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# Glomus Caroticum Tumors: A case report of an operated giant carotid body tumor with a review of our experience in 47 patients

**Review Article**

Sureyya Talay[1], Mustafa Abanoz[1], Mehmet Ali Kaygin[1], Ozgur Dag[1], Umit Halici[1], Derih Ay[1], Semih Murat Yucel[1], Mehmet Esref Kabalar[2], Buket Tasmacioglu[3], Bilgehan Erkut[4*]

[1] *Erzurum Regional Training and Research Hospital, Department of Cardiovascular Surgery, 25020, Erzurum, Turkey*

[2] *Erzurum Regional Training and Research Hospital, Department of Pathology, 25020, Erzurum, Turkey*

[3] *Erzurum Regional Training and Research Hospital, Department of Anaesthesiology, 25020, Erzurum, Turkey*

[4] *Erzurum Regional Training and Research Hospital, Associated Professor for Cardiovascular Surgery, 25020, Erzurum, Turkey*

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**Abstract:** Glomus caroticum tumors, usually used as an alternative term for carotid body tumor, are of neuroectodermal origin and a part of the extra adrenal neuroendocrine system pathologies. These abnormalities are the most frequently detected paraganglioma in the localization of the head and neck. In our report, we present a giant tumor mass on the left side which was operated on successfully with a review of our experience retrospectively. Between the dates of June 1995 and October 2009, 47 patients, all of which had a glomus caroticum tumor, underwent to surgery. Tumor presented a wide variety of size and clinical presentations.

**Keywords:** *Glomus caroticum tumor • Carotid body tumor • Paraganglioma • Excision surgery*



## 1. Introduction

We report a case of a giant carotid body tumor on the left side of the neck which has been successfully carried out via operation. No complications occured during the early follow-up period postoperatively. The patient was discharged after eleven days of hospital care, the end result was complete excision of the tumorous mass.

In this study, a brief summary of pathophysiology, frequency, mortality and morbidity, clinical presentations with diagnostic evaluations, surgery and treatment results of this neoplasm is to be presented. In our opinion, the decision for excision surgery is a predictive choice for a treatment with the best outcome among these patients.

## 2. Case Report

In October 2009, a 65 year old female patient with a history of a neck pain and a slowly growing bulging mass on the left side for several years was admitted to our department with complaints of worsening pain. The patients's medical history included hypertension and hyperlipidemia. No symptoms of hoarseness, dysphagia, tinnitus, hearing loss or hiccups were noted. The patient did not mention any family history for tumors, especially not for carotid body. There was no prior medical evaluation for the mass lesion. Results of physical examination were within normal limits excluding the immobile mass lesion on the left side of the patient's neck along the anterior surface of the sternocleidomastoid

* *E-mail: bilgehanerkut@yahoo.com*

**Figure 1.** In preoperative term, ultrasonography images of the tumor mass.

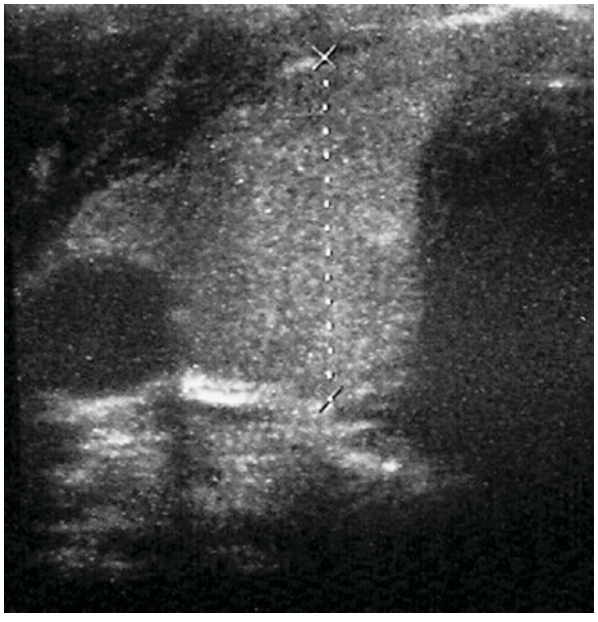
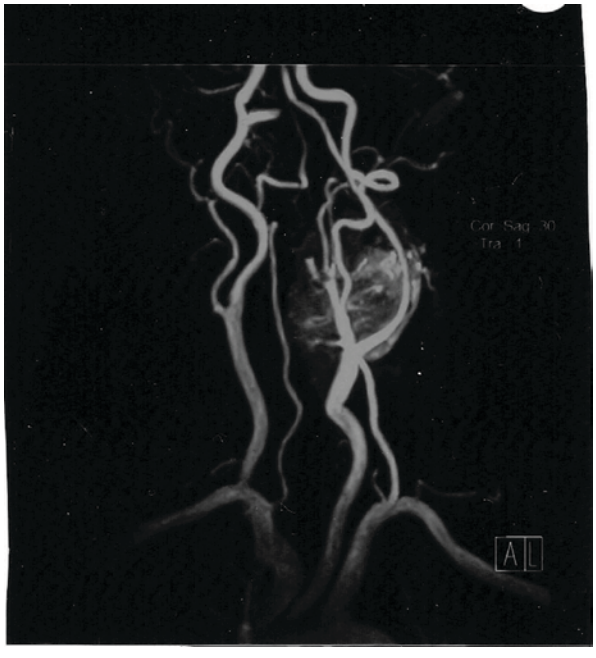
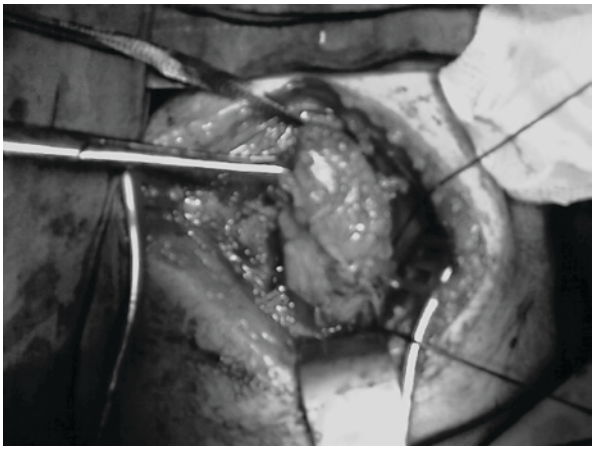


**Figure 2.** In MRI angiography, vascular tumor in carotid bifurcation.



muscle region. The electrocardiogram showed a normal sinus rhythm for 80/min. Arterial blood pressure was 110/90 mmHg. Lungs were clear to auscultation. There were no cardiac murmusr and S1-S2 were normal and rhythmic upon cardiac examination. Laboratory blood tests showed no abnormalities including the cardiac panel. Arterial vascular pulses were palpable on both sides of upper and lower extremities. There were no

**Figure 3.** Intraoperative view of the carotid body tumor mass.



neuromuscular abnormalities which may accompany to a compression or invasion for facial nerve, hypoglossal nerve at the region. The chest radiograph and M-mode echocardiograms showed no abnormalities. An ultrasonography and color Doppler ultrasonography evaluation for the mass lesion and the neighbouring carotid arteries were performed prior to operation (Figure 1). Ultrasonograhy images revealed a giant left glomus caroticum tumor with a smooth contouring mass in diameters of 9x5x7 centimeters. No observations of central calsifications or degenerations in the mass. The tumor showed vascularization pattern in all areas. Carotid artery and its branches were interestingly free of mass compression in all parts and the blood flows were within normal parameters in the Doppler ultrasonography report. Preoperative detailed information of the mass was derived by magnetic resonance imaging (MRI) angiography. T1 weighted tomographic slices demonstrated a carotid body tumor 75 millimeters in diameter located at the left carotis bifurcation (Figure 2). The MRI morphology and paramagnetic agent accumulation suggested an hypervascularization, as seen below at the figures, typical for glomus caroticum tumor. Surgical strategy was determined following the MRI scans with a consensus in our clinic.

The patient underwent surgery with general anaesthesia. Surgical incision took place along the anterior border of the sternocleidomastoid muscle on the left side of the neck as a transcervical approach. Following the subcutaneous tissue and platysma, surgical exposure reached the mass lesion of the carotid body tumor. Sternocleidomastoid muscle deviated laterally from the surgical area. Tumor mass observed around 6x5x7 centimeters, immobile and tightened to surrounding tissue and vascularities (Figure 3). Tumor mass was arising from the bifurcation and expending anterior and posteriorly. Special care showed to the

**Figure 4.** After operation, the carotid arterial system.

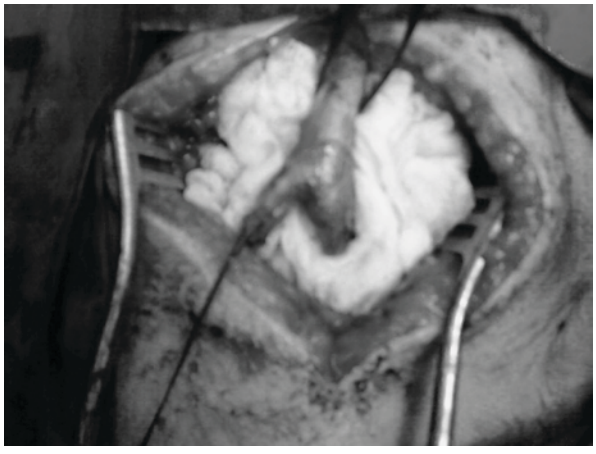
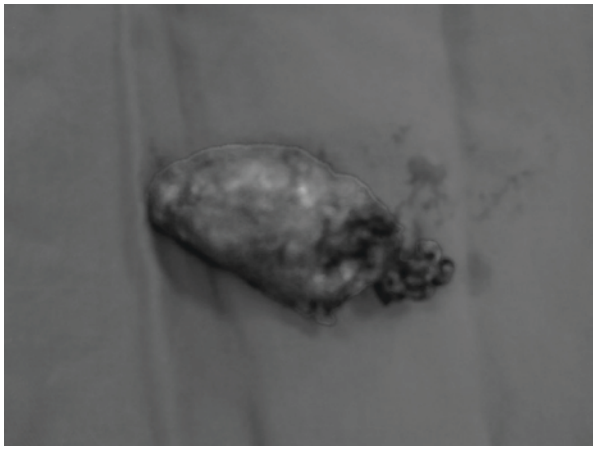
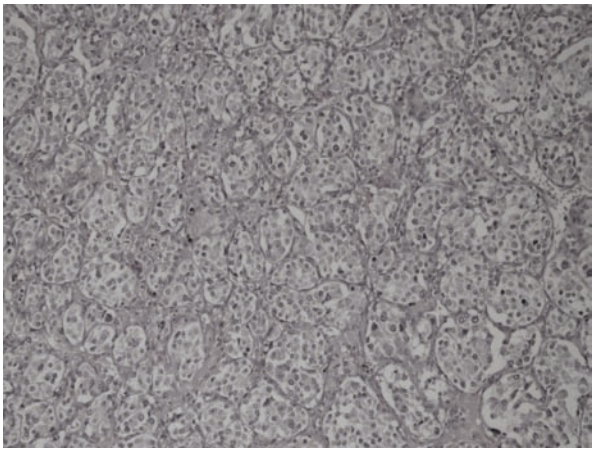


**Figure 5.** The carotid body tumor after excision.



nervous structures such as hypoglossal, vagal and facial nerves during the procedure. The internal juguler vein removed gently from the area. Common carotis artery and its branches turned around with vesselloops and secured. Total mass excision was achieved with multiple ligations of the collateral tumor vascularities and gentle dissection (Figure 4-5). No external carotid artery sacrifice was necessary in our series. In total, 1.2 liters of surgical bleeding accumulated through the suction system during the operation which was a proof of the degree of hypervascularization of the tumorous mass. The surgical loss of intravascular fluid replased with transfusions intraoperatively. The operation was carried out free of any major vascular injuries and complications. Following the hemostasis maneuvers, a drain placed along through the surgical area. Closure of the anatomical layers was carried out safely. The patient was transported to our cardiovascular intensive care unit from the operation room. There were no neurological complications which could be associated and caused by

**Figure 6.** Histopathological examination of the tumor mass.



a damage of the neighboring nerves in the surgical area on the related neuromuscular structures, observed after the extubation of the patient. The early postoperative period passed uneventful and the patient was discharged at the postoperative day of eleven in a tumor free condition. The patient's postoperative histopathological evaluation for the excision material reported a glomus tumor. Multiple characteristics as thick walled vascular channels in H.E application, Zellballen structures, immune reactivity with S-100 and Chromogranin A application was observed. These findings defined a glomus tumor histopathologically (Figure 6).

## 3. Review

Carotid body tumor are reported to have a partiality for females among patients. There exists a female to male ratio of 4:1. On the contrary, 32 of our patients were male and 15 were female. In our apprehension, the number of the patients that are studied in our publication may not be enough to come to a conclusion, respectively. Patient ages were between 17 to 68 years, with an average of 47.4 years which overlaps with the literature on this topic.

In the literature, glomus tumors are frequently reported unilaterally, on the left side or the right side of the neck [1]. Unlike the unilateralization, these neoplasms are mostly solitary lesions. Our study shows a concordance with these publications. Fourty-one cases of our series were unilateral. In detail, 30 cases were on the right side and 11 cases were on the left side. Only 6 of our patients had bilateral lesions. Only 1 case presented with multiple lesions in the latter group. No multiplicity was recorded in the first group of patients with unilateral tumor localization.

Our patients frequently complained about hypertension, headache, palpitations and pain at the lesion side. Elder patients suffered from loss of hearing unspecifically but a distinctive etiology was not possible between the possibilities of nerve compression depending on the effect of tumor mass and aging. No evidence of dysphagia, vertigo, paresis and deficits of facial nerve realted to mass lesion were detected. One male patient manifested a familial component with the age of 17. It is reported that the 11q23 gene abnormality in a dominant pattern is responsible for hereditary paraganglioma cases. The hereditery form is 7-10% of all cases and has a tendency to be multicenrically [1].

Our preoperative preferred examinations included routine all system's physical examination, laboratory studies, a combination of ultrasonography with Doppler, MRI and coronary angiograhy for the cases in which the patient was over 55 years old. No surgical decision occured for coronary artery bypass surgery after coronary angiography in any case. Coronary stens implanted in 17 patients preopertively. Surgical strategy was determined with a consensus of our surgeons preoperatively in all cases. An ultrasonography was never thought to be enough to determine surgery in our hospital but an hypervascular lymphadenopathy, schwannomas, neurofibroms are possible to becloud the diagnose and mimic a carotid body tumor for MRI [2-4].

Surgical mortality and morbidity may vary on the localization of the paraganglioma. Brown et al. [5] reported their experience of resection for mediastinal paraganglioma between 1973 and 2007 in 14 patients. The median age of patients at operation was 39 years (range, 27 to 68 years), 71% were female, and all had a history of hypertension. They report that the tumor was adjacent to the heart or great vessels in all patients and was resected through a median sternotomy (n = 10), or with posterolateral thoracotomy (n = 4). In 6 of their patients, cardiopulmonary bypass was used to facilitate dissection of the paraganglioma from the heart or great vessels. There was one intraoperative death in this series due to blood loss. In our cases no sternotomy was necessary due to the localization of the tumors. Functional paragangliomas can be diagnosed with measurements of fractionated catecholamines and metanephrines; mediastinal localization is determined with appropriate scanning techniques. These tumors can be treated successfully by surgical resection with modest surgical risk, often necessitating cardiopulmonary bypass, with good long-term survival. Hypertension may persist even after complete resection of the tumor. One of this study's patient died intraoperatively due to uncontrollable bleeding from the tumor and tumor bed; the patient had normal coagulation studies prior to operation. Bleeding requiring transfusion occurred in 6 patients (43 %) with a median of 5 units of packed red blood cells (range, 1 to 20 units). The median postoperative hospital length of stay was 6 days (range, 5 to 10 days). Among the 13 survivors, the mediastinal tumor was completely removed in ten patients. Two patients had positive gross margins, and one had microscopic evidence of tumor at the margin of resection and, in addition, had a positive lymph node; this patient elected to undergo radiation therapy. Late recurrence of a paraganglioma was noted in 2 of the 10 patients who were thought to have had complete excision. One patient had subsequent resection of bilateral adrenal pheochromocytomas and also had metastatic gastric leiomyosarcoma. This patient had the Carney triad and treatment was considered palliative, but has survived nearly 20 years after the initial operation for mediastinal paraganglioma. The second recurrence developed 3 years after resection of the mediastinal tumor; wide-spread skeletal metastases were treated with chemotherapy. Depending on the results of Brown et all., mediastinal paragangliomas tend to develop more operative and postoperative morbidity and mortality compared with our study.

In a case of glomus caroticum surgery a preferred definitive treatment for the patient is open excisional surgery [5-7]. Eldery patients with unacceptable motality and morbidiy who are poor surgical candidates can benefit from gamma knife irradiation, sclerotheraphy, laser applications [8], percutaneous embolization inducing necrosis to tumor mass. In some cases a clinical close follow-up without any intervention relying on the benign character of the tumor may be chosen by physicians as a decision. In 1971, Shamblin described a surgical classification system [9]. Patients were divided into three groups; Group I (small and easily removeable tumors), Group II (medium sized tumors posiible to remove totally with a certain difficulty of dissection), Group III (Large and complicated masses). In our group of study, there were 14 Goup I, 23 Group II and 10 Group III patients. We preferred to operate all of our patients.

Operation times were between 95 to 260 minutes, mean 135 minutes. Operation times seemed to be shortened by our experience in the second half of our study's period of time. Glomus caroticum tumors are highly vascularized structures. Arterial blood supply usually flows through pharengeal, tympanic and occipital arteries arising from external carotid artery, internal carotid arter or vertebral arteries [10,11]. Surgical bleeding depending on the mass size of the tumor were between 1 to 1.5 liters on average. Care is taken to replace the bleeding volume intraoperatively by transfusions in all cases between 2 to 15 units.

In 46 cases, a total excision to tumor mass was succeeded during the surgery. In 1 case, we were unable to remove the tumor totally, consequence of the wide spread and invasion to neighbouring nerves and connective tissue.

In 4 of our cases, we needed to practice patchplasty for common carotid artery. There were no major neurological complications in these group of patients postoperatively. No surgical deaths occured. Reported surgical mortality is around 9 % [12]. 1 patient developed a major neurological deficit at the postoperative day of 4 at the cardiovascular intensive care unit resulting a stroke with permanent hemiparesis of the right side of the body. This patient undertook surgery on the left neck at the age of 61. 4 patients were partially disabled with worsened and blury vision. These 4 patients recovered before leaving the hospital. Almost in half of the patients, in several degrees of neurocognitive dysfunction from vertigo to loss of orientation occured. These observations were not permanent. Mean durations for hospitalization was 24 days and intensive care unit for 9 days. 11 patients returned back to operation room for a revision and exploration because of early postoperative serious amount of bleeding. No prolonged entubation experienced in our patients, extubations achieved untill the first postoperative day.

## 4. Discussion

First medical reports related to glomus caroticum tumor are revealed in the sixteenth century. Haller, in 1762 decribed a mass at the carotid bifurcation area in a glomus body-like structure. In 1812 Wood published an article about the clinical presentations associated with glomus tumor. In 1840 Valentin described ganglia tympanica as a variation. A glomus jugulare tumor case was reported by Rosenwasser in 1945. The patient of Rosenwasser survived untill 1987 and died of natural causes associated with his age. Approximately two hundred years after Haller, in 1950 Mulligan introduced this tumor as a chemodectoma arising from the chemoreceptor cells. Glenner renamed the tumor on a basis of its anatomic localization a paraganglioma. A classification described by Glenner and Grimley according to the localization, innervation and histopathological features of the tumor [13,14].

Glomus caroticum tumors arise from part of the extraadrenal neuroendocrine system, especially in glomus caroticum. Glomus tumors take part among all of the neoplasms in a percentage of 0.03%. These neoplasms are rather infrequent. In between the head and neck tumors, glomus caroticum tumors are in 0.6% [12,15]. Glomus tumors have an annual incidence of 1 case in 1.3 million people [16]. Glomus caroticum is chemosensitive tissue which plays an important role in the algorithm of oxygen and carbon dioxide arterial partial pressure and cardiopulmonary system regulation [17,18]. Sporadically, paraganglionic cells of the extraadrenal neuroendocrine system may occur in various localizations of the body. For instance; orbit oculi, pterygopalatine fossa, larynx, pharynx and dermis [19]. These cells in these localizations seem to involve the different steps of human body development except the accumulation in glomus caroticum. Malignant potential is related to large size, deep location, infiltrative growth, mitotic activity, nuclear pleomorphism and necrosis [20]. Carotid body tumors which are autonomic nervous tissue tumors consist of two different histological celluler components; type I (chief-chemoreceptive cells) and type II (supportive cells). Glomus tumors replace in four different localizations in the head and neck region; jugular bulb, middle ear cavity, carotis body and vagus nerve. The most frequent subtype is the glomus jugulare tumor and the rare subtype is glomus tympanicum tumors [21,22]. In our experience of 47 patients, all cases were diagnosed as carotid body tumors but a distinctive diagnostic effort had been performed between glomus jugulare and glomus caroticum tumor meaning as distiguishing the rooting origin of the mass. Clearly our cases were neither glomus tympanicum nor glomus vagale tumors. Postoperative histopathological reports corrected the preoperative macroscopic diagnose as glomus caroticum tumor in all cases. In our opinion, a differential diagnose is imperative for chondrosarcoma, Giant Cell Tumor, osteoblastoma, schwannoma and metastasis postoperatively with the histopathological investigations and aberrant carotid artery, exposed juguler bulb or hypervascular lymphadenopathy preoperatively with the imaging studies.

As a conclusion, we believe that a glomus caroticum tumor for a patient in a good physical status must be cured with open excisional surgery. In the preoperative period, a detailed physical examination, ultrasonography and MRI evaluations are essential. In elderly patients, a preoperative existance of coronary artery disease may be advisable to consider and investigate. Preoperative precaution for possible surgical bleeding would be atmost wisely before entering to the operation room. In most of the cases a total excision is possible by a gentle and patient dissection and major surgical complications occur in an acceptable rate. However, a long term postoperative follow-up is required to determine recurrences in our study as a future aspect to define curative excisional surgery propectively.

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