Radiology Case Reports 16 (2021) 2530–2533

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Case report

Prenatal diagnosis and postnatal management of congenital unilateral hydrocephalus for stenosis of the foramen of Monro ☆,☆☆

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ARTICLE INFO

Article history:
Received 27 May 2021
Revised 5 June 2021
Accepted 6 June 2021

Keywords:
Neurosurgery
Prenatal diagnosis
Hydrocephalus
Ultrasound
MRI

ABSTRACT

Congenital hydrocephalus and ventriculomegaly can be diagnosed reliably with prenatal ultrasound and Magnetic Resonance Imaging (MRI). Unilateral hydrocephalus is uncommon, and the prognosis depends on etiology and postnatal management. Here, we present a case of a 32-years-old woman with prenatal diagnosis of unilateral hydrocephalus associated with stenosis of the foramen of Monro. Unilateral hydrocephalus is an uncommon feature that can be detected prenatally on ultrasound and MRI. In case of isolated stenosis of the foramen of Monro prognosis is good.

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Introduction

Ventriculomegaly is one of the most common brain abnomally diagnosed during the fetal life [1]. It is characterized by dilatation of the lateral ventricles of the brain greater than 10 mm at the level of the atria [2]. Ventriculomegaly is classified according to the degree of dilatation as mild (10-12 mm), moderate (12-14 mm) or severe (≥15mm) [2].

There are several causes of ventriculomegaly, with four main factors that can cause enlargement of the ventricles: a problem that prevents cerebrospinal fluid (CSF) from cir-

☆ *Acknowledgement*: No financial support was received for this study
☆☆ *Competing interests*: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.
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https://doi.org/10.1016/j.radcr.2021.06.011
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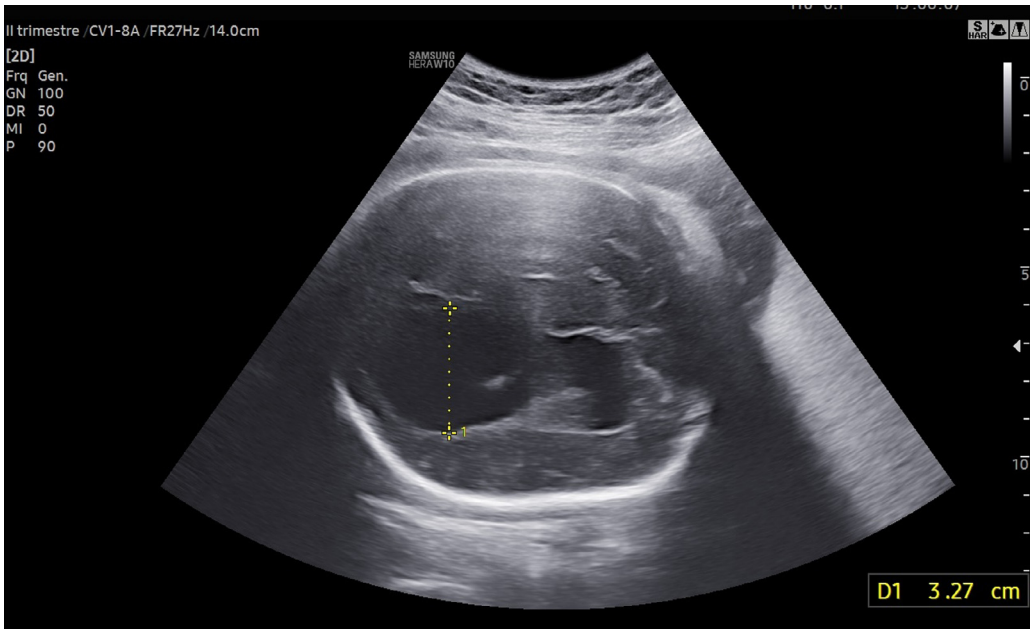

Fig. 1 – Prenatal ultrasound scan.

culating, which causes hydrocephalus; a minor imbalance in fluid circulation and absorption; defects in brain development; or damage or loss of brain tissue. According to the number of ventricles involved, the hydrocephalus can be classified as unilateral or monoventricular (one ventricle involved); biventricular (two ventricles involved); triventricular (three ventricles involved); or tetraventricular hydrocephalus (all four ventricles involved).

The term unilateral hydrocephalus or monoventricular hydrocephalus specifically defines a single ventricle obstruction that by anatomical reasons can only indicate the involvement of the lateral ventricle, given that an obstruction of the third ventricle or fourth ventricles will necessarily cause biventricular and triventricular hydrocephalus, respectively [3]. Unilateral hydrocephalus is uncommon, and the prognosis depends on etiology and postnatal management

Here, we present a case of prenatal diagnosis of unilateral hydrocephalus associated with stenosis of the foramen of Monro. Postnatal management is also reported.

Case report

Prenatal diagnosis and antenatal management

A 23-year-old prima gravida woman was referred to our institution at 31 weeks of gestation for suspected fetal brain anomaly. Second trimester ultrasound scan was regular. Ultrasound examination showed severe unilateral hydrocephalus of 32.7 mm (Fig. 1). Multiplanar ultrasound examination through axial, coronal and sagittal views of the fetal brain using high-resolution transvaginal probe showed no associated anomalies [4]. Stenosis of the foramen of Monro was suspected and confirmed by intrauterine Magnetic Resonance Imaging (MRI).

The woman underwent planned cesarean delivery at 34 weeks and 4 days, and a 2,650 g-sized male infant was delivered with APGAR score of 8 and 8, at 1 and 5 minutes, respectively. The mother postoperative course was uncomplicated, and she was discharged at day 3 after delivery.

Postnatal management

The neonate was referred to pediatric neurosurgeons. The infant was awake and spontaneous breathing; he presented bulging of the anterior fontanelle and enlargement of cranial sutures. No motor deficit was evident. Neonatal MRI confirmed unilateral hydrocephalus with enlarged right lateral ventricle, secondary to agenesis of the foramen of Monro and contralateral shift of the septum pellucidum (Fig. 2). On the second day of life the infant underwent surgery. A neuroendoscopic approach, through the right angle of the anterior fontanelle, was attempted. The enlarged right lateral ventricle was entered. Under magnetic neuronavigation guidance the two layers of the septum pallucidum were recognized and fenestrated, allowing communication between the two lateral ventricles. Furthermore, the membrane between the right lateral ventricle and the third ventricle was also fenestrated, creating a new foramen of Monro (Fig. 3). The procedure was uneventful, with disappearance of signs of intracranial hypertension. Post-operative MRI showed initial reduction of the size of the lateral ventricle and adequate communication between the two lateral ventricles at the level of the septostomy (Fig. 4). On 17th post-operative day, the patient was discharged with normal neurological examination.

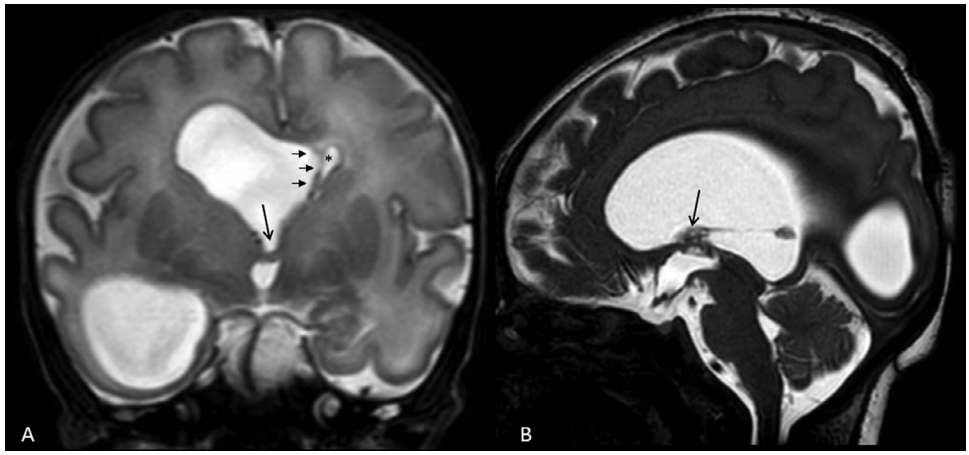

Fig. 2 – Preoperative T2 weighted MR images in the coronal (A) and sagittal (B) planes, showing right monolateral hydrocephalus, with absence of the foramen of Monro, replaced by a thick membrane (arrow). The septum pellucidum is deviated (small arrows) and the left ventricle is compressed (*).

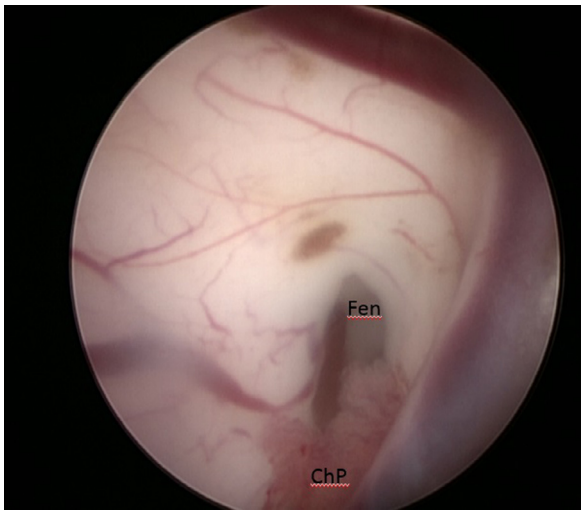

Fig. 3 – Intraoperative image following fenestration (*Fen*) of the thick membrane separating the right lateral ventricle and the third ventricle, just in front of the choroid plexus (*ChP*).

Discussion

The two foramina of Monro, also called interventricular foramina, connect the paired lateral ventricles with the third ventricle at the midline of the brain. Foramina of Monro allow CSF to reach the third ventricle and the rest of the ventricular system. If they are narrowed or blocked, the ipsilateral ventricle enlarges. Obstruction is usually due to acquired conditions, such as thalamic and intraventricular tumors, colloid cysts, ventriculitis, vascular malformations and inflammation [5]. However, the foramen of Monro, might be congenitally atretic, occluded by a membrane or malformed, resulting in congenital unilateral hydrocephalus [6]. This condition is very uncommon, and very few cases have been published in the literature [3]. Several associated anomalies have been described, such as agenesis of corpus callosum, hemihypertrophy and frontoethmoidal encephalocele. If no associated anomalies are present, unilateral hydrocephalus has a good prognosis even when discovered in utero: adequate treatment of CSF disturbance usually allows good neurological outcome [7,8].

Clinically, progressive dilatation of only one lateral ventricle may be associated with unilateral long-tract signs or focal brain symptoms, such as hemiparesis and aphasia. However, in small children, symptoms and signs of diffuse intracranial hypertension may be the only clinical finding.

Treatment options include: shunting, endoscopic fenestration of the septum pellucidum, and foraminoplasty of Monro [6]. Shunting has been used in the past as the classic operative procedure. However, because shunt surgery is burdened by high risk of infective and mechanical complications, now days neuroendoscopic procedures are considered the treatment of choice in all forms of obstructive hydrocephalus, including monolaterl hydrocephalus, caused by Mnor obstruction [9].

An isolated lateral ventricle can be put in communication with the contralateral ventricle, by fenestrating the septum pellucidum (septostomy), or with the third ventricle, by fenestrating the obstructed foramen of Monro (foraminoplasty). Foraminoplasty, restoring the natural flow of CSF from the lateral to the third ventricle, appears, at least theoretically, the procedure to be preferred. However, an obstructed foramen may be difficult to recognize during surgery: thalmostriate vein and the choroid plexus are the most important landmarks. Navigation system may be helpful. Septostomy is a valid alternative, in cases with difficult anatomy for foraminoplasty. Prerequisite for successful septostomy is the patency of the contralateral foramen of Monro. In case of bilateral obstruction of the foramina of Monro, in fact, septostomy should be always associated with foraminoplasty of at least

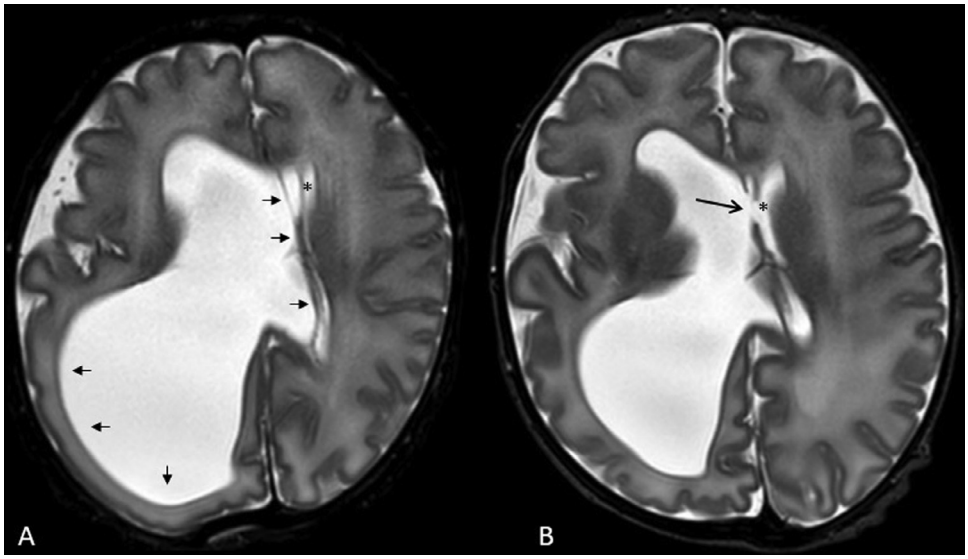

Fig. 4 – Preoperative (A) and post-operative (B) axial T2 weighted MR images. Note the reduction in size of the right lateral ventricle and of the mass effect on the septum pallucidum and brain parenchima (small arrows); the patency of the septostomy (arrow) and the mild expansion of the left lateral venricle (*).

one foramen of Monro. In our patient the foramen of Monro was not recognizable, thus we performed a septostomy; however, because septum pellucidum was very small and deviated toward the contralateral side, the fenestration appeared to be not enough large. Therefore, with the help of neuronavigation we fenestrated also a membrane that separated the lateral from the third ventricle, creating a new Monro foramen.

According to Aldana et al. [10]. endoscopic treatment of unilateral hydrocephalus is effective in 81% of cases. If the septostomy remains patent for 6 months postoperatively, then failure is unlikely to occur.

Conclusions

In summary, unilateral hydrocephalus is an uncommon feature that can be detected prenatally on ultrasound and MRI. In case of isolated stenosis of the foramen of Monro prognosis is good.

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