

Surgical treatment of carotid body tumors

Karotis cisim tümörlerinin cerrahi tedavisi

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Objectives: In this study, we report our clinical experience in a series of patients with carotid body tumors along with diagnosis, treatment and follow-up procedures in the light of related literature data.

Patients and Methods: Between November 2001 and May 2012, 10 patients (5 males, 5 females; mean age 53.2 years; range 27 to 80 years) who underwent surgery due to a carotid body tumor in our clinic were included. Diagnosis was based on ultrasonography, computed tomography, magnetic resonance imaging, magnetic resonance angiography, selective carotid angiography, balloon occlusion test, biochemical tests and preoperative embolization. Complications were also recorded.

Results: Balloon occlusion test was performed in all patients preoperatively, while embolization was implemented in seven patients. All masses were dissected by carotid artery subadventitial approach. Carotid integrity was maintained in nine patients, while a vein graft was used in one patient. Neurological disorder was observed in one patient, whereas transient hypoglossal paresis was in one patient who underwent saphenous vein grafting.

Conclusion: Our study results suggest that (i) carotid body tumors should be handled with multidisciplinary approach; (ii) balloon occlusion test should be performed in all patients undergoing surgery; (iii) a particular attention should be paid to cranial and phrenic nerves, if it is necessary to extend the surgical field while removing the tumor; and (iv) pathological examination should be carried out by an experienced team and in a multi-centered fashion, if necessary.

Key Words: Balloon occlusion; carotid body; embolization; paraganglioma.

Amaç: Bu çalışmada karotis cisim tümörü olan hastalarda tanı ile birlikte uygulanan tedavi ve takip işlemleri, klinik deneyimlerimiz ve konuya ilişkin literatür eşliğinde sunuldu.

Hastalar ve Yöntemler: Kasım 2001 - Mayıs 2012 tarihleri arasında karotis cisim tümörü nedeni ile kliniğimizde ameliyat edilen 10 hasta (5 erkek, 5 kadın; ort. yaş 53.2 yıl; dağılım 27-80 yıl) çalışmaya dahil edildi. Tanı yöntemi olarak ultrasonografi, bilgisayarlı tomografi, manyetik rezonans görüntüleme, manyetik rezonans anjiyografi, selektif karotis anjiyografi, balon oklüzyon testi, biyokimyasal testler ve ameliyat öncesi embolizasyon kullanıldı. Komplikasyonlar kaydedildi.

Bulgular: Balon oklüzyon testi ameliyat öncesi dönemde tüm olgulara uygulanırken, yedi hastaya embolizasyon yapıldı. Tüm kitleler karotis arterden subadventisyal yaklaşım ile diseke edildi. Dokuz hastada karotis bütünlüğü korunurken, bir hastada damar grefti kullanıldı. Bir hastada geçici nörolojik bozukluk, safen venle greftleme yapılan bir hastada ise geçici hipoglossal parezi gözlemlendi.

Sonuç: Çalışma bulgularımız (i) karotis cisim tümörlerine tedavisinin multidisipliner yaklaşım gerektirdiğini, (ii) balon oklüzyon testinin cerrahiye alınacak tüm hastalara yapılması gerektiğini, (iii) tümör çıkarılırken cerrahi sahayı genişletmek gerekirse kraniyal ve frenik sinirlere özellikle dikkat edilmesi gerektiğini ve (iv) patolojik incelemelerin deneyimli bir ekip tarafından ve gerekirse çok merkezli olarak yürütülmesi gerektiğini göstermektedir.

Anahtar Sözcükler: Balon oklüzyon; karotis cismi; embolizasyon; paragangliom.



Carotid body tumors are benign tumors stemming from the carotid body chemoreceptors. These tumors are localized in vascular adventitia. Carotid body tumors which are stimulated due to hypoxia, hypercapnia and acidosis, regulate the blood pressure, heart rate and respiration.^[1] Tumor may be localized in the back of carotid bifurcation as well as between the internal and external carotid artery. The dominant cells of the tumor tissue are of neuroectodermal origin. Although the majority of the cases are sporadic, 10-25% have been reported to be hereditary. Approximately 10% cases are bilateral, including most cases related to hereditary transmission.^[2] Of the cases, in only 5% catecholamine secretion was observed. The incidence of metastatic disease ranges from 2 to 9%.

The most common clinical presentation of carotid body tumors is the lateral neck mass, which is painless and constantly growing. The differential diagnosis of enlarged lymph nodes should be performed from branchial cleft cysts, parotid tumors and vascular tumors. Diagnosis is based on the clinical findings and radiological imaging techniques including ultrasonography (USG), computed tomography (CT), magnetic resonance imaging (MRI), magnetic resonance angiography (MRA) and selective carotid angiography (SCA).^[3]

In this study, we report our clinical experience in a series of patients with carotid body tumors along with diagnosis, treatment and follow-up procedures in the light of related literature data.

PATIENTS AND METHODS

Between November 2001 and May 2012, 10 patients (5 males, 5 females; mean age 53.2 years; range 27 to 80 years), who underwent surgery due to a carotid body tumor in our clinic were retrospectively analyzed. Clinical conditions of the patients, surgical technique applied and pathology results were obtained from medical files. Pathological examination was performed by the same pathologists (Figure 1).

Lateral cervical approach was implemented in all patients. The nerves, vagus and hypoglossus were removed from the tumor in certain patients. Subadventitial dissection was used as a surgical method (Figure 2 a-c).

RESULTS

Three patients had a right-sided tumor, while seven patients had a left-sided tumor. One patient had bilateral glomus tumor. All patients were symptomatic on admission. The most common evident symptom was the neck mass (100%); followed by hearing loss (n=1, 10%) and sore throat (n=1, 10%). Duration of symptoms ranged from two months to four years. Only one patient underwent preoperative tissue biopsy.

While five patients (50%) were diagnosed through CT (Figure 3), eight of them (80%) were diagnosed through MRI. Selective carotid angiography (Figure 4) and balloon occlusion test were implemented in all patients. Using SCA, diagnostic images were obtained due to localization of the tumor between the internal and external carotid. Preoperative embolization was performed in seven patients (70%). Magnetic resonance angiography was applied to three patients (Figure 5). Pathological examination revealed malignancy in one patient (10%).

All patients underwent surgery. Due to the internal and external invasion of the tumor, carotid tumor was completely resected including 1 cm proximal of communis by the vascular surgeons and saphenous vein grafting was used. None of the patients were required for blood transfusion. The vagus and hypoglossal nerves were observed and maintained in all patients. However, postoperative hypoglossal

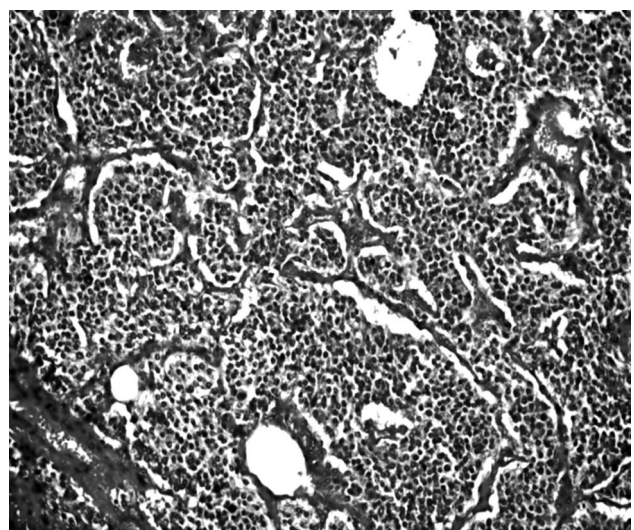


Figure 1. The microscopic image of a paraganglioma, cuboidal cells, septums rich in blood vessel and granular cytoplasm (H-E x 200).

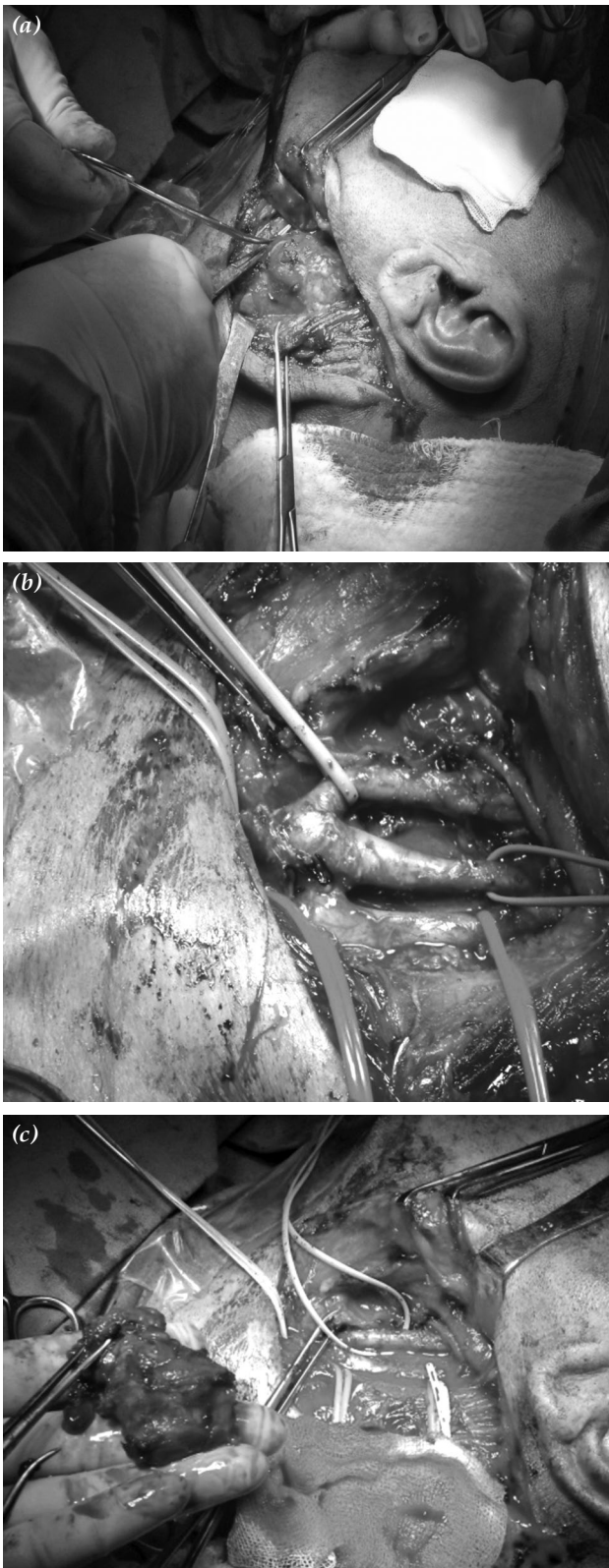


Figure 2. (a) Appearance of preoperative tumor lines during the bifurcation of the carotid in a 54-year-old patient with carotid body tumor. (b, c) During the operation, the carotid bifurcation and the tumor tissue was dissected and removed.

nerve palsy was observed in one patient with saphenous vein grafting after two months of recovery.

Frozen section analysis was not performed in any patient. Paralysis was reported in one of the patients with a carotid body tumor located in the right arm with facial paralysis at six hours postoperatively. No motor deficit was observed at four months postoperatively. No complications were observed in the patients. No procedure-related mortality due to postoperative complications was observed. The mean length of hospital stay was eight (range 5 to 14) days.

Local recurrence or distant metastatic disease was not observed. However, family history of a patient revealed bilateral carotid body tumor. All family members of the patient had a carotid body tumor. The patient and family members were scanned for the multiple endocrine neoplasia (MEN) syndrome. All patients survived. The mean follow-up was five years.

DISCUSSION

Carotid body tumors stem from chemoreceptor tissues of carotid bifurcation. They are usually benign and non-functional.^[4] Since they grow in a slow fashion, they usually remain asymptomatic, until they reach a certain size. They are often neglected until they put a pressure on cranial



Figure 3. Cross-sectional image of preoperative neck computed tomography.



Figure 4. Peripheral angiography images of a carotid body tumor.



Figure 5. Magnetic resonance angiography images of a carotid body tumor.

nerves or vascular structures. They may also lead to dysphasia due to the pressure on the oropharynx. Symptoms may be present due to the release of catecholamines in functional ones. Neck mass was the most common symptom encountered in our cases.

It is more frequent in women and individuals who live in high-altitude regions. Carotid body tumors are likely bilateral and multiple, when they present with other head and neck paragangliomas and malignant tumors (i.e. lung, larynx, breast cancer). Therefore, the whole neck and thorax should be examined carefully in patients with unilateral masses.^[5]

Diagnostic methods and surgical treatment of carotid body tumors are still on debate. The most frequently used diagnostic methods are physical examination, USG, CT, MRI, MRA and SCA. In our study, we used these diagnostic methods; however we did not perform fine-needle aspiration biopsy (FNAB) due to increased risk of bleeding.^[6,7]

The selection of the treatment modality should be done carefully considering age of the patient, symptoms, tumor size, growth rate, and whether it is at the side of dominant hemisphere as well as the potential complications. Due to the slow growth of the tumor, low malignant potential and increased risk for

cerebrovascular complications, radiotherapy (RT) or embolization may be chosen as an alternative to surgical treatment.^[8] Preoperative selective embolization of the feeding arteries is still controversial. During the process of preoperative embolization, neurological deficit including stroke may develop, even it is rare. However, preoperative embolization reduces vascularity and bleeding and hence helping the total resection of the mass.^[9] In our study, we performed selective embolization in seven patients, no embolization-related complications were observed. We also observed reduced bleeding following embolization. It can be challenging to intraoperative dissection due to the leakage of the material extravasation, unless preoperative embolization is applied thoroughly.

Some authors are against RT due to the fact that carotid body tumors are radio-resistant to RT as well as possible side effects.^[10] However, RT has been widely used postoperatively in patients with a malignant glomus jugularis tumor in the middle ear with either local or distant metastases or recurrent disease. Although one tumor was reported as malignant in one patient, RT was not administered, as the whole tumor was removed in our study. Annual follow-up visits were recorded. Since malignant change may develop in 3 to 12%

of the patients, surgical excision is considered the most appropriate treatment for the head and neck paragangliomas.^[11] Pathological examination revealed malignancy in one patient (10%).

Several surgeons routinely recommend the ligation of external carotid artery and its branches to reduce procedure-related complications. However, the internal branch of carotid should be protected or the repair should be performed through saphenous vein or synthetic graft, if not. During surgery, it may be required to apply the clamp to the common carotid artery due to the risk of severe bleeding. If clamping takes less than 10 minutes, the risk of neurological damage is quite low.^[12] In our study, we performed saphenous vein grafting using a clamp for less than 10 minutes and we observed no postoperative neurological problems. However, hypoglossal nerve palsy developed due to the nerve damage which was resolved within two weeks. This may be attributed to the expansion of the surgical site. In addition, we did not perform the ligation of the carotid externa and its branches in our study.

Carotid body tumors which are usually rare, can be treated in reference clinics only, and require a multidisciplinary approach. The pathological examination of the removed tissue is also of utmost importance as treatment. Although it is often benign, the possibility of malignancy should not be overlooked. The growing mass in the anterior triangle of the neck (around the Level 2-3), should be considered carotid body tumor. In the detection of hereditary cases, etiology should be searched by multi-centered genetic studies.

Declaration of conflicting interests

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REFERENCES

1. Qin RF, Shi LF, Liu YP, Lei DL, Hu KJ, Feng XH, et al. Diagnosis and surgical treatment of carotid body tumors: 25 years' experience in China. *Int J Oral Maxillofac Surg* 2009;38:713-8.
2. Netterville JL, Reilly KM, Robertson D, Reiber ME, Armstrong WB, Childs P. Carotid body tumors: a review of 30 patients with 46 tumors. *Laryngoscope* 1995;105:115-26.
3. Pacheco-Ojeda LA, Martínez-Viteri MA. Preoperative imaging diagnosis of carotid body tumors. *Int Surg* 2010;95:242-6.
4. Bernard RP. Carotid body tumors. *Am J Surg* 1992;163:494-6.
5. Lee JH, Barich F, Karnell LH, Robinson RA, Zhen WK, Gantz BJ, et al. National Cancer Data Base report on malignant paragangliomas of the head and neck. *Cancer* 2002;94:730-7.
6. Dickinson PH, Griffin SM, Guy AJ, McNeill IF. Carotid body tumour: 30 years experience. *Br J Surg* 1986;73:14-6.
7. Köhler HF, Carvalho AL, Mattos Granja NV, Nishinari K, Kowalski LP. Surgical treatment of paragangliomas of the carotid bifurcation: results of 36 patients. *Head Neck* 2004;26:1058-63.
8. McCaffrey TV, Myssiorek D, Marrinan M. Head and neck paragangliomas: physiology and biochemistry. *Otolaryngol Clin North Am* 2001;34:837-44.
9. Zhang TH, Jiang WL, Li YL, Li B, Yamakawa T. Perioperative approach in the surgical management of carotid body tumors. *Ann Vasc Surg* 2012;26:775-82.
10. Rodríguez-Cuevas S, López-Garza J, Labastida-Almendaro S. Carotid body tumors in inhabitants of altitudes higher than 2000 meters above sea level. *Head Neck* 1998;20:374-8.
11. Trimas SJ, Mancuso A, de Vries EJ, Cassisi NJ. Avascular carotid body tumor. *Otolaryngol Head Neck Surg* 1994;110:131-5.
12. Matticari S, Credi G, Pratesi C, Bertini D. Diagnosis and surgical treatment of the carotid body tumors. *J Cardiovasc Surg (Torino)* 1995;36:233-9.