Perioperative Management of Catecholamine-Secreting Glomus Jugulare Tumors

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Introduction

Glomus tumors are benign vascularized tumors that arise from paraganglion cells throughout the body. These tumors are also referred to as paraganglioma or chemical receptor tumors, and they often develop in the tympanic cavity and jugular bulb. Early reports of excision of glomus tumor include those by Guild in 1941,¹ Rosenwasser in 1945,² and Winship et al in 1948,³ but surgical treatment of this tumor is still considered challenging because of its association with surrounding structures. Catecholamine-secreting glomus tumors account for ~1 to 8% of all cases of glomus tumor.⁴–⁷ Perioperative catecholamine hypersecretion causes the most problems in the treatment of a catecholamine-secreting glomus tumor and may lead to intraoperative death. Therefore, screening of catecholamine secretion is necessary for all patients with a glomus tumor.⁴,⁷–⁹ Preparation for surgery follows that for pheochromocytoma surgery and requires assistance of the departments of endocrinology, anesthesiology, and endocrine surgery departments. We describe our perioperative management for catecholamine-secreting glomus jugulare tumor. The patient was a 31-year-old woman with a 50-mm glomus jugulare tumor and a significantly elevated plasma noradrenaline level of 21,165 pg/ml. Before the surgery, oral α-blocker administration was initiated for ~3 months, and her body weight increased from 52 kg at the time of examination to 54.2 kg. Coil embolization of the tumor vessel was performed 1 week before surgery, and the intense tumor stain was reduced by 90%. The patient underwent almost total removal of the tumor via mastoidectomy with high cervical exposure via the transsigmoid approach. Postoperatively, plasma noradrenaline decreased markedly. Preoperative pharmacologic stabilization and peri- and postoperative anesthetic management are essential for the treatment of a catecholamine-secreting glomus jugulare tumor.

Keywords
► glomus jugular tumor
► pheochromocytoma
► catecholamine
► surgical strategy
► skull base surgery

Abstract

To treat patients with a catecholamine-secreting glomus jugulare tumor, perioperative management is important. Perioperative catecholamine hypersecretion causes severe problems in the treatment of a catecholamine-secreting glomus tumor. Therefore, a precise therapeutic strategy and perioperative management are required through collaboration of the endocrinology, anesthesiology, and endocrine surgery departments. We describe our perioperative management for catecholamine-secreting glomus jugulare tumor. The patient was a 31-year-old woman with a 50-mm glomus jugulare tumor and a significantly elevated plasma noradrenaline level of 21,165 pg/ml. Before the surgery, oral α-blocker administration was initiated for ~3 months, and her body weight increased from 52 kg at the time of examination to 54.2 kg. Coil embolization of the tumor vessel was performed 1 week before surgery, and the intense tumor stain was reduced by 90%. The patient underwent almost total removal of the tumor via mastoidectomy with high cervical exposure via the transsigmoid approach. Postoperatively, plasma noradrenaline decreased markedly. Preoperative pharmacologic stabilization and peri- and postoperative anesthetic management are essential for the treatment of a catecholamine-secreting glomus jugulare tumor.
Case Presentation

History and Examination

The patient was a 31-year-old woman who visited a university hospital for suspected extra-adrenal pheochromocytoma based on symptoms of headache, weight loss, and juvenile hypertension. She was diagnosed with glomus jugulare tumor and referred to our hospital for surgery. On admission, a swallowing disorder and immobilization of the right vocal cord were noted on physical examination. Laboratory tests showed a plasma noradrenaline level of 21,165 pg/ml (normal: < 100 pg/ml), a urinary noradrenaline level of 2,849 μg/ml (normal: 48.6–168.4 μg/ml), and a urinary vanillylmandelic acid (VMA) level of 10.4 mg/day (normal: 1.5–4.3 mg/day).

Contrast magnetic resonance imaging (MRI) revealed a 50-mm dumbbell-shaped tumor extending from the right jugular foramen to the right parapharyngeal space (Fig. 1A).

Bone window computed tomography (CT) scans showed destruction of the jugular foramen and carotid canal (Fig. 1B). Angiography showed an intense tumor stain that was mainly fed by the right ascending pharyngeal artery, and the internal carotid artery was narrowed and translocated anteriorly (Fig. 2). The right sigmoid sinus-interna jugular vein was also occluded by the tumor. The lesion was a dumbbell-type tumor located mainly in the jugular foramen and extending to the right parapharyngeal space, with the upper and lower ends present in the internal auditory meatus and C4, respectively. The tumor did not extend into the middle ear space. The Fisch classification was De1, the Glasscock-Jackson classification was glomus jugulare type II.

Preoperative Preparation

Preoperative preparation was performed by the departments of endocrinology, surgery, and anesthesiology, with reference to perioperative management of pheochromocytoma.
Administration of an oral α-blocker (doxazosin mesylate) was initiated 3 months before surgery, and the resting blood pressure decreased from 180/100 to 120/60 mm Hg. Body weight increased against the reduced relative circulating plasma volume from 52 kg at the time of examination to 54.2 kg before surgery. Use of contrast medium is contraindicated for pheochromocytoma, but contrast MRI and preoperative endovascular embolization are essential for surgery for a glomus tumor. Thus we performed the examination with continuous infusion of magnesium sulfate intravenously, with the goal of inhibiting catecholamine oversecretion induced by contrast medium injection. Coil embolization of the tumor vessel was performed 1 week before surgery, and the intense tumor stain was reduced by 90% (Fig. 2). The plasma catecholamine level was also reduced after embolization (Fig. 4B).

Surgical Intervention and Intraoperative Management
Surgery was performed via a mastoidectomy with high cervical exposure via the transsigmoid approach, and almost total tumor resection was achieved. Total intravenous anesthesia was used for both of our patients, and a central venous line was inserted in our case. Because the preoperative catecholamine level was very high, blood pressure elevation before tumor resection was treated with isosorbide dinitrate and nicardipine. The blood pressure elevated when the tumor was directly manipulated, and cardiac arrest occurred for ~5 seconds. Blood pressure reduction started when >60% of the tumor was resected, for which isosorbide dinitrate was withdrawn and noradrenaline was initiated. Stable anesthesia management was possible using this approach.

Postoperative Course
After the operation, the patient was managed in the intensive care unit until the following day. There was no hypotension or hypoglycemia due to reduction of the plasma catecholamine level. Mild lower cranial nerve symptoms developed, and several days were necessary before extubation was possible. Postoperative MRI showed that the tumor was almost totally resected. No facial palsy or hearing impairment occurred after surgery (Fig. 3). The swallowing disorder remained but was improved after ~5 months of rehabilitation, and the patient was discharged to home. A postoperative endocrine test showed marked reductions of the plasma noradrenaline level and urinary VMA and normetanephrine levels, and hypertension was also improved (Fig. 4A, B).

Discussion
Preoperative Preparation
The relative circulating plasma volume is reduced due to vascular constriction caused by excess catecholamines in plasma in patients with a catecholamine-secreting glomus tumor. When surgery is performed without correcting the circulating blood volume, and the blood catecholamine level decreases after tumor resection, hypotension can be caused even by a small volume of hemorrhage and may result in shock, in which maintenance of circulatory dynamics is difficult. This is an important difference between this tumor and normal pheochromocytoma, and volume loading to treat decreased blood pressure after tumor excision should be carefully performed to avoid intracranial pressure elevation. In surgery for pheochromocytoma, an α-blocker, selective α1 receptor blocker, and α- and β-blockers are administered for >2 weeks before surgery to dilate peripheral blood vessels and increase the circulating plasma volume to prevent intraoperative abnormal hypertension and collapse of circulatory dynamics after tumor excision.11,12

Preoperative Embolization of Tumor Vessels
Preoperative embolization of tumor vessels is mandatory in surgery for a glomus tumor, but caution is necessary when the tumor is the catecholamine-secreting type. Blood pressure variation is common in patients with a catecholamine-secreting tumor during angiography and tumor vessel embolization, and cardiac arrest cases have been reported. This shows that embolization is not necessarily safe. However, in our case, it was interesting that the embolization of tumor
Fig. 3 Postoperative neuroradiologic findings in case 1. Upper left: bone window computed tomography scans. Upper right and lower: contrast-enhanced T1-weighted magnetic resonance images.

Fig. 4 Serial changes of plasma noradrenaline and urinary vanillylmandelic acid levels. POD, postoperative day.
vessels decreased the plasma noradrenaline level (► Fig. 4B). For the perioperative management, the reduction of a plasma noradrenaline level obviously makes the surgery safer. As long as the embolization is performed with informed consent and careful management, it may be useful management for these tumors.

**Intraoperative Management**

Anesthesia in surgery for a catecholamine-secreting glomus tumor carries a high risk of catecholamine oversecretion—associated fatal arrhythmia and blood pressure instability induced by surgery, similarly to surgery for pheochromocytoma. The several other risks include intracranial pressure control for cases with intracranial extension, air embolism accompanying ligation of the internal jugular vein and sigmoid sinus, and management of a hemorrhagic tumor. Vasopressor administration is also needed because the blood pressure tends to decrease after resection due to reduction of the catecholamine level, although the blood pressure tends to elevate with tumor manipulation during resection.

For intraoperative blood pressure management, drugs with a rapid effect, such as sodium nitroprusside and calcium channel blockers, have been used in many cases. Use of magnesium sulfate has been reported in anesthesia management for cases of pheochromocytoma and catecholamine-secreting glomus tumor, but the number of reports is limited. Magnesium sulfate inhibits catecholamine secretion from both adrenal medulla and peripheral sympathetic nerve endings, directly inhibits catecholamine receptors, and prevents vascular constriction by acting on vascular smooth muscle. It may also have a neuroprotective effect by acting as an N-methyl-D-aspartate antagonist. Goutcher et al found that magnesium sulfate was appropriate for anesthesia in patients with a catecholamine-secreting glomus tumor that has advanced into the skull because it minimizes intraoperative blood pressure instability, but has a favorable effect on intracranial surgery.

**Postoperative Management**

In management after surgery for a catecholamine-secreting tumor, treatment is required for hypotension and hypoglycemia induced by a rapid decrease in catecholamines. Because surgery for glomus jugulare tumor may also induce lower cranial nerve symptoms, extubation should be performed after careful evaluation.

**Conclusion**

To treat catecholamine-secreting glomus jugulare tumors safely, a precise perioperative management requires preparation similar to that for surgery for pheochromocytoma, with cooperation among departments of endocrinology, anesthesiology, and endocrine surgery.

**References**