Surgical management of a symptomatic extracranial internal carotid artery aneurysm and coexisting carotid body tumor

Qusai Al-Jarrah, MBBS, MD, Mohammed Ashrafi, MB ChB, BSc, Jordan Oldbury, MB ChB, Steven Rogers, BSc, Mohammed Baguneid, MB ChB, MD, and **Leszek Wolowczyk, MB ChB, MD,**
Manchester, United Kingdom

Treatment of carotid body tumors and extracranial carotid artery aneurysms are well documented in the literature as separate entities. As distinct pathologies, they present technical difficulties with high complication rates. No patients with simultaneous carotid body tumors and extracranial internal carotid artery aneurysms have been reported. We report, to our knowledge, the first and subsequent surgical management of such a patient. (J Vasc Surg Cases 2015;1:134-7.)

Carotid body tumors (CBTs) are rare neoplasms arising from the carotid body and presenting technical difficulties in excision, with high complication rates. Similarly, carotid artery aneurysms present technical difficulties in excision, with high complication rates. No patients with simultaneous CBTs and extracranial internal carotid artery (ICA) aneurysms have been reported. We review the current management of these two distinct pathologies and relay our surgical experience of this unique patient. The patient consented to publication of this report.

CASE REPORT

A 76-year-old woman presented with sudden-onset left-sided facial weakness and sialorrhea. Her medical history included chronic obstructive pulmonary disease and hypertension. The clinical examination revealed a small, firm, and smooth mass in the right carotid triangle. The neurologic examination was unremarkable. Vocal cords were normal on laryngoscopy.

Results of blood tests, including serum vanillylmandelic acid levels, were unremarkable. An electrocardiogram and echocardiography did not reveal any significant cardiac abnormalities.

A three-dimensional carotid duplex examination revealed a 1.68-cm × 1.3-cm well-vascularized echogenic mass splaying the carotid bifurcation (Fig 1, *A*). Magnetic resonance imaging did not demonstrate an ipsilateral ischemic stroke but did identify a distal cervical segment ICA aneurysm measuring 2.5 × 2.3 × 2.1 cm that had not previously been visualized on duplex imaging (Fig 1, *B*). Symptoms on initial presentation were most probably caused by distal embolization from thrombus within the ICA aneurysm. Sialorrhea is sequelae of left-sided facial weakness.

Initially, a catheter angiogram in view of carotid stenting using a Viabahn (W.L. Gore and Associates, Flagstaff, Ariz) covered stent graft was performed via a right common femoral artery puncture to treat the symptomatic ICA aneurysm. Angiography confirmed the presence of a CBT and a large aneurysm of the distal cervical ICA. Because of the tortuosity of the proximal ICA, the catheter could not be advanced further and the procedure was abandoned (Fig 1, *C*).

Intravenous heparin (5000 IU) was given, and a balloon occlusion test was performed in the proximal ICA using a 5-mm angioplasty balloon. There was no evidence of left-sided neurologic deficit 5 minutes after inflation of the balloon. A balloon occlusion test was conducted to evaluate for adequate cerebral collateral circulation in case we needed to clamp the ICA intraoperatively without shunt insertion, or if the ICA aneurysm repair was deemed to be technically impossible, an alternative would be to ligate the ICA. A prolonged balloon occlusion test was avoided due to the risk of thrombus formation around the balloon and the risk of distal embolization.

Initial plans were to treat the CBT surgically as a second procedure after the patient recovered from the ICA aneurysm stenting. Because the aneurysm could not be stented through an endovascular intervention, open repair was necessary, and a surgical plan was made to tackle both entities simultaneously.

The patient underwent open repair of the ICA aneurysm under general anesthetic, with invasive blood pressure and intraoperative transcranial Doppler monitoring, using an antejugular approach. The common carotid artery, external carotid artery, and ICA were dissected sequentially, and proximal control was achieved. The CBT was attached to the ICA and external carotid artery, splaying the carotid bifurcation (Shamblin type I).

The hilar-like configuration of the ICA aneurysm made dissection of the distal cervical ICA challenging. To facilitate exposure of the ICA aneurysm, we firstly extended the skin incision curving cranially toward the anterior border of the right earlobe. Secondly,

From the Department of Vascular Surgery, University Hospital of South Manchester.

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Reprint requests: Dr Qusai Al-Jarrah, MBBS, MD, Department of Vascular Surgery, University Hospital of South Manchester, Southmoor Rd, Wythenshawe, Manchester M23 9LT, UK (e-mail: q_jarrah@yahoo.com).

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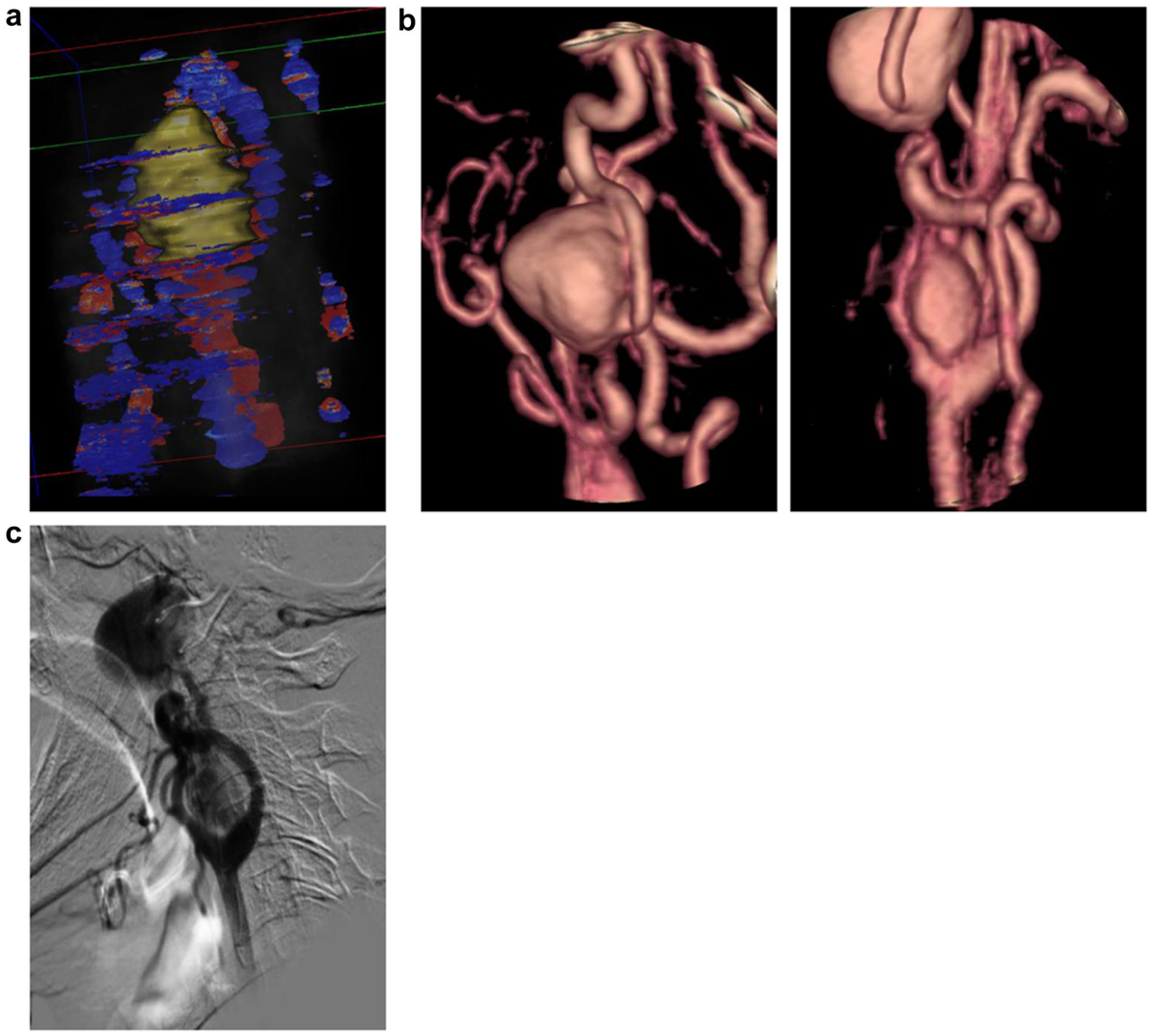

Fig 1. a, A three-dimensional (3D) carotid duplex shows a 1.68-cm × 1.3-cm well-vascularized echogenic mass splaying the carotid bifurcation. **b,** A 3D reconstruction of a magnetic resonance image shows a distal cervical segment internal carotid artery (ICA) aneurysm and carotid body tumor (CBT). **c,** A catheter angiogram shows the tortuosity of the ICA.

we incised the parotid fascia and mobilized the parotid gland medially. Lastly, the digastric muscle was divided, and the CBT was excised (Fig 2, *A*).

Despite undertaking these steps, we were still not able to obtain adequate distal ICA control. After CBT excision, the ICA proximal to the aneurysm was mobilized. The ICA aneurysm had a saccular morphology projecting from the lateral surface of the distal ICA (Fig 2, *B*). This conformation allowed for the proximal ICA to be clamped and the aneurysm sac to be opened without the initial need to achieve distal control. After the aneurysm sac was opened, minimal back bleeding was encountered due to a large amount of thrombus occupying the sac. After evacuation of the thrombus, good back bleeding was allowed, and with the aid of low-flow suction and caudal traction of the aneurysm, the distal ICA was dissected off the sac and safely clamped under direct vision. The use of a shunt was considered but was technically impossible due to the lack of distal segment ICA length to insert the shunt safely.

Ttranscranial Doppler monitoring after cross-clamping showed <60% reduction in flow velocity through the ipsilateral middle cerebral artery. Had flow in the middle cerebral artery dropping significantly, our only strategy would have been to increase the mean arterial pressure to improve the cerebral perfusion pressure. Both ends of the ICA were refashioned, and a wide spatulated end-to-end primary anastomosis was performed (Fig 3). Total clamp time was 30 minutes. A hand-held continuous-wave Doppler probe was used intraoperatively to insonate the distal ICA after the anastomosis.

The patient's recovery was uneventful, apart from transient dysphagia and right-sided facial droop that resolved after 48 hours.

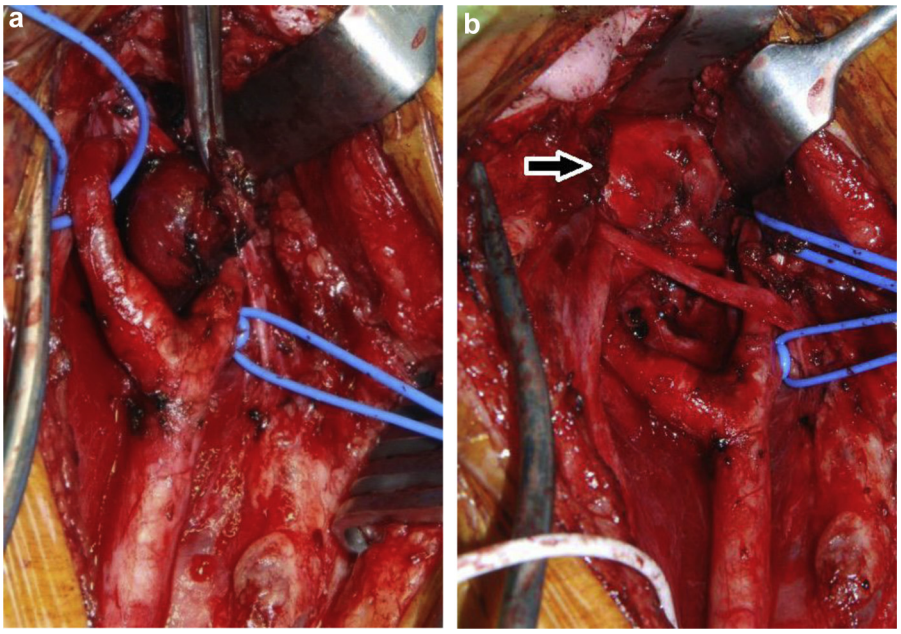

Fig 2. a, Carotid body tumor (CBT) splaying the carotid bifurcation. **b,** Internal carotid artery (ICA) aneurysm. The *arrow* highlights the ICA aneurysm.

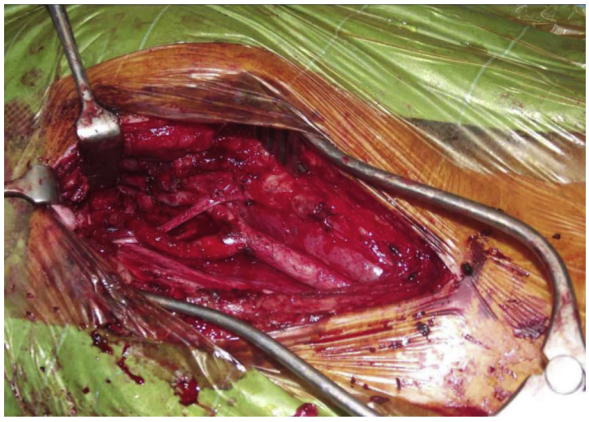

Fig 3. The internal carotid artery (ICA) was repaired primarily using an end-to-end spatulated anastomosis.

She was discharged 6 days postoperatively. The patient was reviewed in the outpatient clinic 3 months later and had made an excellent recovery. However, she was noted to have persistent mild dysphagia, which probably reflects glossopharyngeal nerve neurapraxia.

The histologic evaluation showed a well-circumscribed richly vascular tumor with a thin to focally indistinct fibrous capsule composed of tumor cells showing a medium to large polygonal cell morphology with arrangement in distinct nests. Mild nuclear pleomorphism was present, and a single mitotic figure was noted. There were no overt features suggestive of malignancy and no lymphovascular invasion. The morphology was consistent with a CBT.

DISCUSSION

Inci and Betan[1] reported a patient with a CBT associated with an intracranial aneurysm; however, to our knowledge, this is the first reported case of simultaneous CBT and an extracranial ICA aneurysm. The common management of these two separate entities is surgical intervention. There are no current guidelines or expert consensus on the size of carotid aneurysms to be treated; however, any symptomatic aneurysm needs to be managed promptly. We have outlined above our surgical management of this unique case. Below we discuss the two pathologies as separate entities and then discuss a possible etiologic link between them.

CBTs. CBT, a rare neoplasm with an incidence of <1:30,000, arises from paraganglion cells of the carotid body.[2] Approximately 5% to 10% of CBTs progress to malignancy.[3] CBTs are subclassified as sporadic, familial, or hyperplastic. Chronic hypoxic conditions, such as in chronic obstructive pulmonary disease (as in this patient), can overburden the carotid bodies and subsequently lead to hypertrophy, hyperplasia, and neoplasia.[4]

The diagnosis is generally made through clinical findings and various imaging modalities. Duplex ultrasound imaging is the first-line imaging modality in diagnosing a CBT, although its ability to provide sufficient information to plan management is limited. Angiography has been the gold standard for the diagnosis and management of CBTs for many years. However, a combination of magnetic resonance imaging and magnetic resonance angiography provides significant diagnostic and surgical planning

information, making the use of angiography primarily for preoperative embolization or stenting.

CBT resection is considered the primary treatment option and the only one that allows a definitive cure.[2] Surgical resection of CBTs is challenging because they are adherent to the carotid adventitia, highly vascularized, often involve the cranial nerves, and are in a limited field of exposure.[2,5] Sajid et al[6] demonstrated that CBT resection is associated with cranial nerve injury (18%), stroke (1%), death (1%), and bleeding (8%).

Carotid artery aneurysms. Carotid artery aneurysms have various etiologies and represent a taxing pathology with limited expertise and familiarity. The most common cause of extracranial carotid artery aneurysms is atherosclerosis. These aneurysms tend to be fusiform and are almost always associated with arterial hypertension.Extracranial carotid aneurysm repair comprises 0.1% to 2% of all carotid procedures.[7,8] The high incidence of cranial nerve compression and cerebrovascular events through embolization in untreated patients (68%) justifies invasive management for both symptomatic and asymptomatic carotid aneurysms.[9] Hemorrhage has also been seen as a complication of these aneurysms, although rupture remains uncommon.[10] The surgical techniques applied to the management of extracranial carotid aneurysms are ligation (or angiographic occlusion), endoaneurysmorrhaphy, resection with primary anastomosis, resection with graft replacement, or stent grafting. In the era of endovascular surgery, techniques used for occlusive disease are extrapolated as minimally invasive alternatives to conventional surgical treatment of extracranial carotid artery aneurysms. The combined stroke and mortality rate of surgical reconstruction reported by the Texas Heart Institute was 9%.[11] They described a partial aneurysmectomy and patch closure of a large fusiform aneurysm involving the carotid bifurcation. This approach avoids extensive dissection of the posterior wall of the aneurysm, reducing the rate of cranial nerve dysfunction to 6%.[12] McCann et al[13] showed a stroke risk of 25% and a mortality rate of 20% in the follow-up of ICA ligation. A meta-analysis by Roset et al[14] revealed a 1.2% mortality risk and a 6% stroke risk.

Long-term results for carotid aneurysm repair using stent grafts have not been established. Zhou et al[12] reported a decrease in the 30-day stroke/death rate from 14% to 5% during a 19-year period with the introduction of endovascular techniques. Endovascular interventions reduce the risk of cranial nerve injury, can be done without general anesthesia, are useful for distal lesions where surgical exposure is difficult, are associated with shorter hospital lengths of stay, and have lower morbidity and mortality rates.[12]

Etiologic link. Whether there is a pathophysiologic association between the CBT and ICA aneurysm remains unknown. CBTs have been shown to cause hypertension, and carotid artery aneurysms are almost always associated with hypertension. We agree with Inci and Betan,[1] who postulated a CBT-induced hypertensive crises could be the cause of aneurysm formation.

CONCLUSIONS

This is the first reported patient with a simultaneous CBT and extracranial ICA aneurysm. The two entities separately pose significant surgical challenges, which are exponentially increased when presenting together. We have reported our successful surgical management of this rarity.

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