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Neurocytoma Accompanied by Intraventricular Hemorrhage: Case Report and Literature Review

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Neurocytomas are a rare subset of primary cerebral neuroblastomas. They occur within the ventricles of young adults, may calcify, and demonstrate a better prognosis than do their parenchymal counterparts. Of the 21 patients reported in the literature who appear to meet most of the criteria for this diagnosis, 18 had symptoms referable to increased intracranial pressure [1–10]. The lesions were discovered incidentally in the other three patients [5, 7]. The patient who is the

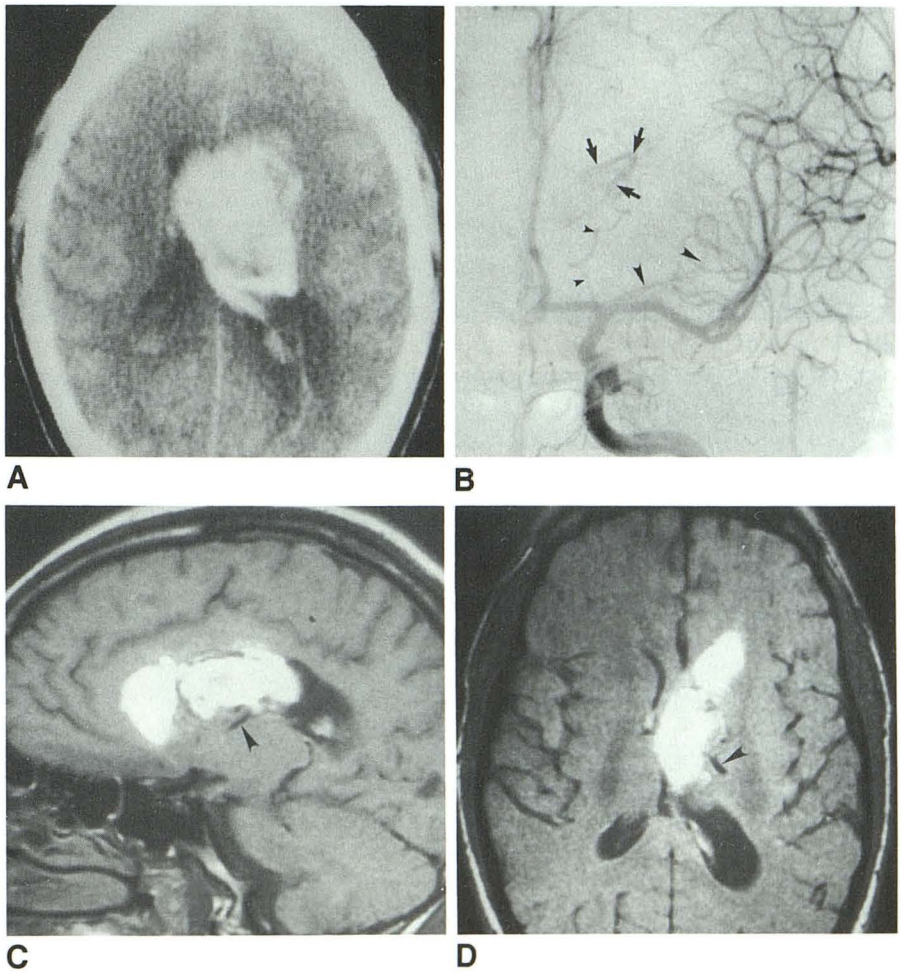

Fig. 1.—*A*, Unenhanced CT scan at time of admission reveals intraventricular hemorrhage and acute hydrocephalus.

***B*, Left internal carotid artery angiogram, anteroposterior view, shows slight enlargement of a lenticulostriate artery (*small arrowheads*), early opacification of thalamostriate vein (*arrows*), and avascular mass effect bowing other lenticulostriate vessels laterally (*large arrowheads*).**

***C* and *D*, Sagittal (*C*) and axial (*D*) T1-weighted MR images (600/20/1) obtained 5 days after presentation. High signal intensity within left lateral ventricle is compatible with subacute hemorrhage. Flow void from early draining left thalamostriate vein is quite prominent (*arrowheads*).**

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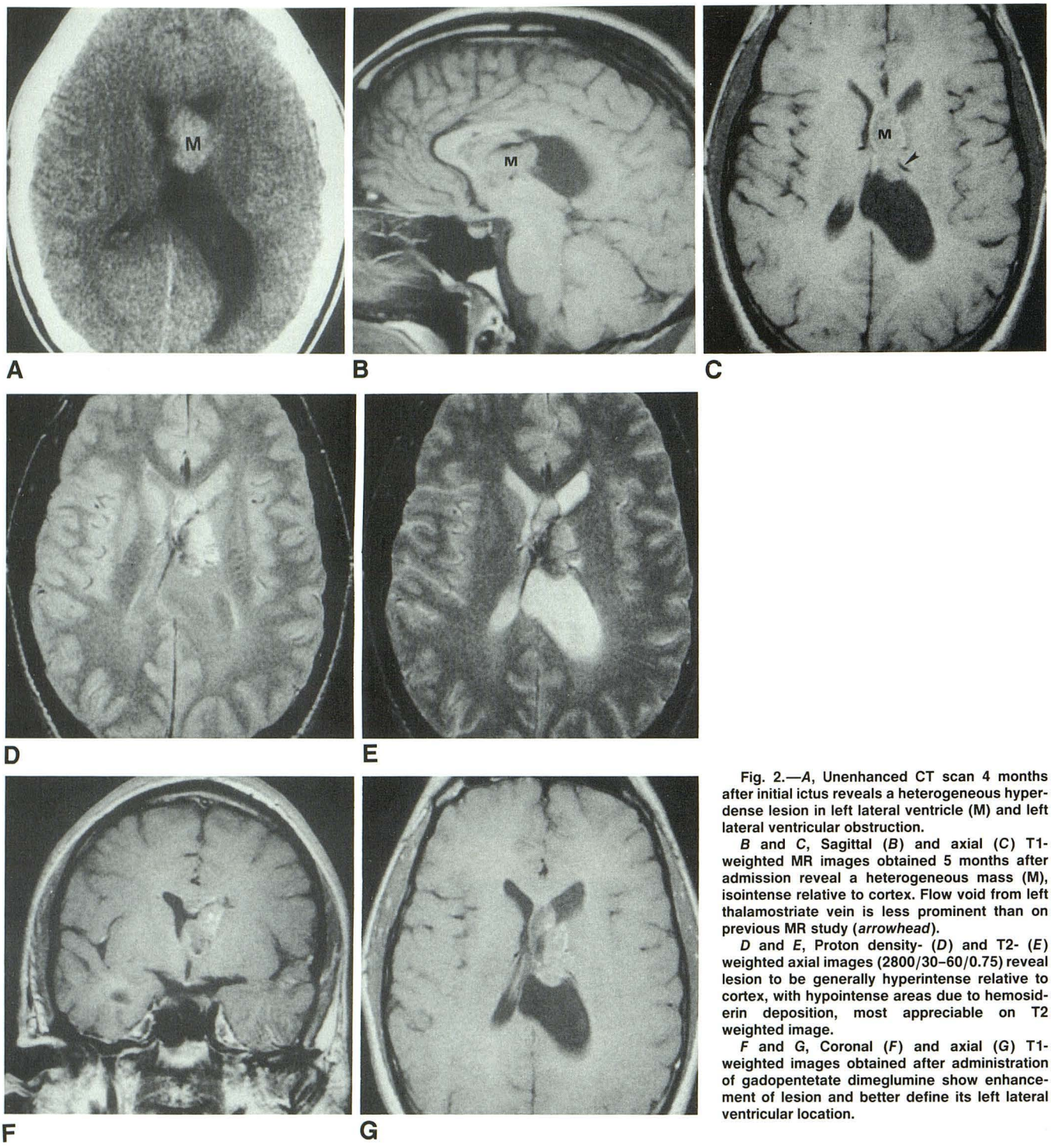

Fig. 2.—*A*, Unenhanced CT scan 4 months after initial ictus reveals a heterogeneous hyperdense lesion in left lateral ventricle (M) and left lateral ventricular obstruction.

***B* and *C*, Sagittal (*B*) and axial (*C*) T1-weighted MR images obtained 5 months after admission reveal a heterogeneous mass (M), isointense relative to cortex. Flow void from left thalamostriate vein is less prominent than on previous MR study (*arrowhead*).**

***D* and *E*, Proton density- (*D*) and T2- (*E*) weighted axial images (2800/30–60/0.75) reveal lesion to be generally hyperintense relative to cortex, with hypointense areas due to hemosiderin deposition, most appreciable on T2 weighted image.**

***F* and *G*, Coronal (*F*) and axial (*G*) T1-weighted images obtained after administration of gadopentetate dimeglumine show enhancement of lesion and better define its left lateral ventricular location.**

subject of this report presented with intraventricular hemorrhage.

Case Report

A previously healthy 26-year-old man was seen at an outside emergency department following a single grand mal seizure. His Glasgow coma scale score was 7. Unenhanced CT demonstrated a large intraventricular hemorrhage with acute obstructive hydrocephalus and subarachnoid hemorrhage (Fig. 1A). After transfer to our institution and bilateral ventriculostomies, cerebral angiography was performed, which demonstrated a small smooth aneurysm in the distal right vertebral artery, prominence of a left lenticulostriate artery, and early opacification of the left thalamostriate vein (Fig. 1B). There

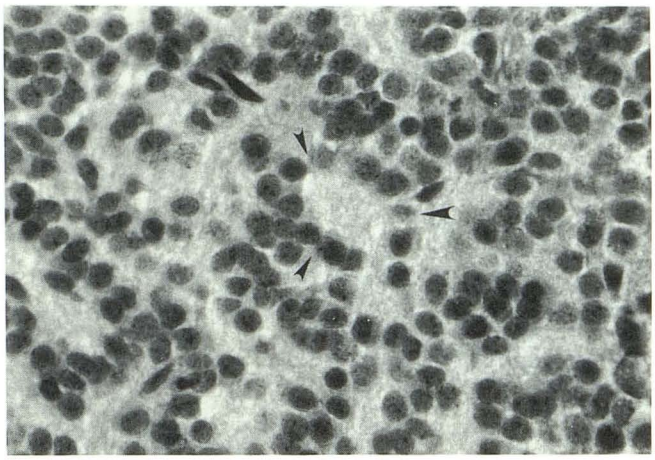

Fig. 3.—Light microscopic section reveals small, round, uniform nuclei within fibrillary background. Well-formed ring of nuclei is seen surrounding acellular fibrillary center. Wright rosette (*arrowheads*). (H and E, original magnification ×750)

was no angiographic evidence of neovascularity or late venous tumor blush. T1-weighted MR imaging, 600/20/1 (TR/TE/excitations), performed 5 days after the seizure, demonstrated intraventricular hemorrhage and provided no additional information (Figs. 1C and 1D). The diagnosis following these studies was a partially thrombosed intraventricular arteriovenous malformation.

Over the next 3 months the patient slowly made a complete neurologic recovery with the exception of a mild deficit in short-term memory. Serial follow-up unenhanced CT scans demonstrated progressive resolution of the intraventricular hemorrhage and suggested a left lateral ventricular mass (Fig. 2A). Angiography no longer demonstrated the prominent left lenticulostriate artery or the early draining thalamostriate vein. There was no change in the size or configuration of the vertebral artery aneurysm.

An MR study 5 months after initial presentation revealed a mass within the body of the left lateral ventricle (Figs. 2B–2G). The lesion was slightly heterogeneous and isointense relative to the cortex on the T1-weighted study and slightly hyperintense on the intermediate- and T2-weighted images. Tumor enhancement was seen after the administration of gadopentetate dimeglumine (Figs. 2F and 2G).

The patient underwent transcallosal biopsy and resection of the tumor. Light microscopic examination demonstrated small, round, symmetric nuclei forming multiple Wright rosettes (Fig. 3). Hemosiderin was present, but there was no calcification and mitoses were absent. Ultrastructural studies revealed numerous cell processes with neurofilaments and neurotubules, occasional dense core granules, and synaptic vesicles. Although well-formed junctional synapses were not present, the diagnosis of neurocytoma was believed to be appropriate because of the benign appearance of the cells.

Discussion

The terms neurocytoma, intracranial neuroblastoma, and differentiated neuroblastoma have been a source of confusion in recent literature. In 1982, Hassoun et al. [1] described two men who had calcified intraventricular tumors composed of small regular cells with clear cytoplasm that were grouped in clusters in a fibrillary stroma. Striking neuronal differentiation with numerous synapses was present on electron microscopic examination, and the term central neurocytoma was applied to these unusual tumors [1]. We were able to find only three other cases in the literature that satisfied both the light and electron microscopic criteria of neurocytoma (presence of synapses) as defined by Hassoun; those cases were reported by Townsend and Seaman [5], Poon et al. [6], and Bolen et al. [9]. Nishio et al. [7] reported six cases designated as neurocytoma and Patil et al. [10] reported one, even though well-formed synapses were not present on electron microscopy in any of these patients. Patil et al. described a second case of neurocytoma in which ultrastructural verification of synapses was not possible because electron microscopy was not performed. To add to the nosologic confusion, Jerdan et al. [2] reported two cases pathologically similar to those described by Hassoun et al., but, because they lacked synapses, they were termed differentiated neuroblastomas. Wilson et al. [4] reported a similar lesion under the same name in 1985. Ferreol et al. [8] discussed both parenchymal neuroblastomas and intraventricular neurocytomas in their review. The tumor in their case, designated as a neuroblastoma/neurocytoma, lacked synapses but was similar in other respects to the other neurocytomas reported. The three cases reported by Pearl et al. [3], under the heading primary cerebral neuroblastoma, were tumors with characteristics similar to those reported as neurocytomas by other authors.

It would appear from our review (see Table 1) that neurocytomas are primarily tumors of the lateral and third ventricles in patients over the age of 15 who have symptoms of increased intracranial pressure. These lesions may or may not be calcified. Of the 18 patients listed in Table 1 in whom CT was performed, calcification was demonstrated in 10. Angiographically, these lesions may be vascular or avascular in nature. On light microscopic examination, neurocytomas are composed of mature, small, regular, round neuronal cells with scant cytoplasm, not mature ganglion cells. Ultrastructural examination has revealed the absence of definite synapses in most of the reported cases. Follow-up studies in the majority of these cases have confirmed the benign nature of these tumors, whether or not synapses were present ultrastructurally. We agree with Nishio et al. [7] that these tumors with small, regular, benign-appearing cells should be termed neurocytomas, whether or not synapses are found in the ultrastructure.

Only three tumors reported before our present case were studied by MR imaging [9, 10]. Two were isointense relative to cortex on T1-, proton density-, and T2-weighted imaging while the third demonstrated slight hyperintensity on the proton density- and T2-weighted images. Two demonstrated large vascular flow voids. In our patient, who had an intraventricular hemorrhage, hyperintensity, as well as areas of prominent hypointensity due to the presence of hemosiderin, were seen on proton density- and T2-weighted images obtained 5 months after the ictus. Mild enhancement was seen after administration of gadopentetate dimeglumine.

The most impressive characteristic of these rare lesions is their benign biological activity. A review of the follow-up

TABLE 1: Demographic and Imaging Features of Neurocytomas Reported in the Literature

Reference: Term Used/ Case No.	Age (yr)	Sex	Symptoms at Presentation	Location	CT Findings	Angiographic Findings	MR Findings	Pathologic Findings	Treatment	Follow-up and Outcome
Hassoun et al. [1], 1982: central neurocytoma										
1	32	M	Progressive memory loss, lack of initiative, disorientation	Lateral and third ventricles	Huge calcified mass; no enhancement	Not performed	Not performed	Striking neuronal differentiation; numerous synapses	Total resection; radiation therapy (brain and cord)	Died from meningitis at 4 months; no tumor present
2	39	M	Progressive memory loss, increased ICP	Lateral and third ventricles	Huge calcified mass; no mention of enhancement	Not performed	Not performed	Striking neuronal differentiation; numerous synapses	Total resection	Alive at 24 months; no recurrence
Jerdan et al. [2], 1983: differentiated cerebral neuroblastoma										
3	23	M	Increased ICP	Lateral ventricle	No mention of calcification or enhancement	Not performed	Not performed	Dense core granules; no well-formed synapses	Biopsy only; treatment not described	Follow-up not reported
4	48	M	Increased ICP	Lateral ventricle	No mention of calcification or enhancement	Not performed	Not performed	Dense core granules; no well-formed synapses	Biopsy only; treatment not described	Follow-up not reported
Pearl et al. [3], 1985: primary cerebral neuroblastoma										
5	23	M	Increased ICP	Lateral ventricle	No mention of calcification or enhancement	Not performed	Not performed	No definite synapses	Subtotal resection; radiation therapy (whole brain)	Alive at 4½ years; no recurrence
6	52	F	Increased ICP	Lateral ventricle	Calcified mass; contrast enhancement	Little vascularity; blush into venous phase; enlarged posterolateral, posteromedial, and anterior choroidal arteries	Not performed	No definite synapses	Biopsy; radiation therapy (whole brain)	Alive at 6 months; no tumor growth but tumor cells in CSF
7	19	M	Increased ICP	Lateral ventricle	Calcified mass; contrast enhancement	No neovascularity, enlarged vessels, or tumor stain	Not performed	No definite synapses	Biopsy; radiation therapy (whole brain)	Alive; follow-up interval not reported
Wilson et al. [4], 1985: differentiated cerebral neuroblastoma										
8	25	M	Increased ICP	Lateral and third ventricles	No calcification; no mention of enhancement	"Large midline vascular tumor filling both lateral ventricles"	Not performed	No junctional synapses	"Debulking"	Died postoperatively from bleeding
Townsend and Seaman [5], 1986: central neurocytoma										
9	25	F	History of headache (dead on arrival)	Lateral and third ventricles	Not performed	Not performed	Not performed	LM: small, round nuclei, Wright rosettes, no mitoses; EM: unsuccessful	Not applicable	Dead on arrival
10	42	F	Drug overdose	Lateral ventricle	Calcification and contrast enhancement	Posterior choroidal artery supplying	Not performed	Well-defined synapses with junctions	Total resection	Alive at 12 months; no recurrence

						mass; no evidence of increased vascularity				
Poon et al. [6], 1988: primary cerebral neuroblastoma										
11	17	F	Increased ICP	Third ventricle	Calcification and contrast enhancement	Not performed	Not performed	Synaptic complexes with well-formed junctions seen throughout	Total resection	Alive at 7 months; no recurrence
Nishio et al. [7], 1988: central neurocytoma[a]										
12	15	M	Head trauma (incidental)	Lateral ventricle	–	Not performed	Not performed	–	Total resection; radiation therapy	Alive at 29 months; no recurrence
13	22	F	Increased ICP	Lateral and third ventricles	–	Not performed	Not performed	–	Partial resection; radiation therapy	Alive at 19 years; no residual tumor
14	22	F	Increased ICP	Lateral ventricle	–	Not performed	Not performed	–	Total resection	Alive at 4½ years; no recurrence
15	24	M	Increased ICP	Lateral and third ventricles	–	Not performed	Not performed	–	Total resection; radiation therapy	Alive at 15 months; no recurrence
16	30	M	Increased ICP	Lateral and third ventricles	–	Not performed	Not performed	–	Partial resection; radiation therapy	Alive at 15 months; no residual tumor
17	39	F	Incidental head trauma	Lateral ventricle	–	Not performed	Not performed	–	Partial resection; radiation therapy	Alive at 25 months; residual tumor persisted
Ferreol et al. [8], 1989: cerebral neuroblastoma (neurocytoma)										
18	66	M	Increased ICP (headache, dementia, ataxia)	Third ventricle	Calcification (hyperdense on unenhanced scan); dense enhancement	Not performed	Not performed	No mention of synapses on EM	Total resection	Alive at 4 years; no recurrence
Bolen et al. [9], 1989: central neurocytoma										
19	32	F	Increased ICP (severe headache)	Lateral ventricle	Large calcified mass; homogeneous enhancement	Avascular	T1: isointense relative to brain with areas of hypointensity (calcium); PD and T2: inhomogeneous increased signal intensity	Occasional synaptic terminals, numerous neuronal cell processes with microtubules and dense core vesicles	Subtotal resection	Follow-up interval not reported
Patil et al. [10], 1990: neurocytoma										
20	34	M	Increased ICP (nausea, vomiting, dizziness, intermittent visual loss), seizure	Lateral ventricle	Calcification; minimal enhancement	Not performed	T1: isointense relative to cortex; PD and T2: isointense relative to cortex, large vascular flow voids	Numerous neuritic processes with microtubules and dense core granules; no synapses present	Total resection	Follow-up interval not reported

21	27	F	Increased ICP (headache and blurred vision)	Lateral ventricle	No calcification or enhancement	Not performed	Isointense relative to cortex on all sequences; large serpiginous flow voids	EM not performed	Subtotal resection; radiation therapy	Alive at 12 months; no residual tumor
Present case, 1991: neurocytoma										
22	26	M	Intraventricular hemorrhage	L lateral ventricle	No calcification or enhancement	First study: prominent lenticulostriate artery and early opacification of thalamostriate vein; second study: neither finding was seen	T1: isointense relative to cortex; PD and T2: slight hyperintensity, inhomogeneous, mild contrast enhancement	Numerous cell processes with neurofilaments and neurotubules; occasional dense core granules and synaptic vesicles; no well-formed synapses	Total resection	Alive at 10 months; no recurrence

Note—ICP = increased intracranial pressure; T2 = T2-weighted images; T1 = T1-weighted images; PD = proton density-weighted images; LM = light microscopy; EM = electron microscopy; L = left.
[a] CT findings in the cases of Nishio et al. included calcification in one patient and slight to moderate tumor enhancement. Pathologic findings comprised a homogeneous population of cytologically bland cells, parallel bundles of microtubules, variable dense core vesicles, some simple dermosomes, and no well-formed synapses.

studies in the patients summarized in Table 1 revealed that none had died from tumor growth or recurrence, and survival time was as long as 19 years. This is in contrast to the typical parenchymal neuroblastoma occurring in young children, for which the overall 3-year survival rate is only 60% [11]. Neurocytomas usually arise from the septal region, to which they may be adherent [7]. Complete surgical resection, if possible, appears to be curative [5]. The benefits of adjunctive radiation therapy or chemotherapy in the treatment of these lesions is unclear.

Neurocytomas may be difficult to distinguish from other intraventricular tumors, such as ependymomas, oligodendrogliomas, or meningiomas, which may have similar characteristics on imaging studies. Although neurocytomas are uncommon intraventricular tumors and rarely bleed, they should be considered in the differential diagnosis in patients with intraventricular hemorrhage.

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