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) 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 anomaly, a developmental arrest of the common carotid artery, or an acute embolism. MRI and MR angiography (MRA) can be used to diagnose this condition. The lack of flow signals in the expected location of the ICAs on either side confirmed the absence of the ICAs.

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

**Figure 2:** Diffusion weighted image showing areas of restricted diffusion (white arrows) in the right high frontal and parietal lobes representing acute nonhemorrhagic infarcts
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.[8] 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.[3] Bilateral absence of the ICA is a rare abnormality.[5] 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 vertebral-basilar system.[11]

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]

References


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