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Meningioma Resection in a Patient with High-Risk Obstructive Hypertrophic Cardiomyopathy: Preoperative Mavacamten Optimization and Multidisciplinary Management

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Case Report

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Abstract

Background

The clinical presentation of meningioma combined with obstructive hypertrophic cardiomyopathy (HOCM) is extremely rare, and perioperative management is exceptionally challenging. High-risk HOCM without optimization is generally regarded as an absolute contraindication for craniotomy, and there is a profound lack of established experience regarding its successful management.

Case presentation:

A 50-year-old man presented with headache and dizziness. Magnetic resonance imaging showed a large anterior skull base meningioma abutting the internal carotid and anterior cerebral arteries. Preoperative cardiac evaluation diagnosed obstructive hypertrophic cardiomyopathy with a resting left ventricular outflow tract gradient of 133 mm Hg and a right ventricular ejection fraction of 28%. After multidisciplinary team discussion, mavacamten was started. The gradient fell to 79 mm Hg after 5 days. The tumor was then completely resected under goal-directed hemodynamic management. The perioperative course was uneventful. Postoperative brain swelling resolved with conservative treatment. At 1-month follow-up, the patient had recovered well without cardiac or surgical complications.

Conclusions

Unoptimized obstructive hypertrophic cardiomyopathy is an absolute contraindication to craniotomy. However, selected patients whose hemodynamics are stabilized by medical therapy can undergo surgery safely within a multidisciplinary framework. Maintaining preload, avoiding tachycardia, and preserving afterload are essential for perioperative safety.

Introduction

Meningioma is the most common primary benign intracranial tumor, with an annual incidence of approximately 2.3–9.7 per 100,000. [1]The prevalence of HOCM is approximately 1/500, with about 66% of cases exhibiting left ventricular outflow tract obstruction.[2] The coexistence of meningioma and HOCM is extremely rare in clinical practice. A search of the PubMed database by the authors revealed no reports of patients with both conditions undergoing craniotomy.Currently, there is no standardized protocol for the perioperative management of such patients, and there is little successful experience to draw upon.

The core pathophysiology of HOCM is dynamic LVOT obstruction.Any factor that reduces preload, decreases afterload, or increases heart rate can exacerbate the obstruction, leading to a sudden drop in cardiac output, insufficient coronary perfusion, and even sudden cardiac death.Neurosurgical

procedures, however, require preoperative fasting, intraoperative dehydration with mannitol, and controlled hypotension, which directly conflict with the management requirements for HOCM. The 2024 AHA/ACC guidelines for the first time list the cardiac myosin inhibitor mavacamten as a Class I recommendation for symptomatic oHCM, providing a new preoperative optimization strategy for such patients undergoing non-cardiac surgery. [3] Furthermore, craniocerebral surgery itself can induce neurogenic stress cardiomyopathy via the "brain-heart axis," which may pose a cumulative insult to patients with HOCM. [4,5,6]

This paper reports a case of a patient with a large anterior skull base meningioma complicated by high-risk HOCM. Following preoperative optimization with mavacamten, a gross total resection of the meningioma was performed within a multidisciplinary team (MDT) framework, aiming to explore the surgical decision-making logic and perioperative management strategies for such high-risk patients.

Case Report

Medical History and Physical Examination

A 50-year-old male was admitted due to dizziness, headache, and fatigue for over one month.There was no history or family history of cardiomyopathy, and no cardiac-related symptoms.Physical examination upon admission: blood pressure 139/97 mmHg, heart rate 71 beats/min, and a 2/6 grade systolic murmur was audible at the 3rd–4th intercostal space of the left sternal border.Neurological examination revealed no focal deficits.

Investigations

Contrast-enhanced cranial MRI revealed a space-occupying lesion at the base of the anterior cranial fossa, measuring approximately 4 cm × 5.4 cm × 3.8 cm. The lesion showed marked and homogeneous enhancement, was supplied by the ipsilateral internal carotid artery, and was closely adjacent to the right internal carotid artery and the A1 segment of the anterior cerebral artery, accompanied by peritumoral edema and midline shift.Transthoracic echocardiography showed an interventricular septal thickness of 2.42 cm, a positive SAM sign, a resting LVOT-PGmax of 133 mmHg, and a mean pressure gradient of 67 mmHg. The left atrial diameter was 4.41 cm, and the LVEF was 69%. Cardiac MRI demonstrated diffuse left ventricular wall thickening with late gadolinium enhancement at the right and left ventricular insertion points. The right ventricular ejection fraction was 28%. NT-proBNP was 331.6 ng/L.HeartCoronary CTA revealed a myocardial bridge in the mid-segment of the left anterior descending artery.The right ventricular ejection fraction is only 28%, which is significantly lower than the normal value.[7] Since right ventricular dysfunction is an independent predictor of cardiovascular death in patients with HCM, this further increases the perioperative risk in this case. [8]

MDT Assessment and Preoperative Optimization

The MDT consists of specialists from neurosurgery, cardiology, anesthesiology, and critical care medicine.The cardiology department determined that the patient's resting LVOT-PGmax reached 133 mmHg, indicating high-risk HOCM, and that craniotomy posed an extremely high risk of sudden cardiac death and acute heart failure.However, the meningioma is large with a significant mass effect; if managed conservatively, tumor progression could lead to cerebral herniation.The critical decision lies in whether to treat the meningioma or the HOCM first. If septal reduction therapy is performed first, postoperative recovery would require at least several weeks, during which the risk of tumor progression is high.Ultimately, the MDT decided to prioritize the resection of the meningioma.

The Department of Cardiology initiated oral mavacamten at 2.5 mg once daily on the fifth day before the procedure. After five days of treatment, the LVOT-PGmax decreased from 133 mmHg to 79 mmHg. Recent real-world studies indicate that in patients with a resting LVOT gradient > 100 mmHg, one week of mavacamten treatment can reduce the gradient from 121 mmHg to 87 mmHg. TreatmentRecent real-world studies indicate that in restingThe response in this case was superior to the average level, suggesting that the patient may be relatively sensitive to mavacamten.[9]

The Department of Anesthesiology assessed the patient as ASA physical status IV, recommending the establishment of invasive arterial blood pressure and central venous pressure monitoring, strict control of heart rate, blood pressure, and fluid volume, and the preparation of extracorporeal circulation support technology.The Department of Neurosurgery indicated that the tumor is adjacent to critical blood vessels, posing an inherent risk of intracranial hemorrhage during surgery; the cardiomyopathy necessitates restrictive fluid management, which conflicts with the neurosurgical requirement for cerebral edema prophylaxis.

Table 1
Timeline of hemodynamic changes before and after mavacamten administration

time point	LVEF (%)	LVOT-PGmax (mmHg)	NT-proBNP (ng/L)	heart rate(bpm)
Before medication administration (day 5 prior to surgery)	69	133	331.6	79
Day 5 after medication administration (preoperative)	—	79	—	—
Day 1 postoperatively	56	28	—	—
Day 5 postoperatively	—	—	52.4	—
1 month postoperatively	68	28	—	—

Note: "—" indicates that no corresponding data were collected at that time point. The failure to re-evaluate LVEF and NT-proBNP after preoperative medication constitutes a limitation of the data in this case.

Surgery and Postoperative Management

The patient underwent total resection of the meningioma under general anesthesia.The core intraoperative objectives were: strict heart rate control to avoid tachycardia exacerbating obstruction; maintenance of appropriate systolic blood pressure to ensure coronary and cerebral perfusion; and implementation of fluid balance management.Anesthesia induction was performed using etomidate, sufentanil, remifentanil, dexmedetomidine, and propofol. Norepinephrine was infused intraoperatively to maintain afterload, and no inotropic agents were used.[10] Intraoperative systolic blood pressure was maintained between 110–130 mmHg, heart rate 55–70 beats/min, central venous pressure 6–10 cmH2O. Mannitol was administered only 162.5 mL,Did not result in hypovolemia.Intraoperative blood loss was approximately 600 mL; no blood transfusion was performed.Gross total resection of the tumor was achieved, classified as Simpson grade I.

Postoperatively, the patient was transferred to the neurosurgical ICU.On the first postoperative day, LVEF was 56% and LVOT-PGmax was 28 mmHg; one month postoperatively, LVEF was 68% and LVOT-PGmax remained stable at 28 mmHg.On the first postoperative day, a cranial CT scan revealed hematoma, fluid collection, pneumocephalus, and brain parenchymal swelling, with a leftward shift of midline structures; on the seventh postoperative day, a CT scan showed increased fluid collection, necessitating vigilance for intracranial herniation.Postoperative brain swelling directly conflicts with the fluid management requirements for HOCM: neurosurgical protocols typically require fluid restriction and the use of dehydrating agents, whereas HOCM requires the maintenance of adequate preload.In this case, dual monitoring of cardiac function and neurological status was conducted in the ICU, with mannitol administration adjusted in real-time based on dynamic data; the patient gradually improved following conservative treatment, and no cerebral herniation occurred.Furthermore, given the postoperative placement of an epidural drainage tube, vigilance is required for intracranial hypotension syndrome resulting from excessive cerebrospinal fluid drainage.[11] Regarding thrombosis prevention, HOCM patients are prone to thrombosis due to left atrial enlargement and blood stasis; in this case, prophylactic anticoagulation was initiated after confirming the absence of active intracranial hemorrhage.[12]

At the 1-month postoperative follow-up, the patient's symptoms of dizziness and headache had basically disappeared, with a KPS score of 90 and NYHA functional class II.Contrast-enhanced cranial MRI confirmed gross total resection of the tumor, with no evidence of recurrence.Pathology report: skull base meningioma, WHO grade I.

A: Preoperative axial contrast-enhanced T1-weighted MRI revealed a large, uniformly enhancing mass in the anterior skull base (meningioma), closely adjacent to the right internal carotid artery and the A1 segment of the anterior cerebral artery, accompanied by peritumoral edema and leftward displacement of the midline structures. B: On the first postoperative day, axial non-contrast head CT showed hematomas, effusions, and pneumocephalus in the operative area, swelling of the right cerebral hemisphere parenchyma, compression and narrowing of the right lateral ventricle, and leftward displacement of the midline structures. C: On the fifth postoperative day, CT revealed reduced hematomas, effusions, and pneumocephalus compared to the previous scan, but liquefactive necrosis

and swelling were observed in the adjacent brain parenchyma. D: On the seventh postoperative day, CT demonstrated decreased density of the hematomas, resolution of pneumocephalus, and increased effusion volume; vigilance for the development of intracranial herniation was warranted.

Discussion

The core educational value of this case lies in the fact that surgical contraindications for HOCM complicated by meningioma are not fixed, but rather depend on the state of cardiac decompensation and the potential for pharmacological optimization.Unoptimized HOCM represents an absolute contraindication; however, patients who have achieved hemodynamic stability following optimization with mavacamten fall into a category of high-risk candidates eligible for screening.

The key difference between this case and previous reports of HOCM combined with brain tumor surgery lies in the preoperative use of mavacamten for heart-specific pharmacological optimization.[13] The standard treatment course recommended by guidelines ranges from several weeks to several months; however, in this case, due to the urgent mass effect of the meningioma, it was administered for only 5 days preoperatively. The EXPLORER-CN trial has already confirmed the short-term efficacy and safety of mavacamten in Chinese patients with oHCM.Postoperatively, the LVOT-PGmax decreased to 28 mmHg, a reduction of 105 mmHg compared to the preoperative value.[14] This significant improvement may not be solely attributable to mavacamten; factors such as reduced sympathetic tone following the relief of surgical stress, changes in volume status, and residual inhibition from anesthetic agents may also have contributed.Furthermore, the early postoperative LVEF decreased from 69% to 56%, which is consistent with the negative inotropic effect of mavacamten. This decline in LVEF is an expected pharmacodynamic effect rather than a deterioration in cardiac function, though continuous monitoring remains necessary to prevent excessive inhibition.

What warrants further discussion is the stratified management of drug-sensitive versus drug-insensitive patients.For instance, the patient in this case responded well to mavacamten, with the LVOT gradient decreasing from 133 mmHg to 79 mmHg after 5 days of treatment, classifying them as drug-sensitive; such patients may undergo surgery cautiously after medication optimization.However, approximately 30% of oHCM patients in clinical practice exhibit a poor response to mavacamten.[15] For drug-insensitive patients, one should not wait indefinitely for the medication to take effect, as this may delay tumor surgery.[16] In such cases, switching to the second-generation cardiac myosin inhibitor aficamten or combining disopyramide with beta-blockers may be considered; [17] if these remain ineffective, septal reduction therapy, including surgical septal myectomy or alcohol septal ablation, should be evaluated as early as possible.[16] Surgical septal myectomy remains a definitive treatment in experienced centers, but postoperative recovery requires several weeks; if the meningioma has already caused a significant mass effect, prioritizing tumor resection is more reasonable.[18] For patients with indications for pacing, DDD pacing may serve as an adjunctive option.It is evident that the timing and sequence of surgery for drug-sensitive and drug-insensitive patients should be managed differently, with the core objective being the timely completion of drug response assessment to formulate an individualized treatment plan.

Clinical Significance and Implications of This Case

First, redefine the boundaries of surgical contraindications.Unoptimized HOCM is an absolute contraindication, but this contraindication is not permanent.This case demonstrates that through pharmacological optimization (mavacamten significantly reduced the LVOT gradient in just 5 days), patients previously considered to have absolute contraindications can be converted to a surgical candidate status.Clinicians should not directly rule out the possibility of surgery based solely on the diagnostic label of 'HOCM', but should instead evaluate the decompensated state of the heart and the potential for pharmacological optimization.

Second, establish a replicable framework for MDT decision-making.The MDT model in this case comprises four core components: the Department of Cardiology completes risk stratification and medication optimization; the Department of Anesthesiology develops hemodynamic monitoring and anesthesia administration protocols; the Department of Neurosurgery assesses the surgical difficulty and hemorrhage risk of the tumor; and the Department of Critical Care Medicine plans postoperative monitoring strategies in advance.This framework can be generalized to other neurosurgical cases involving severe heart disease.

Third, identify the focal points of therapeutic contradictions.This case reveals three core contradictions: the conflict between mannitol-induced dehydration and the maintenance of preload, the conflict between the management of postoperative brain swelling and the volume management of HOCM, and the balance between hemostasis and anticoagulation.Clinicians must maintain vigilance regarding the aforementioned contradictions throughout the perioperative period and adjust strategies in real-time based on dynamic monitoring data.

Fourth, early postoperative identification of cardiac-neurological combined complications.In this case, brain swelling and midline shift occurred on the first postoperative day. It is necessary to distinguish between midline shift caused by exacerbated cerebral edema and that caused by cerebrospinal fluid hypovolemia, as the management strategies for these two conditions are diametrically opposed. This suggests that rigorous dual monitoring of cardiac function and neurological status is required postoperatively.

Fifth, stratified management of drug-sensitive and drug-insensitive patients.For drug-sensitive patients, surgery may be cautiously performed after medication optimization; for drug-insensitive patients, the feasibility of SRT should be evaluated in a timely manner.In clinical practice, drug response assessment and genetic testing should be completed as early as possible to guide individualized treatment decisions.[19]

Conclusion

Meningioma complicated by high-risk obstructive hypertrophic cardiomyopathy represents an extremely high-risk state in neurosurgery.In the absence of multidisciplinary assessment and pretreatment, it

should be considered an absolute surgical contraindication.However, for a small number of patients with clear surgical indications for the tumor and stable cardiomyopathy following optimization, meningioma resection may be cautiously performed after comprehensive multidisciplinary team (MDT) consultation, precise intraoperative hemodynamic management, and postoperative combined cardiac and neurological protection.Clinical decision-making must be individualized, with strict adherence to the boundaries of surgical patient selection.For drug-sensitive patients, surgery may be cautiously performed after optimization with medications such as mavacamten; for drug-insensitive patients, the feasibility of septal reduction therapy should be evaluated in a timely manner rather than indefinitely delaying surgery.

Declarations

Ethics approval and consent to participate

This case report was conducted in accordance with the ethical principles of the Declaration of Helsinki. Ethical approval was obtained from the Ethics Committee of The First People's Hospital of Yunnan Province. Written informed consent for participation was obtained from the patient.

Consent for publication

Written informed consent for publication of this case report and any accompanying images was obtained from the patient. A copy of the consent form is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare that they have no competing interests.

Clinical trial number

Not applicable.

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Author Contribution

CJ and SZ conceived and designed the case report. CJ and DZ collected and analyzed the clinical data. YZ and SL performed the literature review. LX provided critical revision of the manuscript. CJ drafted the manuscript. SZ supervised the project and revised the manuscript. All authors read and approved the final manuscript.

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Availability of data and materials

The datasets generated and analyzed during the current study are not publicly available due to patient privacy, but are available from the corresponding author on reasonable request.

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Figures

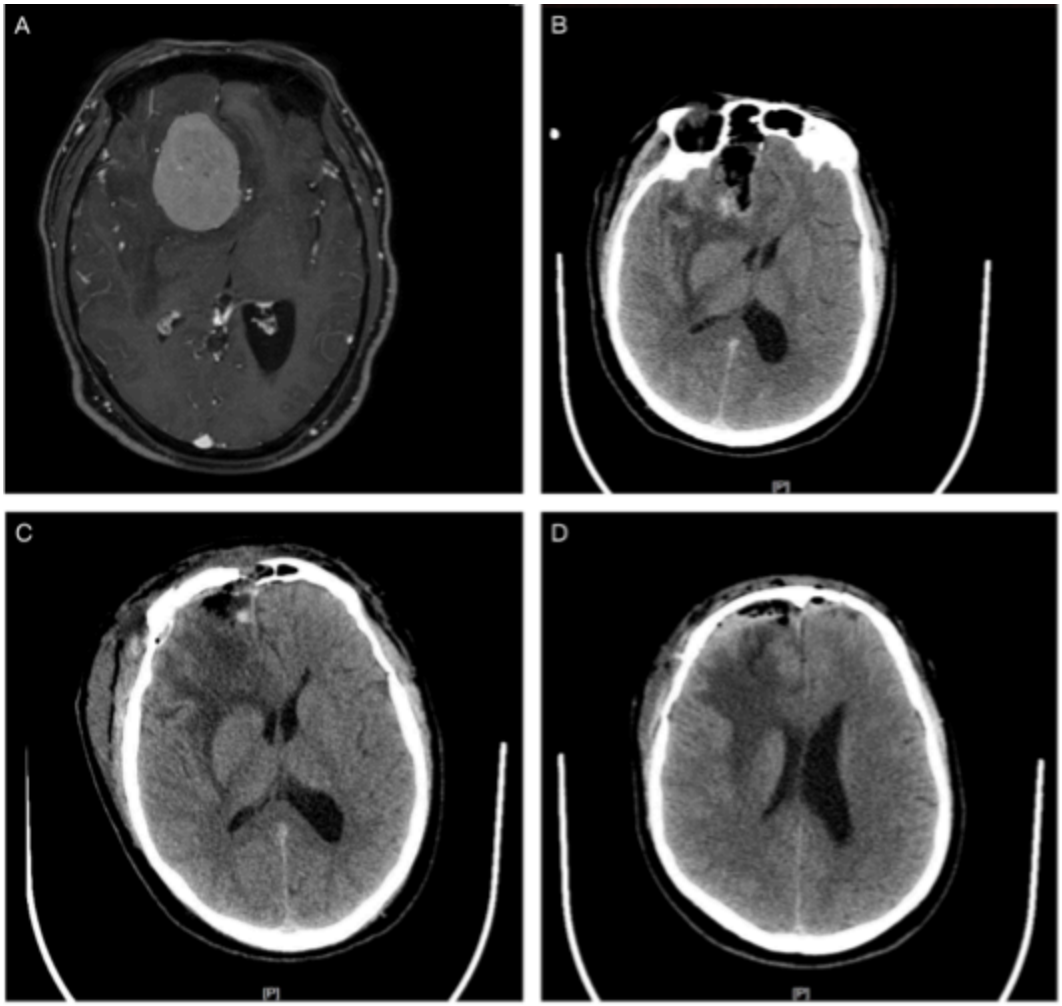


Figure 1

Perioperative imaging evolution in a patient with meningioma and obstructive hypertrophic cardiomyopathy.

A: Preoperative axial contrast-enhanced T1-weighted MRI revealed a large, uniformly enhancing mass in the anterior skull base (meningioma), closely adjacent to the right internal carotid artery and the A1 segment of the anterior cerebral artery, accompanied by peritumoral edema and leftward displacement of the midline structures. B: On the first postoperative day, axial non-contrast head CT showed hematomas, effusions, and pneumocephalus in the operative area, swelling of the right cerebral hemisphere parenchyma, compression and narrowing of the right lateral ventricle, and leftward displacement of the midline structures. C: On the fifth postoperative day, CT revealed reduced hematomas, effusions, and pneumocephalus compared to the previous scan, but liquefactive necrosis and swelling were observed in the adjacent brain parenchyma. D: On the seventh postoperative day, CT demonstrated decreased density of the hematomas, resolution of pneumocephalus, and increased effusion volume; vigilance for the development of intracranial herniation was warranted.