Cavernoma of the foramen of Monro

Case report and review of the literature

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✓ The authors report on a patient who presented with an intraventricular mass located at the level of the foramen of Monro. The clinical presentation and neuroimaging appearance of the mass led to an initial diagnosis of colloid cyst. A neuroendoscopic approach offered a direct view of the ventricular lesion, which was found to be a cavernous angioma partially occluding the foramen of Monro. The lesion was then removed using microsurgery. In this report the authors highlight possible pitfalls in the diagnosis of some lesions of the third ventricle, and the possible advantages of using a combined endoscopic and microsurgical technique when approaching such lesions.

KEY WORDS • cavernous angioma • colloid cyst • third ventricle • foramen of Monro • neuroendoscopy

INTRAVENTRICULAR cavernomas are very rare vascular malformations that account for 2.5% of all brain cavernomas.¹⁶ The lateral ventricles are the most common locations for these lesions, followed by the third and fourth ventricles.¹² Within the small group of third ventricle cavernomas, those located at the foramen of Monro are of particular interest because they may present with foraminal obstruction and biventricular hydrocephalus (a dramatic clinical condition), and must be differentiated from other lesions originating in that region.^{1,5,6,8,13,14}

We report a clinical case in which this diagnostic dilemma is well exemplified: because the results of neuroimaging were consistent with the presence of a colloid cyst, a neuroendoscopic approach was attempted at first.⁸ Under direct endoscopic view of the ventricular lesion, however, the diagnosis of cavernoma was made and a definitive microsurgical treatment of the vascular malformation was ultimately performed.

Case Report

History. This 35-year-old man was admitted to our neurosurgical unit with a 6-month history of recurrent, short episodes of postural headache that were precipitated by head position changes. During the week before hospital admission his symptoms became exacerbated, culminating in a severe headache with cervical, radiating pain and vomiting the day before he was admitted.

Examination and Neuroimaging. Clinical examination showed only a mild papilledema with no neurological deficit. Blood tests revealed a coagulation factor V deficiency. Initial CT scans demonstrated a 12-mm hyperdense lesion within the third ventricle that was partially occupying the right foramen of Monro and causing biventricular asymmetrical hydrocephalus. Admission MR images demonstrated an iso- to hypointense lesion with mild and nonhomogeneous contrast enhancement on T₁-weighted images (Fig. 1) and a hyperintense lesion on proton density-weighted images. A cerebral angiographic study revealed no pathological vascularization. Based on its clinical presentation and imaging appearance, the lesion was diagnosed as a colloid cyst, and an endoscopic approach was selected for its treatment.⁸

Operation. Access was precoronal, through a 12-mm bur hole made 2 cm lateral from the midline. A flexible endoscope with a 3.9-mm diameter was used. After introduction of a peel-away cannula, some xanthochromic CSF spilled out. At first, visibility inside the lateral ventricle was limited and copious washing was necessary to clear the yellow CSF. The foramen of Monro was completely occupied by a blackberry-like formation (Fig. 2) that was recorded as a cavernoma, with its turgid venous capillaries pushing the choroid plexus backward. The lesion apparently originated from the wall of the pars libera of the column fornicis, which was white in the anterior part (as in healthy individuals), but brown toward the posterior border (Fig. 2A).

The procedure was abandoned after we had performed a septostomy to resolve the asymmetrical hydrocephalus, and temporary CSF drainage was assured with an intra-

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computed tomography; MR = magnetic resonance.

FIG. 1. Preoperative MR images obtained in the patient described in this report. A: Coronal T₁-weighted MR image obtained without contrast, demonstrating an asymmetrical dilation of the lateral ventricles and an isointense cystic lesion. B and C: Coronal and sagittal T₁-weighted MR images obtained with contrast, demonstrating mild uptake of contrast medium; spots of hyperdensity were interpreted as shifted internal venous system.

ventricular catheter connected to an Ommaya reservoir. Three days later microsurgical removal of the cavernoma was performed, in which the surgical approach followed the trajectory of the previous transventricular endoscopic procedure. Microsurgery allowed fine dissection of the cavernous angioma from the fornix, plexus, and thalamostriate vein, resulting in a complete and uneventful removal of the lesion. Histopathological findings confirmed the diagnosis of cavernous angioma.

Postoperative Course. No complication was observed during the postoperative period. The patient was discharged 1 week after surgery with no neurological impairment.

Discussion

Cavernous angiomas primarily involving the third ventricle can be divided into four subgroups in relation to the region from which they originate.⁷ Early symptoms of cavernomas involving the suprachiasmatic region may be visual field restriction and endocrine dysfunction, whereas masses involving the lateral wall or floor of the third ventricle can affect short-term memory. In contrast, hydrocephalus (with sudden signs of raised intracranial pressure) is a common presentation for lesions located in the region of the foramen of Monro.^{1-3,5,6,11,15}

Growth patterns and clinical manifestations of cavernous angiomas may be influenced by the fact that they are intraventricular: when these lesions develop within the ventricles, rapid growth is observed, resulting in giant malformations.⁹ Such behavior may be explained by the low mechanical resistance offered by CSF to the expanding cavernoma, or alternatively by the frequent occurrence of intralesional bleeding when cavernomas arise within cerebral ventricles. Our case differed from the aforementioned tendency of third ventricle cavernomas to present as giant lesions.

The data published on this subject are scarce, and are mainly limited to case reports (Table 1). The first report of a cavernoma documented with a CT scan in the foramen of Monro was by Britt, et al.,¹ who described a lesion simulating a colloid cyst. A second case, reported by Pozzati, et al.,¹¹ shared some similarities with ours; a 31-year-old man presented with a 3-month history of headache, and CT scans revealed an asymmetrical dilation of the lateral

ventricle and a large lesion in the foramen of Monro. Harbaugh, et al.,⁵ reported on a patient who presented with an ictal onset and minimal intraventricular hemorrhage. The lesion was surgically removed via a transcallosal approach, and it was identified as hemangioma calcificans. Macroscopically it appeared as a well-demarcated, multilobulated, purple-red mass positioned at the level of the foramen of Monro. Suess, et al.,¹⁵ described one case in which an unsuspected large lesion presented with progressive hydrocephalus and signs of increased intracranial pressure, including headache, nausea and vomiting, short-term memory deficits, and psychic changes. An analogous

FIG. 2. Endoscopic views and graphic illustrations of the intraventricular cavernoma occupying the foramen of Monro. A: Complete view of the Monro region, with all its typical anatomical structures; note that the posterior part of the fornix (just anterior to the septal vein) appears faintly brown due to previous bleeding. B: A closeup view and drawing showing details of the turbid protrusion of the cavernoma with its blackberry-like appearance. Cav = cavernoma; f = foramen of Monro.

TABLE 1
Literature review of cavernous angioma of the foramen of Monro*

Authors & Year	Age (yrs), Sex	Clinical Presentation	Hydrocephalus	Tumor Size (mm)	Appearance on MRI & CT			Extent of Removal
					T ₁	T ₂	C/h	
Britt, et al., 1980	11, F	hydro, N & V, S, pap	lat vents	NR	NR	NR	hyper (CT)	total
Pozzati, et al., 1981	31, F	hydro, N & V, pap	lat vents & quadr cistern	NR	NR	NR	iso (CT)	total
Harbaugh, et al., 1984	44, F	hydro, N, malaise	lat vents & hem	NR	NR	NR	iso (CT)	total
Katayama, et al., 1994	50, F	NR	NS	NR	NR	NR	NR	NR
	45, F	coma	NS & hem	NR	NR	NR	NR	NR
Kaim, et al., 1997	64, M	hydro, G, mem	lat vents	15	hyper	hyper	hyper	total
Crivelli, et al., 2002	38, M	hydro, N & V, mem, G, pap	lat vents	NR	mixed	NR	mixed	total
Suess, et al., 2002	36, F	hydro, N & V, mem	rt vent	30	mixed	NR	hyper (CT)	total
Chen, et al., 2005	51, F	hydro, mem, V, G	lat vents	15	hyper	hyper	hyper	total
present study	35, M	hydro, V, pap	lat vents	12	hypo		mixed	total

* C/h = contrast enhancement; G = gait ataxia; hem = hemorrhage; hydro = hydrocephalus; hyper = hyperintense; hypo = hypointense; iso = isointense; mem = memory disturbance; mixed = mixed signal; N = nausea; NR = not reported; NS = not specified; pap = papilledema; quadr = quadrigeminal; S = syncopal episodes; V = vomiting; vent = ventricle.

clinical presentation was detailed in the case described by Crivelli, et al.,³ in which a small lesion was identified on neuroimaging that could easily have indicated a possible cavernoma because of an associated venous angioma.

Characteristically, cavernomas are hyperdense on CT scanning, whereas on MR imaging they may appear hyper- to isointense on T₁-weighted, and mixed (with rings of low intensity) on T₂-weighted images.^{10,12,14} Some authors have pointed out that third ventricle cavernomas may have a unique appearance on MR imaging when compared with cavernomas located elsewhere.¹⁴

Proton-density MR imaging acquisition excluded signs of bleeding in our case, despite endoscopic evidence of previous blood leakage (xanthochromic CSF and brown discoloration of the fornix). Considering both clinical history and neuroimaging appearance, our patient's lesion resembles a colloid cyst more than a cavernoma, although a careful review of MR images revealed a honeycomb-like appearance, which retrospectively might have cast some doubt on the diagnosis of colloid cyst. A T₂-weighted image, which was not available in this case, probably could have led us to the right diagnosis.

Misdiagnosis of lesions involving the third ventricle is not that rare. Britt, et al.,¹ reported a third ventricle choroid plexus arteriovenous malformation simulating a colloid cyst and producing similar symptoms. Other masses such as meningiomas, plexus papillomas, ependymomas, and germinomas have also been described as possibly simulating a colloid cyst.¹⁴ Furthermore, there are reports of hemorrhagic colloid cysts, a condition that makes the pre-operative differential diagnosis even trickier.¹³ In any case, although MR imaging can provide the initial diagnosis for such an unusually localized malformation, it should be confirmed histopathologically.²

Given this peculiar and ambiguous presentation, the endoscopic approach initially attempted in our patient (albeit not appropriate for a radical removal) was possibly the least invasive and a more definitive way to reach the correct diagnosis. To our knowledge, this is the first time a cavernoma of the foramen of Monro has been endoscopi-

cally viewed; previous reports of endoscopic approaches to intraventricular cavernomas involved a hypothalamic lesion¹⁷ and a cavernoma of the lateral ventricle.⁴ Endoscopic removal was attempted in one patient only, but that one experienced relevant cognitive sequelae. The dense vascularization of the periventricular areas in which the malformation is rooted, and the intimate relationship that the lesion itself has with the normal vascular bed explain why only a microsurgical approach may ensure sufficient precision and care during the excision procedure.

Conclusions

This case of an intraventricular cavernoma of the foramen of Monro, with its clinical and imaging presentation, underlines the diagnostic difficulties posed by these lesions. The combined use of neuroendoscopy and microsurgery resulted in a correct diagnosis, an unprecedented endoscopic view of this type of lesion, and possibly in the best treatment for the patient.

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