Case report

CORTICAL DYSPLASIA WITH SUBCUTANEOUS ANGIOMA AND DILATED DURAL VENOUS SINUSES

K. FUKINO, K. UMEOKA, T. KITAMURA, H. TAKAHASHI, A. TERAMOTO
Department of Neurosurgery, Nippon Medical School, 1-1-5, Sendagi, Bunkyo-Ku Tokyo, Japan

SUMMARY

We report a rare case with dilated dural venous sinuses, cortical dysplasia, and a subcutaneous angioma in the forehead. These lesions may be derived from some factors in the certain period of gestation, during which dural venous sinuses dilate due to increased intracranial pool of blood.

Key words: dural venous sinuses, development, cortical dysplasia, angioma, forehead.

RÉSUMÉ

Dysplasie corticale associée à un angiome sous-cutané et une dilatation des sinus veineux duraux
Nous présentons un rare cas de dilatation des sinus veineux duraux associée à une dysplasie corticale et un angiome sous-cutané du front. Certains facteurs lors de la période de développement où les sinus veineux se dilatent sous la pression d’une augmentation du volume de sang intracrânien peuvent être à l’origine de ces manifestations.

Mots-clés : sinus veineux duraux, développement, dysplasie corticale, angiome, front.

INTRODUCTION

The dural venous sinuses around the confluence of sinuses dilate from the 4th to the 7th month of gestation in the human fetus [1-5]. This dilatation results from the increased intracranial pool of venous blood from the rapidly growing cerebral hemisphere in the absence of the development of the sigmoid sinuses and the superior bulbs of internal jugular veins [6]. Besides the dilatation of the dural venous sinuses, the emissary veins also develop as the drains of the intracranial venous blood [6]. After the 7th month of gestation, the development of the sigmoid sinuses decreases the intracranial pool of venous blood and reduces the dural venous sinuses and the emissary veins. And the formation of the jugular bulbs from the jugular sinuses begins after birth [6].

We report a rare case of the combined abnormalities, that seem to be derived from some factors in the gestational age; the dilated dural venous sinuses thought to be the rests of the fetal dilatation of the sinuses, the cortical dysplasia in the right cerebral hemisphere, and a subcutaneous angioma in the right forehead that seems to result from the enlarged fetal emissary veins.

CASE REPORT

A 19-month-old male had a subcutaneous angioma in the right forehead since his birth. At 12 months old, left hemiparesis and slight mental retardation were pointed out, and magnetic resonance (M R) imaging demonstrated some abnormal findings. So he was admitted to our hospital for further examinations and treatment at 19 months old.

On admission, he could speak no word and showed slight motor weakness of his left upper and lower extremities. The subcutaneous angioma in the right forehead (figure 1) showed neither pulsation nor bruit. The skull under the angioma showed the irregular surface due to dilatation of the grooves for vessels there. Craniogram showed the striated shadows that coincided with the grooves. M R imaging revealed disorder of the gyral formation (cortical dysplasia) in the right cerebral hemisphere, dilatation of the dual venous sinuses, abnormal
Fig. 1. — The photo of the patient’s face. The subcutaneous angioma, seen in the right forehead, is becoming smaller than at birth.

Fig. 1. — Le patient vue de face. L’angiome sous-cutané du front a diminué depuis la naissance.

Fig. 2. — Magnetic resonance (MR) findings in a 19-month-old male with the combined abnormalities in the head. a) T1-weighted MR image, and. b) T2-weighted MR image, showing the cortical dysplasia in the right cerebral hemisphere. c) axial gadolinium-enhanced MR image showing dilated dural venous sinuses and abnormal vessels on the right frontal surface of the brain.

Fig. 2. — IRM d’un enfant de 19 mois ayant une association d’anomalies de la tête. (a) IRM T1, et (b) IRM T2, montrant une dysplasie corticale de l’hémisphère droit. (c) L’IRM axiale après injection de gadolinium met en évidence une dilatation des sinus veineux duraux et des anomalies vasculaires sur la surface cérébrale frontale droite.

Fig. 3. — Three-dimensional computed tomography (3D-CT) scan showing the remarkable dilatation of the confluence of sinuses and the right transverse sinus (a) and the subcutaneous angioma on the right frontal and parietal bones (b).

Fig. 3. — TDM tridimensionnelle montrant une dilatation importante de la confluence des sinus et du sinus transverse droit et a) un angiome souscutané au niveau de la partie droit du front et (b) des os pariétaux.

Fig. 4. — Cerebral angiograms showing no abnormal finding in the arterial phase and dilatation of dural sinuses and dilated emissary veins in the venous phase (a) lateral view, (b) anteroposterior view.

Fig. 4. — Pas d’anomalie au temps artériel de l’angiogramme cérébral et dilatation des sinus duraux et des veines émissaires au temps veineux. (a) de profil, (b) de face.
vessels on the right frontal surface of the brain (figure 2). Three-dimensional computed tomography (3D-CT) scan revealed the remarkable dilatation of the confluence of sinuses and the right transverse sinus, and the subcutaneous angioma on the right frontal and parietal bones (figure 3). Cerebral angiography showed no abnormal finding in the arterial phase, and dilatation of dural sinuses and emissary veins, and abnormal vessels in the venous phase (figure 4). A nd chromosomal analysis revealed no abnormal finding.

During 2 months after the admission no neurological deterioration was seen, while he came to speak some words and stand for several minutes. Therefore, he left the hospital, and he was being followed up as an outpatient.

DISCUSSION

According to the several previous studies on the development of the dural venous sinuses of the human fetus, the venous sinuses around the confluence of sinuses relatively dilate from the second half of the 4 th to the 7 th months of gestation [2-5]. This is the « physical » dilatation that results from the increased intracranial pool of venous blood due to the undevelopment of the sigmoid sinuses and the superior bulbs of internal jugular veins [6]. The increased intracranial pool of venous blood develops the emissary veins as well as the dural venous sinuses, for the emissary veins drain much of intracranial blood in the « physical » dilatation that results from the increased intracranial pool of venous blood due to the undevelopment of the sigmoid sinuses and the superior bulbs of internal jugular veins [6].

Because the development of the sigmoid sinuses and the superior bulbs of internal jugular veins decreases the venous pool after the 7 th month of gestation, the dural venous sinuses relatively narrow again and the emissary veins are also reduced. In this case, the dilatation of the dural venous sinuses and the subcutaneous angioma in the right forehead that seems to be derived from the developed emissary veins are thought to be the rests of the fetal period when the sigmoid sinuses and the superior bulbs of internal jugular veins were underdeveloped. A nd the disorder of the gyral formation derives from the neuronal-cell migration disorder at 3 rd to the 7 th months of gestation [7].

The abnormal finding in the chromosomal analysis, and the family members showed neither neurologically abnormal finding nor apparent anomalies. Therefore, no hereditary factor might operated on this case. No history of drug application was seen that might cause any anomaly in nervous system. The abnormalities of this case are right-sided, namely the right transverse sinus is larger than the left one, the developed emissary veins and the subcutaneous angioma are in the right forehead, and the cortical dysplasia is restricted in the right cerebral hemisphere. Therefore, some local, perhaps « physical », factors would have acted on this case, instead of systemic ones, that disordered the formation of the right head. However, nothing has been known as yet that corresponds to such factors.

No case has been reported that shows such dilated dural venous sinuses much less the combined abnormalities (dilated dural venous sinuses, cortical dysplasia and a subcutaneous angioma).

Now, the subcutaneous angioma is becoming smaller than at birth. It is interesting and important to observe whether the enlarged venous sinus system will become narrower or not after birth. A nd the neurological findings of this case were improving. So it seems to be better to observe the course of such a case than to try some treatment in haste.

REFERENCES