Co-occurrence of agenesis of internal carotid artery with contralateral arterio venous malformation

Sir,

A 19-year-old boy presented with history of recurrent episodes of right focal seizures with secondary generalization and postictal weakness of the right upper limb. On examination, there were no focal neurological deficits. Magnetic resonance (MR) imaging of brain with MR angiography [Figure 1], revealed an arterio venous malformation (AVM) in the left parietal lobe. Right internal carotid artery (ICA) was absent with dilatation of right posterior communicating artery (P Com). Digital subtraction angiography [Figure 2] showed that the right common carotid artery was arising from the innominate artery, which was continuing as the external carotid artery (ECA) with no internal carotid artery. In addition, angiography also revealed an AVM with a nidus measuring approximately 3 cm, in the left parietal region with feeders from M3 segment of the left middle cerebral artery (MCA) and draining into the left transverse sinus and superior sagittal sinus. The right MCA was filling through hypertrophied P Com. The right anterior cerebral artery (ACA) was filling through the anterior communicating artery (A Com). High-resolution CT (HRCT) of the skull base showed absence of the right carotid canal [Figure 3]. Patient underwent stereotactic radiosurgery for the treatment of AVM and he is on regular follow-up.

Absence of the carotid artery was first reported by Tode during postmortem examinations in 1787. First documentation of the absence of ICA on cerebral angiography was by Verbiest. Approximately 100 cases of congenital absence of ICA have been reported in the literature. The incidence of unilateral agenesis of ICA is 0.36%. If the embryonic primordium of ICA fails to develop before three to five weeks of embryonic life, the
ICA and carotid canal cannot develop. Agenesis and hypoplasia can be reliably diagnosed by HRCT of the skull base by noting the absence or narrowing of the carotid canal respectively. There are three types of collateral circulations associated with this entity for completion of the circle of Willis. The commonest is of the fetal type– P Com is dilated and supplies the MCA and A Com supplies the ACA. The second variety is of the adult type, –both ACA and MCA are supplied by the A Com. The third variety-distal ICA - is present with transcranial anastomoses that develop from ECA or contralateral ICA or primitive vessels.

Agenesis of ICA is most commonly associated with cerebral aneurysms at the circle of Willis due to hemodynamic alterations with an incidence of 25-35%. Other associations described are oculomotor and trigeminal nerve palsies, spasmatic torticollis, hypopituitarism, ear malformations with facial nerve palsy, congenital Horner’s syndrome. Infarctions are uncommon, in fact, due to development of adequate collateral circulation.

The etiology of AVM is considered to be dysregulated angiogenesis that occurs in the neuronal proliferation and histiogenesis phase of the brain development. Fukui et al earlier described a case in which there was anomalous origin of the ICA from the CCA, with anomalous course of ICA and an associated ipsilateral AVM. They proposed that the AVM would be due to maldevelopment of the vascular network including the aortic arches on the same side. In our case the AVM is on the side contralateral to the agenesis of ICA. To our knowledge this is the first documented case of this type. We cannot determine whether the agenesis of ICA and contralateral AVM occurred in our patient by an associated pathogenic mechanism or just simply by chance.

In these cases of agenesis of ICA diagnosed by MRA or HRCT, it is mandatory to search for associated vascular malformations like aneurysms and very rarely like our present case, an AVM. Digital subtraction angiography is important for the evaluation of the type of collateral circulation before planning any intervention.

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References


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