

Ultrasonographic findings and collateral circulation in hypoplasia of the internal carotid artery

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Introduction

Hypoplasia of the internal carotid artery (HICA) is a rare congenital anomaly caused by incomplete development of the organ, and only a few cases are reported in the literature (1). The prevalence of HICA (including agenesis and aplasia) is estimated to be about 0.01% (1,2). Detection of this disease has important clinical implications. Ultrasonography (US) is usually the first examination performed to detect suspected HICA. In our opinion, HICA should be suspected in all patients who have a diffuse luminal narrowing of the internal carotid artery (ICA).

Case Report

A 66-year-old man with hearing loss on the left side and no other symptoms or signs of vascular impairment underwent examination of the extracranial cerebral vessels. Approval for this study was granted by our hospital's Medical Research Ethics Committee, and informed consent was obtained from the patient. Extracranial duplex sonography of the cerebral vessel showed kinking and diffuse luminal narrowing of the left ICA with significantly reduced flow velocity (PSV 53.5 cm/sec; PEDV 15.0 cm/sec; Fig. 1), which contrasted with the contralateral side (PSV 89.7 cm/sec; PEDV 45.6 cm/sec). The left external carotid artery showed increased diastolic ("internalised") blood flow, and the left ophthalmic artery showed inverted flow and a high diastolic component similar to that of a brain-supplying artery. No other abnormalities were

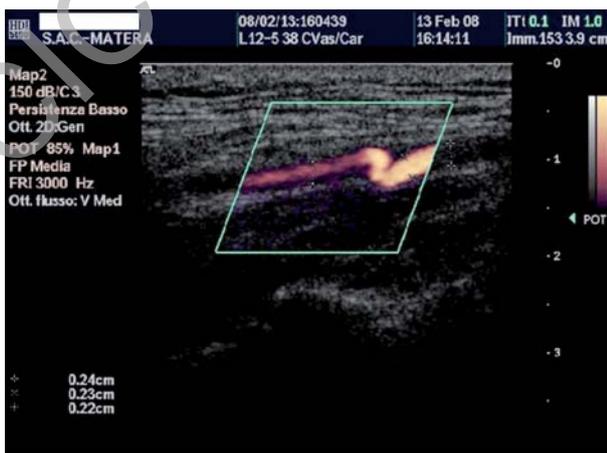


Fig. 1 - Extracranial duplex, longitudinal plane. There is kinking and diffuse luminal narrowing of the left internal carotid artery.

found. Transcranial duplex sonography revealed a difference in flow velocity between the two sides with reduced flow velocity in the left carotid siphon, in the left M1 MCA segment, as well as in the left A1 ACA segment, and absence of the A1 segment of the right anterior cerebral artery (ACA). Assessment of the posterior circulation was unremarkable.

Angio MRI of the cerebral vessels showed reduced signal intensity in the left ICA (left ICA diameter 1.9 mm; right ICA diameter 5.2 mm), which was in accordance with intracranial and extracranial US findings. Axial CT scan of the skull base showed a normally developed right carotid canal and a diminutive left carotid canal, which is a sign of congenital ICA abnormalities.

Discussion

Underdevelopment or absence of the ICA is an uncommon disease (1). Six pathways of collateral circulation in association with aplasia or hypoplasia (a/hypoplasia) of the ICA are described (3). Type 1: a/hypoplasia of the ICA is associated with anterior communicating artery (ACOM) and posterior communicating artery (PCOM) hypertrophy. The ipsilateral ACA is supplied through a patent ACOM, while the middle cerebral artery (MCA) is supplied through a patent hypertrophied PCOM. Type 2: a/hypoplasia of the ICA is associated with ACOM hypertrophy, and the MCA and homolateral ACA are supplied through a patent ACOM. Type 3: a/hypoplasia of the ICA is bilateral. Carotid-vertebrobasilar anastomosis allows anterior circulation of the brain. There is often PCOM hypertrophy. Type 4: there is unilateral underdevelopment of the cervical portion of the ICA associated with the transcavernous communication. The transcavernous collateral vessels may be located behind the clivus, or run above, through, or in the floor of the sella turcica (4). Type 5: there is a bilateral hypoplasia of the ICA. The ACAs are supplied through hypoplastic ICAs while the MCAs are supplied through enlarged PCOMs. Type 6: a/hypoplasia of the ICA is associated with anastomosis (rete mirabilis) from the homolateral external carotid artery. Our case showed a type 6 collateral circulation.

In our opinion, HICA warrants some important considerations. Patients with HICA may be completely asymptomatic due to collateral blood supply to the affected hemisphere, or they may present symptoms due to cerebrovascular insufficiency or compression by enlarged collateral cerebral vessels.

In these patients, the prevalence of intracranial aneurysm is estimated to be about 24%-34%, while in the general population it is 2%-4% (5,6).

Recognition of HICA is important in the assessment of cerebral ischaemia. Emboli in one cerebral hemisphere may be explained by an atherosclerotic disease of the contralateral carotid system or vertebrobasilar system.

Recognition of HICA is also very important in the planning of surgical procedures such as transsphenoidal hypophyseal surgery (7) and carotid endarterectomy.

HICA constitutes a diagnostic challenge for the sonographer. Diffuse luminal narrowing of the internal carotid can be caused by carotid dissection, high grade stenosis, arteritis, fibromuscular dysplasia and radiation-induced angiopathy. The haemodynamic findings in these diseases do not allow a differential diagnosis, but some signs may help to avoid US misdiagnosis.

Carotid dissection is a major cause of stroke in young patients. Although diagnostic confirmation using neuroradiological diagnostics is mandatory, US may be helpful, particularly in the initial investigation and in the follow up. Detection of direct signs of carotid dissection is often possible: a tapering occlusion, a pseudoaneurysm resulting from the rupture of the artery and subsequent encapsulation of the paravascular haematoma, a double lumen with an intimal flap, a flat, hyperechoic structure floating in the lumen or dividing the true and the false lumen. Hypoechoic thickness of the wall, which is a sign of haematoma and intraluminal thrombus, is not pathognomonic of carotid dissection, as it could also be a sign of anechoic plaques or mural thrombus. Also indirect US signs, essentially haemodynamic changes due to high grade stenosis are not pathognomonic of carotid dissection.

Fibromuscular dysplasia is a non-atherosclerotic, non-inflammatory segmental vascular disease of unknown origin. The extracranial cerebral vessels are frequently affected, particularly the distal ICA. US diagnostic findings are a segmental string-of-beads pattern with alternating regions of luminal narrowing and vessel dilatation (8). The patient's anamnesis may cause suspicion of radiation-induced carotid angiopathy. Irradiation leads to fibrosis of the tunica media and tunica adventitia. US shows plaques, which are usually midechoic and non-calcified.

Patients with high grade arteriosclerotic lesions usually present vascular risk factors. Atherosclerotic vessel wall changes are found in several arteries. The atherosclerotic plaques are irregular and with inhomogeneous echogenicity because the lesions are often calcified.

Particularly in Takayasu disease, arteritis may be confounding, but some aspects have to be considered. Takayasu disease occurs mainly in young women and is characterised by progressive stenoses and occlusions of the arteries arising from the aortic arch. US findings are very typical. A severe, diffuse wall thickening is observed especially in the internal and common carotid arteries. The wall deposit is typically homogeneous, midechoic, circumferential and has been described as the “macaroni sign” by Maeda et al. (9). Large-vessel giant cell arteritis may appear in several vessels other than the temporal arteries, so we recommend considering the possibility of carotid artery involvement. The US sign of giant cell arteritis is a dark halo around the artery lumen which may be due to oedema of the artery wall. In clinical practice it is important to bear this in mind, too, as the hypoechoic circumferential halo typical of large-vessel giant cell arteritis, like the midechoic homogeneous wall thickening of the Takayasu arteritis, may disappear with corticosteroid therapy (9,10).

In our opinion, colour duplex US allows diagnosis of underdevelopment of the ICA although it is sometimes difficult to distinguish between underdevelopment of the ICA and other diseases causing diffuse luminal narrowing of the vessel. However, some US signs may help to avoid diagnostic error. In unclear cases and in cases in which US outcome is considered unreliable, angio MRI or CT of the skull base may show absence of the carotid canal as a sign of congenital ICA abnormalities. Angio MRI or CT should therefore be performed to confirm the diagnosis. Recognition of HICA may be important in the assessment of cerebrovascular diseases and in the planning of carotid and transsphenoidal hypophyseal surgery, and it should prompt further evaluation to rule out the presence of a potentially life-threatening intracranial aneurysm, even in asymptomatic patients.

References

1. Clarós P, Bandos R, Gilea I et al. Major congenital anomalies of the internal carotid artery: agenesis, aplasia and hypoplasia. *Int J Pediatr Otorhinolaryngol* 1999;49:69-76
2. Chen CJ, Chen ST, Hsieh FY, Wang LJ, Wong YC. Hypoplasia of the internal carotid artery with intercavernous anastomosis. *Neuroradiology* 1998;40:252-254
3. Lie TA. *Congenital Anomalies of the Carotid Arteries*. Amsterdam; Excerpta Medica 1968:35-41
4. Janicki PC, Limbacher JP, Guinto FC Jr. Agenesis of the internal carotid artery with a primitive transsellar communicating artery. *AJR Am J Roentgenol* 1979;132:130-132
5. Afifi AK, Godersky JC, Menezes A, Smoker WR, Bell WE, 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
6. Quint DJ, Boulos RS, Spera TD. Congenital absence of the cervical and petrous internal carotid artery with intercavernous anastomosis. *AJNR Am J Neuroradiol* 1989;10:435-439
7. Kishore PR, Kaufman AB, Melichar FA. Intrasellar carotid anastomosis simulating pituitary microadenoma. *Radiology* 1979;132:381-383
8. Arning C, Grzyska U. Color Doppler imaging of cervicocephalic fibromuscular dysplasia. *Cardiovasc Ultrasound* 2004;2:7
9. Maeda H, Handa N, Matsumoto M et al. Carotid lesions detected by B-mode ultrasonography in Takayasu's arteritis: “macaroni sign” as an indicator of the disease. *Ultrasound Med Biol* 1991;17:695-701
10. Nicoletti G, Ciancio G, Tardi S, Olivieri I. Colour duplex ultrasonography in the management of giant cell arteritis. *Clin Rheumatol* 2003;22:508-509