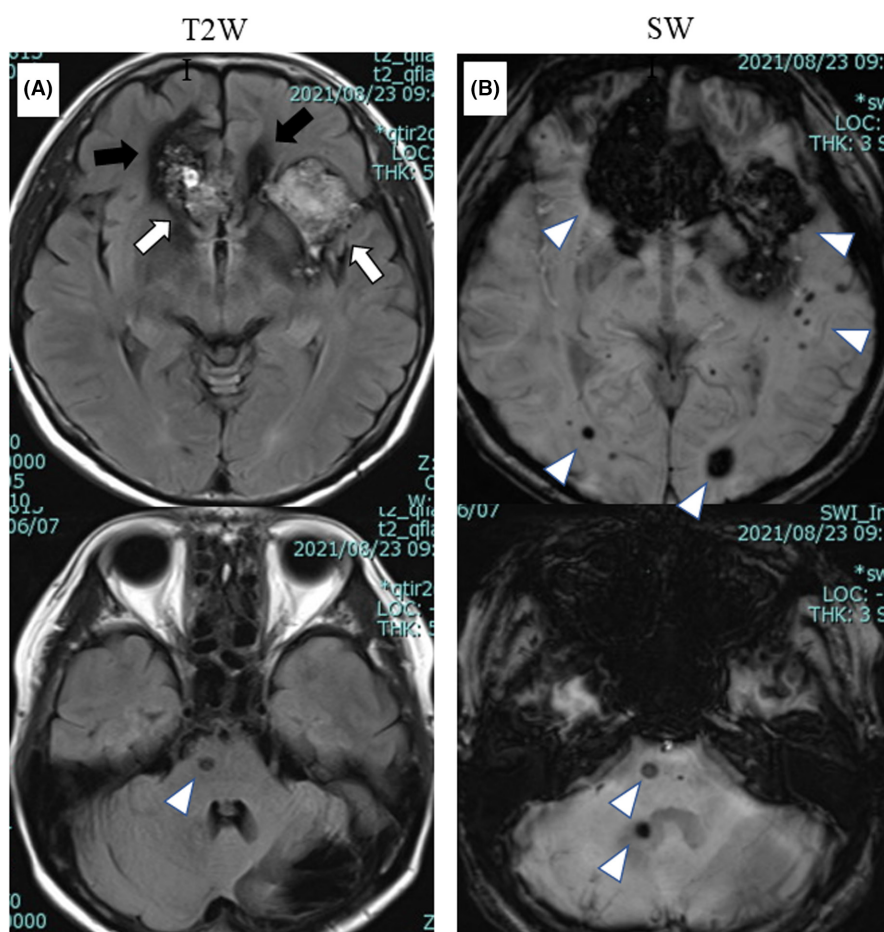


FIGURE 2 Magnetic resonance imaging of the head. (A). T2WI showed a nodule with heterogeneous low to high signal (white arrow), hemosiderin deposition around the nodule in the cerebral hemispheres (black arrow), and a low-signal nodule on the pons (white arrowhead). (B). SWI showed a more pronounced low signal and many other small post-microhemorrhagic lesions (white arrowhead). SWI: susceptibility-weighted imaging; T2WI: T2-weighted imaging.

AHA) guideline recommended a class I indication for MVP.⁹

Because the patient had multiple CCMs, there was a risk of perioperative cranial neurological complications during cardiac surgery with CPB. In this regard, risk factors for hemorrhagic complications in CCM included a

history of hemorrhage, presence of focal neuropathy, site of origin (infratentorial, brainstem), and female sex.¹⁰ Our patient had all of these factors, except for focal neuropathy, and was at a high risk for hemorrhagic complications. Therefore, percutaneous Mitraclip without the use of CPB represented a viable treatment option. After a thorough

discussion at a multidisciplinary heart team conference, cardiac surgery was decided as appropriate, considering the patient's age and long-term outcome.

Surgery was performed under general anesthesia through a median sternal incision. Intraoperative CPB management has been performed according to the guidelines.¹¹ In this case, in addition to the standard CPB management from the guidelines, strict management was required, particularly because of the risk of cerebral hemorrhage, cerebral infarction, and cerebral edema. The measures we strictly adhered to were as follows. To prevent plasma osmolality reduction due to excessive hemodilution at the start of CPB, CPB was primed with 280 ml of red blood cells, 240 ml of fresh frozen plasma, 268 ml of 20% mannitol, 40 ml of 8.4% sodium bicarbonate, 5 ml of heparin, and 500 mg of methylprednisolone sodium succinate. After systemic heparinization, dexamethasone (4 mg) was administered intravenously. CPB was established by ascending aortic cannulation and bicaval drainage, and the patient's deep body temperature was cooled to 30°C. The PaCO₂ levels were strictly regulated to 30–40 mmHg by adjusting the ventilator and CPB. Activated coagulation time (ACT) was measured every 30 min to avoid excessive prolongation. The intraoperative target range of ACT was 400–450 s. As a result, ACT was managed between 473 and 592 s during the use of CPB. The intraoperative mean blood pressure (mBP) was controlled at 60–80 mmHg. Since hyponatremia associated with the use of CPB decreases plasma osmolality and might cause cerebral edema,¹² the target range of serum sodium concentration was 130–140 mmol/L, and the actual serum sodium levels were controlled within a range of 137–139 mmol/L. Observation of the mitral valve via a transseptal approach revealed that the posterior leaflet (P2) had prolapsed significantly because of tendon rupture. The prolapsed leaflet was resected and sutured in a triangular manner. The MVP was completed by suturing a semi-rigid full-ring to the mitral valve annulus. CPB weaning was uneventful, and dexamethasone (4 mg) was administered intravenously. Postoperatively, the patient was managed with an mBP of 60–80 mmHg by continuous infusion of nitroglycerin and carperitide, and from the second postoperative day with oral enalapril and carvedilol. Frequent blood examinations were performed and serum sodium levels were strictly controlled to achieve our target level. As a consequent, her postoperative serum sodium levels ranged from 132 to 142 mmol/L. Oral tolvaptan was administered for approximately 1 week postoperatively to prevent hyponatremia. Neurological complications were not observed. Postoperative TTE revealed no residual MR or abnormal LV contractility. The patient was discharged on the 14th day.

3 | DISCUSSION

CCMs are vascular malformations, and common symptoms include convulsions and focal neurological symptoms due to compression of the mass with a low bleeding rate.¹³ Reports of cardiac surgery with CPB for patients with complicated brain tumors are limited to a small number of case series. Large studies were lacking, and data on appropriate intraoperative and perioperative management are also insufficient. There is no established information on risk stratification or prophylactic perioperative management for this patient population. Therefore, it is necessary to devise perioperative management while conducting preoperative risk assessment for each case. Accurate risk stratification through the accumulation of similar cases in the future will lead to improved outcomes. The present case showed no evidence of focal neurological deficits; however, there was a risk of cerebral edema or bleeding during and after CPB.¹⁰ Therefore, we followed the management recommended in previous reports^{1–8}: (1) maintain a PaCO₂ of <40 mmHg during surgery; (2) steroids (dexamethasone, 4 mg) should be administered before and after CPB; (3) maintain intraoperative and postoperative mBP at 60–80 mmHg; (4) maintain serum sodium concentrations at 130–140 mmol/L to stabilize serum osmolality; and (5) maintain a central venous pressure of <8–10 mmHg.

On the other hand, Aleksic et al.⁸ reported cardiac surgery in 16 patients with intracranial meningiomas and concluded that the presence of intracranial meningiomas was not a risk factor during cardiac surgery, as they found no cranial nerve complications related to meningiomas. The authors consulted a neurosurgeon preoperatively and used steroids if necessary. They also had a policy of maintaining moderate hypocapnia as anesthetic management, adding hypertonic saline or mannitol for cardiopulmonary priming, and maintaining sodium levels of 132–138 mmol/L. Ultimately, good outcomes were achieved possibly because of such perioperative management. Further, it is unlikely that strict management is not necessary during cardiac surgery for patients with brain tumors.

In several case reports of cardiac surgery complicated by brain tumors, cranial nerve complications have occurred relatively late during the postoperative period. These observations indicated the importance of both intraoperative management and management during the first few days after surgery. Therefore, continuous control of blood pressure, blood sodium level, and anticoagulation therapy are necessary during hospitalization. In the present case, blood pressure was well controlled with nitroglycerin and carberytide continuous infusion while the patient was in the intensive care unit (ICU). Thereafter, enalapril and carvedilol were taken orally. Sodium levels

in the blood were controlled by oral tolvaptan for several days with appropriate fluid replacement management. Kinugawa et al.¹⁴ reported that tolvaptan can be used safely in patients with normal sodium levels. In this case, the blood sodium level was well controlled at a dose of 7.5 mg/day, and no postoperative cranial nerve complications occurred.

Cerebral edema generally occurs with the use of CPB, even in the absence of brain tumors.¹⁵ This phenomenon may result from a decrease in plasma osmolality (hemodilution and increased anti-diuretic hormone levels due to CPB stress), increased permeability of the blood–brain barrier (BBB), and hemodynamic changes in the perioperative period. Capillaries in brain tumors do not have normal BBB function and are susceptible to edema around the tumor, increasing the risk of neurological complications.¹⁶

There are many cases of asymptomatic brain tumors, and it is difficult to detect the presence of brain tumors based on physical examination alone. Therefore, imaging examinations, such as head MRI, are necessary to detect brain tumors and are recommended routinely for patients scheduled for surgery using CPB. Head MRI is also effective in detecting asymptomatic cerebral infarctions or stenosis of cerebral arteries in addition to brain tumors and is considered cost-effective, because it can avoid major postoperative neurological complications.

In this case, because the presence of a brain tumor was known preoperatively, the aforementioned measures could be taken, and no major postoperative neurological complications occurred. Previous case studies have reported death or long-term rehabilitation due to neurological complications when brain tumors were not detected before surgery using CPB or when adequate measures were not taken perioperatively.^{1–5} In contrast, when tumors were detected preoperatively and adequate measures were taken, surgery was completed without complications.^{6–8} These findings suggested that intracranial evaluation before surgery using CPB and strict perioperative management are important for patients with brain tumors.

4 | CONCLUSION

In this study, MVP was successfully performed in a patient with MR and multiple CCMs. Strict perioperative management is important to avoid perioperative cerebral complications.

AUTHOR CONTRIBUTIONS

Study conception: S Yamashita. Data collection: all authors. Analysis: all authors. Investigation: S Yamashita,

SY, TD. Writing: S Yamashita. Critical review and revision: all authors. Final approval of the article: all authors. Accountability for all aspects of the work: all authors.

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CONFLICT OF INTEREST

The authors have no conflict of interests to disclose with this study.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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REFERENCES

- Ahmad M, Al-Arifi A, Najm HK. Open heart surgery with intracranial meningioma: case report and literature review. *Heart Lung Circ.* 2015;24(7):e108-e111.
- Sun H, Ross DA. Fatal hemorrhagic infarction of posterior fossa meningioma during cardiopulmonary bypass. *Ann Thorac Surg.* 2012;93(2):653-656.
- Chow HK, Yousafzai SM, Ugurlucan M, Canver CC. Silent meningioma—a rare cause of stroke in post-cardiopulmonary bypass patients. *Arch Med Sci.* 2010;6(2):176-280.
- Kurusu K, Hisahara M, Ando Y, Tominaga R. An unusual manifestation of brain tumor: development of delayed hemiplegia after cardiopulmonary bypass. *J Card Surg.* 2007;22(5):417-418.
- Maeda K, Gotoh H, Chikui E, Furusawa T. Intratumoral hemorrhage from a posterior fossa tumor after cardiac valve surgery—case report. *Neurol Med Chir (Tokyo).* 2001;41(11):548-550.
- Ando Y, Kurisu K, Hisahara M. Aortic valve replacement in a patient with a brain tumor. *Gen Thorac Cardiovasc Surg.* 2008;56(6):295-297.

7. Grigore AM, Grocott HP, Newman MF. Anesthetic management of a patient with a brain tumor for cardiac surgery. *J Cardiothorac Vasc Anesth*. 2000;14(6):702-704.
8. Aleksic I, Sommer SP, Kottenberg-Assenmacher E, et al. Cardiac operations in the presence of meningioma. *Ann Thorac Surg*. 2009;88(4):1264-1268.
9. Otto CM, Nishimura RA, Bonow RO, et al. 2020 ACC/AHA guideline for the management of patients with valvular heart disease: executive summary: a report of the American College of Cardiology/American Heart Association joint committee on clinical practice guidelines. *Circulation*. 2021;143(5):e35-e71.
10. Washington CW, McCoy KE, Zipfel GJ. Update on the natural history of cavernous malformations and factors predicting aggressive clinical presentation. *Neurosurg Focus*. 2010;29(3):E7.
11. Wahba A, Milojevic M, Boer C, et al. 2019 EACTS/EACTA/EBCP guidelines on cardiopulmonary bypass in adult cardiac surgery. *Eur J Cardiothorac Surg*. 2020;57(2):210-251.
12. Leong XF, Cheng M, Jong B, Hwang NC, Roscoe A. Sodium abnormalities in cardiac surgery with cardiopulmonary bypass in adults: a narrative review. *J Cardiothorac Vasc Anesth*. 2021;35(11):3374-3384.
13. Umemura T, Nishizawa S, Miyachi H, Yamamoto J. Removal of double cavernous angioma of the frontal lobe using a three-dimensional printed model: a case report. *J UOEH*. 2020;42(2):217-222.
14. Kinugawa K, Sato N, Inomata T, Yasuda M, Shibasaki Y, Shimakawa T. Novel risk score efficiently prevents tolvaptan-induced hypernatremic events in patients with heart failure. *Circ J*. 2018;82(5):1344-1350.
15. Harris DN, Oatridge A, Dob D, Smith PL, Taylor KM, Bydder GM. Cerebral swelling after normothermic cardiopulmonary bypass. *Anesthesiology*. 1998;88(2):340-345.
16. Heiss JD, Papavassiliou E, Merrill MJ, et al. Mechanism of dexamethasone suppression of brain tumor-associated vascular permeability in rats. Involvement of the glucocorticoid receptor and vascular permeability factor. *J Clin Invest*. 1996;98(6):1400-1408.

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