

ICA diaphragms are a rare cause of ischemic stroke of undetermined etiology in young adults

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Background and purposes

Diaphragms of the IC and VA may cause recurrent cerebrovascular accidents of undetermined etiology, especially in young adults. So far, only few cases of ischemic stroke due to diaphragms have been described and different therapeutic (surgical, endovascular or conservative) strategies have been proposed. Usually, the presence of a diaphragm produces turbulences of the blood flow at the level of the stenosis and this can lead to embolic stroke, often confined to a single arterial territory. We present the case of a woman who experienced an ischemic stroke due to a left ICA diaphragm. We also discuss the best pharmacological strategies for secondary stroke prevention in the presence of endovascular diaphragms.

Case report

D.P., 37 y.o., was brought to the Emergency Department complaining of acute onset frontal headache, global aphasia, right supranuclear facial nerve palsy, numbness and weakness of the right upper limb. She had no major cardiovascular risk factors or history of oral contraceptive use. She underwent head CT scan which showed left fronto-temporal hypodensity due to ischemia of the terminal branches of the middle cerebral artery (MCA, *Figure 1*). CT angiography (CTA) of the supra-aortic trunks and intracranial vessels was performed, showing a lack of blood flow at the M1-M2 junction and in the M3 segment of the MCA.

The woman was admitted to Stroke Unit and prophylactic single antiplatelet therapy was begun. MRI of the brain and study of the intracranial vessels (MR angiography, MRA) confirmed left fronto-temporal ischemia with moderate intralesional hemorrhage (*Figure 2*). Usual investigations (ultrasonography of supra-aortic trunks, echocardiography, thrombophilic and vascular screenings) were performed, but none of them yielded relevant results. As a further step, brain and supra-aortic trunk angiography was performed. Invasive study showed a 50% annular narrowing in the left carotid bulb and the origin of the IC artery, with a post-stenotic saccular dilation, due to the presence of an endovascular diaphragm (*Figure 3*). The diaphragm was not located in a straight segment of the artery, therefore a complete exclusion of the dilatation by means of stenting could not be ensured and stent dislocation was feared. For this reason, dual antiplatelet aggregation with aspirin 100 mg and clopidogrel 75 mg (DuoPlavin®) was started.



Figure 1. Head CT scan showing the left fronto-temporal hypodensity.

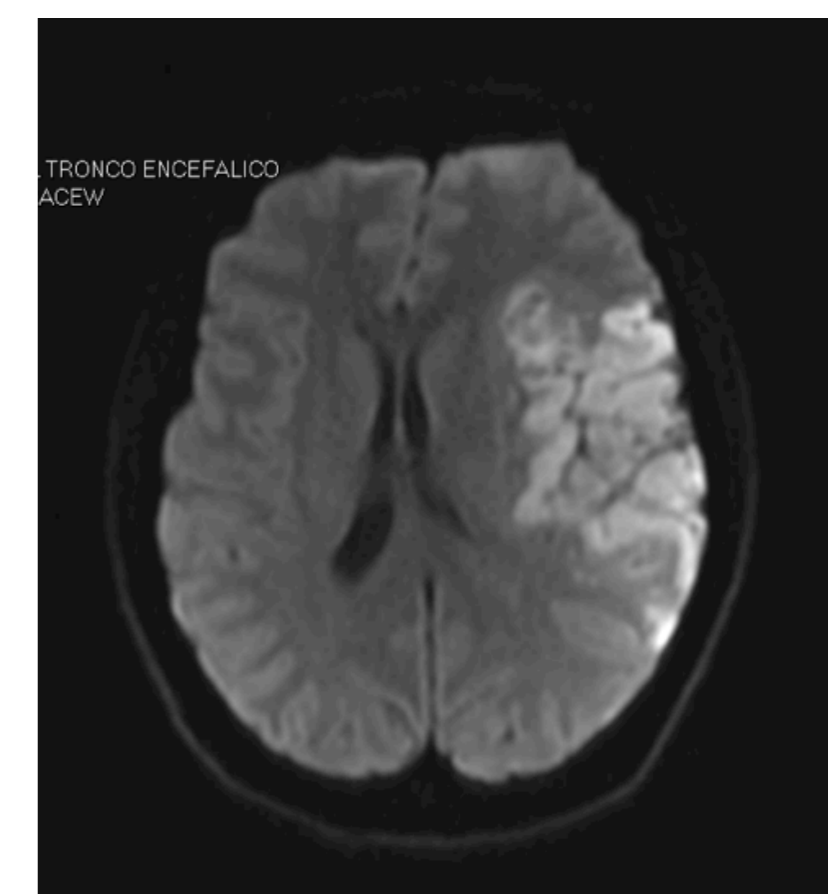


Figure 2. Brain MRI confirmed left fronto-temporal ischemia with signs of moderate intralesional hemorrhage.

Discussion and conclusions

The case described above shows that IC and VA diaphragms may cause stroke of undetermined etiology in young adults. Usual, non invasive cardiovascular investigations may be unhelpful in defining the cause of the ischemic event, as demonstrated by the results of CTA and MRA. Conventional angiography remains an essential diagnostic tool in these conditions. Angiographically, a diaphragm may be defined as a thin translucent endoluminal web that should not disappear after modification of the patient's head position. Pathologically, it is characterized by an abnormal intimal layer which is usually affected by fibrosis, folding, hyperplasia or atherosclerosis.¹

An ICA diaphragm was first reported as a rare cause of ischemic stroke by Ehrenfeld *et al.* in 1967.² In the context of the ICA, diaphragms are more frequently located in the bulb region.³⁻⁴ ICA and VA diaphragms are equally prevalent in men and women with a mean age of stroke of 42. Several therapeutic strategies have been attempted for endovascular diaphragms, and a review of their relative efficacy has been proposed by Lenk *et al.*⁵ When aspirin is used as a monotherapy, there is a high risk of stroke recurrence. On the contrary, stenting, surgery or dual antiplatelet aggregation therapy appear to be more effective for the correct management and the best secondary prevention in these patients.

References

1. So EL, Toole JF, Moody DM, Challa VR. Cerebral embolism from septal fibromuscular dysplasia of the common carotid artery. *Ann Neurol* 1979; 6: 75-78.
2. Ehrenfeld WK, Stoney RJ, Wylie EJ. Fibromuscular hyperplasia of the internal carotid artery. *Arch Surg* 1967; 95: 284-287.
3. Lipchik EO, DeWeese JA, Schenk EA, Lim GH. Diaphragm-like obstructions of the human arterial tree. *Radiology* 1974; 113: 43-46.
4. Morgenlander JC, Goldstein LB. Recurrent transient ischemic attacks and stroke in association with an internal carotid artery web. *Stroke* 1991; 22: 94-98.
5. Lenk S, Labeyrie M-A, Saint-Maurice J-P, Tarlov N and Houdart E. Diaphragms of the carotid and vertebral arteries: an under-diagnosed cause of ischaemic stroke. *European Journal of Neurology* 2014; 21: 586-593.

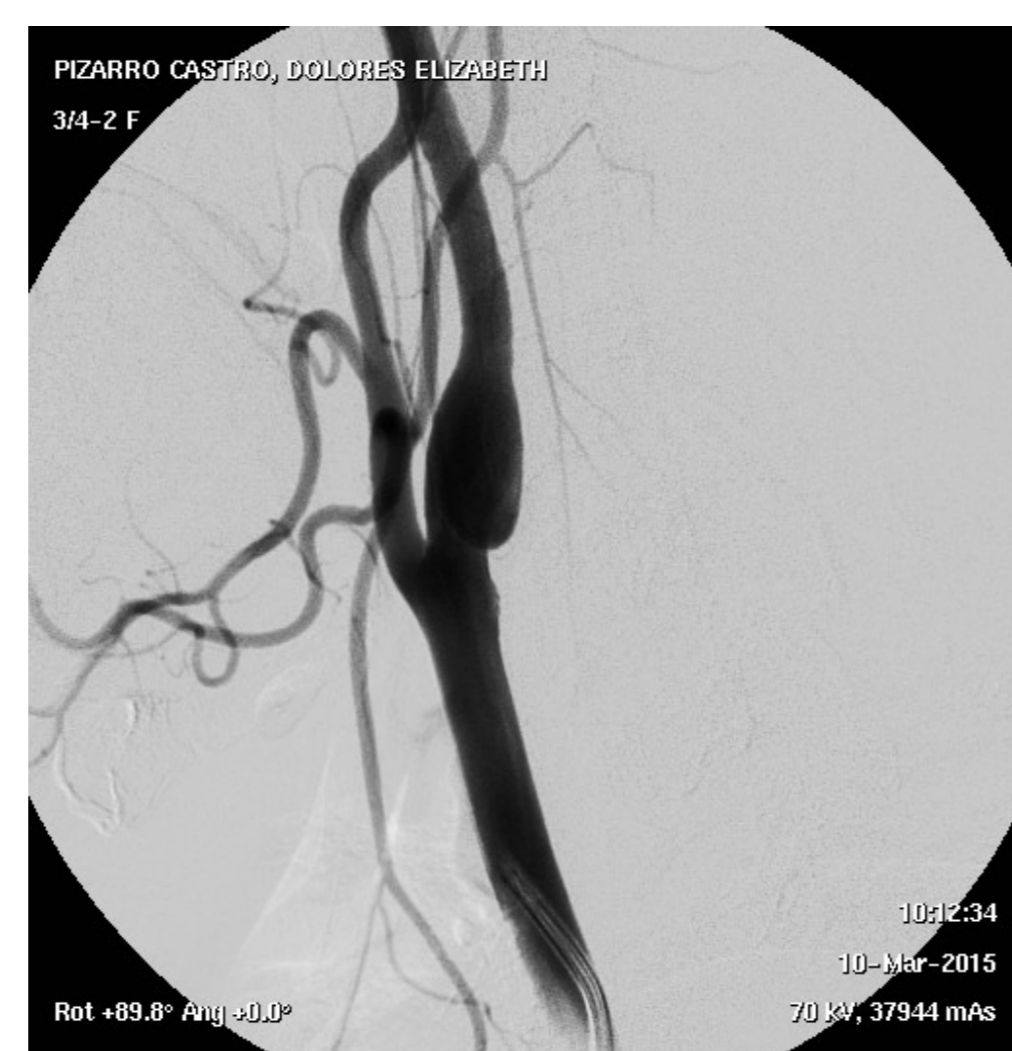


Figure 3. a, b. Angiographic images of the endovascular diaphragm causing a 50% annular narrowing in the left carotid bulb and the origin of the IC artery, with post-stenotic saccular dilation.