A Rare Cause of Juvenile Stroke: Extracranial Carotid Artery Aneurysm with Venous Complete Reconstruction of the Carotid Bifurcation

Carotid Bifurcation Reconstruction of ECAA Using the Greater Saphenous Vein
Extracranial carotid artery aneurysms (ECAA) are a rare cause of embolic stroke. The underlying etiology is variable, with atherosclerosis being the most common entity in older subjects. Several treatments have been developed over the last 20 years, but the preferred method remains unknown. Notwithstanding the widespread use of endovascular techniques, surgical reconstruction by means of a bifurcated venous bypass graft should be applied in younger patients. In this way, it is possible to avoid major concerns about the development of long-term intrastent restenosis, and also to spare the external carotid artery which represents the main branch for the ipsilateral cerebral and facial perfusion. We propose ECAA resection and interposition of the inverted great saphenous vein to both the internal and external carotid...
artery by means the use of a tributary, i.e., the Giacomini vein.

**Abstract**

When feasible, after the avulsion of the extracranial carotid aneurysm, the revascularization of both the internal and external carotid arteries should be mandatory in younger patients.

**Key Messages**

**Body**

**Introduction**

The incidence of extracranial carotid artery aneurysms (ECAA) is ranges between 0.09 and 2.0% of all carotid surgical procedures [1, 2], being even rarer in the younger population. ECAA are associated with a stroke prevalence of 50% and a mortality of 70% under conservative treatment. Literature data suggest that medical treatment may be considered in cases of small, asymptomatic aneurysms [3–6]. In contrast, an active surgical approach, by means of open or endovascular treatment, is generally preferred for symptomatic ECAA [4, 7]. Either way, the best method to treat ECAA has not yet been established [8].

In the case in this study, in view of the unfavorable clinical evolution despite anticoagulant therapy, i.e., the development of a voluminous floating thrombus inside the ECAA,
surgical treatment was performed 10 days after the onset of stroke. Further investigation suggested an underlying collagenopathy whose molecular defect has not been identified so far.

**Case Report**

A 17-year-old boy presented to the emergency department of a local hospital for the sudden onset of focal secondary generalized seizures treated with intravenous (i.v.) lorazepam (4 mg). His past medical history was positive for recurrent pharyngitis with tonsillitis, mitral valve prolapse, kyphosis, and recurrent headache. There was no history of previous head trauma.

**Examination**

General examination revealed a Marfanoid habitus. The blood tests and toxicology results were negative. Due to persistent drowsiness, aphasia, and right hemiparesis, the patient underwent cerebral magnetic resonance imaging which showed a deep left middle cerebral artery stroke (Fig. 1a). Computed tomography angiography disclosed an ECAA of 1.5 cm in diameter located at the origin of the left internal carotid artery (Fig. 1b, c). He was then transferred to the Neurology Department of our hospital and started on low-molecular-weight heparin. A full dose was not administered because of an initial hemorrhagic transformation of the stroke. He then underwent serial Doppler ultrasound monitoring.

On day 9, Doppler ultrasound revealed the presence of a floating thrombus inside the ECAA (Fig. 1d). Consequently, we urgently performed the surgical correction of the ECAA under general anesthesia and electroencephalographic monitoring. Before ECAA exposure, a short segment (8.0 cm in length) of the left great saphenous vein (GSV) was harvested at the inguinal groin. During the exposure, lymphoid tissue hypertrophy was observed in the laterocervical space. Enlarged lymph nodes (>2.0 cm in diameter) were collected and sent to the laboratory for examination. After carefully exposing the carotid bifurcation and ECAA (Fig. 2a), i.v. administration of 2,500 IU of heparin, carotid cross-clamping and aneurysm resection...
were then performed. The restoration of the antegrade flow was obtained by means of interposition of the inverted GSV. In order to complete the reconstruction of the whole bifurcation, the Giacomini vein, a proximal tributary of the GSV, was used to revascularize the external carotid artery (Fig. 2b). Total cross-clamping time was 32 min and no shunting was needed. Running sutures were all performed with a 6-0 monofilament (Prolene, Ethicon, Somerville, USA) under ×3.5 optical magnification.

Postoperative histological examination of the specimen (Fig. 3a) disclosed focal thinning of the tunica media associated with an increased intimal thickness, elastic fiber, fragmentation and myxoid alcyanofilic material deposition (Fig. 3b–d). All 3 layers of the artery wall were involved and no inflammatory infiltrates through the different layers were observed. Histological analysis of the arterial specimen demonstrated the presence of a true aneurysm of the carotid artery. Notwithstanding previous episodes of oropharyngeal inflammation, the lymph node analysis result was negative.

None of the findings were consistent with infectious etiology and this suggested the presence of an underlying connective-tissue disease. The patient underwent genetic testing by next-generation sequencing gene panel sequencing which is available at our institution. The analysis of a panel of 19 genes implicated in collagen pathology (TGFBR3, TGFBR2, FLNB, COL12A1, DSE, COL1A2, TGFBR1, NOTCH1, ACTA2, NTM, COL4A1, COL4A2, FBN1, SMAD3, ABCC6, COL1A1, COL6A1, ATP7A, and FLNA) did not reveal any pathogenic mutations.

The patient was then transferred to rehabilitation, and showed consistent improvements in aphasia and right hemiparesis on antiplatelet therapy (clopidogrel, 75 mg daily). At the 3-month follow-up, a significant improvement in the aphasia was observed but he still presented with weakness of the right arm. Today, 3 years later, the carotid vein graft is still patent on Doppler ultrasound monitoring and the patient has not complained of further symptoms.
Discussion

ECAA in children, adolescents, and young adults is a rare event, and its etiology is currently unclear. Atherosclerosis is the most common entity in elderly people [9], but in younger patients, otorhinolaryngoiatric pathologies have been invoked as initiating events [10--14]. About 10% of subjects with posttraumatic extracranial dissection present with dissecting aneurysms on follow-up [15, 16]. Stab wounds [17], congenital pathologies [18], fibromuscular dysplasia [19], Takayasu arteritis [20], Marfan syndrome [21], neurofibromatosis [22], and early atherosclerosis [23] have also been considered in ECAA pathogenesis.

Currently, little is known about the natural course and pathophysiological mechanisms of ECAA. In a series of 13 patients, treated by open ECAA repair, histology disclosed degeneration with a general loss of elastin fibers in the majority of cases, and dissection with abrupt interruption of the media in the minority [6].

Most patients are usually asymptomatic, and ECAA is often discovered by accident as a beating laterocervical mass with bruits. Pain, dizziness, tinnitus, compression of the cranial nerves, dysphagia, and dyspnea have been also described [1, 11, 17, 23].

The natural history of ECAA is poorly defined and evolution towards stroke is less frequent in young patients [12]. A recently published systematic review confirmed a benign course in dissecting aneurysms [16]. ECAA can also rupture, with massive life-threatening epistaxis or hematemesis [24, 25].

Several treatments have been developed over the last 20 years, but the preferred method to treat ECAA remains unknown. The potential risks involved require surgical correction. Several technical solutions have been proposed, such as ligation, clipping, plication, patch angioplasty, and aneurysm resection with extracranial-intracranial bypass [1, 26] but GSV graft has been advocated as the first choice for treatment because of its long-term patency and reduced risk of infection [27--30]. Endovascular treatment of ECAA with coil embolization and
covered stent are popular nowadays and increasingly used due to their minimally invasive approach [31, 32].

In our case, GSV graft interposition was preferred, considering the presence of the floating thrombus, the young age of the patient and the related life-time risk of the development of intrastent restenosis [33, 34]. Moreover, harvesting the proximal segment of the GSV allows the use of the Giacomini vein for the simultaneous reconstruction of the external carotid artery in order to revascularize the major supply branch of the ipsilateral cerebral perfusion and of the hemiface [35, 36]. ECA collateral circulation reduces, or even cancels, the symptoms of cerebral ischemia [37]. We believe that, when feasible, this fundamental pathway of the cerebral collateral perfusion should always be protected. This issue could be even more important when treating younger patients with a long life ahead of them.

Conclusion

ECAA is a rare cause of ischemic stroke in children and teenagers and the underlying etiology is poorly understood, given the limited availability of pathological studies. A number of conditions, such as local infection, early atherosclerosis, and congenital pathology, have been associated with ECAA in younger patients, with the actual pathogenesis often remaining undefined. We believe that, when feasible, the revascularization of both the internal and external carotid arteries should be mandatory in younger patients.

Disclosure Statement

The authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices described in this article.


Appendix after References (Editorial Comments)

Legend(s)

Fig. 1. a Magnetic resonance imaging. Deep left middle cerebral artery stroke. b, c Computed tomography angiography. External carotid arterial aneurysm of 1.5 cm in diameter located at the origin of the left internal carotid artery. d Doppler ultrasound. Evidence of a floating thrombus in the aneurysm sac.

Fig. 2. Intraoperative imaging. a Exposure of the carotid bifurcation and of the ECAA. b Reconstruction of the carotid bifurcation with interposition of the inverted greater
saphenous vein to the internal carotid artery and of the Giacomini vein to the external carotid artery.

Fig. 3. Anatomopathological examination. a ECAA specimen. b Histology: focal thinning of the tunica media associated with increased intimal thickness. HE. ×20. c Histology: elastic fiber fragmentation. Orcein. ×40. d Histology: myxoid alcyanofilc material deposition. Alcian blue (pH 2.5). ×20.

Supplementary Video on Line:
Doppler Ultrasound of the extracranial carotid artery aneurysms: presence of a floating thrombus inside the carotid aneurysm lumen.