Partially Sequestered Fourth Ventricle: CT and MR Diagnosis of an Unusual Entity

V.G. Wagle, D. Melanson and J.G. Villemure

ABSTRACT: Encystment of the fourth ventricle, due to occlusion of the aqueduct as well as the foramina of Magendie and Luschka has been described previously. Partial sequestration such as that encountered in two cases described is a less common entity. In these two cases, the aqueduct of Sylvius was occluded, but the basal foramina (Magendie and Luschka) were patent. We discuss this partial sequestration or communicating hydrocephalus of the fourth ventricle on the basis of MR and CT scan findings. Although computed tomography following water soluble cisternography did diagnose the entity indirectly MRI proved to be a superior tool. It permitted direct visualization of the basal foramina noninvasively.

RÉSUMÉ: Séquestration partielle du quatrième ventricule: Diagnostic par CT et NMR de cette entité rare L'enkystement du quatrième ventricule, dû à l'occlusion de l'aqueduc de Sylvius et des ouvertures de Magendie et Lushka, a été décrit précédemment. La séquestration partielle, comme celle rencontrée dans les deux cas ici décrits, est une entité moins fréquente. Dans ces deux cas en effet, l'aqueduc de Sylvius était fermé mais en ouvertures de Magendie et Lushka étaient ouvertes. Nous discutons cette séquestration partielle ou hydrocéphalie communicante du quatrième ventricule sur la base des données du l'NMR et du CT Quoique la CT-cisternographie avec contraste hydrosoluble ait démontré l'entité de façon indirecte, l[e]NMR s'est avérée un instrument diagnostique supérieur. Elle a permis en effect la visualisation directe des ouvertures de Magendie et Lushka, et ce, de façon non invasive.

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Magnetic resonance imaging (MRI) is proving to be a reliable tool for evaluation of posterior fossa pathology[1,2] without the superimposition of bony artifacts, a drawback encountered in CT scanning. We report two cases of partially sequestered fourth ventricle. In these cases, CT scanning and magnetic resonance imaging demonstrated the partially sequestered fourth ventricle, which differs from the trapped fourth ventricle usually described in that the basal foramina were patent. Sequestered fourth ventricle has been described as a complete encystment of the fourth ventricle. In both our patients, the sequestration was probably a result of previous shunt surgery.[3,4,5] However, the fact that the foramina of Magendie and Luschka as well as the basal subarachnoid spaces seemed normal or moderately enlarged leads us to speculate that incompetence of the subarachnoid spaces over the convexity was the factor causing ballooning of the fourth ventricle.

Case Reports

Patient 1

This 12-year-old girl had progressive quadraparesis following a meningitis. She had a ventriculo-peritoneal shunt. Magnetic resonance imaging was done in the spin echo pulse sequence (SE), and repetition time (TR) and echo time (TE) of 400 msecs and 30 msecs respectively on a 1-5 Testa Philips gyro scan (Figure 1).

MRI revealed a dilated fourth ventricle, bulging into the brainstem and cerebellum. The contents of this dilated ventricle showed characteristic MR features of cerebrospinal fluid.[5,6] Superiorly, it was presumed that the aqueduct was stenosed, with no apparent enlargement of the third or lateral ventricles, because in spite of a head low position, the contrast on cisternography was not seen above the fourth ventricle. The craniovertebral region did not show evidence of cerebellar tonsillar herniation or Arnold-Chiari malformation, and no cavitation of the spinal cord. The foramina of Magendie and Luschka were open. Computed scanning, following injection of water soluble contrast cisternography via C_1-C_2 puncture (previous lumbar myelogram had shown a block at C3-4, probably a result of meningitis), revealed a dilated fourth ventri-

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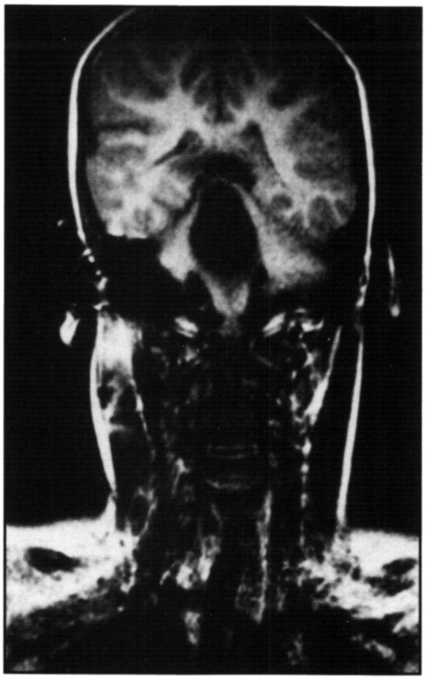

Figure 1 — MR, SE 400/30, in coronal plane showing large fourth ventricle with CSF showing low intensity signal.

Figure 2 — Early post (iopamidol) cisternographic CT scan showing a large fourth ventricle filled with contrast (patient 1).

Figure 3 — Patient 2 showing a ballooned fourth ventricle filled with contrast and basal subarachnoid spaces, six hours after instillation of iopamidol through lumbar puncture. Post surgical changes and droplets of oil based contrast material from a previous myelogram are also seen.

cle filled with contrast, a stenosed aqueduct and dye-filled basal subarachnoid spaces (Figure 2). The convexity subarachnoid spaces, however, did not fill at all, following cisternography. CSF pressure in the fourth ventricle was normal and a fourth ventriculoperitoneal shunt failed to reverse or arrest the patient's clinical problems. The reasons for the deterioration were not clear.

Patient 2

This patient was a 27-year-old male who was operated upon for a hemangioblastoma of the cerebellum eight years previously. Following surgery, he developed hydrocephalus which was corrected with a ventriculoatrial shunt. During the two years that followed, he had frequent episodes of diplopia, headaches, nausea, vomiting and ataxia. He

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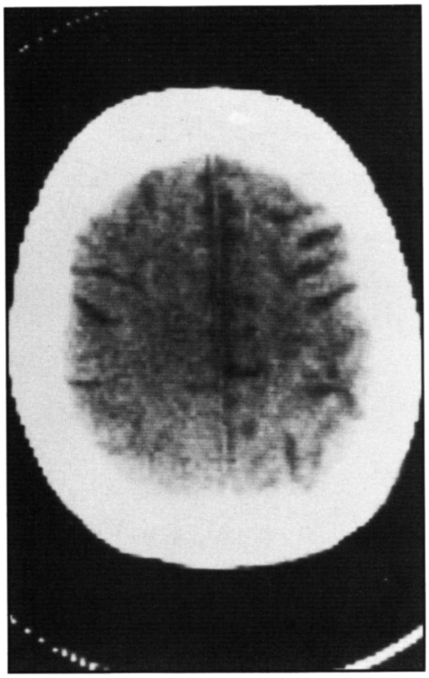

Figure 4 — CT scan showing non-filling convexity sulci six hours after iopamidol cisternography.

underwent exploratory suboccipital craniectomy, and a small nodule of hemangioblastoma was excised. He also had his shunt revised, but the episodic clinical manifestations continued to occur. Air and contrast ventriculography done during an earlier admission had shown an aqueductal stenosis. A post-cisternogram CT scan showed a large fourth ventricle, non-filling of the posterior third ventricle and the ventricles above, and persistent filling of the fourth ventricle and basal cisterns six hours later (Figure 3). However, the delayed scan showed abnormal nonfilling of the convexity sulci (Figure 4). This nonfilling of the posterior third ventricle and convexity sulci was noted in spite of tilting the patient's head from side to side in a head low position. A fourth ventricular shunt was advised, but the patient refused and he was lost to follow up.

Discussion

A completely sequestered fourth ventricle may be seen in children following shunt placement in communicating and non-communicating hydrocephalus. It may present as a posterior fossa syndrome or it may be asymptomatic. In the latter instance it is sometimes discovered as an incidental finding on computed tomography. This condition is rare in adults but is occasionally seen in long-standing aqueductal stenosis.[7] In most cases there is no evidence of raised pressure.[8] A completely sequestered fourth ventricle is never a primary condition and needs to be differentiated from posterior fossa cysts and cystic tumors. Complete encystment of the fourth ventricle, although unusual, has been described in several studies.[3,4,7] Partial encystment, however, has been described only once.[9] In that case report,[9] water soluble contrast cisternography was performed after placement of the fourth ventricle shunt, and thus failed to localize the extraventricular site of the block. In our study, cisternography was performed before any surgical intervention was undertaken, and both early and delayed scanning were performed. The competence of the foramina of Magendie and Luschka was demonstrated by the early filling of the fourth ventricle. Furthermore, MRI clearly demonstrates the open foramina. The convexity sulci on delayed scanning following cisternography generally tend to show ample contrast as they form a major pathway in CSF circulation.[10] Radioactive tracers such as albumin Tc 99m, injected in the subarachnoid spaces during cisternography has been shown to appear at the lateral surfaces of the brain after six hours.[11] This is not the case in our patients, where the filling was poor or nonexistent. In view of

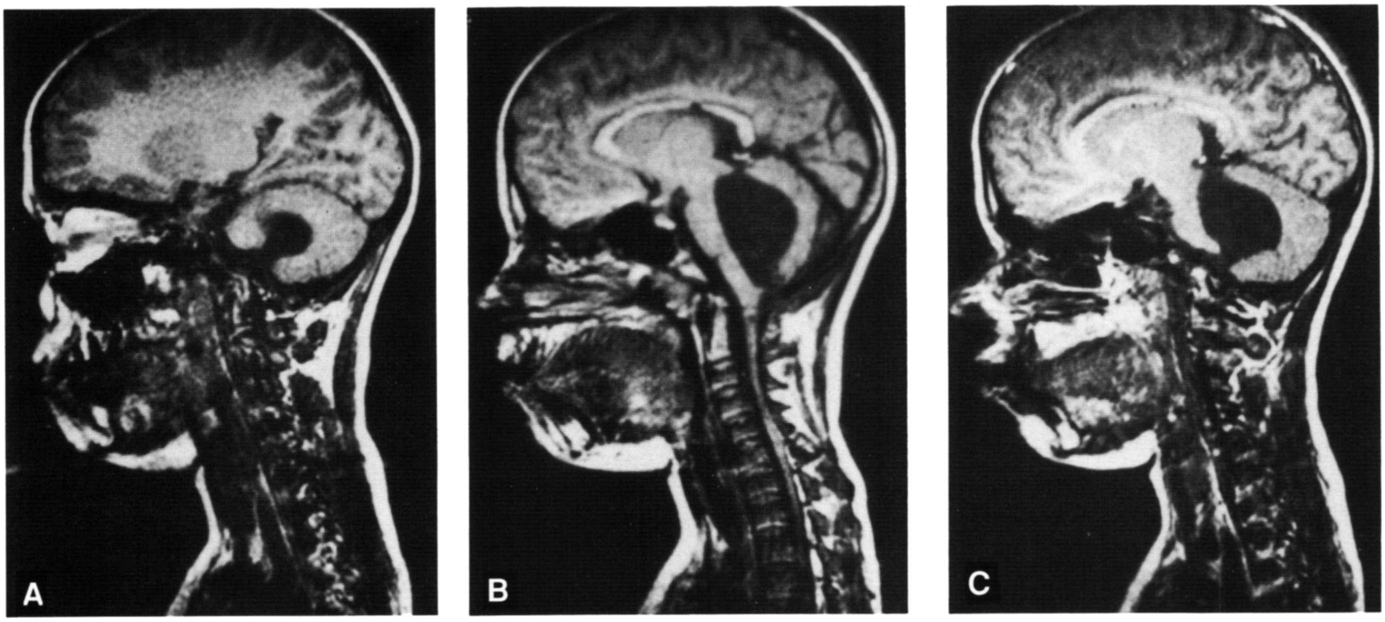

Figure 5 — MR, SE 400/30, in sagittal plane shows an open left Luschka, Magendie and right Luschka, in Patient 1.

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the fact that the ballooned fourth ventricle and basal cisterns filled on the early and delayed scans, and even the delayed scans showed no filling of convexity sulci on tilting the patient's head from side to side in a head low position, seems to suggest that these sulci were the site of obstruction (Figure 4). The block may also exist in other segments of the subarachnoid pathway: the posterior fossa, tentorial notch, the sylvian fissures, or the spinal subarachnoid space.[3] The block is usually symmetrical and the proximal subarachnoid spaces are wide. It is possible that repeated surgical management for generalized hydrocephalus with its inherent risk of subarachnoid hemorrhage together with the severity and chronicity of hydrocephalus may all result in chronic meningitis or leptomeningeal fibrosis, rendering the subarachnoid space incompetent.[12]

Postshunting sequestration of the fourth ventricle, as well as partial sequestration or communicating hydrocephalus of the fourth, are easier to diagnose since the advent of computed tomography. A combination of iopamidol cisternography and CT scanning further facilitates the diagnosis.[13] However, water soluble contrast cisternography is invasive and not without risks.[5] MRI is safe in visualizing CSF pathways, as no ionizing radiation, catheters or contrast materials are required (Figure 5). Fourth ventricular shunting is the treatment of choice in complete sequestration.[4,7] Resultant improvement is probably the result of reduction in CSF pressure or, in cases with normal pressure, due to reestablishment of CSF flow.[13] Treatment in partial sequestration is less certain. Shunt placement reverted the supratentorial hypertension and reduced the size of the fourth ventricle in one patient.[9] In our study, the one patient who underwent shunt surgery showed no improvement, for uncertain reasons. Paucity of case reports describing partial sequestration could be due to reluctance in employing invasive tests such as water-soluble contrast cisternography. Magnetic resonance imaging offers a safe alternative in diagnosing this rare entity.

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