Subtentorial Diverticulum of the Third Ventricle Associated with a Mural Cavernous Angioma in a Child

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The authors describe a case of a large subtentorial supracollicular diverticulum of the third ventricle associated with a cavernous angioma in its wall in a 6-year-old girl who presented with developmental delay and obstructive hydrocephalus. This is the first case in which such association has been diagnosed and successfully treated. The literature is reviewed, and the possible relationship between these two rare lesions is discussed.

KEY WORDS: Cavernous angioma; Computer tomography; Diverticulum; Magnetic resonance imaging; third ventricle

A ventricular diverticulum is a cystic subpial collection of cerebrospinal fluid (CSF) resulting from rupture of the ependymal lining of the ventricular system in cases of advanced hydrocephalus [28]. The condition was first described by Penfield [27] in 1929, and according to Wakai et al [36] 51 cases have been reported in the pertinent literature until 1983. Since then only a few new cases have been added [9,26]. With a few exceptions [7,9,25], the hydrocephalus was secondary to an obstructive lesion, mostly "benign" aqueductal stenosis [2,3,7,8,14,18,22,28,33,37,38]. Less frequently, brain stem tumors [7,14,28,31,32] and third ventricle tumors [5,27] were the cause of obstruction. The association of a ventricular diverticulum with a cavernous angioma has, to the best of our knowledge, never been reported before.

In this report, we describe the first case of a large diverticulum of the third ventricle occupying the quadrigeminal cistern and associated with a cavernous angioma in its wall. This type of vascular malformation has been rarely encountered in the quadrigeminal region, and according to Hahn et al [10], only seven of some 598 (= 1.2%) intracranial cavernous angiomas collected from the literature were located there. The relationship between these two rare pathologic entities is discussed.

Case Report

A 6-year-old girl was referred to the Neurosurgical Division of the King Khalid University Hospital because of progressive difficulty of walking, delayed speech, and abnormal rolling of the eyes since the age of three. Computed tomography (CT) scan of the head revealed a nonenhancing, high attenuation area (measuring 18 x 12 mm) dorsal to the midbrain associated with obstructive hydrocephalus (Figure 1 A). There was also a midline cyst in the posterior fossa (measuring 28 x 31 mm) that communicated with the third ventricle (Figure 1 B). On magnetic resonance imaging (MRI), the cyst gave signal intensities identical with those of the cerebrospinal fluid. The lesion dorsal to the brain stem consisted on both T1- and T2-weighted images of an irregular hyperintense core surrounded by a hypointense peripheral area. The exact anatomic relationship among the cyst, lesion, and surrounding neural structure was best demonstrated on the T1-weighted midsagittal MRIs (Figure 2). It showed an infratentorial and supraquadrigeminal cyst communicating with the third ventricle through a small opening at the level of the pineal recess. At the inferior pole of the cyst wall dorsal to the pons, a lobulated hyperintense lesion was present. The aqueduct of Sylvius was severely compressed and the floor of the dilated third ventricle herniated caudally into the sella turcica and the prepontine spaces.

Examination

The girl was fully alert but showed a marked degree of mental and motor retardation. She could speak only a...
Figure 1. (A) A computed tomography scan showing a slightly hyperdense, nonenhancing lesion dorsal to the midbrain (arrow) with obstructive hydrocephalus. (B) There is a midline cyst in the posterior fossa, communicating with the enlarged third ventricle (asterisk).

few simple words. There was no papilledema, and all cranial nerves were intact. Convergence and, to a lesser extent, upward gaze were, however, impaired. The gait was obviously ataxic with a falling tendency to the right side. The patient was able to walk only with assistance.

Operations
The patient underwent suboccipital craniectomy in sitting position. The fourth ventricle was entered through a midline incision in the cerebellar vermis. A blue-reddish mass with a “bunch-of-grapes” appearance was identified just above the roof of the ventricle (Figure 3 A). The lesion was well-demarcated, and the surrounding tissue was stained intensively yellow. Microsurgical dissection of the lesion was performed. A cystic cavity was entered, and a lot of colorless and clear fluid drained. The third ventricle could be directly viewed through a relatively wide opening in the anterior wall of the cyst (Figure 3 B). No definite cyst wall could be identified. The lesion was totally excised.

Postoperative Course
During the first few postoperative days, the patient remained in a sleep-like state in which she did not open her eyes or obey commands, but she was moving her limbs spontaneously and occasionally crying, laughing, or calling her mother. Her eyeballs were slightly diverging, and pupils were equal and reactive. An immediate postoperative CT scan revealed extensive air inclusions in the dilated lateral and third ventricles and basal cisterns, but there was no hematoma at the operative site. An external ventricular drainage was inserted that was converted 5 days later into a permanent ventriculoperitoneal shunt. Brain stem auditory potentials (BAEP), visual evoked potentials (VEP), and electroretinography (ERG) were all within normal limits. Electroencephalogram (EEG) revealed initially diffuse, high-voltage theta and delta wave activity. However, a second EEG recording a few days later was almost normalized. The patient recovered slowly and gained increasingly full consciousness. On discharge 6 weeks after operation, she was back to her initial neurologic
Figure 2. A sagittal T2-weighted magnetic resonance imaging showing marked obstructive hydrocephalus with the posterior wall of the third ventricle opening into a large infratentorial cyst (asterisk). The cyst shows a lobulated, high intensity lesion at its inferior pole (arrowhead). The floor of the third ventricle is also herniating into the sella turcica and the preoptic subarachnoid space.

state. On follow-up examination 6 months later, the girl was mentally brighter and her speech was improved. She was able to walk independently, and she had no focal neurologic deficits. A CT scan showed a small operative defect in the pineal region, otherwise normal intracranial structures (Figure 4).

Pathological Examination

The gross specimen consisted of an irregular, hemorrhagic soft tissue measuring 20 x 17 x 10 mm and showing bright yellow areas. Histologically, the lesion was composed of irregular and markedly dilated vascular channels lying side by side with no intervening parenchymal tissue (Figure 5 A). The majority of the vessels were thin-walled, but communicating capillaries with thick hyalinized walls were present at some areas (Figure 5 B). At the periphery of the lesion, there were multiple foci showing numerous hemosiderin-laden macrophages.

Discussion

Ventricular rupture is a rare phenomenon that occurs predominantly in cases of advanced obstructive hydrocephalus, but rarely can develop in association with communicating hydrocephalus [7,9,25]. Ventricular rupture may result either in a communication between the ventricular system and the subarachnoid space (spontaneous ventriculocisternostomy) [6,15,20,21,23,24,31], or, if the pia mater is preserved, it may lead to the formation of a subpial cystic collection of CSF (ventricular diverticulum) [2,3,5,7-9,14,18,22,25-29,31,33,36,37]. Whereas the first condition may lead to spontaneous remission of the hydrocephalus [6,21,32], the second can give rise to additional obstruction or compression of the surrounding neural tissues [5,18,31,36].

The exact pathogenesis of ventricular diverticula is
not clear. The condition has been observed in all age groups from infancy to middle age, and it seems that extreme ventricular distension is not a sine qua non. The fact the ventricular diverticula are by far more commonly associated with obstructive than with communicating hydrocephalus suggests that an elevated transventricular pressure gradient is probably the main contributing factor. There are four areas of physiologic attenuation and weakness in the ventricular wall that tend to "blow out" or herniate under elevated pressure due to CSF obstruction [24,29]: the lamina terminalis [21,31], the floor of the third ventricle [23,24], the posterior recesses of the third ventricle [2,3,8,14,18,33], and the medial wall of the trigone of the lateral ventricle [2,5,7,22,25,27,28,31,36]. Although spontaneous ventriculocisternostomy may occur at any of these sites, ventricular diverticula arise almost exclusively in the last two locations. They extend subsequently through the tentorial hiatus down to the posterior fossa where they typically present as subtentorial supracollicular CSF cysts [2,3,7,8,14,18,22,25,26–29,31,33,36–38]. No explanation has been given in the literature for the affinity of ventricular diverticula to this location. Moreover, it is interesting to note that in nearly all reported cases of ventricular diverticula, the ventricular obstruction was either at the aqueduct or in the third ventricle and that the pathology did not develop when the posterior fossa was full either with a dilated fourth ventricle or a large tumor [28,29]. Thus, it is reasonable to presume that following ventricular obstruction proximal to the fourth ventricle, the CSF pressure in the infratentorial compartment may not rise simultaneously to the same extent as in the supratentorial compartment. As a result, the transventricular pressure gradient is likely to be highest at those areas of the ventricular system neighboring the inlet of the infratentorial compartment, i.e., posterior wall of the third ventricle and the postero-medial wall of the lateral ventricle. The relatively wide quadrigeminal cistern allows these parts of the ventricular system to "blow out." With progressive expansion, both the ependymal lining and the thin glial layer rupture leaving the more stretch-resistant pia mater intact. A CSF pial cyst develops that soon thereafter receives an almost complete circumferential external support by the surrounding neural structures (lamina quadrigemini, splenium coporii callosi, and cerebellar hemispheres), thus, counteracting its early rupture. In other basal cisterns, such external support is obviously incomplete, probably accounting for the early rupture of the pia and the formation of a spontaneous ventriculocisternostomy.

The rarity of ventricular rupture is quite surprising considering that obstructive hydrocephalus is a common neurosurgical problem. Some authors believe that this phenomenon occurs more frequently than is recorded [24,25]. Although this may be true for the spontaneous ventriculocisternostomy, it is hardly imaginable that ventricular diverticula, with their fairly impressive appearance on CT and MRI scans, would escape attention. A plausible explanation for the rarity of these lesions is that ventricular dilation and increased transventricular pressure gradient would lead to ventricular rupture only under certain circumstances, such as the presence of focal structural abnormalities of the ventricular wall, e.g., following hemorrhage, inflammation, or tumors.

The clinical picture in patients with ventricular diverticula is almost entirely dominated by the symptoms and signs of the obstructive hydrocephalus. A few patients may additionally show focal neurologic deficits caused by the obstructing tumors [14,31]. Symptoms and signs resulting from direct pressure of ventricular diverticula on the surrounding neural structures are relatively rare and consist mainly of cerebellar and dorsal midbrain deficits [8,14,18]. Commonly, however, they are overlooked or masked by the more prominent clinical picture of ventricular obstruction.
The association in our patient of a ventricular diverticulum with a cavernous angioma in its wall may well be incidental. This vascular hamartoma may occur in any location within the central nervous system, though very rarely in the quadrigeminal region. Obstructive hydrocephalus is a well recognized complication of quadrigeminal cavernous angiomas being present in five of the seven cases reported in the literature up to date [4,12,34]. However, none of these cases had an associated ventricular diverticulum. In our patient, the primary cause of the aqueductal occlusion is unknown. The ventricular diverticulum is more likely to be the result rather than the cause of the ventricular obstruction. It is possible that the vascular lesion in our patient was originally located somewhere in the posterior wall of the third ventricle, thus leading to the ventricular occlusion. It could have also resulted in attenuation of the ventricular wall, either directly by virtue of its location or indirectly because of recurrent local bleedings. As the diverticulum enlarged progressively, the lesion migrated inferiorly.
References


