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Postnatal development and enlargement of primary middle cranial fossa arachnoid cyst recognized on repeat CT scans

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Abstract. The etiology and mechanism of expansion of primary intracranial arachnoid cysts have been much debated. A rare case of an 8-month-old boy is reported, in which postnatal development and enlargement of a middle cranial fossa arachnoid cyst was detected on follow-up CT scans. Based on intraoperative and histological findings, the cyst was found to be intra-arachnoid. The wall was excised completely, and the lobe adjacent to the cyst appeared normal apart from signs of atrophy. Histological study of the excised cyst revealed a common arachnoid membrane with neither ependymal nor inflammatory cells; the cyst fluid was similar to CSF. The etiology of the lesion remains unclear, but it was considered that the expansion of the cyst might have occurred through a ball-valve mechanism of the membrane in communication with the general subarachnoid space.

Key words: Arachnoid cyst – Computed tomography – Ball-valve mechanism – Middle cranial fossa – Postnatal occurrence.

The etiology and enlargement of primary intracranial arachnoid cysts have been discussed sporadically and remain unclear. According to previous investigations, it is believed that the cyst is caused by congenital malformation of the arachnoid membrane, except in those cases suffering from infection, hemorrhage, or trauma.

Recently, an 8-month-old boy, after repeated follow-up CT scans, was found to have an arachnoid cyst, which developed postnatally, occupied the right middle cranial fossa and expanded.

Case report

An 8-month-old boy, born of a normal delivery on 24 November 1983, following 39 weeks of gestation and weighing 2,450 g at birth, was admitted to our hospital on 16 July 1984, because he was suffering from episodes of general convulsions and exhibited a slight bulging of the right temporal region. There were no physical abnormalities at birth, but 2 days later general tonic convulsions occurred abruptly. At that time, a CT scan (Fig. 1, upper row) and an electroencephalogram were performed at our affiliated hospital, even though no abnormal neurological deficit was seen, and were normal. During the following 4 months, he appeared in good condition until a slight rigidity of the four extremities was noticed. There had been no episodes of obvious head trauma or infection. A repeat CT scan (Fig. 1, middle row) was performed at the same hospital, and a surprisingly large hypodense area of the right side, occupying the middle cranial fossa, was found. There was no compression of the surrounding brain. His physical development was good. A follow-up CT scan was recommended 4 months later.

On admission to our hospital, there were no abnormal neurological or physical findings. Plain skull radiographs revealed a slight bulge in the right temporal region. The next neuroradiological examination and CT scan (Fig. 1, lower row), at the age of 8 months, revealed a hypodense area in the right middle fossa that was larger than before. The slight bulging of the skull, corresponding to the hypodense area of the CT scan, was also larger. As in the previous examination, there was no displacement of the cerebral midline structures.

Right carotid angiography (Fig. 2) showed upward displacement of the M1 to M2 portion of the middle cerebral artery, suggesting an avascular space between the cortical vessels and the surface of the deep sylvian vein, which delineated the surface of the inner membrane of the cyst and drained directly into the cavernous sinus. The results of metrizamide CT cisternography are shown in Fig. 3. Three hours after intrathecal administration, the metrizamide reached the subarachnoid space surrounding the cyst, and the value of the CT number in the small area provided within the cyst was higher (an average of 16.3 compared to a control average value of 6.5). By observing the CT scans in sequence, it was possible to see a progressive increase in the value of the CT number in the same area within the cyst, and the value of the CT number at 6 and 24 h averaged 17.8 and 19.7, respectively. With ^{111}In-DTPA cisternography, the RI tracer could be noted later in the right middle fossa and remained in the cyst more than 48 h after the intrathecal injection. Electroencephalogram reexamination revealed only a range train of low voltage, limited to the right temporal region, with no spike discharge.

On 27 July 1984, a right frontotemporal craniotomy and total removal of the cyst were performed. Upon opening the dura, a large cyst was found, which occupied the entire right middle cranial fossa and contained clear, watery fluid. The cyst wall was composed of both outer and inner membranes, resembling normal arachnoid. The inner membrane mainly covered the exposed insula and the rudiments of the frontal and temporal lobes, while the outer membrane made two cavities because it was folded

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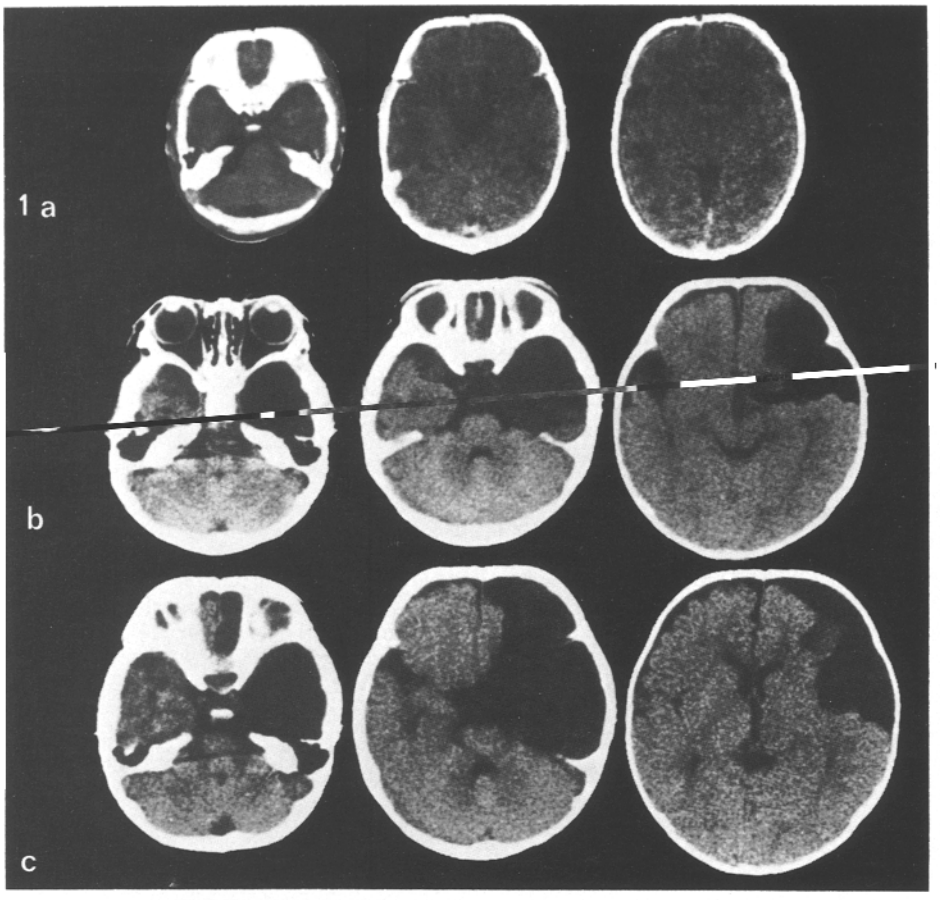
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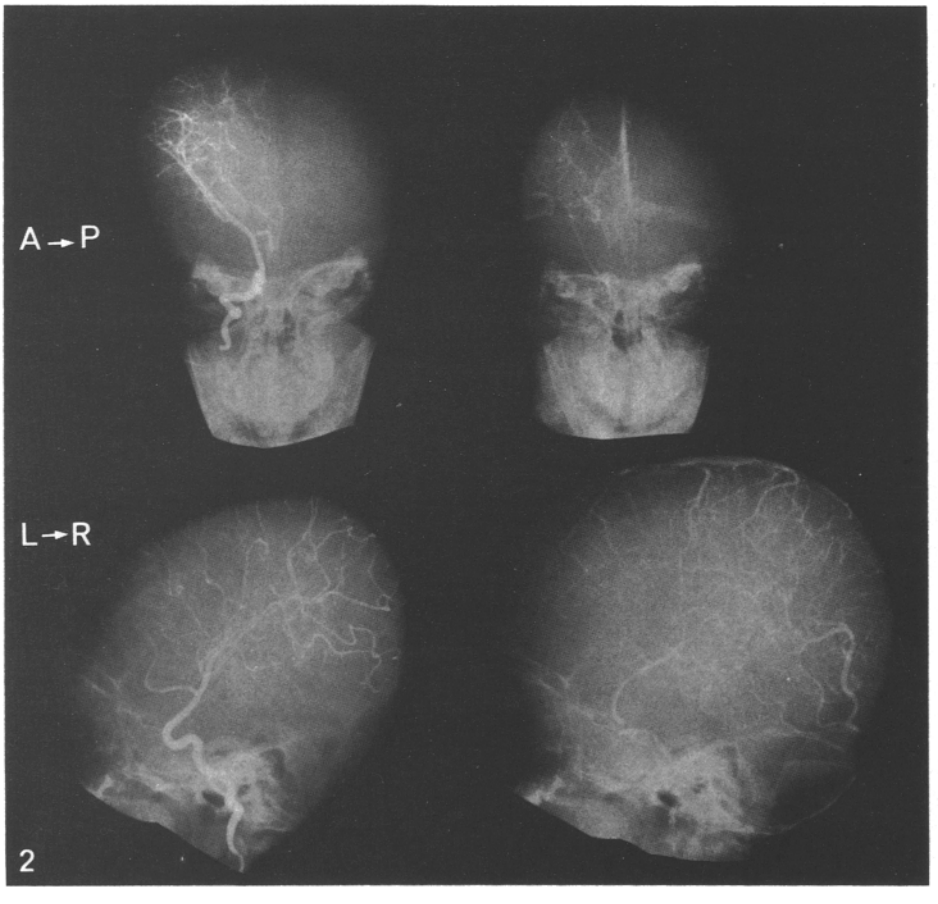
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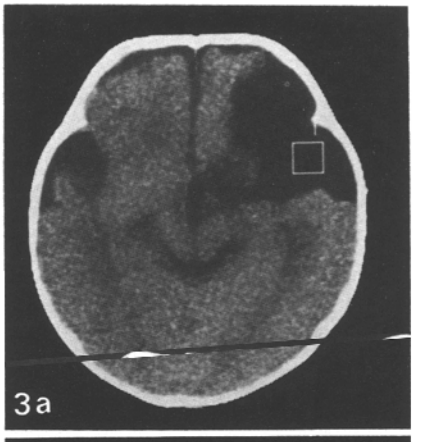
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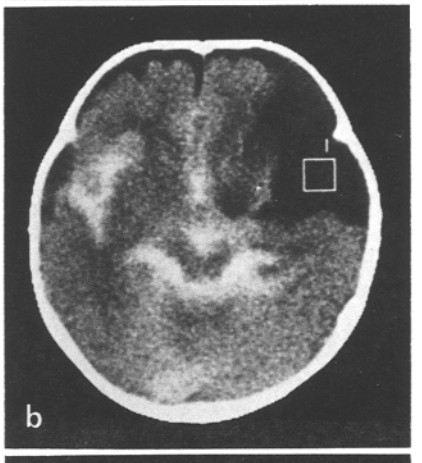
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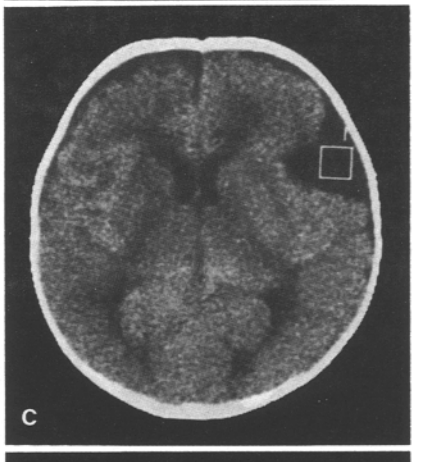
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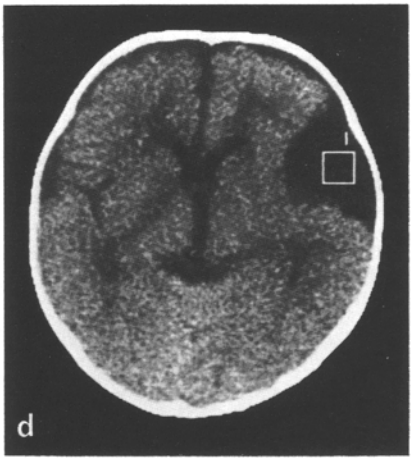
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Fig. 1a–c. The follow-up CT scans, chronologically showing enlargement of the hypodense area in the right middle cranial fossa and slight frontotemporal bulging on the same side 8 months later. **a** 2 days old; **b** 4 months old; **c** 8 months old

Fig. 2. Right carotid angiography (*R-CAG*) demonstrating an elevation of the middle cerebral artery and a single deep sylvian vein entering the sphenoid sinus to drain directly into the cavernous sinus

Fig. 3. CT cisternograms obtained **a** 0 h, **b** 3 h, **c** 6 h, and **d** 24 h after intrathecal administration of metrizamide. Scant increase of CT density, which was measured within the square mark on the CT scans, was observed 6 h and 24 h later

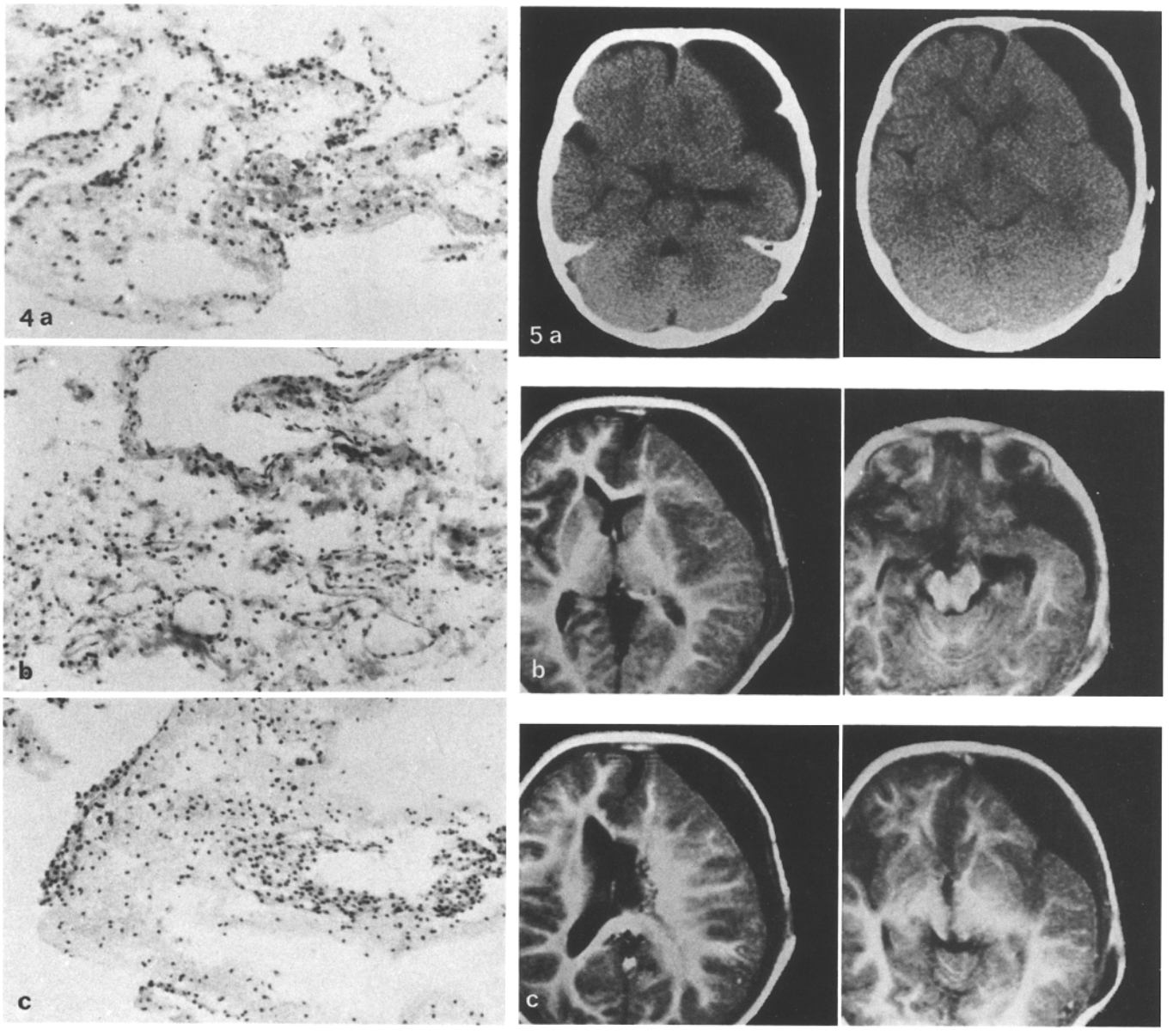

Fig. 4a–c. Photomicrographs show cyst wall consistent with slightly thickened arachnoid membrane (HE, × 100). **a** Surface arachnoid membrane over the brain; **b** outer layer of the cyst wall; **c** fused area of both membranes

Fig. 5a–c. Postoperative CT scans (**a**) showing subdural effusion in the right frontotemporal region, and MRI (**b, c**) showing absence of temporal lobe agenesis

toward the sylvian fissure where it connected to the inner membrane. Communication was achieved by fenestration of the membrane, separating the cyst from the adjacent frontotemporal subarachnoid space and basal cisterns. Shunting was not performed. The osmotic pressure and protein in the cyst fluid were almost identical to those of CSF. Microscopic study of the excised cyst membrane revealed no ependymal, glial or inflammatory cells. Its entire appearance was consistent with slightly thickened arachnoid membrane, as shown in Fig. 4, in which the upper, middle and lower photographs are of the inner membrane, outer membrane, and the fused part of both membranes, respectively. From the intraoperative and histological findings, the cyst was therefore diagnosed as an intra-arachnoid cyst. Postoperative recovery was uneventful, but CT scan and magnetic resonance imaging (MRI) 2 months later (Fig. 5) showed a subdural effusion in the right frontotemporal region and reversion of the foreshortened temporal lobe. There was no evidence of right temporal lobe agenesis on MRI. Consequently, a subduroperitoneal shunt was performed. The child is now 29 months old and healthy.

Discussion

Recently, the usefulness of CT for the diagnosis of arachnoid cysts has been reported, but there have been no reports demonstrating the postnatal occurrence and expansion of a primary arachnoid cyst using repeat CT scans. Regarding the etiology of arachnoid cyst, Robinson reported a developmental defect with unilateral temporal lobe agenesis in 1955 [8], although in a later review he recognized that this condition actually represented a primary malformation of the arachnoid membrane [9]. In 1958, Starkman and Brown pointed out that the occurrence of an intra-arachnoid cyst and atrophic temporal lobe due to pressure from the cystic mass resulted in an arachnoid cyst [12]. Ghatak and Mushrush [4], Galassi and Piazza [2], and Rengachary and Watanabe [7] have hypothesized that the developmental origin of an arachnoid cyst is secondary to prenatal malformation of the arachnoid membrane and, therefore, it seems that the common morphology is an intra-arachnoid cyst. In our case, when the first CT scan was performed two days after birth, because of general convulsions, both sylvian regions were visualized as very small, hypodense areas (predominantly on the left side). Four months later, he again had a generalized seizure and when a repeat CT scan was done, it showed a surprisingly large hypodense area in the right middle cranial fossa. For another 4 months, the hypodense area in the right middle cranial fossa began to expand, eventually creating a slight bulge in the skull, which corresponded to the hypodense area on the follow-up CT scan. During his first 8 months, there was no episode of evident head trauma and/or infection.

In explaining the expansion mechanism of a primary intracranial arachnoid cyst, recent literature has pointed to three main factors [1, 3, 6, 11]. The first is secretion of fluid by ependymal cells, the second is fluid ingress due to an osmotic gradient, and the third to trapping of fluid by a ball-valve mechanism. Our histological study of the cyst membrane revealed no ependymal and glial cells, making the secretion theory of the arachnoid cyst membrane unlikely. The fact that osmotic pressure, proteins and electrolytes of the cystic fluid had similar values to those in CSF does not completely rule out the osmotic gradient theory, because it was impossible to measure osmotic pressure continuously of both intra-arachnoid and CSF cavities with nonsurgical techniques. Metrizamide CT cisternography resulted in slow filling and delayed clearing of the contrast agent within the arachnoid cyst, lending strong support to the ball-valve mechanism theory. In addition, the imbalance between the inflow and outflow volumes of CSF across the cyst wall, as visualized by metrizamide CT cisternography, may evoke retention of the fluid inside the arachnoid cyst, causing the cyst to expand, as stressed by Sato and Shimoji [10]. Also, Williams, in addition to Sato and Shimoji, pointed out that the fluid pulsation of the intra-arachnoid cyst is an important factor relating to cyst expansion [10, 13]. However, the factor causing the anomalous arachnoid split is still unclear, even if a congenital arachnoid malformation is present. On our first CT scan, two small hypodense areas in both sylvian regions were visualized, predominantly on the left side. Eight months later, a large arachnoid cyst occurred in the right middle cranial fossa, despite the absence of extrinsic factors. Furthermore, the change of the hypodense area, as shown in our pre- and postoperative CT scans, cannot assist in determining whether the lesion was secondary to cerebral destruction, histogenetic anomaly, or compression and cyst formation. It is also difficult to explain how the smaller hypodense area, located in the right sylvian region, had already formed a large arachnoid cyst with an intra-arachnoid cyst (as seen during surgery), which has been shown histologically and schematically by Smith and Smith [11] or Dyck and Gruskin [1]. However, the postoperative MRI of this case revealed no temporal lobe agenesis. We wish to emphasize that MRI is the most important examination to analyze cerebral structure. Moreover, histological investigation of the cyst wall revealed a common arachnoid membrane, and the outer layer fused with the surface arachnoid membrane over the brain, similar to observations reported by Starkman and Brown [12], Geissinger and Kohler [3], and Go and Houthoff [5]. Thus, the cyst was diagnosed as intra-arachnoid cyst secondary to compression and cyst formation. Rengachary et al. postulated that an anomalous splitting of the arachnoid membrane occurred during the process of the complex folding of the primitive neural tube and the formation of normal subarachnoid cisterns. Additionally, in relation to the mechanism of the expansion of the arachnoid cyst, Rengachary and Watanabe have reported that the arachnoid cyst wall has unique characteristics that distinguish it from the normal arachnoid membrane [7]. More recently, Go and Houthoff have demonstrated that the structural organization of the arachnoid cyst membrane supports fluid transport toward the cavity; this observation is based on a morphological and enzyme ultracytochemical study of the cyst wall [5].

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