

A Rare Case of Bilateral Carotid Glomus Jugulare with Partial Responses to Radiotherapy

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Abstract

Glomus jugulare is known as a benign tumor that could involve in different parts of the body. The most prevalent site of involvement is head and neck area. This disease is rare and few of the cases are bilateral. However, in familial cases bilateral or multicenter lesions are even rarer, in which there is an autosomal dominant pattern of inheritance. The most efficient treatment is believed to be surgery or radiotherapy depending on the location and certain other factors, such as age and performance status. Sporadic bilateral lesion is rarely seen and most bilateral cases are familial. Herein, we reported a case of bilateral glomus jugulare of carotid with no history of this tumor in family or any other familial diseases. Our subject was a 67-year-old man. The diagnosis was made via Tru-cut biopsy. He was treated successfully by 3-dimensional conformal radiotherapy up to a dose of 60 Gy. The follow-up computed tomography scan revealed partial tumor responses. He had no family history of any systemic disease related to functional glomus jugulare, such as uncontrolled hypertension or mass in the neck. He also had no history of catecholamine excretion-related symptoms.

Keywords: Glomus jugulare, Radiotherapy, Chemodactoma, Carotid

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Introduction

Glomus jugulare (GT) or chemodactoma or paraganglioma are benign and rare tumors that may

occur in different parts of the body. Head and neck are at a higher risk than other sites.¹ The origin of these lesions is the neural crest and they

arise from paraganglia cells. These tumors might be functional or non-functional and few of them are familial. Multiplicity is reported to be more commonly seen in familial cases, rather than sporadic ones.^{2,3}

The main applied treatments are surgery or radiotherapy (RT), yet surgery is not always trouble-free. Even though GTs are radio-resistant tumors and complete tumor disappearance is rare, RT is an effective treatment.^{1,2}

In the present paper, we aimed to share our experiences regarding a case of bilateral carotid glomus jugulare with no familial history and partial response to RT.

Case Presentation

The patient was a 67-year-old man with bilateral neck mass for about 20 years; the mass had enlarged since two years ago. He had no other symptoms and the reviews of systems were normal. He had no family history of GT or any other systemic diseases except for diabetes mellitus, Alzheimer, and ischemic heart disease. Physical examination demonstrated that he had bilateral neck masses in anterior sides of his neck and the masses were firm and non-mobile. Primarily, FNA of the lesion was done, which was not conclusive. Subsequently, Tru-Cut biopsy was carried out; it illustrated the tumoral process with a nesting pattern architecture separated by

a vascular stroma. The tumoral cells were uniform with salt and pepper nuclei. Immunohistochemical study showed immunoreactivity for chromogranin, synaptophysin with KI-67 labeling of about 2%; therefore, the diagnosis of paraganglioma was confirmed (Figures 1 and 2). Fortunately, after Tru-Cut biopsy, no bleeding or complication occurred. Neck computed tomography (CT) scan showed a bilateral lesion in his neck while no other lesions were found in his chest, abdomen, or pelvic CT scan (Figure 3A). Afterwards, we carried out RT with 3-dimensional conformal radiotherapy (3D-CRT) up to 60Gy. Following six months, both lesions decreased concerning the size (Figure 3 B).

Discussion

GTs are benign and slow growing tumors that are rarely observed. Only 0.03% of human neoplasms and 0.6% of head and neck tumors are GT.¹ This neoplasm is more common among women and is usually reported in the fourth to sixth decades of life.² These tumors are usually asymptomatic and present with neck mass. On a number of occasions, pressure effect leads to certain symptoms, including dysphonia hoarseness stridor and odynophagia.⁴ Certain cases of GT are familial with an autosome dominant pattern.¹

These lesions may be functional and produce catecholamines that could lead to symptoms like

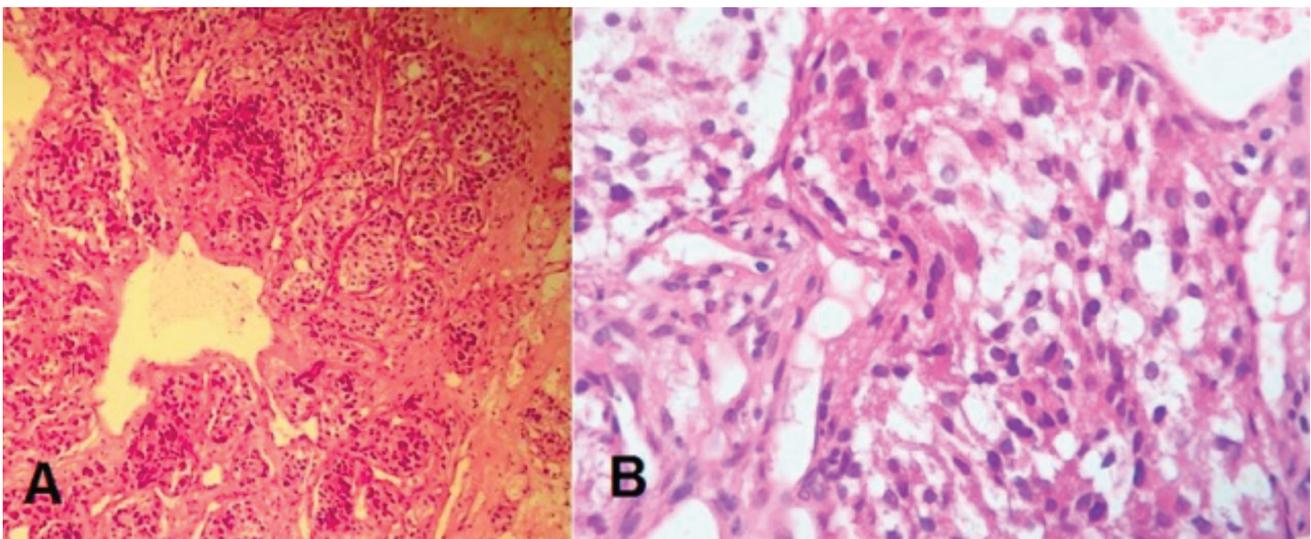


Figure 1. A & B: H&E stain, the tumors show a nesting architecture separating with a vascular stroma. The cells are uniform with minimal nuclear atypia, 100 \times , and 400 \times , respectively.

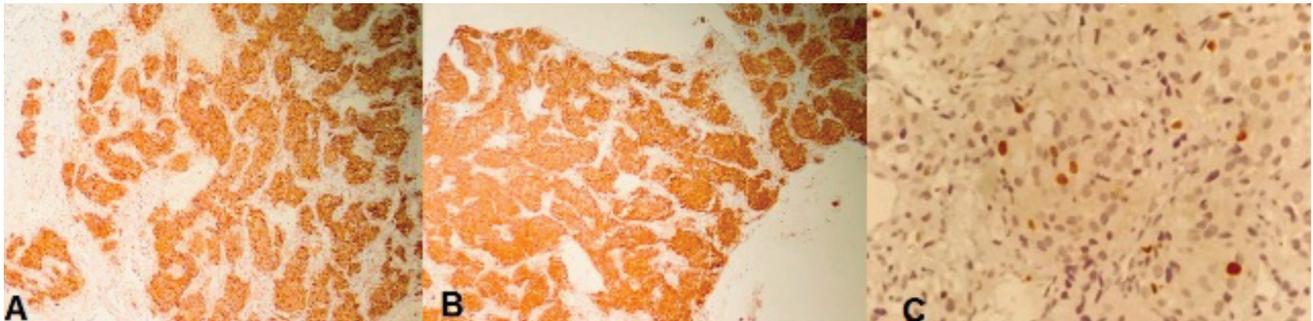


Figure 2. A, B and C: immunohistochemical stain of tumoral cells: A: chromogranin, X100, B: synaptophysin, 100 \times , and C: KI-67 index about 2%, 400 \times .

diarrhea or hypertension. Despite the necessity of treatment in patients with symptomatic biologically active tumor, in other non-functional lesions, treatment is usually not urgently needed.^{2,5} In the absence of these symptoms, age of the patient, neurologic deficit, tumor size, general condition, and location of the tumor are determinant factors in choosing the proper treatment.^{2,6} Our patient had no signs and symptoms of any functional tumors and needed treatment due to a large mass.

Regarding hypervascularity of GT, biopsy is not usually recommended and radiologic modalities are employed for diagnosis.^{3,7} Magnetic resonance imaging may show the classic features

of this tumor and anatomy and adherence to adjacent structures.^{4,7,8} In physical examination, particularly in carotid GT, a rubbery mass is usually found, which moves laterally rather than vertically; it is called Fontains's sign.⁹ The masses in our patient's neck were fixed bilaterally.

Bilateral GT lesions are rare. Most cases of bilateral GT are familial with mutation in succinate dehydrogenase gene that has an autosomal dominant pattern of inheritance. Familial cases present in younger ages. Among familial cases, up to 30% may have bilateral or multicenter GT.^{2,5,8} Wong et al. investigated 15 patients with GT of jugular area during a 15-year period. The only bilateral lesion belonged to a 14-year-old

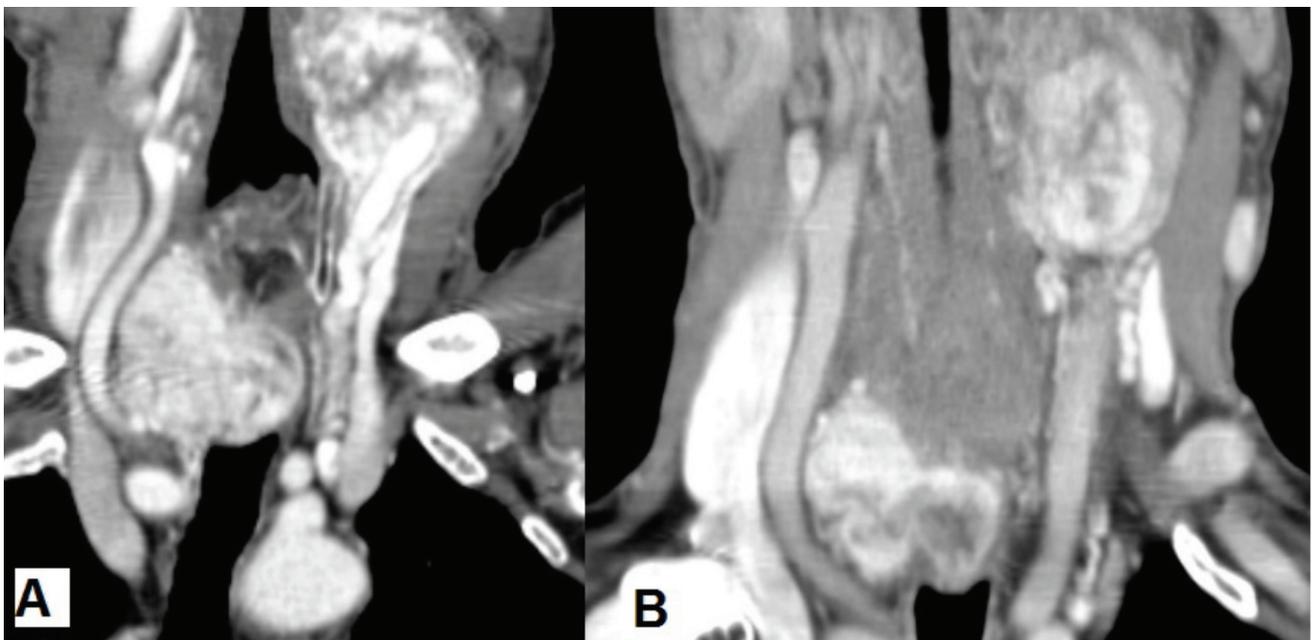


Figure 3. A: large mass just superior to the left carotid bulb, between internal carotid artery and external internal carotid artery, with severe enhancement. Another mass in the right tracheoesophageal recess, just posterior to the right thyroid lobe. B: the same lesions after radiotherapy with partial responses.

male patient with a family history of GT.⁶ Our patient had no history of GT.

Treatment of GT is controversial. The main therapeutic options are surgery, RT, or both.² Surgery may result into severe complications.⁴ RT is employed for old patients and those with severe comorbidity, inoperable masses, and malignant features in pathology. Chemotherapy does not play a role in this treatment.¹⁰ It seems that both surgery and RT are effective treatments. In studies that have reported outcome of patients, RT has been found to be successful in 75%-100% of patients and in surgery reports, the success rate has been 54%-100%.³ In addition, in rare cases of bilateral GT, surgery may lead to damage to baroreceptors and lead to lots of problems in daily activity of patient.^{2,11} Surgery is reserved for patient with progressive neurologic deficit or catecholamine excreting lesions.¹⁰

GTs are radioresistant tumors that partially response to treatment.² Complete tumor disappearance occurs rarely and stopping tumor enlargement is considered as successful treatment.^{1,2,8} Our patient demonstrated partial response in right and left sides of his neck.

Conclusion

Bilateral GT is a rare tumor and could be treated successfully via RT.

Informed Consent

The patient signed the informed consent prior to the treatment.

Conflict of Interest

None declared.

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