BILATERAL DYSGENESIA OF THE INTERNAL CAROTID ARTERIES – CASE REPORT

Cristina Tiu, Alina Poalelungi, Carmen Gavan, Elena Terecoasa

Neurology Department, University Hospital, Bucharest

ABSTRACT

Dysgenesia of the internal carotid artery (ICA) is a rare vascular congenital anomaly with a variety of different grades (agenesia, aplasia and hypoplasia). We report a case of 46 year old Caucasian women with history of asthma, who presented, 1 month after a neck trauma in a car accident, headache and blurred vision in the left eye. Also, she described, intermittent weakness of both hands ("she has been dropping objects"). Neurological examination was normal. Brain MRI showed no ischemic lesion, absence of the left ICA, hypoplastic right ICA. The ultrasonography examination and angiogram revealed agenesis of left ICA, hypoplasia of right ICA, right ACA from a patent anterior communicating artery, aneurysm of right subclavian artery, left vertebral megadolichoartery.

Key words: internal carotid artery, agenesia, aplasia, hypoplasia, megadolichoartery, congenital abnormalities

INTRODUCTION

Developmental anomalies of ICA (agenesia, aplasia and hypoplasia), are rare, and aetiologies are not well known. Most frequently, they are asymptomatic due to collateral formations, but they may also present as cerebrovascular events (1).

CASE REPORT

A 46 year old Caucasian women with history of asthma, presented, 1 month after a neck trauma in a car accident, headache and episodes of blurred vision in the left eye. Also, she described, intermittent weakness of both hands ("she has been dropping objects"). After the minor car accident she complained of headache and she performed a brain CT scan. The radiologist raised the suspicion of a brain tumor due to a compressive lesion isodense with the brain parenchyma with areas of calcification localized in the right cavernous area (Fig. 1). She was sent to an MRI, which showed a T2 hyperintense round mass, with a vascular traject (Fig. 2) and the absence of the flow in the left ICA and a low flow in the right ICA. She was referred to our department with the suspicion of occlusion/high grade stenosis in both carotid arteries, possible due to a dissection (taking into account the neck trauma) and suspicion of brain tumour.

Neurological examination was normal.

Subsequent ultrasonography of the neck arteries (Fig. 3, Fig. 4) revealed no left ICA (agenesia), small calibre of right ICA (hypoplasia, diameter 2.2 mm), right ECA very tortuous, with a coiling and diameter of 3.5 mm, larger than right ICA, left vertebral artery dilated – megadolichoartery (5 mm diameter), left ACA and MCA supplied from the posterior circulation and right ACA from a patent anterior communicating artery. Flow direction in both ophthalmic arteries was anterograde (positive).

We performed an echocardiography to rule out possible associated cardiac anomalies. Transtorachic echocardiography had a normal aspect.

The angiogram revealed agenesia of left ICA (Fig. 5); hypoplasia, tortuous traject of the intrapetrous segment, and a coiling in the cavernous segment of the of right ICA (Fig. 6); right ACA from

Author for correspondence:
Cristina Tiu, Neurology Department, University Hospital, Bucharest

a patent anterior communicating artery; aneurysm (with a diameter of 2cm) of right subclavian artery (Fig. 7); left vertebral megadolichoartery (Fig. 8).

All these vascular anomalies were considered to be incidental and congenital. There was no sign of dissection and no atheromatous lesions.

Due to blurred vision in the left eye with agenesis of left ICA (the ophthalmic artery is supplied from the vertebrobasilar arterial system) we put the patient on Clopidogrel (primary prevention).

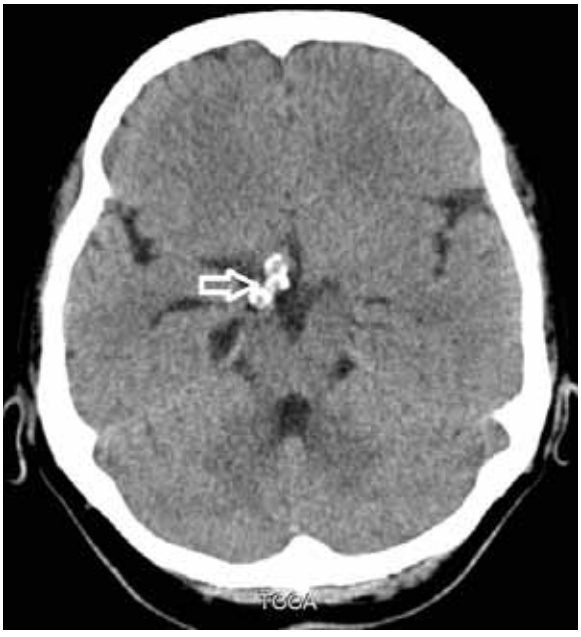

FIGURE 1. *Right ICA areas of calcification (white arrow) localized in the right cavernous region (misinterpreted as a brain tumour)*

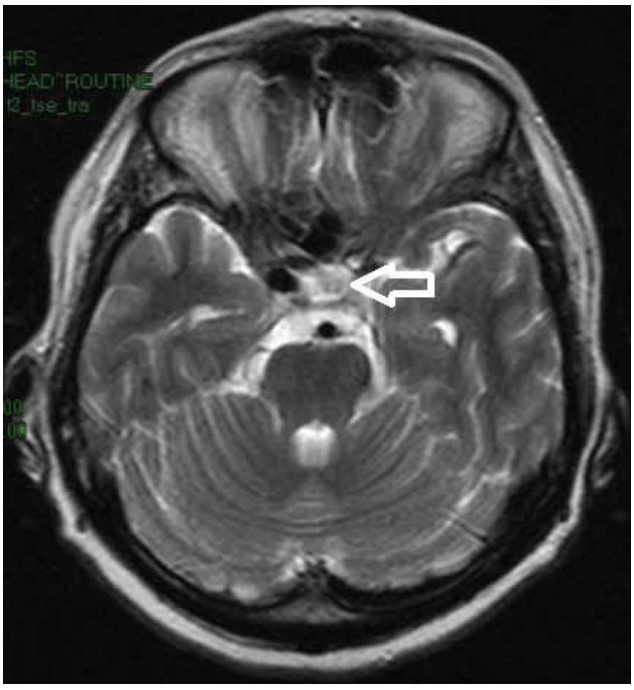

FIGURE 2. *T2 Hyperintense round mass, with a vascular traject and calcifications, intracavernous and interpeduncular corresponding to the area observed on the brain CT scan*

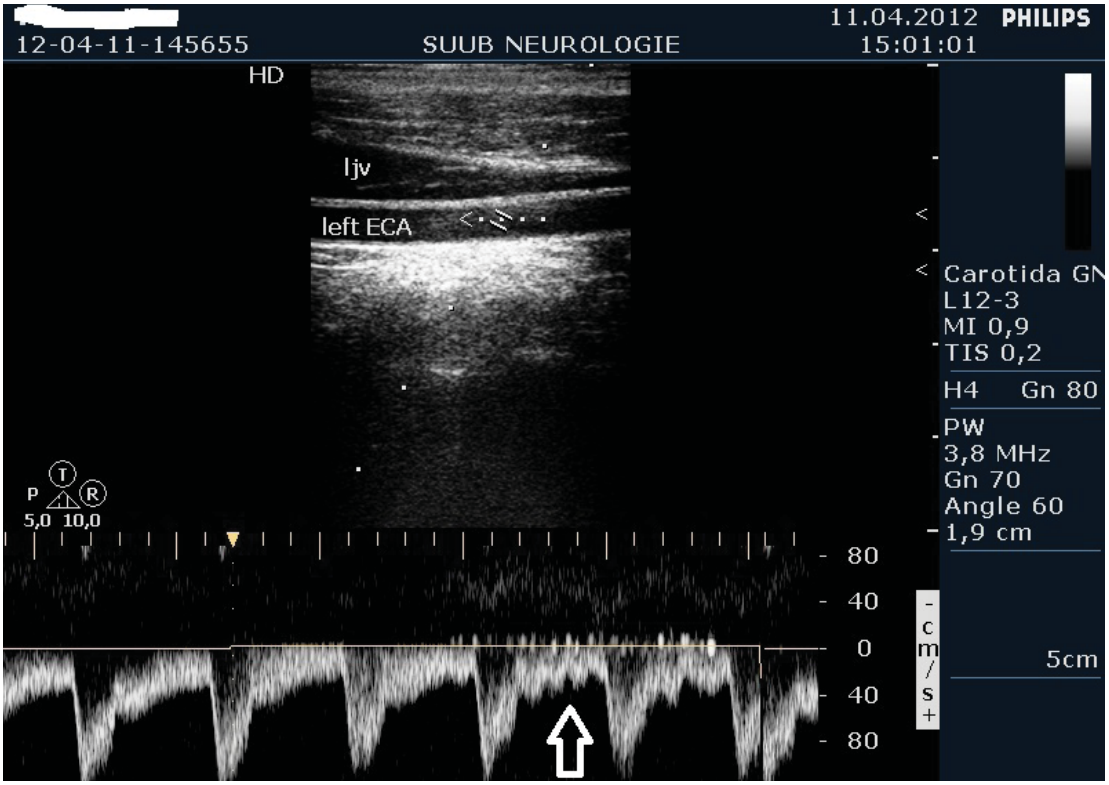

FIGURE 3. *Longitudinal view of the neck vessels. Absence of the left ICA. There is a single artery, in which the spectral analysis of the complexes shows typical aspect of ECA, with disturbance of the flow after temporal artery tapping (white arrow)*

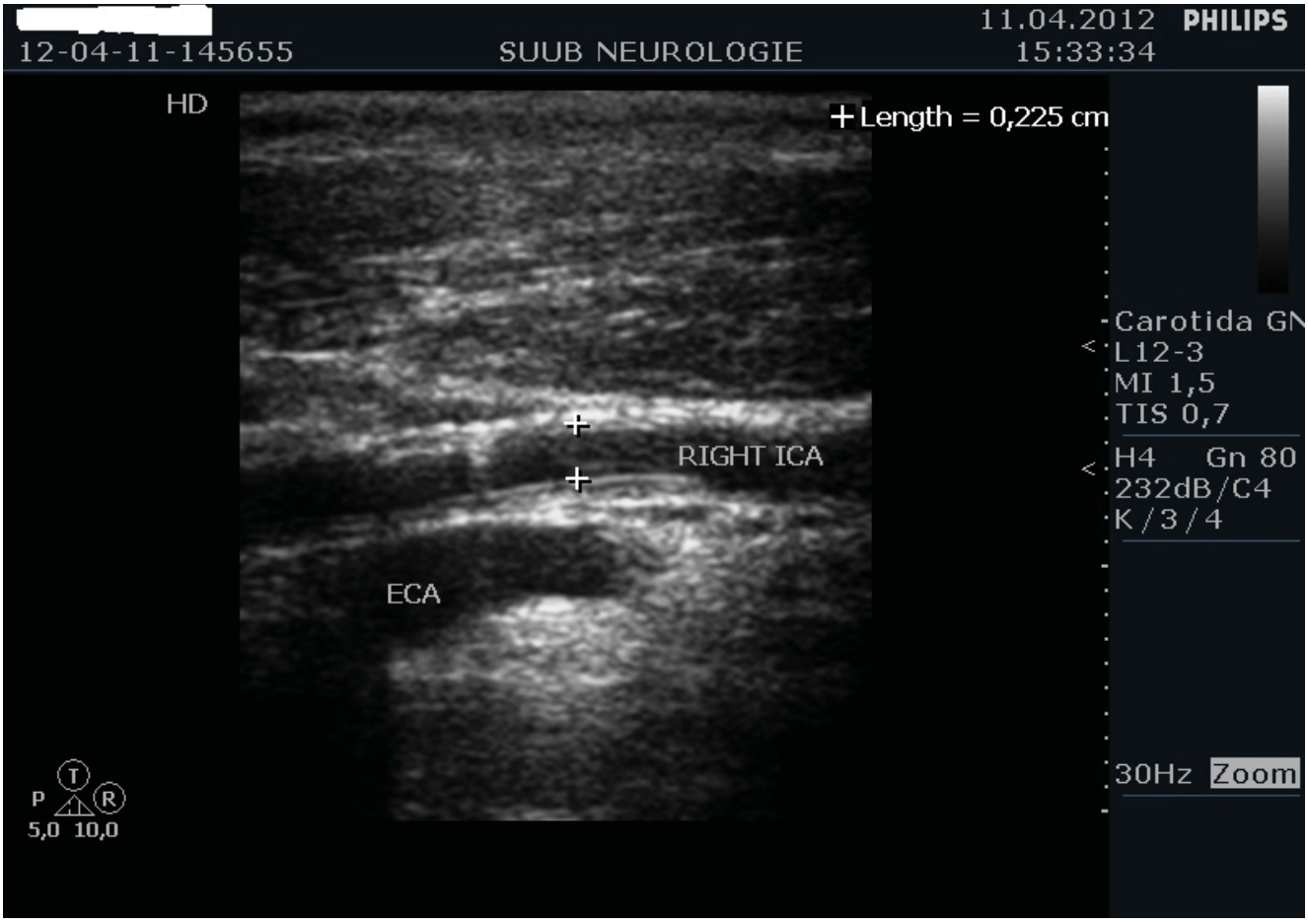

FIGURE 4. *Right ICA and ECA*

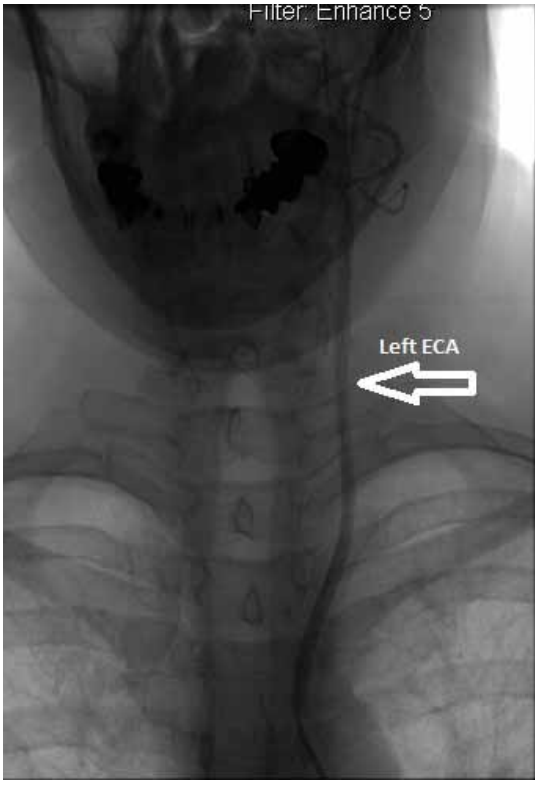

FIGURE 5. *Left ECA and agenesia of left ICA*

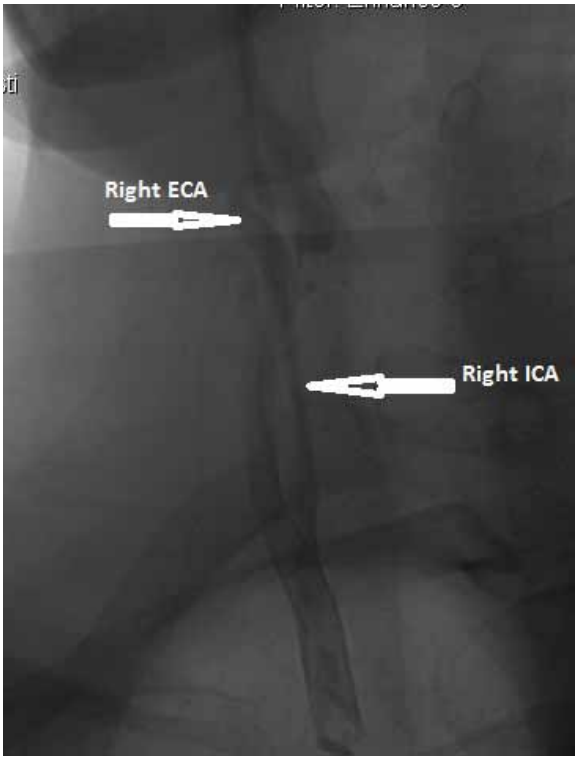

FIGURE 6. *Hypoplasia of right ICA and tortuous right ECA*

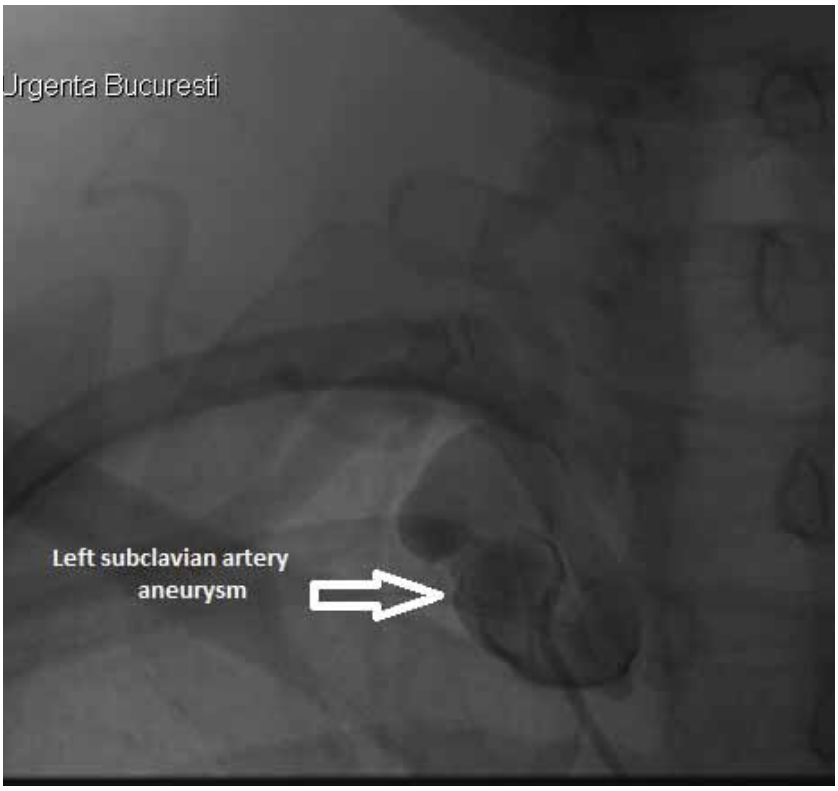

FIGURE 7. *Aneurysm of right subclavian artery*

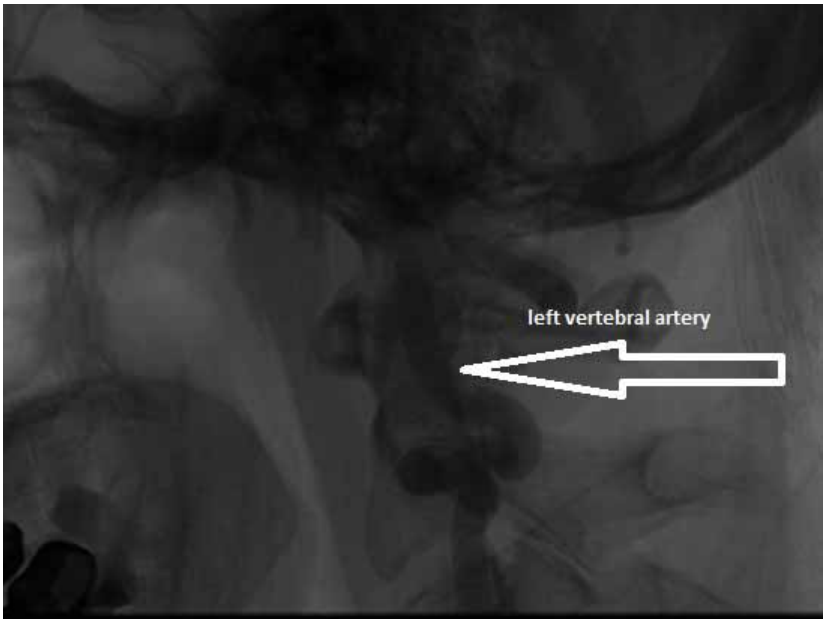

FIGURE 8. *Left vertebral megadolichoartery*

DISCUSSION

Dysgenesia of the ICA is a rare vascular congenital anomaly with a variety of different grades (agenesia, aplasia and hypoplasia). This is more commonly seen unilaterally (2).The left ICA is reported to be affected by dysgenesia three times more often than the right one. Most of the patients with dysgenesia of the ICA are asymptomatic (3).

Agenesia refers to the complete failure of development of the ICA; hypoplasia indicates that the development of the ICA has taken place, but the

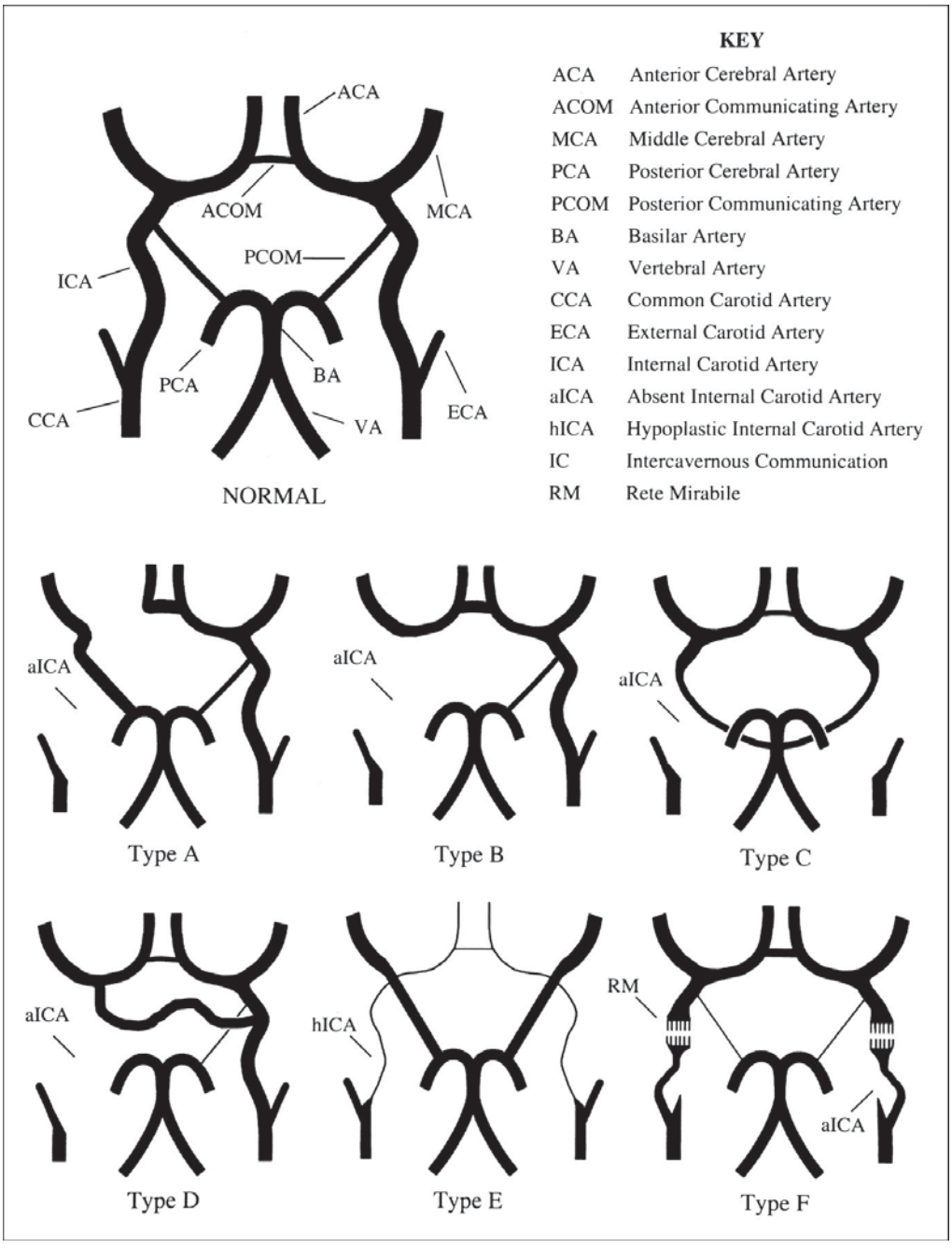

FIGURE 9. *Absence of the ICA: pathways of collateral blood flow. After Curtis A et al (11)*

artery is of a very small calibre; the term aplasia is used when only vestiges of the ICA are presented (4, 5).

Tode documented the first case of carotid agenesia, discovered on post-mortem examination in 1787(6). In 1954, the first case of ICA agenesia at cerebral angiography was reported by Verbiest (7).

Padget's analysis revealed insult sustained by the growing embryo during the 4th-8th weeks of gestational life. The ICA develops from the terminal segment of the dorsal aorta and the third aortic arch arteries before the embryologic stage of 3 mm (24 days).

The skull base starts to develop around the 5th week of gestation. If the ICA is absent, the carotid canal fails to develop. The cause of the unilateral hypoplasia of the ICA has been postulated to be exaggerated folding of the embryo to one side or constriction by amniotic bands. The cause of bilateral hypoplasia has not been identified (1, 7).

There are six pathways of collateral circulation in association with dysgenesia of the ICA. In type A, unilateral absence of the ICA is associated with collateral circulation to the ipsilateral anterior cerebral artery (ACA) through a patent anterior communicating artery (ACOM) and to the ipsilateral middle cerebral artery (MCA) from the posterior circulation through a hypertrophied posterior communicating artery (PCOM). In the type B pattern of collateral flow, the ipsilateral ACA and MCA are supplied across a patent ACOM. Type C represents bilateral agenesis of the ICA with supply to the anterior circulation via carotid – vertebrobasilar anastomoses. Type D represents unilateral agenesis of the cervical portions of the ICA with an intercavernous communication to the ipsilateral carotid siphon from the contralateral cavernous ICA. In type E, diminutive ACAs are supplied by bilateral hypoplasic ICAs, and the MCAs are supplied by enlarged PCOMs. Type F pattern provides collateral flow to the distal ICA via transcranial anastomoses from the internal maxillary branches of the ECA system, the rete mirabile(5). Our case is type B.

These collaterals do not prevent completely the occurrence of TIAs or even stroke. Despite the widespread use of antiplatelet drugs for the prevention of cardio vascular disease, they are not recommended for the prevention of stroke in people without a history of vascular disease (primary prevention), as the risk of having a major bleed may outweigh any potential vascular benefit. Nevertheless, due to the episodes of blurred vision in the correspondent side of the ICA agenesia we considered it is appropriate to treat the patient with Clopidogrel for three months, with a further reevaluation for long term treatment. The patient does not have an indication for long term use of antiplatelet drugs in primary prevention since she does not have other vascular disease excepting the developmental anomalies of the vessels, but she has to be monitored carefully; the addition of atheromatosis lesions to the hypoplastic and tortuous vessels can exponentially increase the risk of stroke.

We have to consider also the increased risk of aneurysm formation (24%-34%) and the occurrence of subarachnoid haemorrhage. Increased flow through collateral vessels and altered flow dynamics are cited as plausible explanation for this increased prevalence. Our patient had aneurysm of right subclavian artery (8, 9).

Congenital unilateral narrowing of the ICA must be differentiating from acquired causes: chronic dissection, severe atherosclerosis, fibromuscular dysplasia or moya- moya disease. Tortuous traject of the intrapetrous segment of right ICA was missinterpreted as a tumoural brain mass; Doppler ultrasonography examination of the neck arteries revealed the etiologic diagnostic, and digital substraction angiography confirmed it. Magnetic resonance angiography was not sufficient for the diagnosis since irt revealed only the absence of the flow, which was misinterpreted as an occlusion due to a dissection.

This could have been avoided by the radiologist if he/she would have been looked on transverse neck sections, where the absence of the arterial wall of the left ICA was obvious.

Recognition of the absence of carotid artery is important when planning carotid endarterectomy, as both cerebral hemispheres may be dependent on a single carotid artery (10).

CONCLUSION

Agenesis, aplasia and hypoplasia of the ICA are rare anomalies. Many of these cases remain asymptomatic and patients go on undetected. Our case report suggests that ultrasound examination is very reliable in anatomical and developmental anomalies, being superior to magnetic resonance angiography. Doppler ultrasound examination can be very useful for differentiating developmental anomalies from atherosclerotic lesions, and for monitoring these patients in order to prevent cerebrovascular events.

REFERENCES

1. **Wassim Farhat, Rechdi Ahdab, Hassan Hosseini** – Congenital agenesis of internal carotid artery with ipsilateral Horner presenting as focal neurological symptoms. *Vascular health anf risk management* 2011:7 37-40
2. **Mellado J.M., Merino X., Ramos A., Salvado E., Sauri A.** – Agenesis of the internal carotid artery with a trans- sellar anastomosis: CT and MRI findings in late onset congenital hypopituitarism. *Neuroradiology* 2001; 43:237-41
3. **Olga Kiritsi, George Noussios, Konstantinos Tsitas, Dimitrios Lappas** – Unilateral agenesis of the internal carotid artery presented as transiet ischemic attack: a casa report. *Surg Radiol Anat* april 2011
4. **Ito S., Miyazaki H., Iino N., Shiokawa Y., Saito I.** – Unilateral agenesis and hypoplasia of the internal carotid artery: a report of the three cases. Neuroradiology. 2005; 47:311-5
5. Lie T.A. – Excerpta Medica; 1968. Congenital anomalies of the carotid arteries; 35-51
6. **Midkiff R.B., Boykin M.W., McFarland D.R., Bauman J.A.** – Agenesis of the internal carotid artery with intercavernous anastomosis. *AJNR Am J Neuroradiol* 1995; 16: 1356- 1359
7. **Claros P., Bandos R., Gilea I. et al.** – Case report: major congenital anomalies of the internal carotid artery- agenesis, aplasia and hypoplasia. *Int Pediatr Otorhinolaryngol* 1999; 49:69-76
8. **Afifi A.K., Godersky J.C., Menezes A., Smoker W.R., Bell W.E., Jacoby Cg.** – Cerebral hemiatrophy, hypoplasia of internal carotid artery , and intracranial aneurysm: a rare association occurring in an infant. *Arch Neurol* 1987; 44:232-235
9. **Quint D.J., Boulos R.S., Spera T.D.** – Congenital absence of the cervical and petrous internal carotid artery with intercavernous anastomosis. *AJNR An J Neuroradiol* 1989; 10: 435-439.
10. **Luk Y.S., Man E.M., Sy A.N.L.** – Bilateral hypoplasia of the internal carotid arteries. *Singapore Med J* 2010; 51(9) 163-165
11. **Curtis a et al.** – Congenital absence of the internal carotrid artery: case reports and review of the collateral circulation. *AJNR Am J Neuroradiol* 22: 1953-1959, Nov/Dec 2001