

## CASE REPORT

### Glomus Faciale: Primary Paraganglioma of the Facial Canal

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Primary glomus tumor of the facial canal is a rare entity. We report the glomus tumor originating from the facial nerve, which is the largest intratemporal glomus faciale and the first with demonstration of the pathologic evidences. The patient had an 8-year history of facial fasciculation followed by facial weakness and pulsatile tinnitus. After complete surgical removal of the tumor, the pathologic finding revealed a paraganglioma which was encasing nerve tissues without a penetration into the jugular bulb. Although the incidence is very low, glomus faciale tumor should be considered in the differential diagnosis of progressive facial weakness with pulsatile tinnitus.

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#### Introduction

Glomus faciale is a rare tumor arising from the glomus bodies in the temporal bone, which is found within the facial canal.<sup>[1]</sup> Glomus complex tumors, also known as nonchromaffin paragangliomas or chemodectomas, are the second most common neoplasm seen in the temporal bone after vestibular schwannomas.<sup>[2]</sup> They arise from the glomus body, which is composed of chemoreceptor cells derived from developing neural crest tissue. Glomus bodies are usually found in the adventitia of the jugular bulb and along the Jacobson's and Arnold's nerves. At least one branch of the Arnold's nerve ascends the fallopian canal through a mastoid canaliculus. Guild observed glomus formations along this branch of vagus nerve within the vertical portion of the facial canal, which explains the occurrence of primary facial canal paragangliomas.<sup>[1]</sup>

Since Dutcher et al. reported the first case in 1986, only seven cases of glomus tumors primarily originating from the fallopian canal have been reported.<sup>[2-7]</sup> We report a case of large paraganglioma strongly suggesting glomus faciale with supporting radiologic and pathologic findings.

#### Case Report

A 45-year-old woman presented with a 4-year history of right-sided pulsatile tinnitus. The patient had had intermittent fasciculations of her right lower eyelid for the past 8 years. She visited a local clinic 7 years ago, and laboratory work and a computed tomography (CT) scan of brain failed to reveal any abnormal findings except for iron-deficiency anemia. Two years later, the patient noticed right-sided facial weakness which had been gradually worsening regardless of acupuncture treatment. At the time of visit to our clinic, she complained of loud pulsatile tinnitus which was severely annoying. There was no one in her family who had similar symptoms or had been diagnosed with a familial disease.

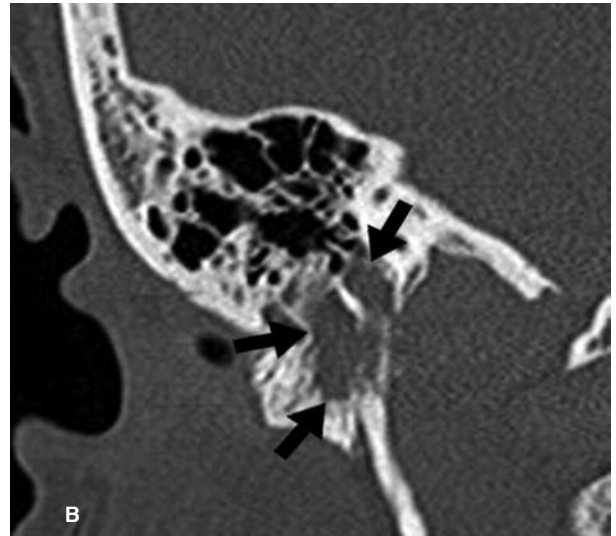
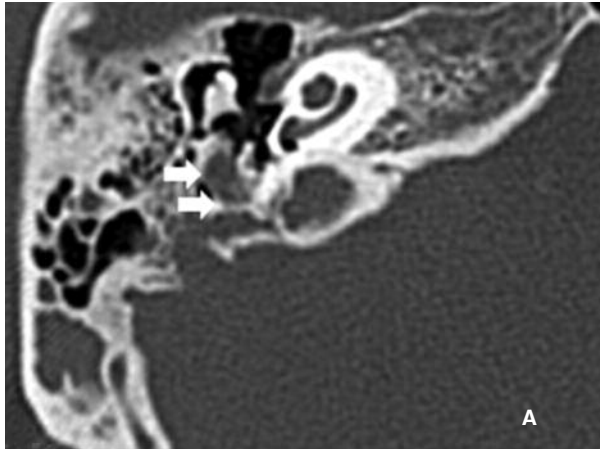
Physical examination showed right-sided facial nerve palsy of House-Brackmann grade III. Her face was not disfiguring and showed complete eye closure and asymmetric mouth movement. Pulsatile tinnitus was not alleviated by carotid or internal jugular vein compression. Otoendoscopic examination and routine head and neck examinations showed no abnormal findings. Pure tone audiometry revealed a normal hearing.

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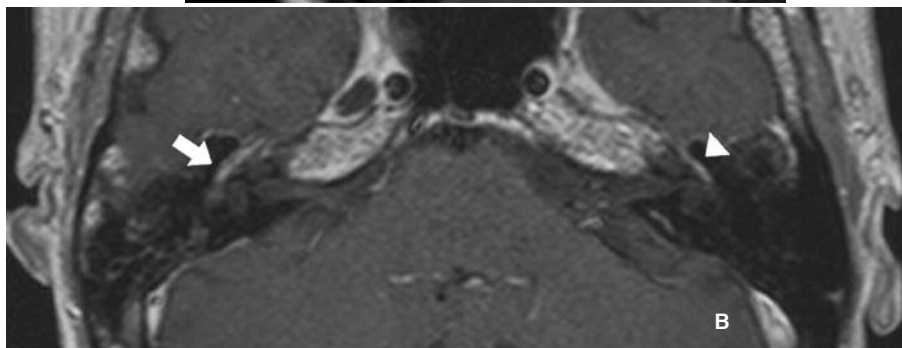
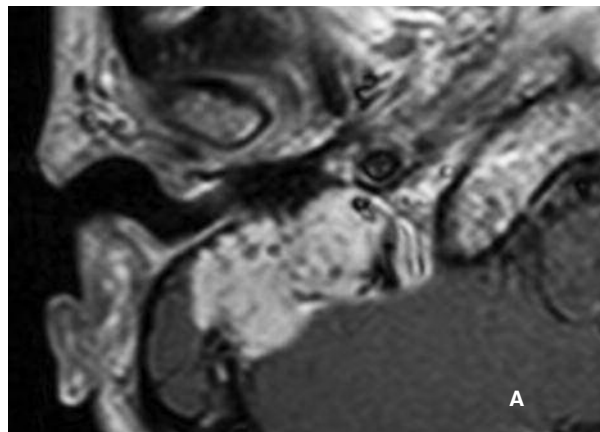
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A CT scan of the temporal bone showed an enlargement of the right facial canal from the tympanic portion to the stylomastoid foramen (Figure 1). The middle ear cavity and the ossicles exhibited unremarkable findings. A contrast-enhanced CT scan of the temporal bone demonstrated a well-enhancing and ill-margined destructive mass with a size of 2.5

x 1.5 x 2.5cm. The tumor occupied the right mastoid air cells, and, inferiorly, partially compressed the adjacent jugular bulb. Magnetic resonance imaging (MRI) of the temporal bone revealed a hypervascular tumor along the course of the right intratemporal facial nerve (Figure 2).



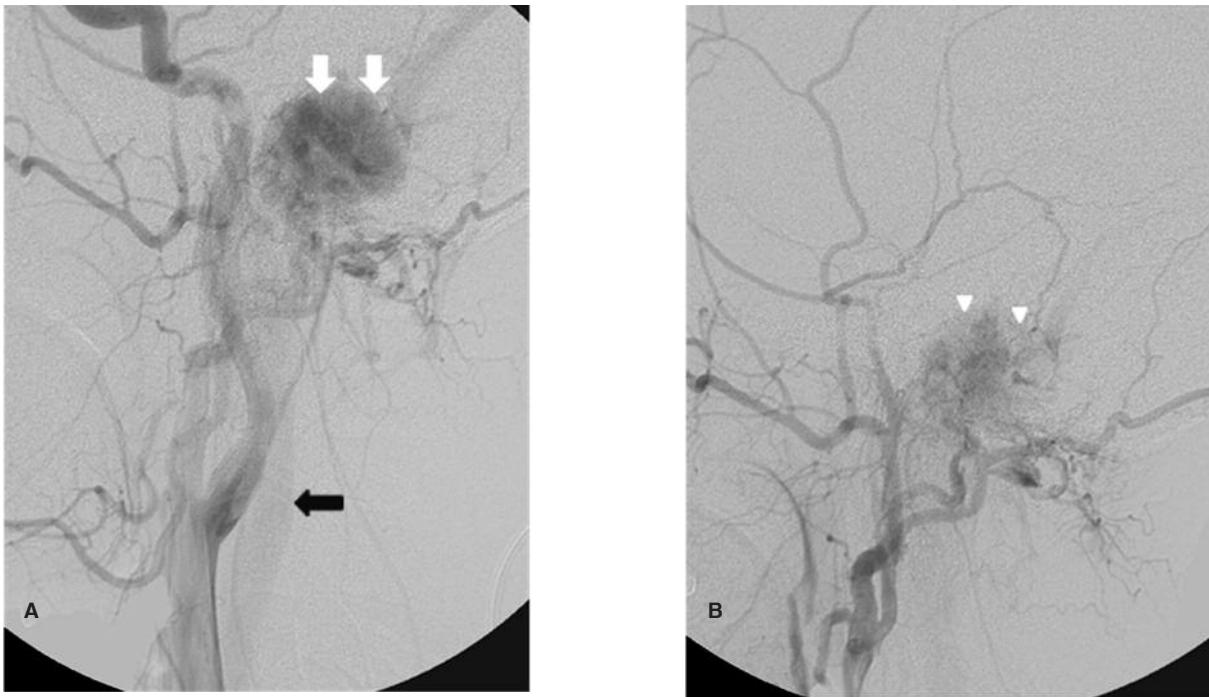
**Figure 1.** Computed tomography (CT) image of temporal bone. **(A)** Axial scan shows widening of the descending portion of facial canal (white arrows). **(B)** Coronal image shows the destructive mass expanding the descending portion of facial canal (black arrows).



**Figure 2.** Axial magnetic resonance (MR) scan of the temporal bone. T1-weighted postcontrast MR image demonstrates **(A)** well-enhancing tumor of the right mastoid air cell and jugular fossa, and **(B)** enlarged intratemporal segments of facial nerve on the right side (white arrow) compared to the left (arrowhead).

Laboratory work, including serum catecholamine levels, revealed no abnormal findings except for anemia. Preoperative angiography of the common carotid artery identified a hypervascular tumor fed from small branches of the right external carotid artery (Figure 3A). A branch of the right occipital artery was

selected as a main feeding artery and was embolized successfully (Figure 3B). There was early filling of the right internal jugular vein due to arteriovenous shunting, which is a characteristic of glomus tumors. The left carotid artery had a normal blood flow.



**Figure 3.** Angiography of the right common carotid artery. **(A)** A densely staining mass (white arrow) was fed by the right occipital artery. Note the early filling of the right internal jugular vein (black arrow). A branch of the right occipital artery was selected as a main feeding artery. **(B)** After angiographic embolization, vascularity of the tumor markedly decreased (arrowheads)..

The patient subsequently underwent tumor resection via a retroauricular approach to the posterior fossa and the jugular foramen, including blind-sac closure of the external auditory canal, mastoidectomy, identification of the proximal and distal facial nerve and preservation of the lower cranial nerves. It was impossible to separate the tumor from the facial nerve, so the facial nerve segment from the tympanic segment to the proximal extracranial segment was removed along with the tumor. The jugular bulb was also removed with the tumor. The resection margins of the proximal and distal ends of facial nerve were free of tumor in frozen sections. There was a moderate amount of bleeding during the procedure, despite the successful preoperative embolization. The facial nerve was

reconstructed with a 4-cm cable graft using the ipsilateral greater auricular nerve. A free abdominal fat graft was harvested and packed in the operative cavity, and the posterior part of the temporalis muscle was rotated to cover the defect.

Histological examination of the specimen showed a typical appearance of paraganglioma (Figures 4A, 4B) with findings which are highly suggestive of a primary facial nerve paraganglioma with extension into the jugular fossa. The tumor was encasing the nerve tissues (Figure 4C). The margin between the tumor and the jugular bulb was clearly discrete, though the tumor cells were adjacent to the jugular bulb. Areas of chronic inflammation with necrotic change were found between the tumor cells and the jugular bulb wall



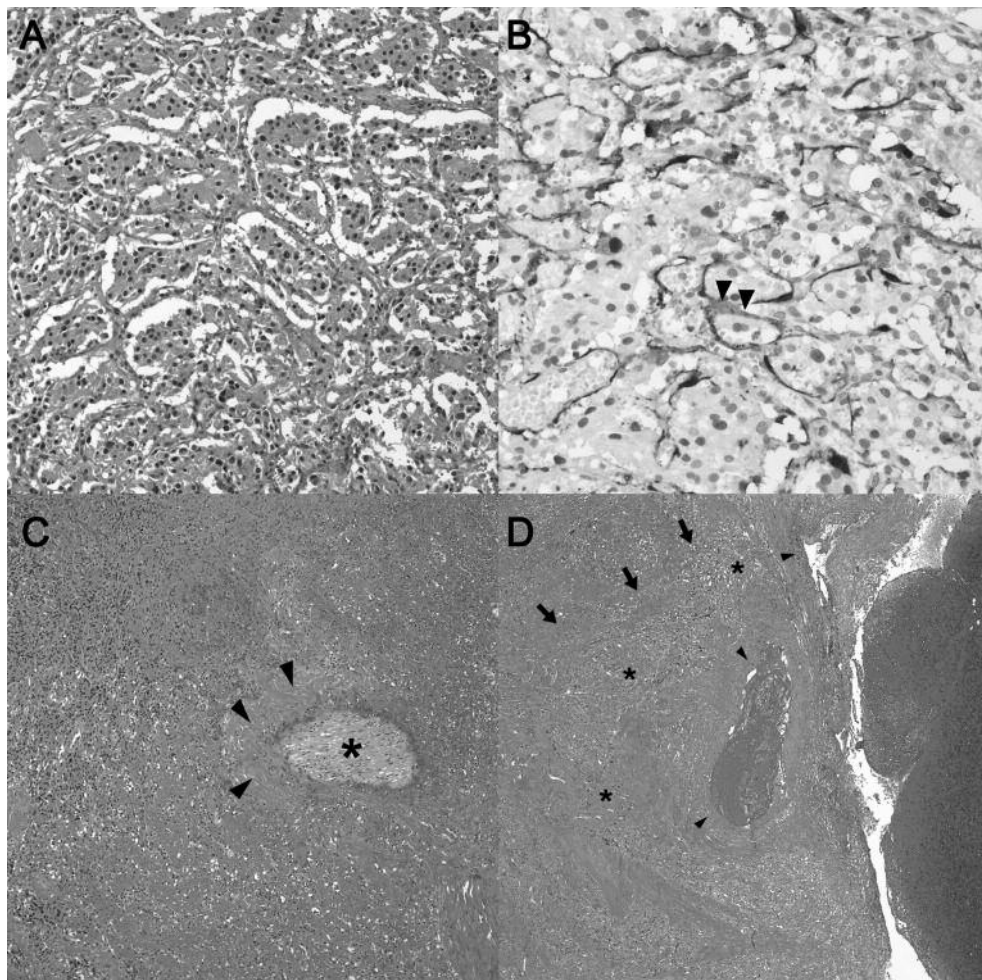
(Figure 4D). Postoperatively, pulsatile tinnitus disappeared and cranial nerve functions showed no deficits except for large conductive hearing loss and complete facial nerve paralysis.

### Discussion

Guild described paragangliomas of the temporal bone as vascular tumors arising from the glomus bodies which are ovoid shaped chemoreceptor cells derived from developing neural crest tissue of the autonomic nervous system.<sup>[8]</sup> Glomus bodies ordinarily follow the path of the Jacobson's and Arnold's branches of the

glossopharyngeal and vagus nerves, respectively.<sup>[1,4]</sup> According to the Guild's observation, a branch of Arnold's nerve ascends to the facial canal, and glomus formations were found along this branch of vagus nerve within the vertical portion of the facial canal, which explains the occurrence of primary facial canal paragangliomas.<sup>[1]</sup>

In the presenting case, imaging studies revealed a hypervascular tumor widening the right facial canal from the tympanic to the mastoid segments, occupying the right mastoid air cells with permeable osteolysis,



**Figure 4.** Histologic findings of the surgical specimen. **(A)** H&E staining showing a cell component of paraganglioma represented by the nest of small cells with round, sometimes pleomorphic nuclei and homogenous, eosinophilic cytoplasm (x 400). **(B)** Immunohistochemical staining for S-100 protein showing strongly reactive sustentacular cells forming a peripheral coat, "Zellballen" configuration (arrowhead) (x 400). **(C)** H&E staining showing the paraganglioma cells are surrounding the facial nerve (asterisk) with necrotic change (arrowheads) in-between them (x 40). **(D)** The folded jugular bulb wall (arrowheads) is seen completely separated from the tumor cells (black arrows). There are areas of chronic inflammation with necrotic change between the tumor cells and the jugular bulb wall.(x 40).

and partially compressing the adjacent jugular fossa. Since the facial nerve was admixed with the mass from the mastoid segment, the distal facial nerve could not be traced in the imaging. The tumor extended extratemporally with further widening of the stylomastoid foramen. The middle ear cavity and the parotid gland were free of lesions. These findings could explain the patient's symptom of pulsatile tinnitus and also the objective findings of facial weakness, normal hearing and a normal tympanic membrane. Based on these findings, glomus faciale tumor is considered as the primary possibility.

Jugulotympanic paragangliomas usually derive their blood supply from the ascending pharyngeal arteries.<sup>[9,10]</sup> However, in this case, preoperative arteriography showed a hypervascular mass principally fed by a branch of the occipital artery. In a review of two previous reports of facial canal paragangliomas, their feeding arteries were the posterior auricular branches of the occipital artery and the posterior auricular artery.<sup>[2,7]</sup> Therefore, the arteriographic findings may be helpful in the differential diagnosis of paraganglioma.

Pathologic findings of the specimen showed that the facial nerve was surrounded by the tumor cells and necrotic debris. Inferiorly, although the jugular bulb was compressed by the tumor, microscopic review revealed that the tumor cells were far from the jugular bulb wall. Areas of chronic inflammation with necrotic change were found between the tumor cells and the jugular bulb wall (Figure 4D).

In a review of seven previous reports, all glomus faciale tumors were small (less than 2cm) enough to locate within or around the facial canal. However, in this case with a large intratemporal tumor compressing the adjacent jugular bulb, an alternative possibility of glomus jugulare with subsequent invasion along the facial canal should be considered before the confirmative diagnosis.

There are several evidences in this case favoring a glomus faciale over a glomus jugulare. First, the clinical presentation of the patient is supporting glomus faciale. She complained of 8 years of facial

fasciculation, 6 years of facial weakness, and 4 years of pulsatile tinnitus. The symptom of facial nerve compression preceded the symptom caused by a growth of glomus tumor. Second, the imaging studies showed diffuse expansion of the right facial canal from the tympanic segment to the proximal extracranial segment. Although the mass was compressing the jugular bulb, the main mass was centered on the facial canal, not in the jugular bulb (Figure 1, Figure 2). Furthermore, growth pattern of the tumor which is expanding laterally to the mastoid air cells, vertically along with the facial canal, and sparing the middle ear cavity without any contact with the internal carotid artery is not typical of glomus jugulare. Third, the pathologic review showed no tumor cell contact to the jugular bulb wall, while the facial nerve was indistinguishable from the tumor in microscopic finding. Lastly, as described above, arteriographic finding of tumor feeding vessel may support the diagnosis of glomus faciale tumor.

The recommended treatment for glomus tumor is complete surgical excision with preservation of vital neurovascular structures. Radiation therapy may be considered but still remains controversial.<sup>3</sup> In the presenting case, as the patient was young and was suffering from pulsatile tinnitus, alleviating the disturbing tinnitus by complete tumor removal was her primary concern.

In patients with progressive facial weakness with or without pulsatile tinnitus, glomus faciale should be considered as one of the differential diagnosis. Radiologic studies including CT, MRI and angiography provides indispensable information and the pathologic investigation may be required for the confirmation of a large glomus faciale tumor with extension into the jugular fossa.

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