

Images: Bilateral agenesis of the internal carotid arteries - MRI and MR angiography

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A 55-years-old woman came with sudden onset left hemiparesis for 12h, following an episode of convulsions. On admission, the power was grade 2/5 in the left upper and lower limbs. An MRI brain was performed. The MRI showed hyperintense areas in the right high frontal and parietal lobes on T2W images [Figure 1], with restricted diffusion on diffusion-weighted (DW) images [Figure 2], representing acute non-hemorrhagic infarcts. In addition, the axial spin-echo (SE) T1W and T2W [Figure 3] images revealed absence of the normal flow voids of the internal carotid arteries (ICAs) in the region of the carotid canals in the skull base. The expected regions of the carotid canals were filled with fibro-fatty tissue.

The 3D time-of-flight (TOF) MRI angiography (MRA)

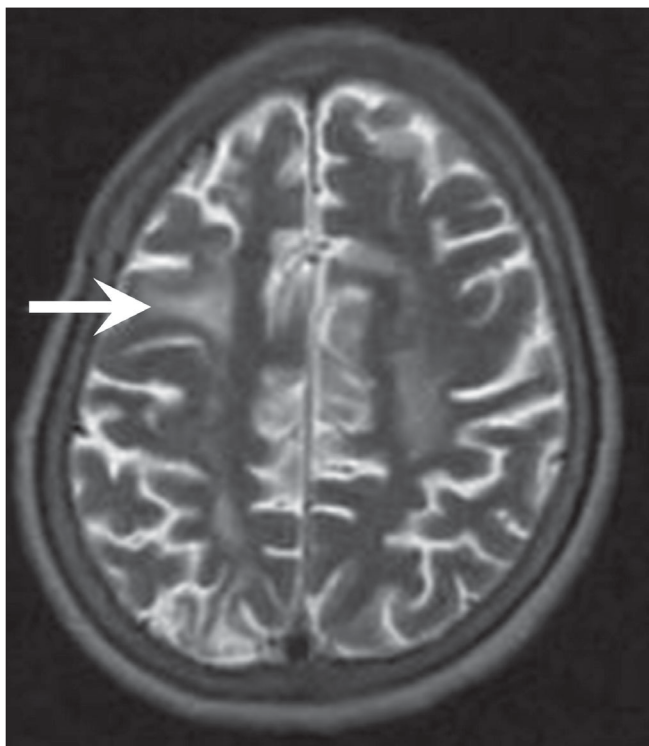


Figure 1: Axial T2W image showing hyperintense signal (white arrow) in the right high frontal and parietal lobes.

images [Figures 4, 5] demonstrated prominent posterior cerebral arteries supplying both the anterior circulations through the posterior communicating arteries. The axial raw data (source images) [Figure 6] confirmed the absence of flow signals in the expected location of the ICAs on either side. No other vascular abnormality was seen in the form of aneurysm or focal stenosis.

Subsequently, the patient was managed with anticoagulant therapy, to which she responded favorably. No complications were encountered during her hospital stay till her discharge after nine days. She left with mild residual weakness in the left upper and lower limbs.

Discussion

Absence of the internal carotid artery (ICA) is a rare congenital anomaly,^[1] which may be the result of an acquired

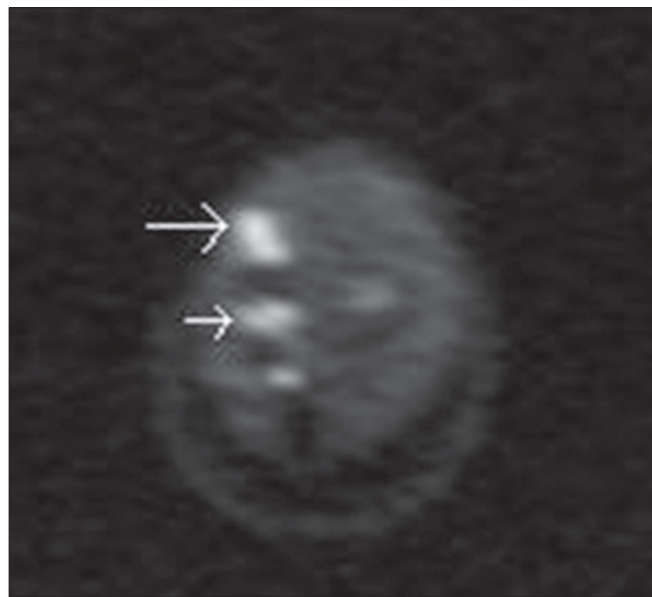


Figure 2: Diffusion weighted image showing areas of restricted diffusion (white arrows) in the right high frontal and parietal lobes representing acute nonhemorrhagic infarcts

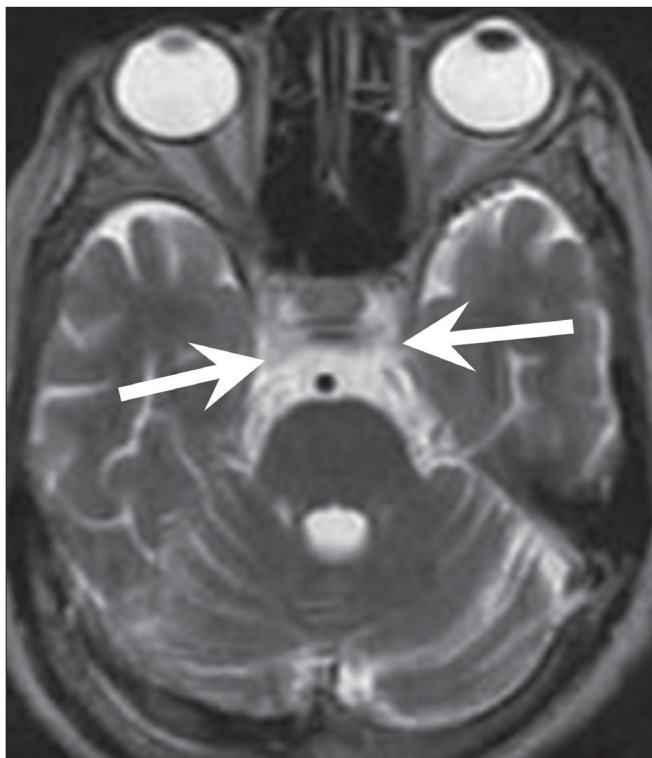


Figure 3: Axial T2W image taken through the skull base at the level of the carotid canals showing absence of flow voids in the expected region of the carotid canals bilaterally (white arrows). The normal flow void of the basilar artery is well seen.



Figure 5: The left lateral projection image shows the absence of internal carotid arteries bilaterally and prominent vertebral (black arrows), basilar (double arrows), posterior cerebral and posterior communicating (white arrow) arteries supplying the anterior circulation.

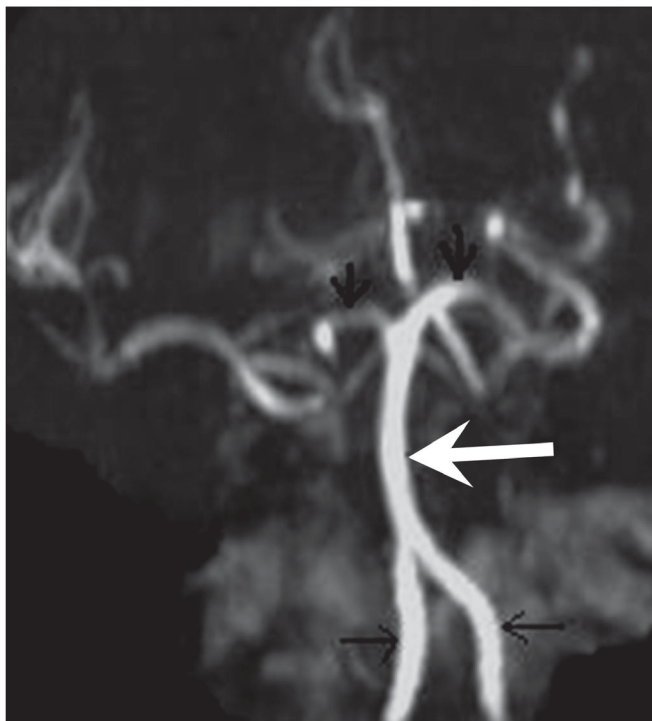


Figure 4: A MIP images of 3D TOF MRA. The oblique antero-posterior projection image shows absence of the internal carotid arteries bilaterally and prominent vertebral (black arrows), basilar (white arrow) and posterior cerebral (short thick black arrows) arteries.



Figure 6: Axial raw data images (source images) showing the prominent posterior communicating artery (arrow) supplying the anterior circulation on the left side. The internal carotid arteries are absent on both sides.

disease (common), congenital hypoplasia (uncommon) or congenital aplasia (rare). This is more commonly seen unilaterally.^[2,3] Most reported cases of bilateral absence have been recorded prior to 1980.^[4] Previous reports have been largely based on catheter angiography.^[5] In 1954, the first case of ICA agenesis at cerebral angiography was reported

by Verbiest.^[1] Earlier case reports relied on dissection studies.^[2]

Lie^[6] defined agenesis as complete failure of an organ to develop, aplasia as lack of development (but its precursor did exist at one time) and hypoplasia as incomplete development of the organ. Although an exact cause of these developmental anomalies has not been established, all three variations are thought to represent the sequelae from an insult to the developing embryo.^[4] To date, an explanation for bilateral absence has not been rendered.^[4] Evaluation of the skull base for the presence or absence of the carotid canal may be required for distinguishing aplasia from agenesis, as presence of the ICA (or its precursor) is a prerequisite for development of the carotid canal at five to six weeks of gestation.^[6] Therefore, demonstrating an absence of the carotid canal with skull base CT will confirm the diagnosis of agenesis.^[7]

Several case reports have discussed the MRI and MRA findings in unilateral aplasia or hypoplasia of the ICA, including children.^[2] Bilateral absence of the ICA is a rare abnormality.^[2] Kidooka *et al* believed the MRA assessment in their case was equivalent to that provided by catheter angiography.^[3] Anderson's case report^[2] also suggests that MRA is able to successfully demonstrate ICA absence and common collateralization in a noninvasive fashion.

Usually, patients with this anomaly are asymptomatic; some may have symptoms related to cerebrovascular insufficiency, compression by enlarged intracranial collateral vessels or complications associated with cerebral aneurysms.^[8]

The estimated prevalence of cerebral aneurysms in the general population is 2-4%, but the reported prevalence of aneurysms in association with absence of ICA is 24% to 34%.^[1] Recognition of this anomaly becomes important in thromboembolic disease, as emboli in one cerebral hemisphere may be explained by atherosclerotic disease in the contralateral common carotid artery (CCA) or vertebro-basilar system.^[1]

Lie^[6] described six pathways of collateral circulation in association with absence of the ICA. Others have simplified Lie's original classification of collateral circulation in absent ICAs into three main types: collateral flow through the circle of Willis (*most frequent and also seen in the present case*); collateral flow via persistent fetal circulation and reconstitution of the ICA through skull base collaterals from the ECA.^[1]

The MRA in this case demonstrated supply to the anterior circulation via enlarged posterior communicating arteries, the most commonly seen collateral arrangement.^[9]

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