

CASE REPORT

Malignant carotid body tumor: a case report

Malign karotis cismi tümörü: Olgu sunumu

M. Zafer UĞUZ, M.D., Seçil ARSLANOĞLU, M.D., Kazım ÖNAL, M.D., Hünkar GÖKÇE, M.D.

Malignant carotid body tumors are rare neoplasms which originate from the chemoreceptor cells at the carotid bifurcation. A 61-year old man presented with a neck mass which was diagnosed as malignant carotid body tumor. Surgical resection of the mass with accompanying lymph nodes was performed. No evidence of local recurrence or distant metastasis were detected during the follow-up period.

Key Words: Carotid body tumor/diagnosis/pathology/ therapy; paraganglioma, extra-adrenal/diagnosis/pathology/ therapy; embolization, therapeutic.

Malign karotis cismi tümörleri, karotis bifurkasyonundaki kemoreseptörlerden kaynaklanan nadir görülen neoplazmlardır. Boyunda kitle şikayeti ile başvuran 61 yaşında erkek hasta malign karotis cismi tümörü tanısı aldı. Eşlik eden lenf nodları ile birlikte kitle çıkarıldı. Ameliyat sonrası izlemde tümör nüksü ya da uzak metastaz gözlenmedi.

Anahtar Sözcükler: Karotis cisim tümörü/tanı/patoloji/ tedavi; paragangliom, adrenal dışı/tanı/patoloji/tedavi; embolizasyon, tedavi edici.

Carotid body tumors (CBT) are rare neoplasms which consist of chemoreceptor cells at the carotid bifurcation. The most common presenting symptom of a carotid parotid paraganglioma is a slow growing neck mass located at the bifurcation of common carotid artery. Symptoms of pressure, cough, hoarseness or dysphagia can be encountered in large lesions. The majority of these tumors are benign whereas 5-10% are malignant.^[1] Diagnosis should be based on radiographic findings, and surgery is the mainstay of treatment.

CASE REPORT

A 61 year old man was admitted to İzmir Atatürk Training and Research Hospital, in December 2001

with an asymptomatic, nontender, mobile, slow-growing, hard mass with a diameter of 4x5 cm in the upper half of the left side of the neck. The mass had been present for 1 year before the admission. A Doppler ultrasound revealed a 4x5 cm solid, highly vascular mass located at the left carotid bifurcation. The arterial structures within the lump showed flow types of reduced resistance. Computed tomography (CT) of the neck showed a mass, 4x5 cm in size at the carotid bifurcation with associated lymph nodes reminding of a malignant CBT. A fine needle aspiration biopsy revealed malignant cytology with pleomorphic and occasional multinuclear cells with hyperchromatic nuclei in a hemorrhagic environ-

- ◆ Department of Otolaryngology, İzmir Atatürk Training and Research Hospital (İzmir Atatürk Eğitim ve Araştırma Hastanesi KBB Hastalıkları Kliniği), İzmir, Turkey.
- ◆ Received - July 29, 2003 (Dergiye geliş tarihi - 29 Temmuz 2003). Request for revision - January 14, 2004 (Düzeltilme isteği - 14 Ocak 2004). Accepted for publication - February 14, 2004 (Yayın için kabul tarihi - 14 Şubat 2004).
- ◆ Correspondence (İletişim adresi): Dr. M. Zafer Uğuz. 108/25 Sokak, No: 4/5, 35350 Esenyalı, İzmir, Turkey. Tel: +90 232 - 231 62 52 Fax (Faks): +90 232 - 243 48 48 e-mail (e-posta): mzuguz@hotmail.com
- * Presented at the 26th National Congress of Turkish Otolaryngology, Head and Neck Surgery (September 22-26, 2001, Belek, Antalya, Turkey) (26. Ulusal Türk Otorinolarenoloji ve Baş-Boyun Cerrahisi Kongresi'nde sunulmuştur (22-26 Eylül 2001, Antalya)).

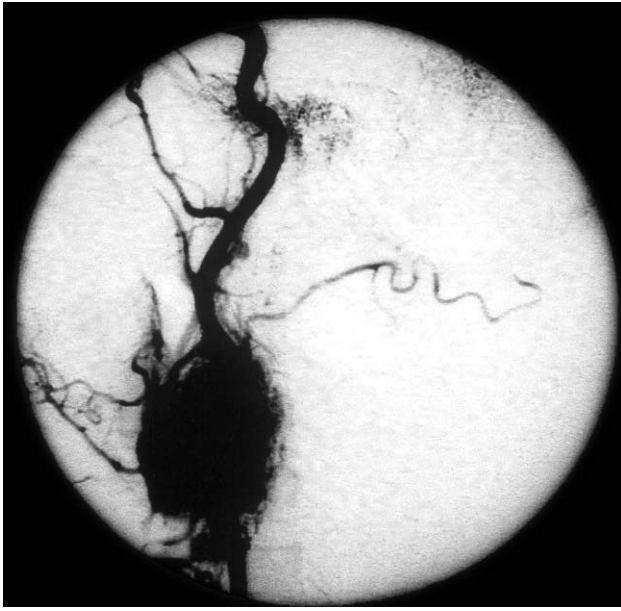


Fig. 1 - Carotid body tumor at the carotid bifurcation demonstrated by angiography.

ment. A carotid arteriogram demonstrated a mass consistent with CBT located between the left internal and external carotid arteries. The blood supply of the tumor was primarily the external carotid artery (Fig. 1).

Preoperative balloon occlusion testing during angiography was performed and no neurologic

deficit was observed. The test revealed that the mass had blood supply from the perforating branches of external carotid artery (ECA). The lesion was embolized with N-butyl cyanoacrylate in Lipiodol solution, 33% reduction in the tumor blush.

Under general anesthesia the mass was removed with the accompanying lymph nodes preserving the carotid vascular structures. At the operation the hemorrhagic mass was seen to originate from the carotid bifurcation and extend along the ECA and internal carotid artery 2 and 2.5 cm, respectively. Four lymph nodes in a cluster, the biggest of which 2x2 cm, were removed from the cervical level II. Another lymph node of 1x1 cm was located inferior to the mass was also included in the dissection. The histopathologic examination revealed the presence of malignant paraganglioma with invasion to the lymph nodes (Fig. 2). Histopathologic examination of the specimen was consistent with the fine needle aspiration biopsy.

In the postoperative period the 9th and the 10th cranial nerve deficits and nominal aphasia were encountered. The cranial CT and magnetic resonance imaging (MRI) performed in this period revealed a wide subacute cortical infarction. Cranial nerve deficits disappeared within 2 weeks of surgery, and aphasia diminished gradually. Radiotherapy (50 Gy) was administered after the

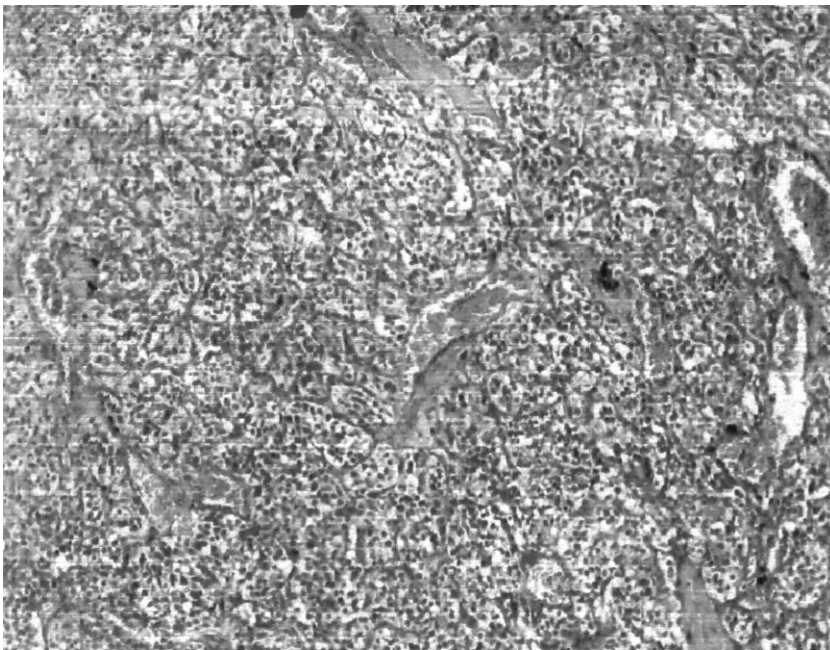


Fig. 2 - Tumor infiltration in a lymph node. Cell nests separated with fibrovascular septae (H-E x 220).

operation. The patient was free of disease 20 months after resection.

DISCUSSION

A carotid body is situated at the bifurcation of the common carotid artery and it may give rise to CBT which is the most common of the paragangliomas. Most carotid body tumors are benign but a small percentage may manifest regional or distant metastases so are classified as malignant. Malignancy in paragangliomas occurs in 5-10% of cases and is defined by the presence of metastases.^[1,2] However, it has been observed that the frequency of metastases show a wide range between 0-20%. Histologic characteristics do not determine malignancy. The diagnosis of malignancy is achieved by the existence of metastases.^[2]

Usually the patients refer with a slow-growing lateral neck mass, sometimes associated with hoarseness and cough. The diagnostic procedures include MRI, MR angiography, yet angiography is still valuable for evaluation of CBT.^[3] An incisional biopsy is not recommended because of the risk of hemorrhage and injury of adjacent neurovascular structures. A balloon occlusion test can be performed to assess the intracranial collateral circulation.^[4] Preoperative transarterial embolization to reduce vascularity of the tumor is a controversial procedure because it carries the risk of cerebral complications, yet there are reports which advocate its benefits.^[2,4] Kafie and Freischlag^[4] recommended to perform the operation within 48 hours of embolization to minimize revascularization edema or a local inflammatory response.^[5] We performed surgery on the 4th day of embolization.

Defining malignancy in CBT has been controversial. Central necrosis of cell clusters, mitosis and invasion of vascular spaces have been described while other authors have accepted only the presence of metastases as the evidence of malignancy.^[3,5] In our patient, the histopathologic examination revealed

clusters of cells with nuclei of pleomorphic vesicles. Mitoses were evident with wide areas of necrosis. Tumor infiltration in the same fashion was notable in a lymph node. The presence of lymph node involvement was the reliable characteristic for malignancy.

Surgical resection is the mainstay of therapy. To minimize the risk of complications subadventitial dissection should be carried out at an early stage. If the tumor is considered malignant, locoregional control is usually obtained with complete primary tumor resection and lymphadenectomy, and eventual radiotherapy. Furthermore, the patient must be observed for the possibility of metastases which might occur after long periods of time (which vary between 20 months and 20 years) following resection.^[5] We resected the tumor with the associated lymph nodes and postoperative radiotherapy was given. Our patient, by the 25th month postoperatively was free of tumor, and we need to follow-up for a long period of time owing to the wide range of time interval between the resection of the primary tumor and the appearance of metastases. Since a wide range of time interval between the resection of the primary tumor and the appearance of metastases is present, patients should be in close contact for the detection of metastasis.

REFERENCES

1. Bernard RP. Carotid body tumors. *Am J Surg* 1992;163:494-6.
2. Liapis CD, Evangelidakis EL, Papavassiliou VG, Kakisis JD, Gougoulakis AG, Polyzos AK, et al. Role of malignancy and preoperative embolization in the management of carotid body tumors. *World J Surg* 2000;24:1526-30.
3. van der Mey AG, Jansen JC, van Baalen JM. Management of carotid body tumors. *Otolaryngol Clin North Am* 2001;34:907-24.
4. Kafie FE, Freischlag JA. Carotid body tumors: the role of preoperative embolization. *Ann Vasc Surg* 2001;15:237-42.
5. Pacheco-Ojeda L. Malignant carotid body tumors: report of three cases. *Ann Otol Rhinol Laryngol* 2001;110:36-40.