Malignant carotid body tumor: A case report

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Carotid body tumors (CBTs) have an unpredictable history with no correlation between histology and clinical behavior. Of reported cases since 1891, local and distant metastases appear in approximately 10% of cases and remain the hallmark of malignancy. Currently, there are not enough data to support a single treatment regimen for malignant CBTs. The reported case demonstrates some unanswered issues with regard to malignant CBTs to include lymph node dissection, the need for carotid resection, and the role of radiation therapy. A 46-year-old pathologist underwent a resection of a Shamblin I CBT, to include jugular lymph node sampling, without complication. There was lymph node involvement, and tumor cells were found on the margins of the pathologic specimen. Subsequent carotid resection with reversed interposition saphenous vein graft and modified neck dissection were performed again without complication. Follow-up at 4 years has been uneventful. Diagnosis of CBTs with the use of magnetic resonance angiography, magnetic resonance imaging, color flow duplex scanning, and the role of arteriography are reviewed. The current treatment options are discussed with reference to primary lymph node sampling, carotid resection, and neck dissection in malignant cases. This case demonstrates that the unpredictable nature of CBTs and their malignant potential warrant aggressive initial local treatment to include jugular lymph node sampling and complete tumor resection. (J Vasc Surg 2000;32:821-3.)

In 1894 a case of a carotid body tumor (CBT) in which there was lymph node involvement was reported for the first time by Kopfstein. Sapegno published the first account of distant metastases from this tumor in 1913.

CBT is a slow-growing tumor, and there is no report of spontaneous regression. It usually presents as a gradually enlarging mass that is incidentally identified. Metastases to lymph nodes or distant sites remain the hallmark of malignancy for CBTs.

Usually there is no hint of malignancy in the histology of the primary tumor. This fact, along with the unpredictable natural history of CBTs, makes the diagnosis and treatment of malignant CBTs difficult. Of 46 malignant CBTs described in the literature after 1980, there were five patients who died of their metastatic disease, but cases of controlled metastatic disease have been described for up to 32 years.[1] Currently, there are not enough data to support a single treatment regimen for malignant CBTs. Should lymph node sampling be routine when resecting CBTs? What should the management of nodal disease noted on the pathologic examination of the primary resection be? A case of a malignant CBT is reported below followed by a discussion of the management options.

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CASE REPORT

The patient is a 46-year-old white male pathologist who incidentally noticed a right anterior neck mass approximately 3 months before admission. The patient had undergone an attempted lymph node biopsy before referral. The mass at the time of biopsy was noted to be confluent to the carotid artery, and the procedure was aborted. The patient's past medical history included hypercholesterolemia. He grew up in Utah in an area of high elevation. There was no familial history of paragangliomas. On physical examination his cranial nerves were normal. The right side of his neck had a 2 × 2 cm mass palpable in the area of the right carotid bifurcation. The mass

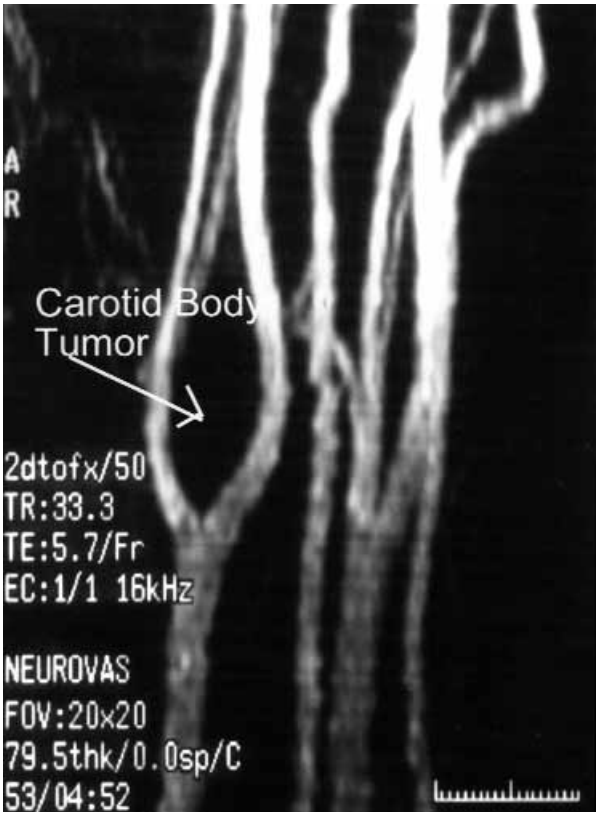

Fig 1. Right carotid tumor splaying the internal and external carotid arteries on MRA.

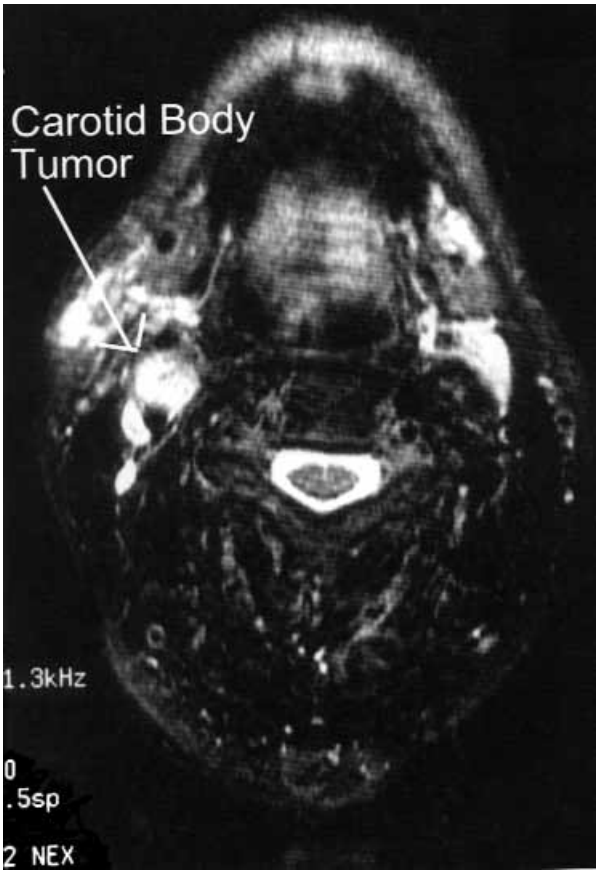

Fig 2. Right CBT on MRI.

was mobile laterally and medially, but not vertically. Color flow duplex, magnetic resonance angiography (MRA) (Fig 1), and magnetic resonance imaging (MRI) (Fig 2) scans revealed a single mass of approximately 3 × 2 cm splaying the right carotid artery bifurcation. Hypervascularity could be shown with power Doppler imaging. No significant carotid artery disease was found. No signs of a functional paraganglioma were found.

A diagnosis of CBT was made, and the patient underwent a surgical resection in a periadventitial plane with sampling of jugular lymph nodes. The patient's postoperative course was unremarkable, and he was discharged on postoperative day 2.

The pathologic results confirmed the diagnosis of CBT and revealed a positive tumor margin on the carotid bifurcation as well as metastatic disease in one of the lymph nodes. An indium In 111 octreotide whole body scan was performed 1 month postoperatively and showed increase uptake in the region of the left anterior neck. Two months later, a follow-up study showed resolution of the prior abnormality, which was probably due to an inflammatory response. The remainder of the neck, chest, abdomen, and pelvis did not show any sign of metastatic disease.

Five months after the CBT removal and after much discussion with consultants and the patient, he was returned to the operating room for carotid resection and a modified neck dissection. There was no evidence of gross metastatic disease in this area. The carotid bifurcation area showed a normal appearance. The external carotid artery was ligated, and the carotid bifurcation was resected with an interposition reversed saphenous vein graft reconstruction. The result of an intraoperative color flow duplex scan was normal. The pathologic examination of the carotid artery and lymph nodes showed no sign of residual disease. The patient recovered without any evidence of neurologic deficit. Four years after the operation, the patient's follow-up has been uneventful with no signs of recurrence or metastatic disease and a patent graft.

DISCUSSION

Since 1891, approximately 152 of 1425 CBTs described in the literature were malignant. The histologic pattern of the primary lesion is not a reliable indicator of malignancy. Rather, lymph node evolvement or the presence of distant metastasis remains the hallmark for the diagnosis of malignant CBTs.[2]

The initial diagnosis of CBT is not uncommonly made at the time of the neck exploration for possi-

ble lymph node biopsy and may account for as many as 30% of the cases. If a CBT is suspected preoperatively, color flow duplex scanning is very characteristic and should be the first screening test. MRI and MRA are complementary examinations that are useful in evaluating for other paragangliomas. The contralateral neck and carotid bifurcation should also be evaluated because bilateral disease is not uncommon, especially in familial cases where they can account for 30% of the cases. Recent reports of radioisotopic scans have demonstrated a possible yet undefined role in diagnosis, follow-up, and treatment. Patients should also be screened for functional paragangliomas because of their association with pheochromocytomas.

Arteriography has been regarded as the "gold standard" for diagnosis. However, with high-definition color flow duplex scanning, MRI, and MRA, arteriography may be reserved for larger CBTs.[3] Preoperative embolization has been shown to diminish the operative blood loss for these larger CBTs.[4]

This case is unique in that the primary lesion was small yet had already metastasized to regional lymph nodes. The primary lesion itself gave little evidence of being malignant. It is possible that the incidence of malignant CBTs is underestimated because formal lymph node sampling may not be a common practice. Late recurrences, which occur in 5% in the largest reported series,[5] may indeed represent malignant primary lesion.

The benefit of radiotherapy combined with surgery in the treatment of paragangliomas is controversial. CBTs have been considered resistant to radiotherapy, but recent studies show local control[6] with regression in some cases.[2,7] Radiotherapy is usually reserved for patients medically unfit for surgery, unresectable disease, recurrence after surgery, or cases of metastases.[1,3,7,8] On the other hand, recurrent tumor growth following radiation,[1] as well as serious complications after radiotherapy, has been described.[2] Radiotherapy is generally accepted for the treatment of inoperable paragangliomas but is rarely used for resectable CBT.

Most of the benign CBTs do not justify a radical lymph node dissection or carotid bulb resection. However, the sampling of regional lymph nodes for metastases has been proposed,[9] and some authors state that radical resection prevents local recurrence and shows the best long-term results.[8] This case demonstrates why initial lymph node sampling with the primary tumor should be performed routinely. In our case the pathologic examination of the resected carotid artery showed no evidence of residual tumor even though that was suspected after the first tumor resection. This may have been due either to the limitations of the pathologic preparation or to the complete removal of the tumor at the time of the original resection. However, without definite evidence of a clear margin on the first pathologic specimen, we were not comfortable leaving the carotid bifurcation behind. As noted, this patient has not shown any evidence of gross recurrence at 4 years.

Metastases may develop many years after original resection; therefore, long-term follow-up is mandatory. When carotid resection with graft reconstruction is necessary, duplex scanning follow-up of the graft is also recommended. Screening of primary relatives is recommended in all cases of CBT.

Even though it was said as early as 1917 that "these curious little tumors have been dissected, studied and described almost ad nauseam et ad infinitum,"[10] there is still a lot to be learned.

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