







Bilateral Hypoplasia of the Internal Carotid Artery with Subarachnoid Hemorrhage and Distal Posterior Cerebral Artery Aneurysm

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Bilateral internal carotid artery (ICA) hypoplasia is a rare congenital anomaly. Their actual incidence is higher than what is reported as majority of them remains asymptomatic during lifetime.¹ It is important to differentiate congenital ICA hypoplasia from acquired causes of ICA stenosis like moyamoya disease. The most conclusive differentiating point is the size of the carotid canal that in congenital cases cannot be clearly made out.² Further unlike the supraclinoid stenosis of ICA in moyamoya disease, ICA hypoplasia here is much more proximal just distal to its point of origin.³

Our case was a 27-year-old primigravida, who presented with severe headache. No other relevant positive points were there in the history. Clinical examination revealed neck rigidity. She was conscious, alert, oriented with no deficits. Computed tomographic (CT) brain showed extensive intra ventricular hemorrhage involving the lateral and third ventricles, specks of blood in the basal cisterns, and mild hydrocephalus. Bone windows revealed bilateral atretic carotid canals (>Fig. 1). CT angiography revealed hypoplasia of bilateral ICA that were only 1 to 1.5 mm in diameter and ended in string like structures on both sides. An aneurysm was seen arising from the distal cortical branches of the right P4 segment of posterior cerebral artery (PCA) (Fig. 2). Angiogram was performed with the help of C arm having the roadmap software and images of the posterior circulation showed extensive collateral circulation arising from the vertebrobasilar system (-Fig. 3). Endovascular coiling was attempted for the PCA aneurysm, but it was not possible to navigate up to the neck of the aneurysm as the aneurysm was very distally located and access to it was difficult. Also, the distal branches of the PCA were of very small caliber and in acute stage they were in spasm. Ultimately, distal parent branch of the PCA wherefrom the aneurysm was arising was occluded with polyvinyl alcohol particles of size 300 nm (>Fig. 4). Recovery was uneventful. Till the time of last follow-up, that is, 12 months following the procedure, the patient is doing fine.

Etiogenesis of ICA hypoplasia can be described as maldevelopment of the dorsal aorta that can explain the reason of hypoplastic vessel being seen at the point of origin itself just above the bifurcation. In some cases, the hypoplastic ICA can also continue as a very small vessel intracranially or may end by giving rise to ophthalmic artery.4

The pattern of bleeding in the CT scan in our case was not conclusive of rupture of PCA aneurysm. We strongly feel that the collateral vessels were flimsy and friable in nature and

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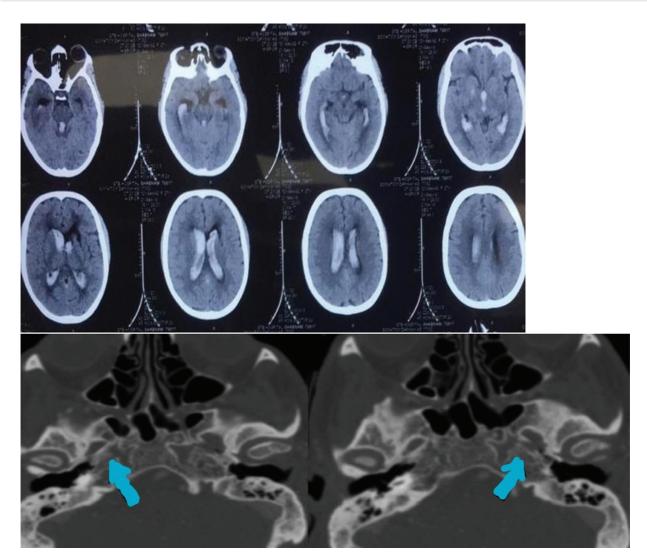


Fig. 1 Computed tomographic brain showing extensive intraventricular hemorrhage and bilateral atretic carotid canals.

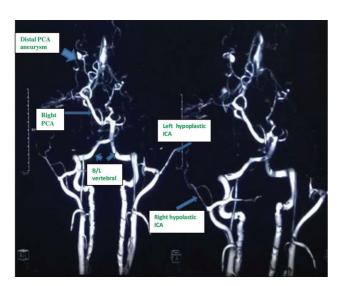


Fig. 2 Computed tomographic angiography of the aortic arch and major vessels. It shows hypoplastic internal carotid artery (ICA) on both sides and an aneurysm arising from the distal branches of the right posterior cerebral artery (PCA).

had abnormal architecture that was prone to rupture owing to hemodynamic stress. We also think that pregnancy additionally increased the risk of rupture because of hemodynamic alterations. Otherwise in symptomatic patients in ICA hypoplasia can present as aneurysmal subarachnoid hemorrhage (SAH), ischemic stroke, or focal deficits. But associated aneurysm with SAH as the presenting symptom was found in only eight cases in our literature search including our case.⁵

Congenital hypoplasia of the bilateral ICA presenting with a bleeding episode is very rarely seen. Detailed evaluation of cerebrovascular anatomy is important for a prompt diagnosis. Whenever aneurysm is detected, it should be treated to prevent rebleeding. Overall management option should take into account acceptable risk-benefit equations.

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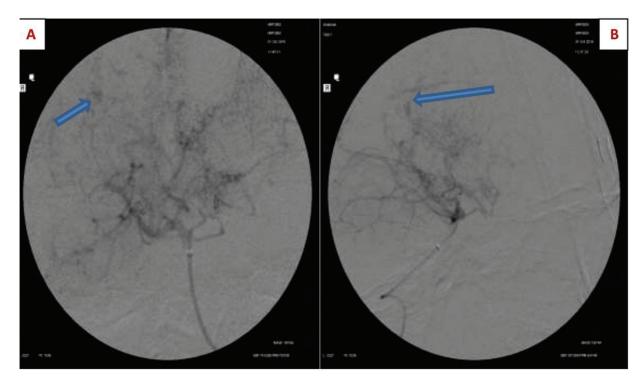


Fig. 3 Anteroposterior and lateral view of the digital subtraction angiography images of basilar artery showing extensive collateral circulation. Blue arrow shows the aneurysm in both views.



Fig. 4 Follow-up digital subtraction angiography of basilar artery showing good filling of the B/L posterior cerebral artery with extensive collateral circulation. Blue arrow shows absence of the aneurysm with the cortical branch which was occluded.

Conflict of Interest None.

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