

Unilateral congenital hypoplasia of the internal carotid artery in a newborn: a rare case report

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Abstract

Hypoplasia of one or both internal carotid artery (ICA) is a rare congenital developmental abnormality. The early neurological presentation of this disorder is rare because many of these cases remain asymptomatic and go undetected due to the presence of collateral vessels. We describe a newborn that presented with seizures at 27 hours after birth. Extended ischemia of the right hemisphere was observed on computed tomography (CT), while the 3D MIP reconstruction showed hypoplasia of right internal carotid artery. After about 3 weeks, the rapid improvement of the newborn's cerebral ultrasound and EEG allowed to discontinue corticosteroid and sedative therapy. The infant was discharged after 40 days of life in good clinical condition.

Keywords

Internal carotid hypoplasia, neonatal ischemic stroke, neurologic signs, congenital abnormalities, brain computed tomography, angio-MDCT.

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Introduction

Hypoplasia of the internal carotid arteries (ICA) is a rare arterial anomaly that can interest one or both arteries. The diagnosis is based on its angiographic characteristics and is often incidentally discovered. In adults, this is due to sufficient cerebral collateral circulation through the circle of Willis [1]. The probable cause is an insult to the embryo during its early development. Agenesis, aplasia and hypoplasia of ICA occur in less than 0.01% of the population [2]. During the embryonic period several developmental anomalies of the ICA can occur. ICA agenesis or hypoplasia are due to an improper development of one of seven different embryologic segments that form the ICA. ICA agenesis is associated to the absence of petrous bony carotid canal, while ICA hypoplasia is associated to a smaller size of the canal [3]. Three major collateral pathways to the anterior cerebral circulation have been described. The most common collateral pathway is through the enlarged communicating artery; the others types of collateral pathways are posterior through anastomosis between the external carotid artery and the internal carotid artery at the skull base [4]. Most patients with this anomaly are asymptomatic and this is due to sufficient collateral circulation [4]; some patients present with recurrent headache, blurred vision and convulsions.

The clinical manifestations in childhood are rare and only one case has been previously reported in the neonatal period [5]. We report a case of severe unilateral ischemic stroke in a full term newborn who was diagnosed hypoplasia of the ipsilateral internal carotid artery with neuroimaging.

Case report

A full term male baby was born at 39 weeks by vaginal delivery to a 39-year third gravida mother. The mother had normal pregnancy and no family history of malformation; the previous two children had not had neonatal problems and were in good health. The baby was born with cord around his neck but he cried immediately after birth. Apgar score was 8 and 9 at 1 and 5 minutes after birth, respectively. His birth weight was 3.650 kg, height 51 cm and head circumference 34 cm. Neurologic examination and reflexes were normal at birth. At 27 hours after birth the baby suddenly developed clonic seizures associated with cry, for this reason he was transferred to the NICU. Laboratory data

showed a normal cell count, abnormal C reactive protein (CRP 25 mg/l); he received ampicillin and gentamicin for suspected infection. Blood culture and cerebrospinal fluid examination were negative for bacterial infection. Seizures were gradually sedated with injectable phenobarbitone and diazepam. The brain echographic imaging showed diffuse cerebral edema. A brain CT scan (**Fig. 1**) and an angio-MDCT of epiaortic and intracranial circle were performed on the third day of life. Moreover 3D MIP vascular reconstructions were obtained from the axial images. Extended ischemia of the right hemisphere with no alteration of frontal and occipital lobes was observed on CT. The 3D MIP reconstruction showed hypoplasia of right internal carotid artery (2 mm lumen diameter), starting from its origin to the intrapetrous segment; there was no opacification of the clinoid segment. The right carotid canal was normal (**Figures 2-5**). A diagnosis of right internal carotid hypoplasia was done and, at the same time, the agenesis was excluded. The intracranial vascular study showed regular contrast opacification on right posterior cerebral artery and the riabilitation of the right cerebral artery from the anterior communicating artery. The right middle cerebral artery supplying the ischemic area was not opacified (**Figures 6-8**).

After this diagnosis corticosteroid therapy (dexamethasone 4 mg/kg/day) was promptly ini-



Figure 1. Axial image: extendend ischemia of the right hemisphere.



Figure 2. 3D MIP reconstructions: hypoplasia of right internal carotid artery (arrow).

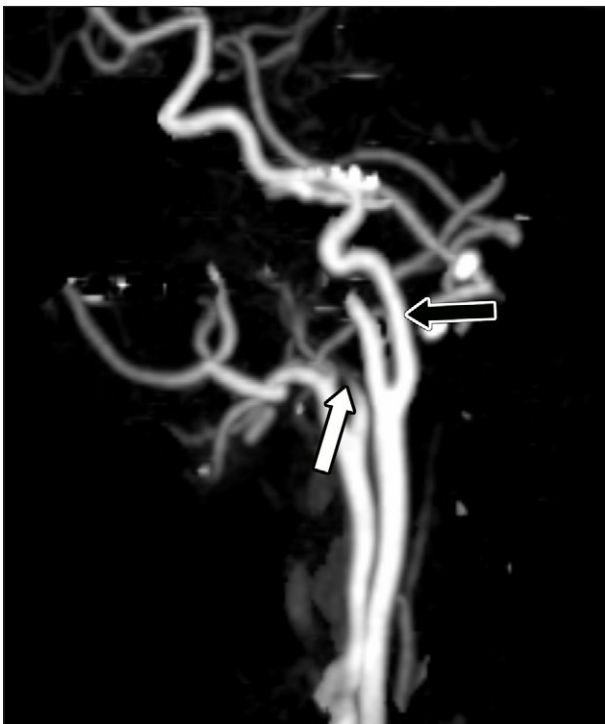


Figure 3. 3D MIP reconstructions: hypoplasia of right internal carotid artery (white arrow) and regular opacification of left internal carotid artery (black arrow).

tiated in order to reduce the cerebral edema and continued with phenobarbital. After about 3 weeks, the rapid improvement of cerebral ultrasound and EEG allowed to discontinue corticosteroids and

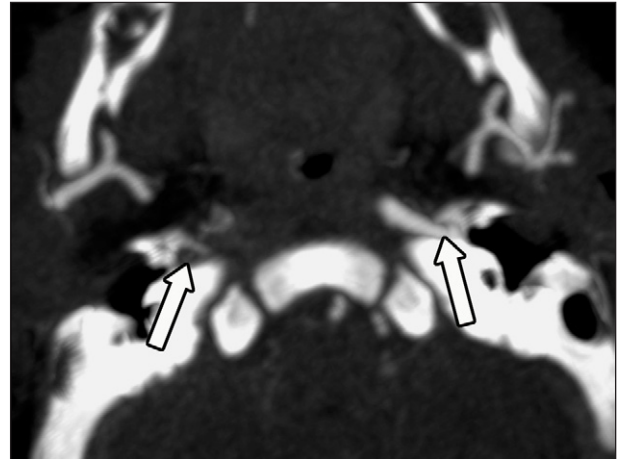


Figure 4. Axial CT scan of the skull base shows a normally developed right and left carotid canal (arrows).

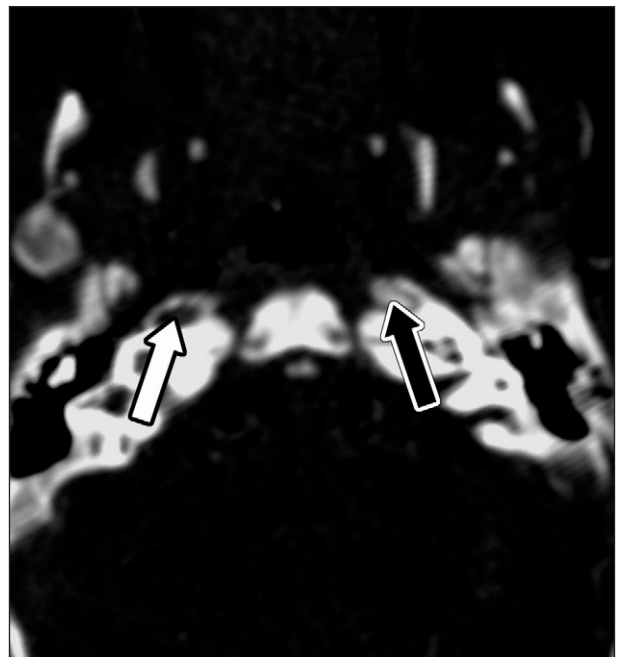


Figure 5. Axial image from a CT angiogram shows hypoplasia of right internal carotid artery (white arrow); regular opacification of left internal carotid artery (black arrow).

sedative therapy. The infant was discharged after 40 days of life in good clinical condition. The neonatal study of thrombophilia profile of the newborn showed *MTHFR* A1298C/A1298C mutations with homocysteine normal level. The echocardiogram was normal and embolism was excluded. The MRI control, performed before discharge, showed an extensive damage of the side of right hemisphere. The follow-up of newborn, currently available up to the age of 6 months, shows a slight hypomobility of the side left already in physiotherapy treatment

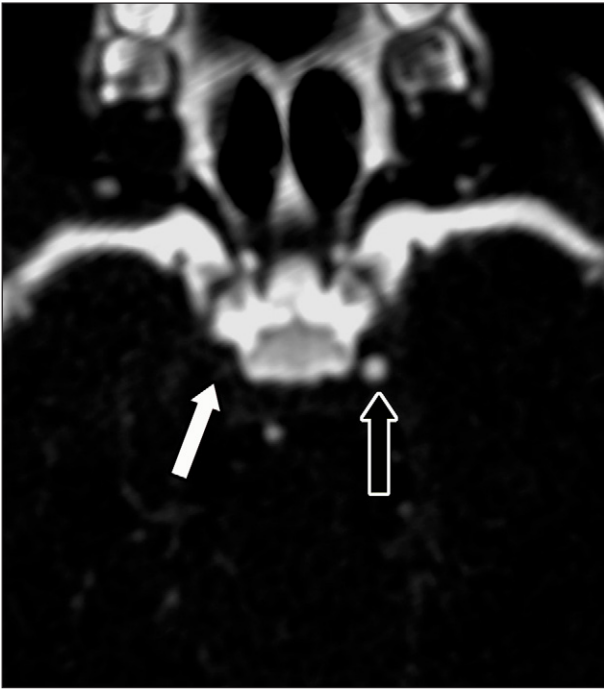


Figure 6. Axial image from a CT angiogram shows no opacification of the suvraclinoid segment of right internal carotid artery (white arrow); left internal carotid artery (black arrow).

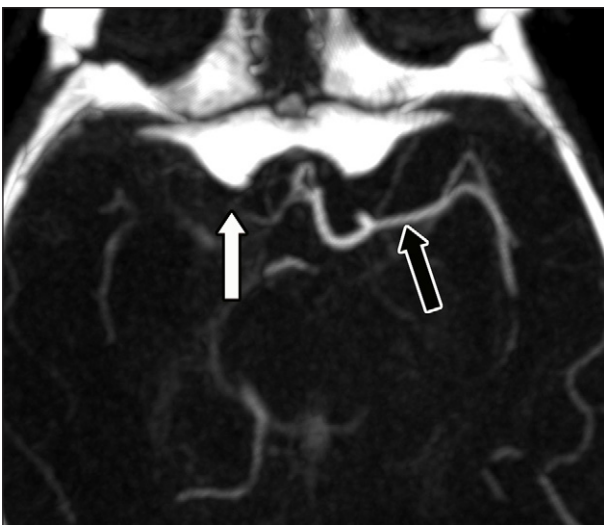


Figure 7. Absent opacification of right middle cerebral artery (white arrow) and regular opacification of left middle cerebral artery (black arrow).

and EEG abnormalities compatible with West syndrome.

Discussion

The hypoplasia of ICA is part of the spectrum of “absence” which includes agenesis, aplasia and



Figure 8. 3D MIP reconstruction shows regular opacification of right posterior cerebral artery (white arrow) and riabilitation of the right anterior cerebral artery (black arrow).

hypoplasia. In agenesis and aplasia the internal carotid artery is completely absent.

In hypoplasia, the development of the internal carotid artery is incomplete. Although an exact cause of these developmental anomalies has not been established, they represent the sequelae of an insult to the developing embryo. Development of the carotid canals occurs in the presence of the embryonic internal carotid artery during early gestation. A small or absent carotid canal indicates a congenital internal carotid abnormality. The acquired causes of a significantly narrowed internal carotid artery (chronic dissection, severe atherosclerosis and fibromuscular dysplasia) were excluded by neuroimaging study.

In our case the CT scan and angio-MDCT of epiaortic and intracranial circle and 3D MIP reconstruction showed hypoplasia of right internal carotid artery (2 mm lumen diameter) starting from

its origin to the intrapetrous segment. Extended ischemia of the right hemisphere was caused by a sudden blood hypoperfusion through a congenitally narrow vessel. The hypoperfusion may have been caused by the cord around the neck at birth, or a significant decrease in blood pressure caused by vigorous crying a few hours after birth. In fact, perinatal arterial ischaemic stroke (PAS) is defined as a group of heterogeneous conditions in which there is a focal disruption of cerebral blood flow, normally secondary to arterial or cerebral venous thrombosis or embolization, between 20 weeks of foetal life through 28th post-natal day and confirmed by neuroimaging or neuropathological studies [6-9]. In this case thrombosis and embolization were excluded because the neuroimaging remained unchanged in the remote control and ischemic stroke may have been caused by an acute arterial hypoperfusion. In addition, the thrombophilia profile was not compatible with the most common thrombophilia traits found in neonatal arterial ischemic stroke or cerebral sinovenous thrombosis [10].

Conclusion

The neurological manifestation of hypoplasia of ICA is a rare condition, especially in newborns. The congenital nature of the pathology is represented by a small or absent carotid canal. The exact mechanism leading to agenesis or hypoplasia of the ICA remain unknown, but some investigators have suggested the occurrence of various disturbances during early development. In the present newborn, hypoplasia of ICA was right sided and caused an ischemic stroke due to an imbalance of arterial blood flow shortly after birth. We can hypothesize that the timing of the embryo injury occurred before 24 mm stage of embryonic development. In fact, only if the embryo injury occurs after this stage collateral flow can develop. This also explains why in this case the collateral circle was not present. Patients with hypoplasia of ICA generally have no neurological symptoms because the cerebral blood flow system can be aided by collateral blood flow. The newborn showed symptoms related to cerebral ischemia, because of decreased blood flow. The damage can not be related to imbalance of fluxes during the delivery because hypoplasia of right internal carotid artery (2 mm lumen diameter) would not be present at the 3D MIP reconstruction. Hypoplasia

of ICA should be part of the differential diagnosis of neonatal cerebral ischemic stroke and should be confirmed by neuroimaging as soon as possible.

Declaration of interest

The Authors declare that there is no conflict of interest.

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