

# CLASSICAL GLOMUS JUGULARE RESULTING IN UNILATERAL TONGUE ATROPHY

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## ABSTRACT

Glomus jugulare tumors arise from the paraganglionic cells in jugular bulb. These are very rare tumors with a reported incidence of 1 in 1 million people. We are presenting a case of 42 years old male with 03 months history of headache and slurred speech with atrophy of right side of tongue. CT, MRI and four vessel angiogram confirmed the diagnosis of right sided glomus jugulare.

## Case

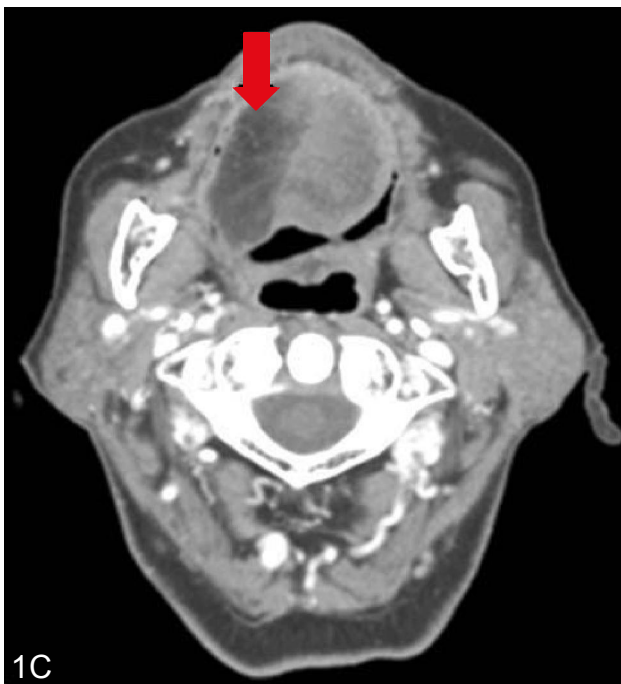
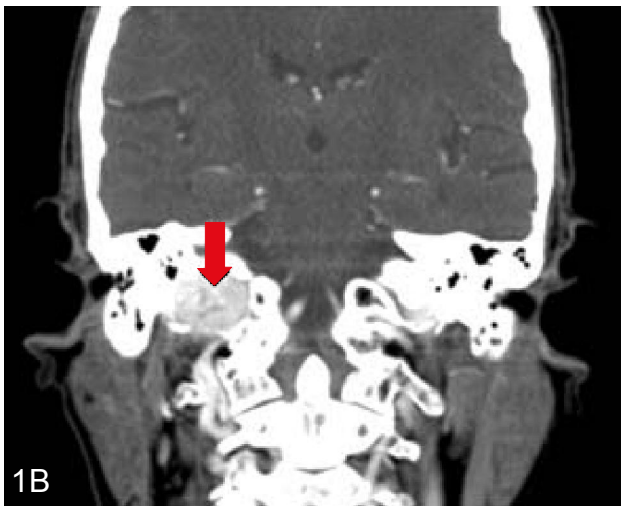
42 year old male patient presented to the emergency department with complaints of severe headache, slurring of speech with difficulty in articulation and hoarseness. No history of otorrhea was given, however patient complained of tinnitus on and off. The overall duration of symptoms was approximately three months. There was no history of otalgia. Blood and urine analysis revealed no significant abnormality. Examination of oral cavity was done as patient complained of difficulty in articulation which revealed atrophy of right half of tongue. Otherwise the neurological examination was unremarkable. On account of tongue atrophy, a mass lesion of jugular fossa was suspected. The patient was further investigated by carrying out CT scan (Fig. 1) and MRI (Fig. 2) of the head which confirmed the presence of a glomus jugulare on the right side.

A four vessel angiogram was later on performed which showed a large highly vascular lesion on right side (Fig. 3). The lesion had dual supply, with external carotid artery (ECA) supplying approximately 70% of the lesion while remaining 30% received its supply from right vertebral artery. Very

minimal supply was also seen from the right anterior inferior cerebellar artery.



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**Figure 1A, B:** CT scan of head and neck region axial and coronal sections showing a, 27 X 23 X 20mm sized intensely enhancing soft tissue mass lesion in the region of the right jugular foramen (Red arrow), causing enlargement and erosion of jugular foramen.

The lesion is abutting the internal carotid artery however not invading it. Jugular vein is compressed by the lesion and indistinct from this mass.

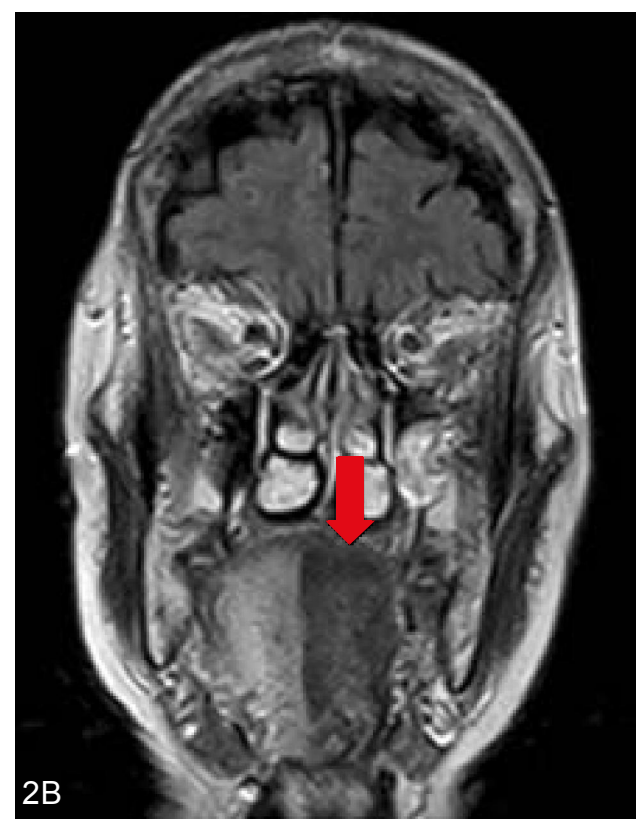
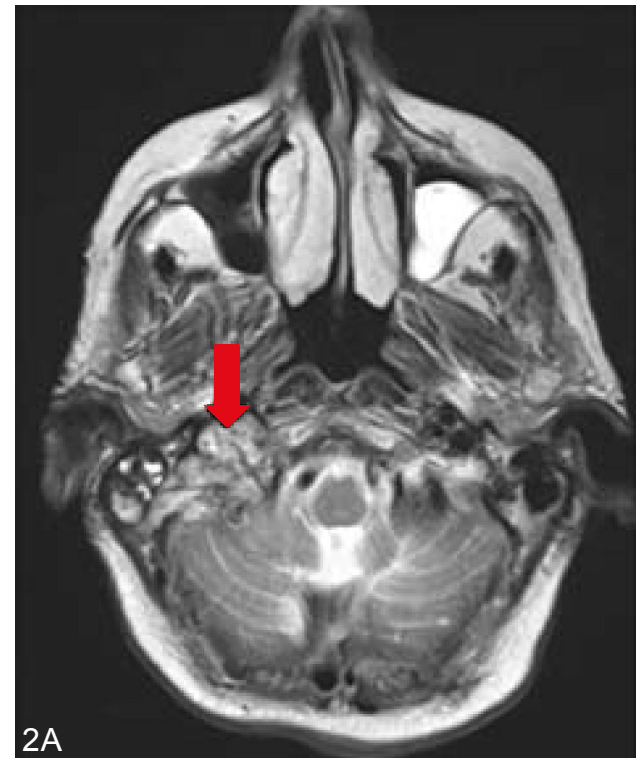
**1C:** Fatty replacement of right half of tongue muscles suggestive of paralysis of right hypoglossal nerve

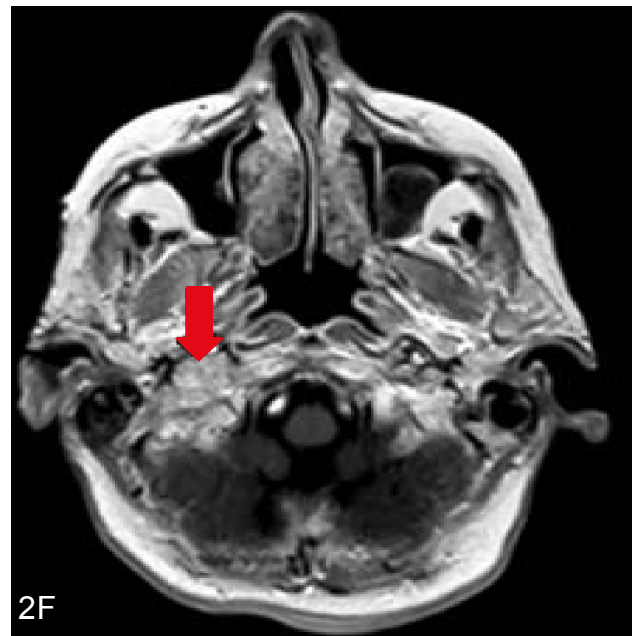
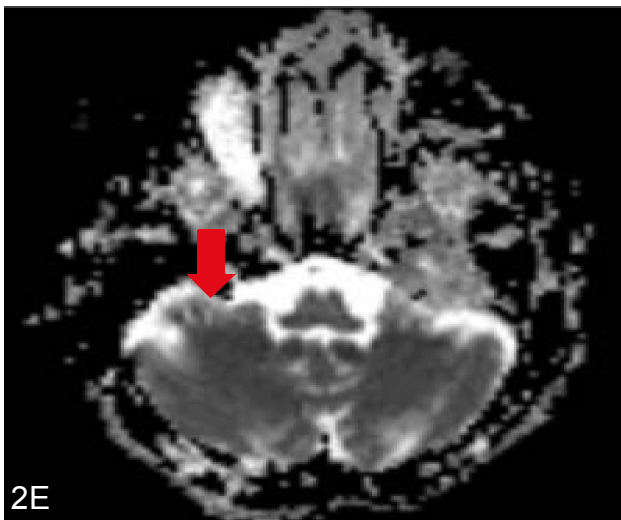
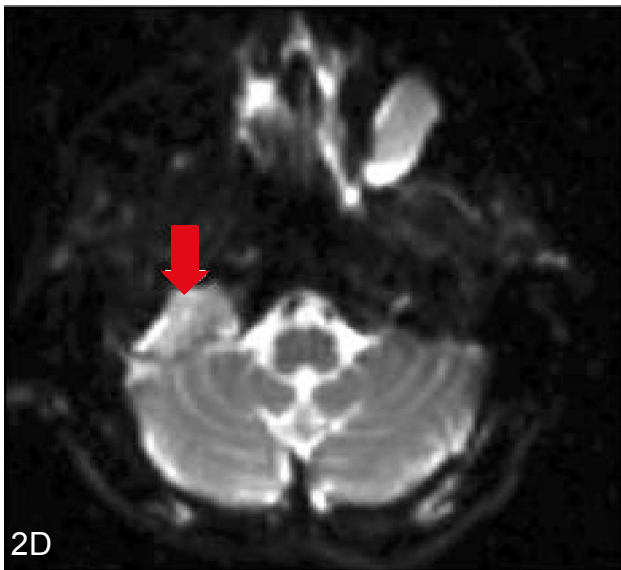
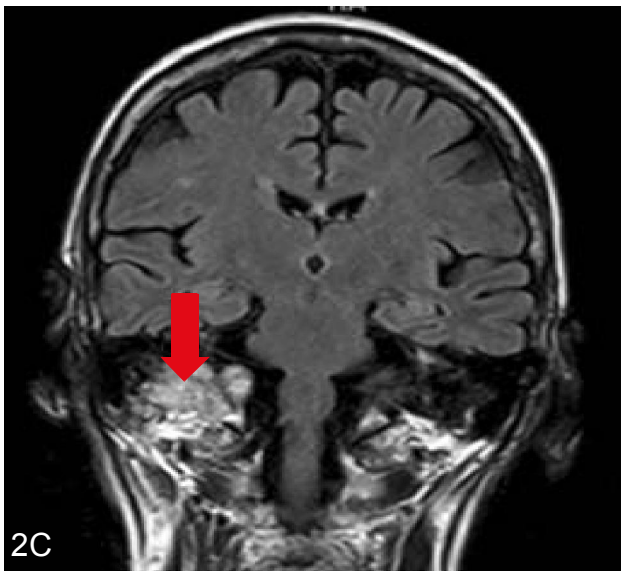
There was no supply from right internal carotid, left external carotid and left vertebral artery.

Venous drainage was via multiple collateral veins into the jugular system.

Two major feeders from right external carotid artery were super selectively cannulated and embolized

with PVA particles and gel foam. Further the patient was planned for surgery.





**Figure 2A:** T2 weighted axial sequence showing abnormal signal intensity lesion seen in the region of the right jugular foramen appearing hyper intense on T2-weighted axial image. The bright appearance representing the high vascularity in the mass, and the hypo intense areas are flow voids. This is the characteristic “salt and pepper” pattern of these lesions.

**2B:** T2 weighted coronal sequence showing hyper intense right half of tongue muscles appearing Hypo intense on coronal image suggestive of fatty replacement due to paralysis of right hypoglossal nerve.

**2C:** FLAIR coronal

**2D:** Diffusion weighted (DW)

**2E:** Apparent diffusion coefficient image (ADC)

**2F:** T1 Post contrast axial



**Figure 3:** Abnormal blush of contrast on the right side consistent with appearances of a highly vascular lesion (White arrow, Glomus jugulare). The catheter (small arrow) is in the branch of external carotid artery.

## Discussion

The term glomus was used to describe the rich presence of vascular channels and nerves seen in these masses.<sup>1</sup> Glomus jugulare tumors arise from the paraganglionic cells in jugular bulb. Valentin and Krause gave first descriptions of temporal bone paraganglionic tissues.<sup>2</sup> The term glomus jugulare was first used by Winship and Louzan.<sup>3</sup> Guild described it in 1941 as being similar to carotid body histologically at a meeting of anatomists in Chicago.<sup>1</sup> Glomus jugulare is part of the parasympathetic paraganglial system, which mediates chemosensory reflexes affecting breathing, blood pressure and pulse frequency. This system includes carotid and aortic bodies as well as glomus tympanicum and ganglion jugulare; the later are found along the 10<sup>th</sup> and 11<sup>th</sup> cranial nerves respectively. (Along Jacobson's and Arnold's nerves). The blood supply is derived from the ascending pharyngeal branch of the external carotid artery.<sup>4</sup> Glomus jugulare tumours are not frequent and their incidence is estimated to be 1 in 1 million people.<sup>5</sup>

Although rare, and benign they still can pose undue confusion to the radiologist and otologist because of their tendency to erode temporal bone and invade intracranially as well as surrounding structures such as middle ear, jugular vein, clivus, internal carotid artery, cavernous sinus and cranial nerves. It commonly affects middle age females. Malignant variations have been reported to occur in about 3% of cases.<sup>6</sup> Furthermore the tumor has also been reported to reoccur locally after resection which mandates follow up of these cases.<sup>7</sup> Although glomus jugulare varies in size and form but a relative uniformity of their appearance with light microscopy is one of their characteristic features.<sup>8</sup>

Histologically, the glomus tissue neoplasm shows epithelioid cells arranged in clusters with surrounding stroma of connective tissue containing multiple vessels of varying caliber interspersed with collagen and reticulin fibers.<sup>9</sup>

Patients with glomus jugulare usually presents with pulsatile tinnitus and hearing loss or either conductive or sensorineural type. The later occurs with invasion of labyrinth. Cranial neuropathy suggests a more extensive process with symptoms such as changes in voice, dysphagia, aspiration, shoulder

weakness, deformity of tongue etc. Unilateral atrophy of tongue was also presently in our case suggesting involvement of hypoglossal nerve by the tumor.<sup>10</sup>

Rarely these tumors may be secretory in nature producing catecholamines. Patient can develop symptoms such as palpitations, labile hypertension, flushing and headache. These signs and symptoms are an indication to estimate the serum catecholamine/VMA levels. A markedly increased urinary excretion of CA and CA-metabolites is usually found in these patients.<sup>11</sup>

The advent of high resolution CT and MRI have made the diagnosis much easier in the early clinical stage of disease, when the tumor causes relatively little problems. CT scan of the skull in bone window settings mostly shows erosion of base of skull with enlargement of the jugular foramen. A permeative or moth eaten pattern of bony destruction of jugular foramen with demineralization of the skull base is a characteristic finding as well. The erosion can lead to destruction of the labyrinth extending into the hypo tympanum, mesotympanum, sinus tympani, jugulocarotid spine and hypoglossal canal in the first instance.<sup>12</sup>

Since these tumors have a rich blood supply as well, MRI can depict the presence of vessels by showing multiple flow voids within the mass on T2-weighted images, as well as hyper intense foci on precontrast T1-weighted sequence, which represents slow tumoral flow or hemorrhagic foci.<sup>13</sup> The appearance of multiple flow voids and has been termed as salt and pepper appearance and is pathognomonic of glomus tumors.<sup>14</sup>

On account of their rich blood supply arteriographically, glomus jugulare tumors show up as hyper vascular masses with an intense characteristic tumor "blush". Commonly large feeding vessels and early draining veins are seen constituting an early arterovenous shunting. The predominant supply comes from external carotid artery system, mainly the ascending pharyngeal artery.<sup>15</sup>

On account of these myriad imaging appearances on various modalities, currently CT scan, along with MRI as well as four vessel angiography are the modalities used in liaison for the diagnosis as well planning the management of these tumors.

CT being helpful for evaluating the erosion of

surrounding structures and MRI delineates the relationship of the intracranial portion to neurological structures. Furthermore Four-vessel angiography is recommended for all patients to evaluate vascular supply of the tumor, and additionally because 10% of cases have been reported to have bilateral or multiple paragangliomas.<sup>16</sup>

Glomus tumors are treated mainly by surgical means however despite all the advances in the microsurgical techniques some of these lesions still prove a challenge to the surgeon due to their close proximity to intracranial nerves and vessels. In cases where complete excision is performed by open surgical approaches, total removal can be followed by severe morbidity and occasionally mortality as well.<sup>17</sup>

Most patients might undergo embolization before surgery which can be of great help in maintaining hemostasis. By means of presurgery embolization the loss of blood can be reduced to about 60%.<sup>18</sup>

Van der May recommends observation for tumors of the glomus jugulare which show no progressive neuropathy of the cranial nerves, brain stem compression or intracranial hypertension.<sup>19</sup>

Whatever the approach a combined otologic / neurosurgical team during the resection is advantageous. Advanced tumors requiring the resection of cranial nerves are better treated with radiotherapy than surgery.<sup>20</sup> Due to the morbidity associated with glomus tumors early diagnosis should be carried out radiologically so that prompt management can be done.

## Conclusion

Careful and meticulous analysis of important differentiating imaging findings may lead to a correct preoperative diagnosis, which allows adequate patient counseling and surgical planning.

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