CASE REPORT

Tandem symptomatic internal carotid artery and persistent hypoglossal artery stenosis treated by endovascular stenting and flow reversal

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SUMMARY

Persistence of the hypoglossal artery into adulthood is a rare vascular anomaly and, when present, provides the predominant vascular supply to the posterior circulation. We describe a case of vertebrobasilar insufficiency associated with severe high-grade stenosis of the persistent hypoglossal artery and tandem stenosis of the proximal ipsilateral internal carotid artery, treated by an endovascular approach. The unique anatomical and technical challenges associated with this case are reviewed in detail.

BACKGROUND

A persistent hypoglossal artery (PHA) is the second most common carotid–basilar anastomosis in adults after the persistent trigeminal artery, failing to obliterate after development of the fetal craniocerebral circulation.¹ It is the only such vascular anomaly that can complicate the performance of carotid endarterectomy, and its intraoperative recognition is fundamental for a successful surgical intervention of the cervical internal carotid artery (ICA).² A PHA can also be a route for embolic showering of the posterior circulation in cases of carotid atherosclerotic disease, therefore mimicking cardioembolism.³ In this paper we describe the unique case of a patient presenting with vertebrobasilar ischemic symptoms associated with severe tandem stenosis of both the ICA and the PHA on the right side, treated endovascularly using a flow-reversal technique and stent placement.

CASE PRESENTATION

A patient aged mid-60s presented with numbness and tingling on the right side of the tongue, lip and fingers as well as dizziness, headaches, nausea and vomiting.

INVESTIGATIONS

A CT scan of the head and CT perfusion images showed no obvious strokes or perfusion deficits (figure 1). The CT perfusion scan was obtained using a 320-slice CT scanner which provides excellent coverage of the posterior circulation and, although it may be difficult to appreciate small or lacunar strokes, any sizeable ischemic area that may increase the risk of intervention is readily evident. An MRI scan—the study of choice to exclude stroke—was not possible given the patient's large body habitus. A CT angiogram of the head and

neck was significant for an occluded or atretic right vertebral artery at its origin, a small left vertebral artery ending in the posterior inferior cerebellar artery and an anomalous vessel arising from the distal right cervical ICA and entering the skull base through the hypoglossal canal, giving rise to the distal right vertebral artery (figure 2). This vessel then continued into a basilar artery of normal caliber, being the only vascular supply to the posterior circulation due to the presence of bilateral fetal posterior cerebral arteries. A digital subtraction angiogram (figure 3A) confirmed that this anomalous vessel was consistent with a PHA, with stenosis of approximately 80% in severity in its mid-segment (figure 3B). In addition, the proximal cervical portion of the right ICA had an atherosclerotic plaque causing a tandem 50% stenosis. Cardioembolic evaluation was negative for other possible embolic sources to explain the patient's symptomatology.

TREATMENT PLAN

The patient was considered a high surgical risk because of medical comorbidities, the presence of 80% symptomatic stenosis at the PHA, the presence of a high cervical lesion (PHA stenosis located at the C1–2 level), the presence of tandem stenosis at the proximal aspect of the ICA and anatomic difficulties associated with PHA during the performance of a carotid endarterectomy. In addition, it was unclear whether one or both lesions were symptomatic and to what extent. On the basis of the degree of stenosis, we were concerned about the PHA; however, the large proximal ICA atherosclerotic plaque could not be ignored. We therefore decided to proceed with stent placement in both the proximal ICA and the PHA during the same treatment setting. The high degree of stenosis in the PHA and the presence of a large atherosclerotic plaque in the proximal ICA were felt to be good indications for proximal embolic protection by a flow-reversal technique using the GORE Flow-Reversal System (W L Gore and Associates, Flagstaff, Arizona, USA) rather than distal embolic protection.

TREATMENT

The patient received a loading dose of aspirin and prasugrel (prasugrel was used instead of clopidogrel because the patient did not develop sufficient platelet inhibition with clopidogrel) and, after

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Figure 1 CT perfusion imaging showing no asymmetry and no evidence of perfusion deficit.

Figure 2 CT angiogram showing an anomalous vessel traveling through the hypoglossal canal with evidence of calcification and stenosis as it traverses the skull base. This is consistent with the diagnosis of persistent hypoglossal artery.

confirmation of therapeutic platelet responses to both agents, was brought into the angiography suite. Moderate sedation was established using midazolam and fentanyl. Access was obtained via the right femoral artery and left femoral vein for the extracorporeal connection allowing flow reversal through the GORE filter device. The patient received a sufficient quantity of heparin to achieve an activated coagulation time of between 250 and 300 s.

Using roadmap guidance, a 9 Fr GORE guiding catheter was advanced over a Slip-Cath (Cook Medical, Bloomington, Indiana, USA) and 0.035 inch exchange wire into the proximal right cervical ICA, proximal to the ICA stenosis. The proximal ICA was chosen because the atherosclerotic plaque was in the mid-cervical segment of the ICA and placing the guide distal to the external carotid artery (ECA) take-off obviated the need to worry about ECA backflow. The distal balloon was then advanced into the ICA, distal to the take-off of the PHA (instead of the ECA because we were concerned about retrograde flow from the ICA releasing embolic debris into the posterior circulation). At this point, both the proximal balloon on the guide in the ICA and the distal balloon in the ICA were inflated (figure 4) and the flow-reversal system was applied with blood being shunted back from the PHA into the cervical ICA and then into the left femoral vein after passing through the filter. A 0.014 inch Spartacore wire (Abbott Vascular, Abbott Park, Illinois, USA) was advanced across the ICA stenosis and then into the PHA, across the stenosis essentially into the proximal vertebral artery. A 6 mm×22 mm self-expanding closed-cell biliary Wallstent (Boston Scientific, Natick, Massachusetts, USA) was then brought over the wire and deployed across the PHA

Figure 3 (A) Digital subtraction angiogram (lateral projection) showing the persistent hypoglossal artery (PHA) communicating the right internal carotid artery (ICA) with the basilar artery. Notice the high-degree stenosis at the PHA and the atherosclerotic plaque with stenosis of the proximal ICA. (B) The same angiographic view showing stenosis of the PHA measuring approximately 80%.

stenosis, covering the lesion successfully. Post-stenting balloon angioplasty was carried out using a 4 mm×20 mm Aviator balloon (Cordis, Warren, New Jersey, USA). A second Wallstent (10 mm×24 mm) was then similarly brought across the ICA stenosis and deployed over the proximal ICA stenosis, also covering this lesion successfully (figure 5). Finally, an intravascular ultrasound probe (IVUS, Volcano Corporation, San Diego, California, USA) was used to confirm no evidence of residual intrastent plaque and/or dissection in either lesion. Aspiration was performed actively for any residual debris. The balloons were then deflated and a final angiographic run confirmed good revascularization without any evidence of dissection, significant residual stenosis or distal emboli (figure 6A,B). The GORE catheter was removed and the arteriotomy was closed using an Angioseal 8 Fr closure device (St Jude Medical, St Paul, Minnesota, USA).

OUTCOME AND FOLLOW-UP

Postoperatively the patient remained with premonitory minimal subjective right-sided numbness (National Institutes of Health Stroke Scale (NIHSS) score 1) but the remaining symptoms resolved. The patient was discharged home on aspirin (325 mg daily), prasugrel (5 mg daily) and rosuvastatin (20 mg daily), in addition to the previous regimen of antihypertensive and oral hypoglycemic medications. There were no complications related to the endovascular procedure. All symptoms had resolved by the 30-day clinic follow-up and this was confirmed at the 6-month clinic follow-up.

DISCUSSION

PHA is the second most common carotid–basilar anastomosis after persistent trigeminal artery, occurring in approximately 0.025% of patients.⁴ The PHA connects the primordial carotid artery with the longitudinal neural arteries which later form the basilar artery. The PHA leaves the ICA as an extracranial branch in the cervical region, enters the skull through the hypoglossal canal and joins the caudal portion of the basilar or distal vertebral artery. PHA is associated with an increased incidence of ischemic strokes, intracranial aneurysms and arteriovenous malformations.⁵ In extremely rare cases a PHA may originate from the ECA.⁶ Because this anomaly is also associated with hypoplasia of the vertebral and posterior communicating arteries⁷—making it the predominant provider of blood flow to the posterior circulation—the management of vascular pathologies affecting this vessel, such as aneurysms or atherosclerotic disease, is fraught with an increased risk of complications and technical difficulties.

We describe the case of a patient who presented with clear symptoms of vertebrobasilar insufficiency including dizziness, hemisensory loss and nausea and vomiting. The patient was found to have severe high-grade (approximately 80%) stenosis of the PHA, which was the only effective afferent vessel supplying the basilar trunk. To complicate the situation further, the right ICA proximal to the take-off of the PHA also demonstrated atherosclerotic disease and a large plaque obstructing access to the PHA. The patient's symptoms could be attributed to hemodynamic insufficiency from the high-grade stenosis at the PHA, embolic phenomenon from the proximal ICA plaque, or both, requiring that both lesions be treated.

Figure 4 Roadmap guidance angiogram demonstrating the proximal (in the proximal internal carotid artery (ICA)) and distal (in the distal ICA) balloons of the GORE Flow-Reversal System. Inflation of the balloons creates anterograde flow arrest and, once the extracorporeal connection to the contralateral femoral vein is established, allows flow reversal through the persistent hypoglossal artery, essentially preventing any embolic debris from moving distally into the cerebral circulation.

Carotid endarterectomy in the setting of persistent carotid-basilar anastomosis is often challenging and the surgeon must be aware of potential technical difficulties and inherent pitfalls.⁸ Moreover, surgical repair of the PHA would be fraught with significant risk, considering this is the only vessel supplying the entire posterior circulation and is typically located in the upper cervical segments of the ICA around the C1 vertebral level. Conversely, endovascular treatment may offer a simpler, faster and better alternative in the management of such cases. In our case, the stenosis in the PHA was found at the C1–2 level, which is considered a high-risk anatomic criterion for carotid endarterectomy according to Medicare treatment guidelines and therefore constitutes an indication for endovascular treatment. Because of the high-degree symptomatic stenosis in the PHA associated with tandem stenosis and atheromatous disease in the proximal ICA, we felt that proximal embolic protection was necessary to minimize the risk of embolic complications during the endovascular procedure. A GORE flow-reversal system was therefore chosen as the optimal embolic protection technique to be used during endovascular stent deployment and revascularization for both lesions.

The GORE Flow-Reversal System allows blood flow reversal through the stenotic segment, preventing released emboli from moving distally towards the brain parenchyma. It consists of a guiding catheter with two balloons, one larger balloon positioned proximally to the stenotic segment and another smaller balloon usually placed in the ECA to prevent backflow, similar

Figure 5 The persistent hypoglossal artery and proximal internal carotid artery Wallstents have been deployed with excellent resolution of the vessel stenosis.

to the temporary occlusion clamps applied during endarterectomy. Once the balloons are inflated, the guiding catheter is connected extracorporeally to a venous sheath placed in the contralateral femoral vein through a filter to collect embolic debris and return intracranial reversed blood flow into the venous circulation. The GORE device was recently tested in the Embolic Protection with Reverse Flow Clinical Study, which revealed one of the lowest stroke and death rates among the various carotid stenting trials and therefore confirmed the safety and effectiveness of the device.⁹ In the present case, the proximal balloon was placed in the ICA proximal to the ICA plaque and the distal balloon in the distal ICA, distal to the take-off of the PHA (figure 4). The flow was then reversed through the PHA stenosis, allowing us to safely deploy a stent through it, perform a post-stent angioplasty, and finally to deploy a second stent through the ICA atherosclerotic plaque (figure 5). The flow-reversal system carries some risk of distal ischemia in this situation, given the unique circulatory pattern associated with the PHA—that is, only one vessel supplying the posterior circulation. On the other hand, the presence of symptomatic high-grade stenosis, such as that encountered in the PHA, constitutes the premier indication for proximal embolic protection strategies. The alternative option of distal embolic protection, even if placing two separate filters in the distal ICA and PHA (distal to the stenotic lesion), would be worse because of the need to cross a high-grade stenosis lesion with a filter device with a high risk of devastating distal embolism to the posterior circulation vascular territory. Further, the limited distal segment of the PHA beyond the stenosis would restrict the ability to safely deploy a distal filter, which essentially would be placed into the intracranial segment of the PHA into the vertebral artery. To minimize risk to the posterior circulation, the GORE device balloons were inflated and the flow reversed for a very limited period of time during which the stents were deployed in the

Figure 6 Final intracranial angiographic run, anteroposterior (A) and lateral (B) projections, demonstrating good filling of the intracranial circulation after successful tandem stenting. The territories of the carotid and basilar arteries are supplied by the right internal carotid artery through the persistent hypoglossal artery.

PHA and ICA as described above. Final angiographic runs demonstrated an optimal anatomic result with no evidence of distal emboli (figure 6A,B). Furthermore, the patient had an excellent clinical result, remaining with premorbid subjective right-sided numbness (NIHSS score 1) but no new deficits and resolution of nausea, headaches and dizziness. All symptoms had resolved at the 6-month follow-up.

To our knowledge, this is the first description of tandem stenosis of the ICA and a PHA treated endovascularly by stenting and a flow-reversal technique. This case represents an example of safe and effective application of endovascular techniques to treat complex extracranial atherosclerotic disease.

Learning points

- Persistent hypoglossal artery (PHA) is the second most common carotid–basilar anastomosis and its presence can complicate the performance of carotid endarterectomy.
- Endovascular techniques can be used as a safe and effective strategy to treat internal carotid artery (ICA) and PHA stenosis.
- Proximal protection with flow reversal is an ideal strategy to perform ICA and/or PHA stenting safely.

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