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Carotid body tumors: challenging complexity of diagnosis and surgical treatment

Karotid body tümörleri: Tanı ve cerrahi tedavideki güçlükler

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Objectives: To evaluate the patients treated for carotid body tumors and the problems encountered during diagnosis and treatment.

Patients and Methods: The study included eight patients (5 females, 3 males; mean age 50 years; range 36 to 68 years) who underwent surgery for carotid body tumors. Diagnosis was confirmed by power Doppler ultrasonography and computed tomography or magnetic resonance imaging in all patients. Angiography was performed in three patients to evaluate vascularity and to perform embolization before surgery. Two patients had previously undergone biopsy elsewhere. The mean follow-up was 15 months (range 2 to 36 months).

Results: The size of the tumors varied from 3 cm to 11 cm. In two patients the tumors were found to extend to the skull base. Ligation of the external carotid artery and the common carotid artery was performed in five patients and in one patient, respectively. Surgery-associated injuries given to the internal carotid artery in two patients were repaired by sutures. The 10th cranial nerve was dissected in two patients who had undergone a prior biopsy. They developed cord paralysis and hoarseness postoperatively. Resection of the 11th cranial nerve in one patient resulted in shoulder pain and drop shoulder. The 12th cranial nerve was repaired end-to-end by neurorrhaphy in two patients. They exhibited significant improvement in nerve functions a year after surgery.

Conclusion: The larger the tumor is, the more difficult the resection is, and the more injuries are caused to the surrounding nerves and vessels.

Key Words: Angiography; biopsy, needle/adverse effects; blood loss, surgical; carotid artery injuries; carotid body tumor/diagnosis/surgery/complications/radiography; cranial nerves/surgery; embolization, therapeutic; paraganglioma.

Amaç: Karotid body tümörü nedeniyle tedavi edilen hastaları ve tanı ve tedavi sırasında karşılaşılan sorunları değerlendirmek.

Hastalar ve Yöntem: Çalışmaya karotid body tümörü nedeniyle ameliyat edilen sekiz hasta (5 kadın, 3 erkek; ortalama yaş 50; dağılım 36-68) alındı. Tüm hastalarda tanı power Doppler ultrasonografi ve bilgisayarlı tomografi ya da manyetik rezonans görüntüleme ile doğrulandı. Ameliyat öncesi damarlanmayı değerlendirmek ve embolizasyon uygulamak için üç hastada anjiografi yapıldı. İki hastaya daha önce başka yerlerde biyopsi yapılmıştı. Ortalama izlem süresi 15 aydı (dağılım 2-36 ay).

Bulgular: Tümörlerin büyüklüğü 3-11 cm arasında değişiyordu. İki hastada tümörün kafa tabanına uzanım gösterdiği görüldü. Beş hastada eksternal karotid arter, bir hastada ana karotid arter bağlandı. İki hastada, cerrahi sırasında internal karotid arterde yaralanma oluştu ve arter dikişle tamir edildi. Başka merkezlerde daha önce biyopsi yapılan iki hastada 10. kranyal sinir kesildi. Bu olgularda ameliyat sonrasında vokal kord paralizisi ve ses kısıklığı gelişti. Bir hastada 11. kranyal sinir kesisine bağlı omuz ağrısı ve düşük omuz ortaya çıktı. İki hastada 12. kranyal sinir uç-uca anastomozla tamir edildi. Bu olgularda ameliyattan bir yıl sonra sinir fonksiyonlarında anlamlı düzelme gözlendi.

Sonuç: Karotid body tümörlerinde tümör büyüklüğü arttıkça rezeksiyon zorlaşmakta ve çevre sinir ve damarlarda hasara neden olma riski yükselmektedir.

Anahtar Sözcükler: Anjiyografi; biyopsi, iğne/yan etki; kan kaybı, cerrahi; karotis arter yaralanmaları; karotis cisim tümörü/tanı/cerrahi/komplikasyon/radyografi; kranyal sinirler/cerrahi; embolizasyon, terapötik; paragangliom.

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Carotid bodies are chemoreceptors located in the carotid bifurcation. They belong to the paraganglial system derived from neuroectoderm.[1] Carotid body tumors (CBT) originating from these chemoreceptors may be sporadic or familial. The treatment of CBTs is important because they may develop in close relationship with important nerves and vessels.[2]

In this study, we reviewed our patients with CBTs and evaluated some difficulties of diagnosis and treatment.

PATIENTS AND METHODS

We treated eight patients for CBTs during the period between 1998 and 2001. Five patients were females, three were males, the mean age being 50 years (range 36 to 68 years). Hospital records of the patients were analyzed, including age, gender, main complaints, duration of symptoms, familial history, location and dissemination of tumors, treatment and follow-up data. All pathologic diagnoses were reviewed. Power Doppler ultrasonography (US) was performed for upper lateral cervical masses located at the level of the carotid bifurcation. Auscultation was carried out in all cases. Suspected cases were studied by computed tomography (CT) or magnetic resonance imaging (MRI) to confirm the diagnosis and to establish the existence of any paraganglioma. Coexisting paragangliomas were also sought by measuring 24-h urinary vanillylmandelic acid (VMA) levels. In three patients with a large mass and extreme vascularization, angiography was performed to determine the relationship with large vessels. The degree of vascular collaterals was assessed with balloon occlusion test and embolization was carried out 24 hours prior to surgery. Two patients whose diagnoses were made at another center through biopsy examination underwent Doppler US and CT or MRI of the cervical region.

Under general anesthesia, a horizontal incision was made starting from the anterior side of the sternocleidomastoid muscle. The carotid sheet was opened from the inferior part of the tumor and the common carotid artery was set free in order to prepare it for ligation. Then, the internal and external carotid arteries were exposed. The tumor was carefully dissected from the surrounding vessels and nerves. The external carotid artery was ligated and cut when necessary for total excision of the tumor. During surgery, blood loss, amount of blood transfusion, and interventions to the cranial nerves were recorded. The patients were followed-up for a mean of 15 months (range 2 to 36 months) postoperatively.

RESULTS

The main patient complaint was the presence of painless unilateral swelling in the neck. The existence of the mass ranged from two to 20 years. Unilateral involvement was detected in all cases. None of the patients had a positive family history. The size of the tumors ranged from 3x2x1.5 cm to 11x8x4 cm (Table I). Three patients had associated vascular bruits on auscultation. On MRI scans, pressure to the surrounding structures and extension to the skull base were noted especially for large tumors. No intracranial involvement was detected. No neurological deficits were induced during angiography and balloon occlusion test. Of three patients, partial and total embolizations were performed in two patients, respectively, whereas embolization was unsuccessful in one patient. Five patients required external carotid artery ligation and one patient with a large tumor required common carotid artery ligation because of the tumors' intimate arterial adherence. In two patients, surgery-associated injuries given to the internal carotid artery were repaired by sutures. No neurological deficits devel-

TABLE I

DEMOGRAPHIC FEATURES OF PATIENTS AND DIAGNOSTIC PROCEDURES EMPLOYED

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7	Case 8
Age	68	57	46	50	65	36	55	45
Gender	M	M	F	F	F	F	F	M
Duration of complaint (year)	9	2	4	20	3	3	9	15
Tumor size (cm)	7.5x5x3	3x2x1.5	4.5x3x2.7	5.5x4.5x1.5	4x3x2	3.5x2.5x1	5.5x3.5x2.5	11x8x4
Power Doppler+CT/MRI	+	+	+	+	+	+	+	+
Angiography	+	–	–	–	–	–	+	+
Embolization	+	–	–	–	–	–	+	+
Preoperative biopsy	+	–	–	–	–	–	–	+

oped in any of the patients. Dissection of the cranial nerves was required in three patients because of the tumoral encirclement of the nerves. The 10th cranial nerve was dissected in two patients who had undergone prior biopsy examinations. These two patients developed cord paralysis and hoarseness during the follow-up period. Resection of the 11th cranial nerve in one patient resulted in shoulder pain and drop shoulder. The 12th cranial nerve was repaired end-to-end by neurorrhaphy in two patients. They exhibited significant improvement in nerve functions a year after surgery.

During surgery four patients required blood transfusion (6, 4, 5, and 8 units in the 1st , 4th, 7th, and 8th patients, respectively). The size of the tumor was larger than 5 cm in these patients.

DISCUSSION

Difficulties in the identification and treatment of CBTs start at the stage of diagnosis. Since CBTs grow slowly and the only symptom is painless unilateral swelling in the upper lateral neck, differential diagnosis should include branchial cleft cysts, metastatic tumors, lymphomas, carotid aneurysms, and parotid gland masses.[3,4] With the extensive growth of the tumor, pressure sensation in the throat, hoarseness, dysphagia, cough, local irritations, pain, and cranial nerve disorders may arise.[1,5] In some cases, tumor growth to the parapharyngeal space may give rise to the displacement of the tonsils medially and to symptoms accordingly.[1,2,4] A high index of suspicion is required for the correct identification of CBTs. Although In some cases, biopsies are performed because of misdiagnosis.[1,6,7]

In some cases, biopsies are performed because of misdiagnosis.[1,6,7] The number of misdiagnosed cases was one, six, and five in three studies including 11,[5] 16,[1] and 18[8] patients, respectively. Of note, surgical exploration was performed in some cases, as well, which was associated with a higher incidence of cranial nerve injuries and an increased amount of bleeding compared to surgical operations performed with correct diagnoses of CBTs.[1,8] The main complaint was the existence of a mass in the neck in our patients. Of these, two patients underwent prior biopsy examinations in another medical center.

Although history and physical examination are helpful, they are not diagnostic in CBTs. Preoperative radiologic studies should be employed to determine the relationship of the tumor with vascular structures, to demonstrate blood supply, and to indicate the need for embolization.[2]

Routine ultrasonograms and power Doppler studies were reported to be diagnostic in CBTs.[9] Doppler US is an inexpensive method without any risks. It demonstrates vascular and solid masses located at the carotid bifurcation. However, its value is limited in the identification of tumoral extension and invasion to the neighboring structures.[10] On the other hand, CT and MRI may show invasion and relationship of the tumor with the neighboring structures with higher sensitivity and specificity.[10,11] Pepper and salt sign is an important finding on MRI scans by which paragangliomas even 0.8 cm in size can be accurately detected.[3]

In addition, magnetic resonance angiography (MRA) is available to evaluate vascularity of the CBTs and the surrounding vascular structures, although it has some limitations in the demonstration of specific vascularization of CBTs.[2] Finally, angiography is the gold standard in the diagnosis of CBTs, demonstrating them as vascular tumors with precise differentiation of the external and internal carotid arteries in the bifurcation of the common carotid artery.[12] It gives valuable information about the distribution of blood vessels, relationship of the lesion with the carotid arteries, the extent of involvement, and on the localization of the collaterals. Moreover, it enables to carry out evaluation tests for preoperative embolization and internal carotid artery occlusion.

We performed power Doppler US to determine whether any mass existed around the carotid bifurcation in all patients. Diagnoses were confirmed by CT or MRI.

Gardner et al.[5] reported that four-vessel cerebral angiography is necessary for both familial and sporadic cases because of high possibility of multiple paragangliomas. In their study, cerebral angiography revealed CBTs in seven patients, vagal paragangliomas in four patients, and glomus paragangliomas in two patients, all of which were not diagnosed before. Leonetti et al.,[1] however, did not support the use of this procedure for one-sided and isolated CBTs.

Total surgical excision is the only treatment modality for CBTs. The role of preoperative embolization is controversial. Some authors do not recommend embolization because of lack of benefit, [1,13,14] while others advocate its need especially for tumors greater

than five centimeters in order to decrease the amount of perioperative bleeding, to cause tumoral shrinkage, and to reduce associated morbidities.[1,5,10,12,13,15-17]

Angiography was performed in three patients with very large lesions to evaluate blood supply to the tumors, cerebral circulation, and the relationship of the tumor with the carotid vessels. In none of them embolization was helpful. This may be related to large amounts of blood supplied to the tumors, the presence of wide collaterals, or failure in embolization.

Adherence to the basic principles of surgery may help facilitate the operation. These include precautions to explore and protect the cranial nerves neighboring the tumor by wide surgical exposure, giving considerable attention to distal and proximal areas of the vessels, correct identification of neurovascular structures, subadventitial dissection of the lesion from the bottom to the top, and, when necessary, performing external carotid artery ligation.[11] The use of bipolar cautery in the surgical area may also help reduce bleeding. Last but not least, having a vascular surgeon available in the operation room is of vital importance for emergent situations especially in the presence of very large tumors encircling the carotid artery.[10]

Irrespective of the extent of care given to follow the above mentioned principles, the larger the tumor is, the more difficult the resection is and the more injuries are caused to the surrounding nerves and vessels.[1] Netterville et al.[7] reported that 65% of patients suffering from postoperative cranial nerve paresis had tumors larger than five centimeters in size. In a study by Anand et al.[8] the size ranged from 3 to 6 cm in 80% of the tumors, exceeded 6 cm in 15%, resulting in a total of 21 cranial nerve deficits in 50% of patients. Similarly, in our patients increase in tumor size was associated both with a higher incidence of nerve paralysis and bleeding.

Carotid body tumors may be accompanied by other paragangliomas. It has been reported that the possibility to detect a coexistent paraganglioma is about 5% to 10% in the sporadic type, and 10% in the familial type.[3] In addition, Gardner et al.[5] reported that the possibility of a coexistent paraganglioma is 100% in patients with a positive family history. Netterville et al.[7] detected associated paragangliomas in 43% of sporadic cases and in 87% of familial cases. Since the presence of multiple paragangliomas is associated with a higher incidence of injuries to the nerves and vessels, we performed Doppler US in all patients and found that all were solitary masses.

The major complication of surgery for CBTs is injury occasioned to the 9th, 10th, 11th, and 12th cranial nerves, the 10th cranial nerve being the most common site.[5] Surgery-associated nerve injuries are more likely to occur in the presence of such conditions as large tumors, tumors encircling the nerve, prior surgical interventions directed to the lesion, excessive bleeding, and a coexistent paraganglioma. In addition, dissection of the cranial nerves during surgery may be required because of the intimate tumor involvement. Whatever the cause of the injury to cranial nerves is, about 32% to 34% of patients undergoing surgery may suffer from postoperative sequelae.[3]

Although injuries to the 9th, 11th, and 12th cranial nerves can be tolerated by the patients,[1,12] injury given to the 10th cranial nerve may result in aspiration and postoperative airway obstruction, so that medialization may be necessary for the paralyzed cords following any injury to the 10th cranial nerve. Moreover, if multiple cranial nerve injuries have been induced, the use of a nasogastric tube and therapy to rehabilitate the patient's speech and swallowing should be considered.

Dissection of the cranial nerves was required in three patients because of the tumoral encirclement of the nerves. The 10th cranial nerve was dissected in two patients in whom intact anatomical structure might have been destroyed by prior biopsy examinations. These two patients developed postoperative cord paralysis and hoarseness. Resection of the 11th cranial nerve in one patient resulted in shoulder pain and drop shoulder. On the other hand, significant improvement was observed in nerve functions after a year in two patients whose 12th cranial nerve was repaired end-to-end by neurorrhaphy.

One important complication that we encountered during surgery was massive bleeding.[8] It mainly resulted from small lacerations in the carotid arteries and from the tumor itself. Although small lacerations in, and trivial injuries to, the carotid arteries can be primarily repaired, larger lesions may require grafting or shunting especially with the use of saphenous vein grafts. We performed ligation of the external carotid artery in five patients and of the common carotid artery in one patient. In two patients, injuries given to the internal carotid artery were repaired by sutures. The size of the tumors was greater than 7 cm in one patient whose common carotid artery was ligated and

in another whose internal carotid artery was repaired by sutures. Of particular note, the two patients who had undergone prior biopsies had large tumors, the removal of which caused profuse bleeding and resulted in significant injuries.

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