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CASE REPORT

Hemorrhage in the cavum septi pellucidi: Description of a newborn with macrocrania

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Abstract:

The Cavum Septi Pellucidi represent the normal transitional finding from 24 to 36 weeks post-conceptional age in prenatal ultrasonography. Usually it closes between the second and sixth month of life. Haemorrhage of CSP is a rare entity. Only two cases of hemorrhage in the abnormal dilated CSP have been previously reported in full term neonate. We describe a case of Cavum Septi Pellucidi and lateral ventricles haemorrhage associated with familiar macrocrania, in a full term neonate. Our baby was discharge at hospital a 16 days of life in good clinical conditions and the MRI at 3.5 months of life showed improvement brain anatomy.

Keywords: *Cavum Septi Pellucidi (CSP); intraventricular haemorrhage; Septum Pellucidum (SP)*

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Case report

A male baby was born at 39 weeks of gestation by elective caesarean section due to macrocrania. Because the mother had experienced two previous spontaneous abortions, she had assumed acetylsalicylic acid for the first two weeks of pregnancy and, after that 4,000 UI of enoxaparin daily until the 13th week. The father reported a family history of macrocrania. The prenatal ultrasonographic exam at 32+4 weeks of gestation showed cephalic diameters at the upper limit of normal range. At birth, Apgar score were 7 and 8 at 1 and 5 min respectively, birth weight was 3520 g (50th percentiles of WHO curves), length 52 cm (75th - 90th percentile) and cranial circumference 37.5 cm (> 97th percentile). The anterior fontanel was full, but not tense. General physical examination was normal for gestational age. Neurologic examination revealed slightly assial hypotonia, partial lacking of stimuli response, normal presence of newborn reflexes. Screening cranial ultrasound (CS) performed through the anterior fontanel on the second day of life demonstrated the presence of large blood clots in the dilated CSP and in the lateral ventricles (grade III hemorrhage) (figure 1). Computed Tomography (TC) scan confirmed the haemorrhage in the CSP and lateral ventricles. Magnetic resonance angiography (figure 2) excluded aneurysm, thrombotic occlusion, arteriovenous malformation and other congenital

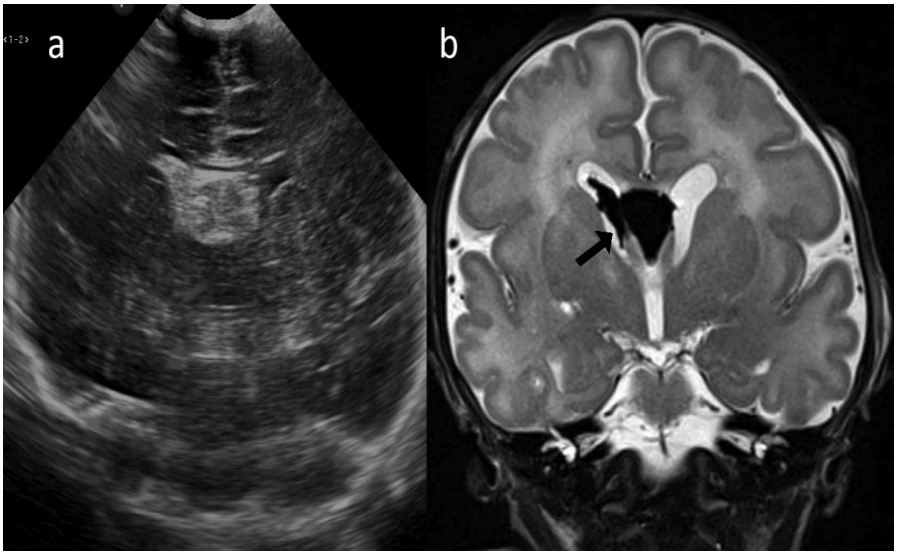

Figure 1. Coronal ultrasound scans of the head (a) and MRI coronal T2w image (b) at diagnosis. The images show an intraventricular hemorrhage and a dilatated cavum septum pellucidum with a large clot inside (arrow).

anomalies. Screening for thrombophilia showed that a heterozygous methylenetetrahydrofolate reductase mutation. Subsequent CS revealed a slight dilatation of the lateral ventricles and a progression of hemorrhage that involved the third and the fourth ventricle. Pre-discharge CS demonstrated progressive resolution of the blood clots. Clinical course was uncomplicated and the baby was discharge at 16 days of life in good clinical conditions. At 3.5 months, Magnetic Resonance Imaging (MRI) showed haemosiderin deposits on the fourth ventricle, on the occipital horns, on the SP and on the ventricular walls. The ventricular spaces were still wide (figure 2). At 4 months of life the occipito-frontal circumference was 45 cm (> 97th percentile), but no obvious neurological abnormalities were present.

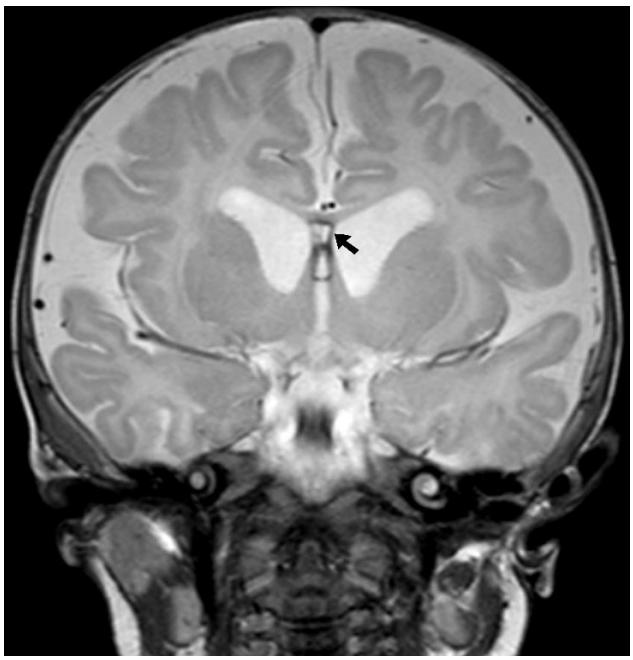

Figure 2. MRI obtained at 3.5 months of life showing the disappearance of the clot inside the cavum septum pellucidum and the decreased dilatation. Notice the persistence of the haemosiderin deposits on the septum pellucidum (arrow) and on the ventricular walls. The thickness of periventricular white matter is reduced, showing ventriculomegaly. The subarachnoid spaces are wide.

Discussion

Septum Pellucidum (SP) is a cerebral midline structure running as a sheet from the corpus callosum to the fornix and it is based on two vertically oriented leaflets, which separate the anterior horn of the two lateral ventricles [1]. The functional role of SP is still uncertain, but it appears to be involved in the limbic system and connected with commissural pathways and diencephalic system [2]. The development of the SP is linked closely to that of the corpus callosum, starts at nine weeks and is complete by approximately 20 weeks of gestation. At approximately the twelfth week of gestation the underlying portion of the commissural plate of the SP becomes thinned to form the two laminae delimitating two cavities called Cavum Septi Pellucidi (CSP) and Cavum Vergae (the

posterior extension of the CSP). CSP represent the normal transitional finding from 24 to 36 weeks post-conceptional age in prenatal ultrasonography and it is gradually reabsorbed starting from the sixth month of gestation. At birth, a small slit might persist in 60% of pre-term [3] and in 50% of full term infants [4]. Usually it closes between the second and sixth month of life [5], although is present in 12% of adults [6]. Generally there is no communication with the ventricular system and the subarachnoid spaces, though in 12% of the pediatric population there is a connection between CSP and lateral ventricles. The septum pellucidum walls of the CSP are richly vascularized [6] and the cerebrospinal fluid in the cavity comes from the ventricles trough the septal laminae. Parietal diameter, gestational age or birth weight do not correlate with the size of the CSP. Postnatal normal values in sonographic exam range from 2 to 10 mm in width and up to 12 mm in height [6]. CSP haemorrhage and dilatation are unusual and rarely reported [7]. We describe a case of CSP haemorrhage in a full term neonate with macrocrania.

Conclusions

Haemorrhage of CSP is a rare entity [8]. Only two cases are reported in the literature in full-term infants (one of them with Down's syndrome) [7]. Its pathophysiology is still unclear; it might be due to an over-distending and rupture of the septal veins of the abundantly vascularized septi pellucidi [9]. The delivery and the macrocrania (familiar origin), could have contributed of the septal vein rupture [6]. We think that macrocrania could be related with hemorrhage in our case, therefore the real explanation of that; nevertheless we have not evidence in literature, and at the same time it could be a chance coincidence. The presence of blood within the CSP and the lateral ventricles could be explained by the communication that might exist between the two structures. An alternative hypothesis is that the blood came from lateral ventricles to the CSP trough fenestration of the septal wall. The prognostic implications of hemorrhage of CSP are still unknown. The CPS is connected to the limbic system, which control emotional reactions, and its presence is associated with neuropsychiatric syndromes in adults [10]. The intraventricular haemorrhage adds a further variable and at present we do not know whether it will have neurological or neuropsychiatric consequences.

Declaration of Conflicting Interests

The authors declare they have no source of financial assistance nor potential conflicts of interest.

Ethical Approval

Written informed consent was obtained from the patient's parents for publication of this Case report and the accompanying images.

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