

A successful treatment with carotid arterial stenting for symptomatic severe internal carotid artery stenosis with ipsilateral persistent primitive hypoglossal artery and aplasia of the A1 segment of anterior cerebral artery: a case report

İpsilateral persistan primitif hipoglossal arter ve ön serebral arterin A1 segmentinde aplazi ile birliktelik gösteren ciddi internal karotis arter darlığının karotis arteriyel stentleme ile başarılı tedavisi: Olgu sunumu

Furuzan Numan,¹ Fatih Gülşen,¹ Oğuz Yılmaz,² Murat Cantaşdemir,¹ Serdar Solak,¹ Bingür Sönmez²

¹Department of Interventional Radiology, Medical Faculty of Cerrahpaşa University, İstanbul, Turkey

²Department of Cardiovascular Surgery, Memorial Hospital, İstanbul, Turkey

We report a 85-year-old male case with persistent primitive hypoglossal artery (PPHA) who experienced vertigo and syncope attacks and had cervical internal carotid artery (ICA) stenosis at the same side. A carotid artery stenting (CAS) procedure was performed without embolic protection devices. The patient's postoperative clinical course was good. Postoperative magnetic resonance imaging showed no ischemic complications. On fifth day of surgery, the patient was discharged with no neurological symptoms. We report, to the best of our knowledge, the second case of ICA stenosis with ipsilateral carotid-basilar anastomosis treated with CAS presented in the literature. Although persistent carotid-vertebrobasilar anastomoses are very rare; awareness of the presence of this rare variation is essential for interventional radiologists, vascular and cardiovascular surgeons and neurosurgeons to avoid iatrogenic injury causing neurologic deficits, and even death. For the treatment of neurovascular pathologies, variations and anomalies which can affect treatment options or modify the procedural technique should be also considered.

Key words: Aplasia of the A1 segment of anterior cerebral artery; carotid artery stenting; persistent primitive hypoglossal artery; symptomatic severe internal carotid artery stenosis.

Cases of the vertebral artery originating from the carotid arterial system that reflect the persistence of primitive carotid-vertebrobasilar anastomoses are very

Vertigo ve senkop nöbetleri geçiren ve aynı tarafta servikal internal karotis arter (İKA) darlığı olan 85 yaşında bir erkek persistan primitif hipoglossal arter (PPHA) olgusu sunuldu. Embolik koruma cihazları olmaksızın karotis arter stentleme (KAS) işlemi yapıldı. Hastanın ameliyat sonrası klinik seyri iyiydi. Ameliyat sonrası manyetik rezonans görüntüleme de iskemik komplikasyon izlenmedi. Ameliyatın beşinci gününde, nörolojik semptomu olmayan hasta taburcu edildi. Bilgimiz dahilinde bu olgu, literatürdeki KAS ile tedavi edilen ve ipsilateral karotis-baziler anastomozlu ikinci İKA darlığı olgusudur. Persistan karotis-vertebrobaziler anastomozları çok nadir görülmekle birlikte, nörolojik defisitlere ve hatta ölüme yol açan iyatrojenik hasarı önlemek için iyatrojenik girişimsel radyologlar, vasküler ve kardiyovasküler cerrahlar ve nöroşirürji uzmanlarının bu nadir varyasyonun varlığını göz önünde bulundurması gereklidir. Nörovasküler patolojilerin tedavisinde tedavi seçeneklerini etkileyebilen veya işlem tekniğini değiştirebilen varyasyon ve anomaliler de göz önünde bulundurulmalıdır.

Anahtar sözcükler: Ön serebral arterin A1 segmentinin aplazisi; karotis arter stentleme; persistan primitif hipoglossal arter; semptomatik şiddetli internal karotis arter stenozu.

rare. The primitive hypoglossal artery is an important anastomosis between the internal carotid artery (ICA) and the vertebrobasilar system in human embryos which



Available online at
www.tgkdc.dergisi.org
doi: 10.5606/tgkdc.dergisi.2013.5162
QR (Quick Response) Code

Received: December 24, 2010 Accepted: April 17, 2011

Correspondence: Fatih Gülşen, M.D. İstanbul Üniversitesi Cerrahpaşa Tıp Fakültesi Girişimsel Radyoloji Anabilim Dalı, 34089 Fatih, İstanbul, Turkey.

Tel: +90 532 - 330 35 24 e-mail: drfgulsen@yahoo.com

usually disappears before birth. When it survives after birth, it is called the persistent primitive hypoglossal artery (PPHA). It is usually observed incidentally while performing carotid angiography, and has an estimated incidence rate of 0.05%.^[1-4]

The presence of the PPHA has clinical implications. Previous reports have suggested that it may rarely cause multi-territorial cerebral infarction when complicated by stenosis of the ipsilateral ICA.^[1-10] Herein, we report the treatment of a patient with PPHA who experienced vertigo and syncope attacks and had cervical ICA stenosis on the same side.

CASE REPORT

An 85-year-old male had complained of severe vertigo and syncope for four months before admission to our hospital. His past medical history revealed a right carotid endarterectomy (RCEA) and coronary artery bypass graft (CABG) surgery. Neurological and physical examinations on admission showed no abnormalities, and his family history was unremarkable. During his hospitalization, a cerebral angiographic examination was performed using the Seldinger technique for the percutaneous puncture of the right femoral artery and selective catheterization of the right and left carotid artery and both vertebral arteries. This revealed severe left ICA stenosis and divergence of the PPHA immediately distal

to the stenosis (Figure 1a, b). The bilateral vertebral arteries exhibited hypoplasia (Figure 2a), and the A1 segment of the left anterior cerebral artery (LACA) also exhibited aplasia (Figure 2b). The posterior communicating arteries could not be visualized. The anterior intracranial circulation and vertebrobasilar system were mainly opacified via the left stenotic ICA (Figure 2b). Furthermore, no incidents of aneurysms or arteriovenous malformation were seen.

A carotid artery stenting (CAS) procedure was performed under local anesthesia without embolic protection devices. A guiding catheter (8F) was placed in the left common carotid artery (LCCA). Over an exchange guidewire, a Precise[®] self-expanding stent (Cordis Corporation, Hialeah, Florida, USA) measuring 8.0x40 mm was successfully deployed in the stenotic segment of the left cervical ICA. Postdilatation was performed due to residual stenosis using an Aviator[™] Plus 6.0x20 mm diameter balloon (Cordis Corporation, Hialeah, Florida, USA). An angiographic examination immediately after the CAS showed good apposition of the stent to the arterial wall with restoration of the luminal diameter (Figure 3). The patient's postoperative clinical course was good, and postoperative magnetic resonance imaging (MRI) showed no ischemic complications. Five days after surgery, the patient was discharged with no neurological

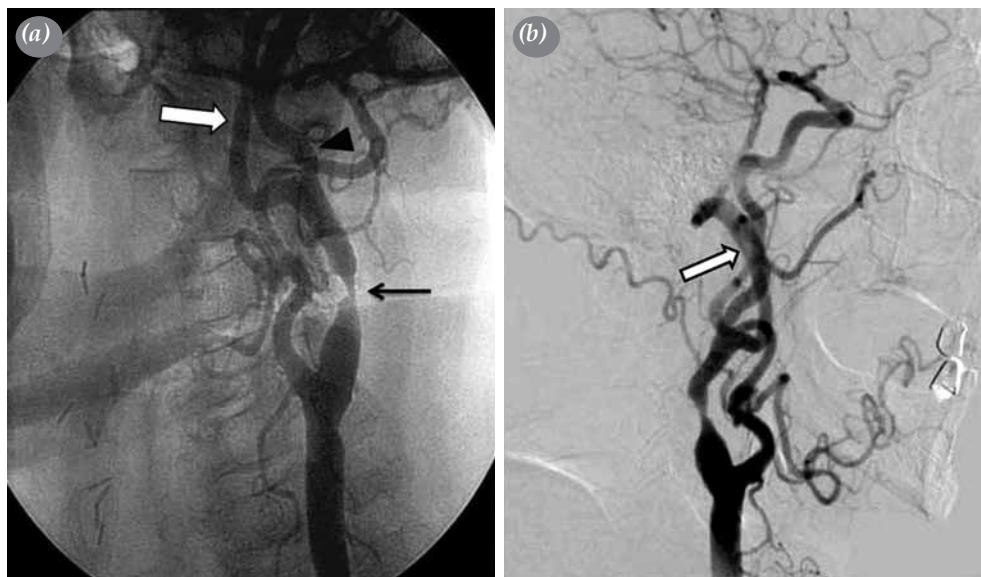


Figure 1. (a) Live image of preoperative left carotid angiography. Cerebral angiography showed severe left ICA stenosis (black arrow) and divergence of the persistent primitive hypoglossal artery immediately distal to the stenosis (arrowhead). The white arrow shows the distal internal carotid artery after the divergence. (b) Lateral view of intra-arterial digital subtraction angiography showing the persistent primitive hypoglossal artery (arrow) connecting the left internal carotid artery with the basilar artery.

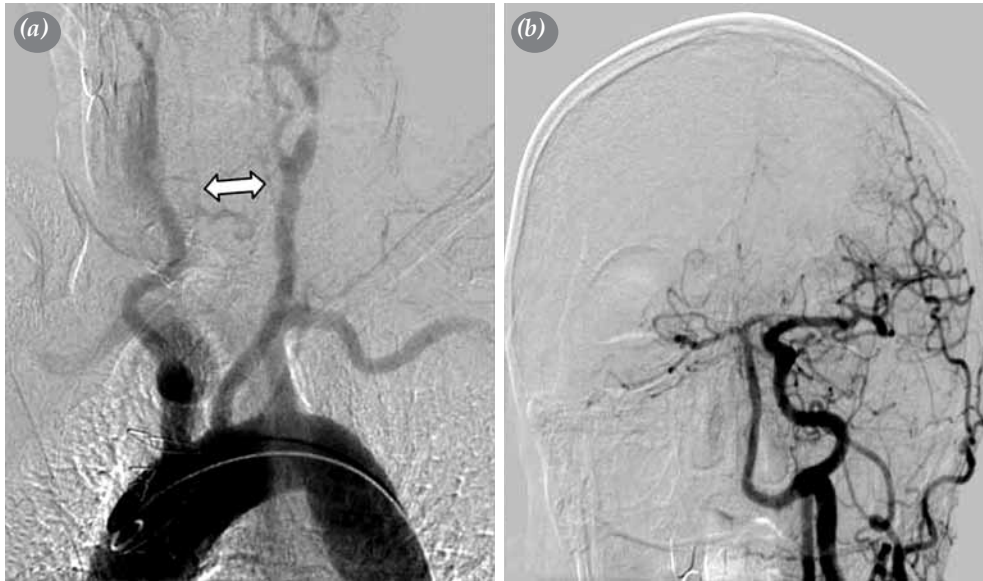


Figure 2. (a) Aortogram demonstrating hypoplasia of the bilateral vertebral arteries (arrow). (b) Anterior view of intra-arterial digital subtraction angiography showing that the left middle cerebral artery and vertebrobasilar system were mainly opacified via the stenotic internal carotid artery. The A1 segment of the left anterior cerebral artery was aplastic.

symptoms, and one year later, a follow-up angiography showed no significant restenosis of the lesion.

DISCUSSION

The PPHA represents a rare carotid-basilar anastomosis, with a reported incidence of between 0.05 and 0.02% on cerebral angiography.^[1-7] It is the second most frequently seen anastomosis after the persistent trigeminal artery,

which makes up the vast majority of persistent primitive connections. Anastomosis of the persistent otic and proatlantal intersegmental arteries are observed less often.

The PPHA is related to alterations to the anatomy of cerebral circulation. Its presence may be completely asymptomatic and can appear as an incidental finding in a cerebral angiogram. The criteria for the diagnosis

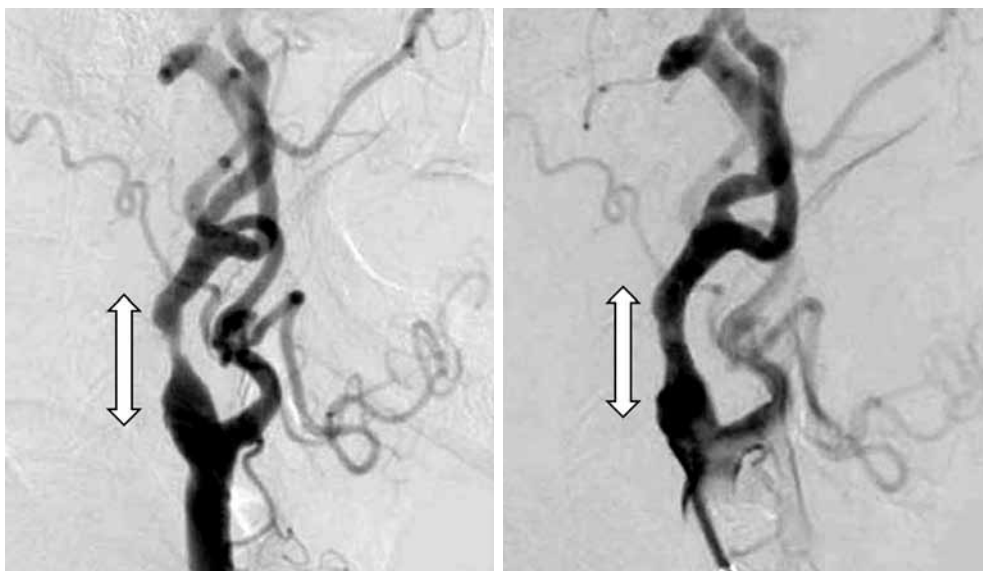


Figure 3. Subtraction images of the preoperative and postoperative left carotid angiogram. Stenosis was restored, and the stent was placed between the internal carotid artery and the divergence.

of PPHA are the following: (i) the artery originates from the cervical ICA at anywhere from the first to the third cervical vertebrae, (ii) the artery passes through the hypoglossal canal to the posterior cranial fossa, (iii) only the part of the basilar artery peripheral to the anastomosis is visualized by angiography, and (iv) angiography indicates deficiency or absence of the posterior communicating artery.^[1,5-8,10] The second criterion is important for differentiating the PPHA from the proatlantal intersegmental artery.^[7] Our case satisfied all of the above criteria.

The vertebral arteries in patients with carotid-basilar anastomosis are usually either hypoplastic or, as seen in our case, aplastic. The clinical manifestations of patients with PPHA can be classified into three groups. The first group is comprised of patients that show no signs or symptoms, with the PPHA being incidentally detected by angiography. The second group is made up of patients with symptoms caused by associated intracranial vascular lesions, such as cerebral aneurysms and arteriovenous malformation, or a brain tumor. Finally, the third group is composed of patients with symptoms caused by ischemia in posterior circulation.^[3,7]

Patients with a PPHA may have various associated lesions. Like other persistent primitive arteries, the main coexisting lesions are cerebral aneurysms, arteriovenous malformation, and atherosclerotic cerebrovascular disease.^[1-3,5,7,8] The latter may present as a plaque extending from the distal carotid artery into the ICA or as an isolated stenosis near the origin of the PPHA. The group of persistent primitive arteries, which includes the PPHA, has been implicated in cases of moyamoya and quasi-moyamoya disease as well,^[3,11] and the high incidence rates may be explained by the contribution of congenital or hemodynamic factors.

In the majority of cases, a PPHA is found incidentally; however, in cases of considerably decreased carotid blood flow, both the anterior and posterior vascular territories can be under ischemia simultaneously. Persistent anomalous carotid-basilar anastomosis should also be recognized as a potential pathway for an artery-to-artery embolism in patients with atherosclerotic stenosis of the proximal ICA.^[2,4-8]

We planned our treatment by considering that aggravation of the stenosis would be catastrophic. Treatment options were CEA and CAS.^[6-10] The CAS procedure was preferred in our case over CEA due to the observed higher position of the lesion and the intraoperative difficulties associated with an endarterectomy, including the prevention of ischemia both in the unilateral hemisphere and the whole

vertebrobasilar system caused by blocking the blood supply from the carotid artery.

To the best of our knowledge, this is only the second case of ICA stenosis with ipsilateral carotid-basilar anastomosis treated with CAS that has been presented in the literature. The first case was published by Kanazawa et al.,^[10] and in that case, the internal CAS procedure was performed via the modified Parodi method. However, in our case, aplasia was observed in the A1 segment of left ACA, and the posterior communicating arteries could not be visualized. Therefore, we did not utilize the modified Parodi method so as to prevent ischemia in the anterior or posterior intracranial circulation that may occur if flow reversal is not achieved during balloon occlusion. Furthermore, considering the presence of variations originating in the complex anatomic structure and because any complications may have catastrophic implications, we performed our procedure without the use of any distal embolic protection device. Besides, the simultaneous use of two embolic filters for the PPHA and the carotid system would not be feasible. The patient was under antilipidemic treatment with both plavix and statins, and the plaque formation was of a calcified and non-vulnerable type.

During the stenting procedure, we used an under-sized stent, following the measurement of the ICA lumen, to avoid plaque fracture and to reduce the potential risk of emboli. Although persistent carotid-vertebrobasilar anastomoses are very rare, awareness of the presence of this rare variation is salient for interventional radiologists, vascular and cardiovascular surgeons, and neurosurgeons in order to avoid iatrogenic injury that can cause neurological deficits or even death. In addition, aggravation of severe ICA stenosis with the ipsilateral primitive artery may result in a fatal outcome due to the high degree of hemodynamic insufficiency derived from immature collateral circulation. In addition, interventional neuroradiological procedures may require modification, such as stenting without the use of a filter, if these anomalies exist.

Today, there are two options for the treatment of cervical ICA stenosis: the surgical approach and the interventional approach. Although the operative indication for patients should be decided on a case-by-case basis, the interventional option should be considered for future patients whenever possible.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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