# GLOMUS VAGALE

**- Case Report -**

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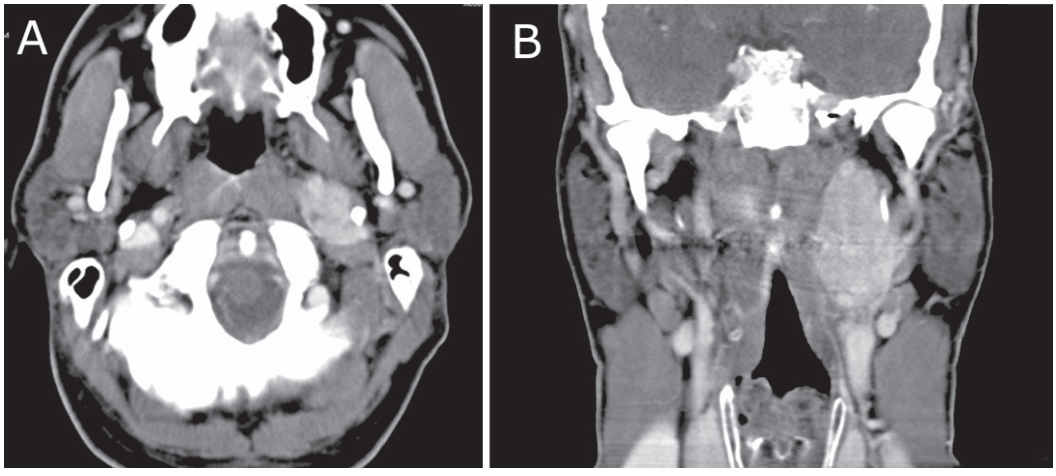
## Introduction

Glomus tumors, also called paragangliomas or chemodectomas, are highly vascularized benign tumors that arise from cells derived from the primitive neural crest. Unlike the chromaffine-paragangliomas of the abdomen and chest, paragangliomas of the head and neck are usually inactive. They occur most commonly at the carotid bifurcation (carotid body tumors), followed by the jugular bulb (glomus jugulare) and vagus (glomus vagale)[1]. The authors of this manuscript would like to present a rare case of glomus vagale with emphasis on the radiologioc findings.

## Case Presentation

A 35 year old male previously healthy presented with history of dysphagia of one year duration. The patient reported also nasal food regurgitation and a change in voice quality. He has noticed the appearance a left neck mass that was growing slowly over the past year. On physical exam he had a 3x3 left neck mass, firm, non tender, well circumscribed with no overlying skin changes together with medialization of the left pharyngeal wall. He had a decreased gag reflex but a preserved pharyngeal sensation. Flexible fiberoptic laryngoscopy revealed normal vocal fold mobility bilaterally. A computerized tomography of the neck with IV contrast was done together with a 3-D reconstruction of the mass showing a highly enhancing mass with signs of increased vascularity, displacing the jugular posterolaterally and the carotids anteriorly. (figure 1 & 2).

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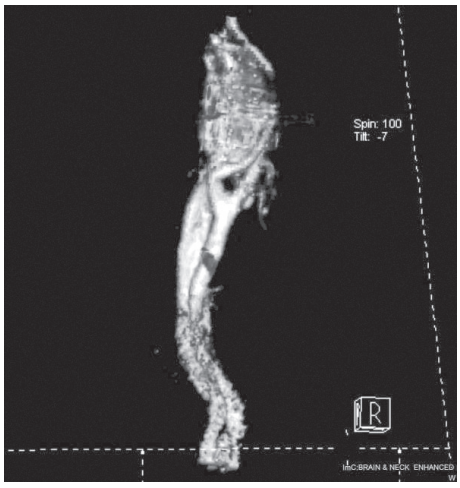


## Discussion

Glomus tumors are rare tumors of the head and neck region. They are 3-4 times more frequent in females with a prediliction to the fourth and fifth decade[1-4]. They are considered benign and 5% may carry malignant potential. Persson et al has reported vagale tumors to be malignant in up to 20% of the cases[2]. The malignant potential is not diagnosed by histological features but by the presence or absence of metastasis. Histologically glomus tumors are composed of nests of neoplastic cells surrounded by multiple tiny blood vessels. Platelet derived endothelial cell growth factor and vascular endothelial growth factor are highly expressed in these tumors which may contribute to their extensive vascularity[3]. In 10% of the cases they are hereditary with an autosomale dominant mode of inheritance. Three different loci were identified for the hereditary form of the disease and these are PGL1, PGL2 and PGL3. Glomus tumors can be multiple in 10% of the cases, occurring bilaterally or on the same side, with the incidence being higher in the hereditary form[5-8].

Clinical presentation includes history of a neck mass and the presence of dysphonia in 50% of cases of glomus vagale tumors. Other symptoms related to cranial nerve involvement include palatal weakeness, tongue weakness and horner syndrome[1].

Radiologic signs are usually diagnostic. Computerized tomography (CT) shows a highly enhancing mass with signs of increased vascularity, displacing the jugular posterolaterally and the carotids anteriorly. These were the findings in our case. Glomus jugulare on the other hand, tends also to displace the jugular posteriorly and carotid arteries anteriorly but with more significant involvement of the jugular foramen. Large tumors cannot sometimes be differentiated as jugulare or vagale. CT scan is also essential to identify the status of the skull base in cases where erosion is suspected. Angiography is an important diagnostic tool for all parapharyngeal space tumors. Arteriography will allow visualization of the feeding vessels, demonstrating an early intense tumor blush and an early venous phase. On Magnetic resonance imaging, glomus tumors show low to intermediate signal intensity on T1 and high intensity on T2. A pathognomonic "salt and pepper" pattern is commonly seen and accentuated after contrast injection. With fat suppression sequences the tumor can be better delineated from surrounding fat or muscle.

The vascularity of these tumors and the close relation to the major vessels makes histologic diagnosis difficult preoperatively and even not recommended[4,9]. The differential diagnosis includes: metastatic renal cell carcinoma, metastatic pheochromocytoma, thyroid carcinoma and hemangiopericytoma.

The main treatment for glomus tumors is controversial. Surgery was believed and still by many authors to be the main modality of treatment. Despite the tailoring of the surgical approach according to the extent of the tumor, dreadful complications such as vocal fold paralysis and other cranial nerve palsies are nearly inevitable following resection of vagale tumors[10]. This has led many to chose another option for treatment mainly radiation therapy. It is keen to know that radiotherapy is not effective in decreasing tumor size. Although it is not a curative, its high control rate and low morbidity compared to surgery, makes it a good alternative that ought to be discussed with the patient. It is mainly advocated as a primary mode of treatment when the tumor is large and its resection entails significant morbidity. A suggested dose of 45 Gy in 25 sessions over 5 weeks has proved to be effective in controlling tumor growth in close to 95% of the cases over more than 10 years of follow up[11]. This dose is believed to be effective and mildly toxic to normal tissue. Embolization has also been reported as an alternative to control symptoms, despite the lack of long term follow up in the literature. It is indicated

either pre-operatively to reduce the amount of blood loss, or as the sole mode of treatment in poor surgical candidates and patients who can not receive radiation therapy. The incidence of cerebral embolization following embolization should always be kept in mind[8].

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