SHORT REPORT

Indication for Surgery in the Presence of Familial Bilateral Carotid Body Tumours

G. De Toma, P. Sapienza, C. Letizia, V. Nicolanti, G. Piccirillo, U. Basile and A. Cavallaro

1Department of Surgery “Pietro Valdoni”, and 2Second Department of Medicine, University of Rome “La Sapienza”, Rome, Italy

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Introduction

In recent years the failure of the baroreceptor reflex after the excision of bilateral carotid body tumours (CBT) has been described with increasing frequency in the world literature.1–5 The baroreceptor reflex is a powerful negative feedback control mechanism that buffers acute changes in cardiovascular function and regulates the fluctuations in blood pressure. Few cases of labile blood pressure after the resection of a second CBT in familial disease have been reported in the literature. We report the cases of two family members affected with bilateral CBT and discuss the indication for surgery in the presence of bilateral familial CBT. In one case a bilateral CBT excision was performed while the other, because of the occurrence in the first family member of a life-threatening loss of the arterial baroreceptor function, was left untreated.

Case 1

A 47-year old female patient was admitted because of a small painless mass in the left cervical region. She had had surgery for a papillary carcinoma of the thyroid gland 2 years previously. A colour Doppler was performed and showed the presence of a solid, hypoechoic mass 2.4 cm in diameter at the level of the left carotid bifurcation. Angiography showed the presence of a bilateral CBT. The left tumour was removed from the carotid vessels in a sub-adventitial plane. The postoperative course was uneventful. The patient was then re-admitted for removal of the right carotid body tumour. The postoperative course was complicated by hypertension (240/130 mmHg) and sinus tachycardia (120 beats/min) accompanied by severe, pounding frontal headaches, nausea, vomiting, and skin flushing. The symptoms were treated with the intravenous administration of clonidine and nifedipine. The patient’s heart rate remained a fairly constant 100–120 beats/min between hypertensive crises. The patient developed marked emotional lability which was treated with benzodiazepine. The presence of an undiagnosed pheochromocytoma was excluded with an ultrasound and 24-hour urine tests for catecholamines. All other investigations were normal. However, the patient continued to have hypertensive crisis twice a day. The patient underwent baroreflex sensitivity evaluation by means of transfer function analysis, which revealed a severe impairment of the baroreceptor reflex system consistent with the clinical manifestations. She was discharged under benzodiazepine, clonidine and β-blockers. At 4-year follow-up the patient still complains of hypertensive crises (once to twice a month) and a disrupted lifestyle.

Case 2

A 38-year-old female patient was admitted because of
a bilateral small painless mass in the cervical region. A colour Doppler showed the presence of a solid, hypoechoic mass $3.5 \times 4.5$ cm in diameter at the level of the right carotid bifurcation, and a similar mass $(2.3 \times 1.8)$ cm on the left side. A CT scan confirmed the presence of bilateral tumours. The left tumour was removed from the carotid vessels in a sub-adventitial plane. The patient’s postoperative course was uneventful. Blood pressure and heart rates remained in the normal range. The patient was felt to be at risk of development of baroreceptor reflex failure so close follow-up was planned to monitor any eventual enlargement of the left CBT. At 3 years follow-up the patient is in good general condition without signs and/or symptoms of recurrence or distant metastases and the untreated carotid body tumour remains the same size.

**Discussion**

Baroreflex failure syndrome is characterized by the loss of buffering of blood pressure with consequent volatile blood pressure and heart rate on both sides of the spectrum. Hypertension usually develops after 24–72 hours after the second CBT excision. The pathophysiology of the baroreflex failure syndrome is poorly understood but damage to Hering’s nerve, the glossopharyngeal, hypoglossal or vagus nerves is believed to be significant. Since CBT excision and carotid endarterectomies are performed through a sub-adventitial plane, the surgical impact at the level of the carotid bifurcation nerves could be similar thus producing damage to the carotid afferent nerves. In our series of carotid endarterectomies we observed the development of this syndrome in one case out of 173 bilateral endarterectomies (0.6%). This patient underwent carotid endarterectomy for a severe bilateral carotid artery stenosis. After the second operation the patient developed a typical baroreflex failure syndrome with volatile blood pressure and heart rate. The patient responded well to medical treatment. The actual incidence of baroreflex failure syndrome is not clear because the diagnosis is not made using tests of baroreflex control of cardiovascular function. In our cases we carried out specific tests able to detect baroreflex impairment. The tests have already been well decribed in our previous study. Based on our experience we recommend unilateral excision of bilateral carotid body tumours with regular follow-up and surgery if the second tumour increases in size.

**References**


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