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Case report

Diagnosis of congenital absence of internal carotid artery by power angio sonography

E. Harps, K. Helmke

Universitätskinderklinik Eppendorf, Martinistrasse 52, D-20246 Hamburg, Germany

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Abstract. This case report describes aplasia of the internal carotid artery (ICA) in a preterm infant. The collateral circulation could be mapped with power angio mode (PAM) and was confirmed by conventional angiography. In the literature, there is no case of ICA aplasia diagnosed at this early age. PAM is a method for imaging infantile cerebral vessels as reliably as angiography.

Key words: Aplasia of internal carotid artery – Premature infant – Power angio mode

Introduction

Congenital absence of one or both internal carotid arteries (ICA) is a rare anomaly. Occasionally, this condition is coincidentally found on CCT or angiography. This anomaly is associated with hemorrhage, aneurysms, steal phenomena, and cerebral hemiatrophies [1–6]. It is mainly diagnosed by imaging techniques when neurological symptoms occur. If ICA lesions secondary to trauma, hypercoagulopathy, or atherosclerosis can be excluded, congenital absence of the ICA is likely [7–9]. Although the manifestation of this condition in infants and children is rare [4] and the onset of neurological symptoms slow because they require additional disease, there is a particular interest in screening patients in order to detect organic manifestations early in their course. Thus, the possibility and benefit of intervention like extracranial-intracranial bypass or endovascular occlusion of aneurysma can be discussed. An easily performed imaging procedure is therefore warranted.

The following case report is of a preterm infant with congenital absence of the ICA. The diagnosis and mapping of the intracerebral circulation with power angio mode (PAM) is described.

Correspondence to: E. Harps

Case report

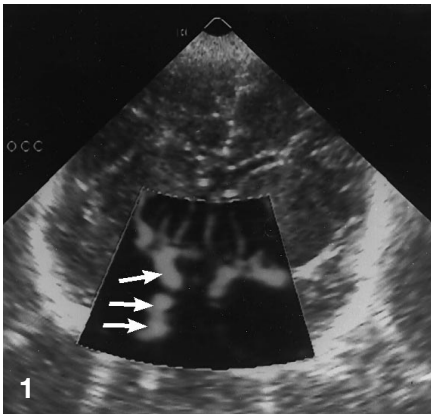
In vitro fertilization led to a triplet pregnancy. The retarded growth of the first triplet was the reason for cesarean section in the 32nd week of gestation. Four days prior to this, betamethasone was administered in order to accelerate lung maturation. In the following report, only the data of the second triplet are reported.

The female preterm infant had an Apgar score of 6, 7 and 8 at 1, 5, and 10 min, respectively, and umbilical artery blood pH of 7.32, birth weight of 1720 g, body length of 44 cm, and head circumference of 30 cm. Amniotic fluid was clear. Increasing respiratory insufficiency required intubation and intermittent mandatory ventilation. Treatment with surfactant was not felt to be necessary. The initial chest X-ray showed findings compatible with second-degree respiratory distress syndrome [10]. Three days post partum, mechanical ventilation was discontinued and theophylline was used to treat the apnea of prematurity.

Physical examination was within normal limits given the gestational age of 32 weeks. Interestingly, there were no stigmata of malformation. Head circumference and body size increased normally.

Ultrasound

To exclude an intracranial hemorrhage following several episodes of apnea, a cranial ultrasound was performed. Ventricular dimensions and parenchyma were normal. Decreased pulsation of the ICA in the left parasellar area, however, was noted. Color-coded imaging and PAM were therefore performed, revealing absence of the infraclinoidal portion of the left ICA. Only the supraclinoidal part of the left ICA together with the left ophthalmic artery could be visualized. The collateral circulation was assessed by PAM. Blood flow from the right ICA to the left anterior cerebral artery (ACA) was shunted via the anterior communicating artery of the circle of Willis (CW). Also, there was retrograde



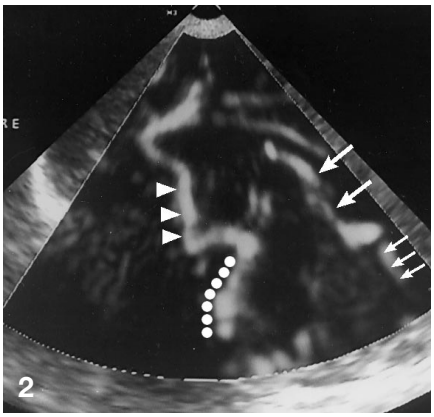
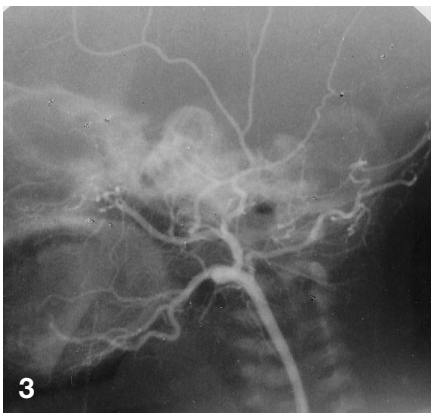


Fig. 1. Coronary scan in PAM: the whole intracranial part of the ICA is present on the right side *(arrows)*. Only the supraclinoidal part of the left ICA is seen

Fig. 2. Sagittal scan in PAM: the basilar artery *(circles)* can be visualized in the typical area, as can the left PCA and the feeding of the left ACA *(triangles)*. Additionally, the inferior sagittal sinus and rectus sinus *(arrows)* are shown



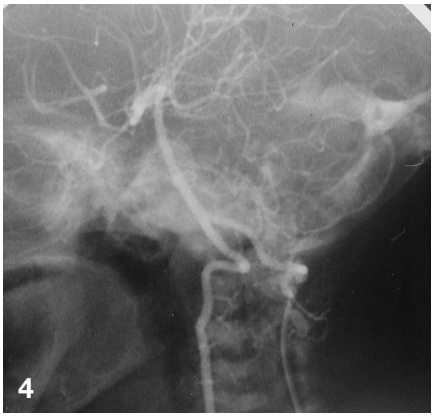


Fig. 3. Left carotid angiography: the contrast medium was injected through a catheter in the umbilical artery, the tip of which was placed in the left common carotid artery. The external carotid artery is also visualized here. However, no origin of the left ICA can be found

Fig. 4. Angiography of the left vertebral artery: after placing the catheter in the left vertebral artery, contrast medium was injected. There was also inverse flow in the right vertebral artery. Blood flow is through the basilar artery into the left PCA and the left MCA and the ACA

blood flow from the basilar artery to the posterior communicating artery (PCA) of the CW, to the left middle cerebral artery (MCA), and also to the left ophthalmic artery (Figs. 1, 2). Flow direction was based upon Doppler spectral analysis.

Cerebral angiography

Arterial access was achieved by catheterization of the umbilical artery. The catheter was placed in the left common carotid artery and contrast medium was injected. The angiogram showed the left external carotid artery, but not the ipsilateral ICA: no rudimentary debranching ICA could be visualized (Fig. 3). Subsequently, the tip of the catheter was placed in the left vertebral artery. The contrast medium filled the basilar artery, both posterior cerebral arteries, and the left PCA. The angiogram also showed the left MCA and the left ophthalmic artery (Fig. 4). There was no sign of aneurysm at this point. According to the classification of Lie [11], this is a case of ICA aplasia type A.

X-ray of the skull base

A malformation of the carotid canal (CC) in the skull base could not be documented by X-ray due to the physiological lack of calcification at this age.

The affected triplet was discharged 41 days post partum in good health. Both of the other triplets appeared to be in stable condition. Cranial sonography and color-coded imaging of the intracranial vessels did not reveal any malformation.

Discussion

Absence of the ICA is a rare vascular malformation. It is rarely diagnosed in childhood. A review of the literature suggests that this malformation has not previously been described in a preterm infant [4, 9, 12].

Agenesis of the ICA is defined as a combination of the absence of ICA, hypoplasia (or aplasia) of the CC, and persistence of fetal arteries. Agenesis is caused by developmental failure during the first month of pregnancy. Aplasia is defined as a disorder of later development, when also the CC is developing.

The left ICA is affected three times as often as the right. Aplasia may be the result of an extreme intrauterine position and local pressure [13, 14]. In this case, a correlation between the absence of the ICA and the triplet pregnancy is possible.

It was not possible to assess the CC in an X-ray of the skullbase because of the age-related lack of calcification. A cranial CT was not obtained, to avoid further radiation exposure, and there was no therapeutic implication. In our experience, structures like the CC only

change size or shape when challenged by growth pressure. Differentiation between aplasia and agenesis can therefore be difficult. In this case, however, an aplasia of the ICA is likely, especially since angiography did not show any persistent fetal arteries.

Absence of the ICA is associated with the development of aneurysms in the CW and consecutive bleeding in 25–34 % of detected cases. However, these complications are not typical in childhood [13]. The literature describes different symptoms due to malperfusion of the auditory system or the pituitary gland [4, 7]. Moreover, cerebral hemiatrophy is possible [1]. Early diagnosis facilitates optimal intervention before neurological disorders become serious.

In this case, the diagnosis of the absence of the left ICA could be established with PAM. The lack of experience with PAM, a very new method, was the reason for additional conventional angiography, which, however, did not add any further information. PAM is a new ultrasound technique, which utilizes only the intensity of reflection of a Doppler signal for imaging. In contrast to color-coded imaging and flow velocity techniques, PAM is not biased by the angle between flowing particles and the Doppler signal [15–17]. With PAM, the diagnosis of absent ICA could be established better than using color-coded imaging, because the latter's dependence on the angle makes examination of the carotid sinus difficult. For a similar reason, it was not possible to detect precisely all branches of the CW using color-coded imaging only. The PCA and its flow direction can be visualised from an occipital view. However, in this case, the angle between the probe and the CW was unfavourable. Knowledge of the reverse flow direction between the PCA and the ophthalmic artery allowed the conclusion that a collateral circulation was present.

PAM is useful for imaging intracranial vessels [18–20]. The thickness of the calvarian bone is the limiting factor. However, it is possible to examine the MCA with PAM using a 2-MH probe in older patients as well.

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