Clinical Brief

Congenital Hypoplasia of the Internal Carotid Artery

Padam Yadav, Ajay Kumar, Arvind Saili and Vikram Datta

Department of Pediatrics Kalawati Saran Children Hospital and Lady Hardinge Medical College, New Delhi, India

ABSTRACT

A newborn presented at 14 hrs of age with right sided clonic seizures and shrill cry. Magnetic Resonance Imaging of the brain showed left cerebral hemiatrophy with cystic changes in left fronto-parietal lobe and parasylvian region. The Magnetic Resonance Angiography revealed hypoplasia of left supraclinoid Internal Carotid Artery and hypoplasia and irregularity of vessel wall affecting the left Middle Cerebral Artery. Such an early presentation of this rare disorder has not been reported previously. Recognition of this anomaly has important implications during carotid and trans-sphenoidal surgery, in thromboembolic disease, and in the surveillance and detection of associated cerebral aneurysms.**[Indian J Pediatr 2009; 76 (10) : 1061-1062]** *E-mail: ajayk5@sify.com, ajayk5@yahoo.com*

Key words: *Carotid hypoplasia; Cerebral hemiatrophy*

CONGENITAL HYPOPLASIA OF INTERNAL CAROTID ARTERY

Hypoplasia of the internal carotid artery (ICA) is a rare congenital anomaly. ICA hypoplasia presenting in the neonatal period has not been described before. We report a newborn with carotid artery hypoplasia that presented with seizures on day one of life.

REPORT OF CASE

A full term male baby was born by normal vaginal delivery to 25-yr third gravida mother. Baby cried immediately after birth (Apgar Score 8 and 9 at 1, and 5 minutes). There was no consanguinity or family history of malformations. His birth weight was 3.0 kg, height 50.5cm and head circumference 35cm. At 14 hrs after birth the baby developed right-sided clonic seizures associated with shrill cry. Seizures were treated effectively with injectable phenobarbitone. He received injection ceftriaxone and amikacin for suspected infection, as the screen for sepsis was positive. Cerebrospinal fluid examination did not reveal any meningitis and blood culture was sterile. There was no neurological deficit. Eye examination including fundus and optic disc was normal. In view of first day onset and focal nature of the seizures the baby was taken up for Magnetic resonance imaging (MRI). MRI of the brain showed evidence of hemiatrophy of the left cerebral hemisphere with focal areas of cystic encephalomalacia in the left frontoparietal lobe and parasylvian region. Sella turcica, pituitary stalk, body and hypophysis were normally visualized. Threedimensional time of flight magnetic resonance angiography (3D TOF MRA) of the intracranial vessels revealed hypoplasia of the left supra clinoid ICA and hypoplasia and irregularity of vessel wall affecting the left middle cerebral artery (MCA) (Fig. 1). There was also non-visualization of insular and opercular branches of left MCA. Baby improved and was discharged on fourteenth day of life on oral phenobarbitone. Follow up at four weeks showed a well thriving baby with no recurrence of seizures.

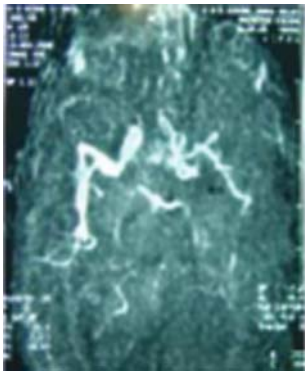

Fig. 1. MRA showing hypoplasia of the supra-clinoid part of the ICA and MCA.

Correspondence and Reprint requests : Dr Ajay Kumar, A-154, Ashok Vihar-1, Delhi-110052, India.

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DISCUSSION

Conventional angiography, CT angiography, MRI, MRA and single photon emission computed tomography (SPECT) could provide excellent description of extra and intracranial vascular abnormalities. In most of the reported cases no evidence of brain lesion or cerebral perfusion defects were shown despite the markedly altered anatomy.[1] Absence of any collateral in the presented case probably was responsible for such early presentation and extensive findings on MRI. To the best of our knowledge such an early presentation has not been reported previously.

While the literature supports a nearly 3:1 left sided preponderance of ICA absence[2] there is no side predilection when absence of the ICA is associated with an intercavernous anastomosis.[3] Congenital absence may be unilateral or bilateral, although the unilateral variety is distinctly more common.[4] The present case had left sided hypoplasia of ICA. There may be complete absence of the ICA or a portion of the ICA may be missing. A tiny fibrous band may be the only remnant of the ICA in case of aplasia and angiography alone may be incapable of differentiating it from agenesis. Evaluation of skull base for the presence or absence of the carotid canal may be required for distinguishing aplasia from agenesis.[5] Carotid canal though small was visible in the present patient on MRI there by ruling out agenesis.

Recognition of this anomaly becomes important in thromboembolic disease, as emboli in one cerebral hemisphere may have their origin from the normal but atheromatous contralateral common or internal carotid artery through collaterals. Similarly while planning surgery it may be possible that both the cerebral hemispheres are dependant upon that single nonhypoplastic carotid. Similarly collateral may have an implication in trans-sphenoidal surgery.[6]

CONCLUSION

Agenesis, aplasia and hypoplasia of the ICA are rare anomalies. Many of these cases remain asymptomatic and go undetected. Their recognition is important, as they may have associated aneurysm and implication in the setting of surgery or thromboembolic disease. It may, though rare, should be considered in the differential diagnosis of cerebral hemiatrophy and seizures in a neonate.

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