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Congenital Aplasia of Right Internal Carotid Artery Presenting as Complex Partial Seizures

KEYWORDS

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ABSTRACT *Agenesis of Internal carotid artery is a rare congenital anomaly and most often they are asymptomatic. We report a case of developmentally normal infant with recurrent left sized seizures. Evaluation revealed a hypo-plastic internal carotid artery. Patient was treated conservatively with Carbamazepine and is under regular follow up with clinically normal neurological examination. As it is a rare case and is symptomatic it is being presented. Abnormal vascular anatomy has to be carefully investigated prior to go for any endovascular surgery for ruptured coexisting aneurysm because any iatrogenic manipulation may disturb compensatory collateral circulation and induce irreversible neurological deficit. Combined use of MRI, MRA and CT scanning of base of skull and head are the necessary imaging studies to diagnose such abnormalities and anomalies*

Introduction: Agenesis, aplasia or hypoplasia of the internal carotid artery is rare congenital vascular anomalies with an incidence less than 0.01% of population and with less than 100 cases reported worldwide. They are asymptomatic and are often detected as incidental findings. Restoration of sufficient cerebral circulation to the defective area will be by communicating arteries of circle of Willis and other anastomosing arteries of carotid territory. We report a rare case of Aplasia of Right Internal Carotid artery. Unilateral absence or hypoplasia of ICA is more common on left side. In the reported cases, prevalence of right to left to bilateral agenesis of ICA has been found to be 1:3:1.

Abbreviations: MRI - Magnetic resonance Imaging, MRA - Magnetic resonance angiogram, CT – Computed tomography, ICA - Internal carotid artery, ACA – Anterior cerebral artery, MCA – Middle cerebral artery

Case report: A one year old girl presented with 4 episodes of left tonic clonic seizures for last 3 months. There was a Gross motor delay in the development and neurological evaluation was normal. Ophthalmological evaluation and EEG done were normal. A provisional diagnosis of complex partial seizures was made. MRI Brain showed aplasia of right Internal carotid artery. MR angiography brain confirmed the aplasia of cervical segment of right ICA with hypoplasia of petrous, lacerum, cavernous and clinoid segments of the right ICA and demonstrated the maintenance of flow related signal in the right ACA and MCA due to collateral supply across the circle of Willis with prominent posterior communicating artery on right side. Origin of the right ICA was visualized with attenuated caliber, so ruling out agenesis. The child remained asymptomatic with pharmacological management (Carbamazepine) of her seizure disorder. No surgical or interventional correction was attempted.

Discussion: In 1987, Tode first described it in a cadaver. Later, ICA agenesis was reported by Verbiest at MR angiography imaging. The definitions of agenesis, aplasia and hypoplasia are often used.

Embryology: Proximal portion of right and left ICA originate from third arch arteries. The distal portion of ICA is derived from cranial extensions of dorsal aorta and is fully developed by 6th week. Involution of third aortic arch and distal part of dorsal aorta in embryonic phase decides agenesis or aplasia of ICA. Only after 5th or 6th week, skull base begins to form, hence if by 3rd-5th week of fetal life, the embryonic precursor of the ICA has not developed, the ICA and the carotid canal will never develop.

Insults during embryonic development result in the variants of ICA anomalies like excessive bending of the embryo to a side or pressure or restriction by amniotic bands.

They are usually asymptomatic as cerebral circulation is compensated to the affected area by communicating arteries of circle of Willis.The associated intracranial vascular anomalies like aneurysms, dilated vascular channels develop secondary to increased hemodynamic load on the normal side and congenital defects of the vessel wall. Tsuruta et al and Miyaziki distinguished 3 types of unilateral agenesis of ICA

Type I: ACA on the ipsilateral side is filled from contralateral ICA and MCA from basilar artery via the p-com artery.

Type II: ACA is filled from the MCA on same side.

Type III: Anastomotic vessels from contralateral ICA supply the ACA.

Clinical features: Though asymptomatic most of the times, they may present as headache, symptomatic epilepsy, intracranial hemorrhage, cerebral ischemia. Associations are cardiac anomaly, arachnoid cyst, Klippel Trenanauy syndrome, cerebral hemiatrophy, neurofibramotosis, hemangi-

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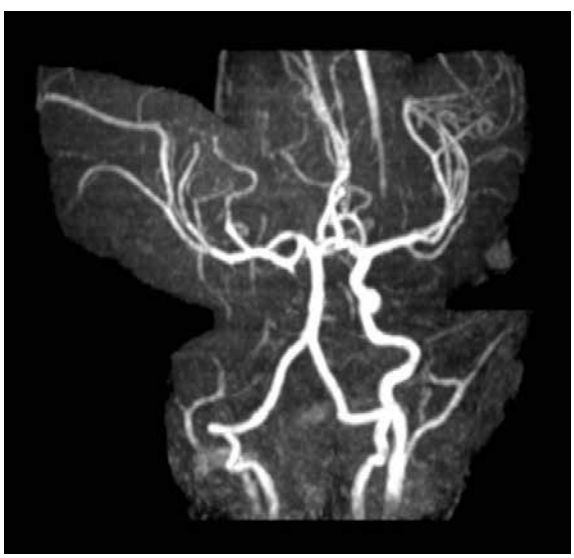
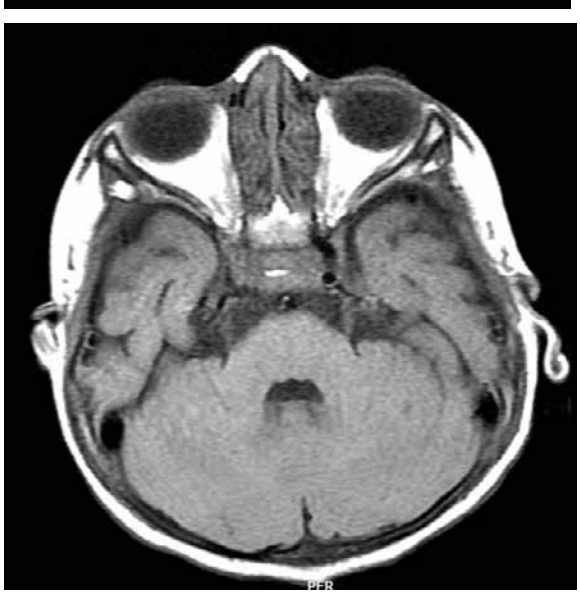
omas, angiofibromas, congenital polycystic kidney disease, external, middle ear malformations, Horner syndrome, and congenital hypopituitarism.

The diagnosis of congenital absence of ICA is confirmed by no visualization of ICA on angiography and absent carotid canals in the skull base on CT. This is to rule out functional/acquired stenosis of ICA.

Arterial lumen, wall thickness were decreased in ICA hypoplasia .But wall thickness is normal in functional stenosis.

Conclusions: ICA agenesis which is an uncommon congenital vascular anomaly is usually a silent condition, which can be associated with other serious malformations and disorders. Strict follow up is advised focusing at early identification of fatal changes. Abnormal vascular anatomy has to be carefully investigated prior to go for any endovascular surgery for ruptured coexisting aneurysm because any iatrogenic manipulation may disturb compensatory collateral circulation and induce irreversible neurological deficit. Combined use of MRI, MRA and CT scanning of base of skull and head are the necessary imaging studies. Diffusion and perfusion weighted sequences are done in search of acute or sub-acute infarctions.





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