

revealed that only light perception in the left eye and hand motion in the right eye remained.

The usefulness of radiation therapy as adjunctive therapy for CNS gangliogliomas has been debated. In general, surgical resection with radiation therapy reserved for recurrence is recommended (8, 12, 13, 19). We recommended radiation to our patient for two reasons. First, only a biopsy was possible, and second, there is some evidence that the prognosis of gliomas of the chiasm and optic tract is poorer than that of gliomas of the optic nerve (1, 20). Our patient remains free of tumor progression 1 year after surgery.

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Intraventricular Neurocytoma: A Clinical and Pathological Study of Three Cases and Review of the Literature

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Three patients with a recently described tumor of neuronal origin, intraventricular neurocytoma, are presented. These tumors occur as intraventricular lesions in young patients, and the prognosis after surgical treatment is favorable. The initial pathological diagnosis of intraventricular neurocytoma may be difficult because of the striking resemblance of these tumors to oligodendroglioma and, to a lesser extent, ependymoma on light microscopic examination. Despite the use of wide-ranging panels of monoclonal antibodies, previous authors have not found any characteristic immunohistochemical staining patterns, but in our three patients, the use of synaptophysin, glial fibrillary acidic protein, and Leu-7 demonstrated staining patterns that may be useful in the diagnosis of this tumor. The monoclonal antibody Ki-67 was used to stain one tumor and showed a low cell proliferation rate. We have reviewed the clinical, radiological, and pathological features of these 3 patients and 17 previously described patients in an attempt to determine the important diagnostic features of intraventricular neurocytoma. Intraventricular neurocytoma should be considered in any young patient with symptoms of raised intracranial pressure and radiological evidence of an intraventricular lesion. Pathological diagnosis requires the use of electron microscopy to show features of neuronal differentiation; however, immunohistochemical demonstration of a neuronal phenotype is also a useful adjunct to diagnosis. Failure to use specialized techniques for pathological diagnosis will lead to misdiagnosis of these lesions as oligodendrogliomas, as was the initial diagnosis in 2 of our patients before review. (*Neurosurgery* 26:1045-1054, 1990)

Key words: Brain neoplasm, Electron microscopy, Intraventricular neoplasm, Immunohistochemistry, Neurocytoma

INTRODUCTION

Intraventricular tumors previously have been considered to arise principally from ependyma or choroid plexus cells and only rarely from neuronal cells (6, 19). Intraventricular neurocytoma, a well-differentiated tumor of neuronal histogenesis distinct from ganglion cell tumors and neuroblastoma, was first described in 1982 by Hassoun et al. (6), who presented two cases. The main features they described were those of a primary intraventricular tumor composed of small uniform cells in a patchy fibrillary stroma, with extensive calcification and showing neuronal differentiation associated with synapse formation on electron microscopic examination. Since then a small number of additional cases have been described in the literature and importantly, this lesion has been associated with a good prognosis. Because of the difficulty in distinguishing these tumors from oligodendroglioma and ependymoma, however, it is possible that cases are missed and that the reported number does not reflect the true incidence (8, 11, 14, 15, 19). There is also discussion about the nomenclature of these tumors with possible overlap between the neurocytoma and neuroblastoma. In an attempt to broaden discussion on these points and provide additional data, we present clinical and pathological details for three patients seen at the Queens Medical Centre, University of Nottingham Medical School, between 1985 and 1989. Two of these tumors were diagnosed on review after having initially been called oligodendrogliomas, which illustrates the importance of specialized techniques for diagnosis.

MATERIALS AND METHODS

Three patients from the University Hospital, Nottingham, who had intraventricular tumors form the basis of this report. After the initial clinical assessment, radiological investigation consisting of computed tomographic (CT) scan and magnetic resonance imaging (MRI) (two patients) scans of the brain was carried out, followed by surgery.

The material for pathological examination obtained at surgery was divided for smear examination, cryostat sectioning, electron microscopy, and routine processing. Smears and cryostat sections were examined at the time of operation. Samples for electron microscopy were fixed in 2% glutaraldehyde and embedded in Epon. Ultrathin sections were stained with uranyl acetate and lead citrate and examined by transmission electron microscopy. The remaining tissue was fixed in 10% formalin and embedded in paraffin. Sections of 5 μ m in thickness were cut and stained with hematoxylin and eosin. For immunohistochemistry paraffin sections were prepared with primary antisera directed against synaptophysin (ICN Biomedicals, Bucks, England), glial fibrillary acidic protein (GFAP; DAKO, Bucks, England), protein gene product 9.5 (Ultraclone, Cambridge, England), Leu-7 (Becton Dickinson, Oxford, England), and neurofilament protein (NFP; Milab, Malmö, Sweden). Cryostat sections were also stained using the monoclonal antibody Ki-67 (DAKO) to determine the cell proliferation index (20, 21).

CASE REPORTS

Case 1

A 17-year-old female adolescent was seen in March 1985 with a 2-month history of headaches and diplopia. The only positive finding on neurological examination was the presence of bilateral papilledema. CT scanning demonstrated a well-defined mass of increased attenuation situated in the body of the left lateral ventricle at the level of the foramen of Monro.

The lesion abutted against the septum pellucidum and bulged across into the right lateral ventricle (Fig. 1, *A* and *B*). Marked lateral ventricular enlargement was present, particularly on the left. Small cystic areas and punctate calcifications were present within the mass. There was moderate heterogeneous enhancement after injection of contrast material.

At operation the patient had subtotal removal of the lesion performed through an anterior transcallosal approach. The tumor was soft and attached to the septum pellucidum. On the basis of initial light microscopic examination only, a diagnosis of low-grade oligodendroglioma was made. Postoperatively she made an uneventful recovery and received postoperative radiotherapy. Four years later she is symptom free and has no signs of neurological disease. On pathological review with ultrastructural studies in 1988, the diagnosis was revised to intraventricular neurocytoma.

Case 2

A 25-year-old woman was seen in 1987 with a 3-month history of headache. As in Case 1, the only positive finding on neurological examination was bilateral papilledema. CT scanning demonstrated a relatively well-defined mass of pre-

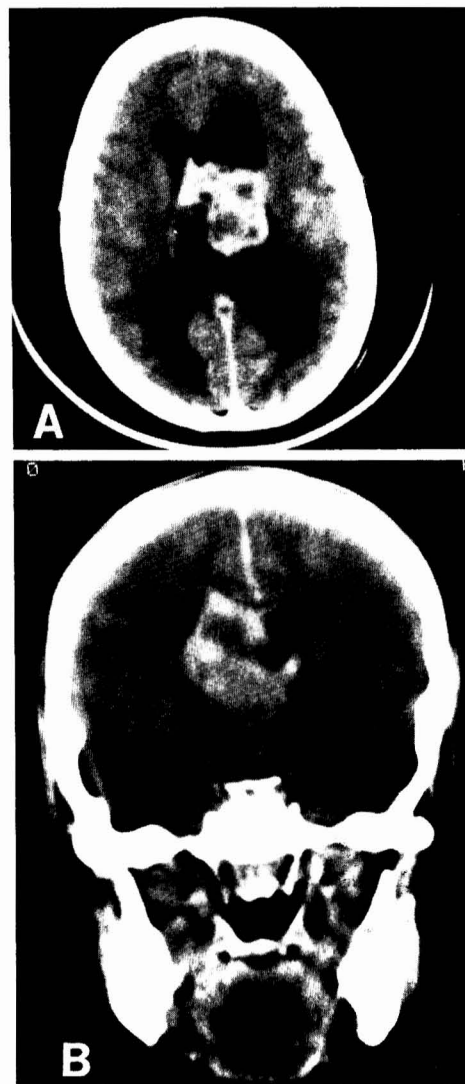


FIG. 1. *A*, axial CT scan. Note cystic areas and dense white calcified areas. *B*, coronal CT scan, which better demonstrates the biventricular involvement.

dominantly increased attenuation, situated in the region of the foramen of Monro and extending into both lateral ventricles, the left more than the right. There were several areas of low attenuation but no calcification within the lesion. Little enhancement was noted after injection of contrast material. There was an associated hydrocephalus.

At surgery subtotal removal of the tumor was performed through an anterior callosal approach. The tumor was seen to be occupying the foramen of Monro and both lateral ventricles. On the basis of initial light microscopic examination only, a diagnosis of low-grade oligodendroglioma was made. The patient's postoperative recovery was uneventful, and she underwent a course of radiotherapy. Twenty months after surgery she remains symptom free. On pathological review with ultrastructural studies in 1988, the diagnosis was revised to intraventricular neurocytoma.

Case 3

A 26-year-old man was seen in 1989 with a 3-month history of visual disturbance, which he described as seeing "white lines crossing the road" while driving. Decreased visual acuity was also noted in the right eye 3 weeks before admission. This strange visual disturbance settled after 10 days of rest, but the reduction in visual acuity persisted.

Neurological examination on admission revealed bilateral papilledema and reduced visual acuity (6/24) in the right eye. The CT scan showed a large, slightly hyperdense mass situated predominantly in the right lateral ventricle with generalized ventricular system enlargement (Fig. 2A). Heavy amorphous calcifications were present centrally with small cystic foci peripherally. The lesion exhibited mild heterogeneous enhancement after injection of contrast material (Fig. 2B). MRI scanning (Picker 0.15T resistive unit) demonstrated more clearly the tumor to be applied to the septum pellucidum and bowing across the midline into the body of the left lateral ventricle (Fig. 3A). The lesion was isointense with brain on the T1-weighted axial spin echo images with internal foci of low signal corresponding to the cystic and calcific areas shown on the CT scan. On the coronal inversion recovery images the lesion intensity was shown to be slightly reduced relative to the adjacent brain (Fig. 3B). In the light of our experience with Cases 1 and 2 above, a diagnosis of neurocytoma was suggested on the basis of the radiology.

At surgery subtotal removal of the tumor was performed through an anterior transcallosal approach. A variegated mass based on the septum pellucidum was found to be occupying the foramen of Monro and the right lateral ventricle with minimal involvement of the left lateral ventricle. The diagnosis of intraventricular neurocytoma was confirmed on the basis of light microscopic and ultrastructural findings. He has not received postoperative radiotherapy so far and 2 months after surgery remains well apart from a reduction in visual acuity in his right eye.

PATHOLOGICAL FINDINGS

Light microscopic and immunohistochemical findings

Smear preparations from the three surgical specimens showed sheets of monotonous cells with small, uniform, rounded nuclei and small inconspicuous nucleoli. No mitoses were seen. The cytoplasm was indistinct and had a vacuolated or fibrillar appearance. Capillary sized blood vessels were a prominent feature and occasional acellular fibrillary areas were seen. Flecks of calcific material were present in smears and sections in two specimens (Figs. 4 and 5).

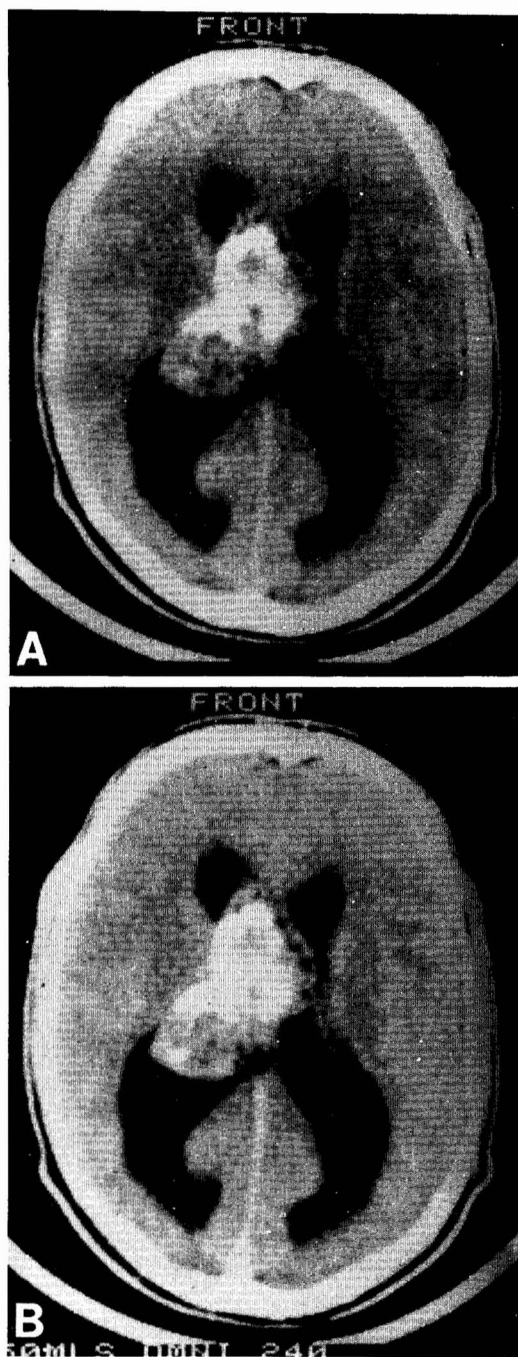


FIG. 2. A, axial CT scan showing heavy calcifications with peripheral small cysts. B, scan after injection of contrast material demonstrating mild enhancement.

Paraffin sections showed a uniform population of closely packed small cells as described above with a fine arborizing vascular supply. Prominent perivascular acellular eosinophilic fibrillar areas, some resembling pseudorosettes, gave a vaguely trabecular appearance on low-power examination and resembled the nuclear free perivascular zones seen in ependymomas (Fig. 5A). Ill-defined true rosettes with central fibrillar acellular material were also present (Fig. 5B). No mitoses were seen, and there was no necrosis. Calcification was confirmed, the deposits being equal in size to or larger than the constituent cells and some had the laminated appearance of psammoma bodies.

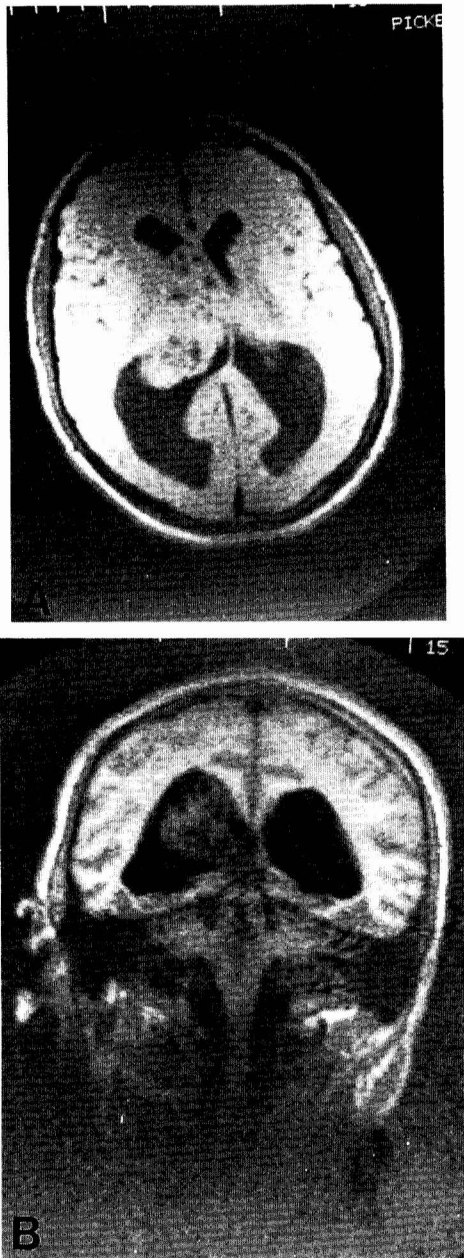


FIG. 3. *A*, axial MRI scan (SE 700/40). Note the relationship of the mass to the septum pellucidum. *B*, coronal scan (IR 1660/400) showing biventricular enlargement. The mass has a reduced signal intensity.

Immunohistochemical reactions were performed as follows. Synaptophysin staining showed diffuse positivity of intercellular fibrillar areas with a punctate and granular pattern at high power in two of the three tumors. Occasional cells showed very dense cytoplasmic staining (Fig. 6). GFAP staining was positive in scattered astrocytes with processes running within the tumor but not within tumor cells themselves (Fig. 7). The intercellular fibrillar areas, including the rosette-like areas, were strongly positive with Leu-7, but the cell cytoplasm was only very weakly positive (not illustrated here). No NFP positivity was detected using the antibody described. Protein gene product 9.5 stained occasional scattered cells in a pattern

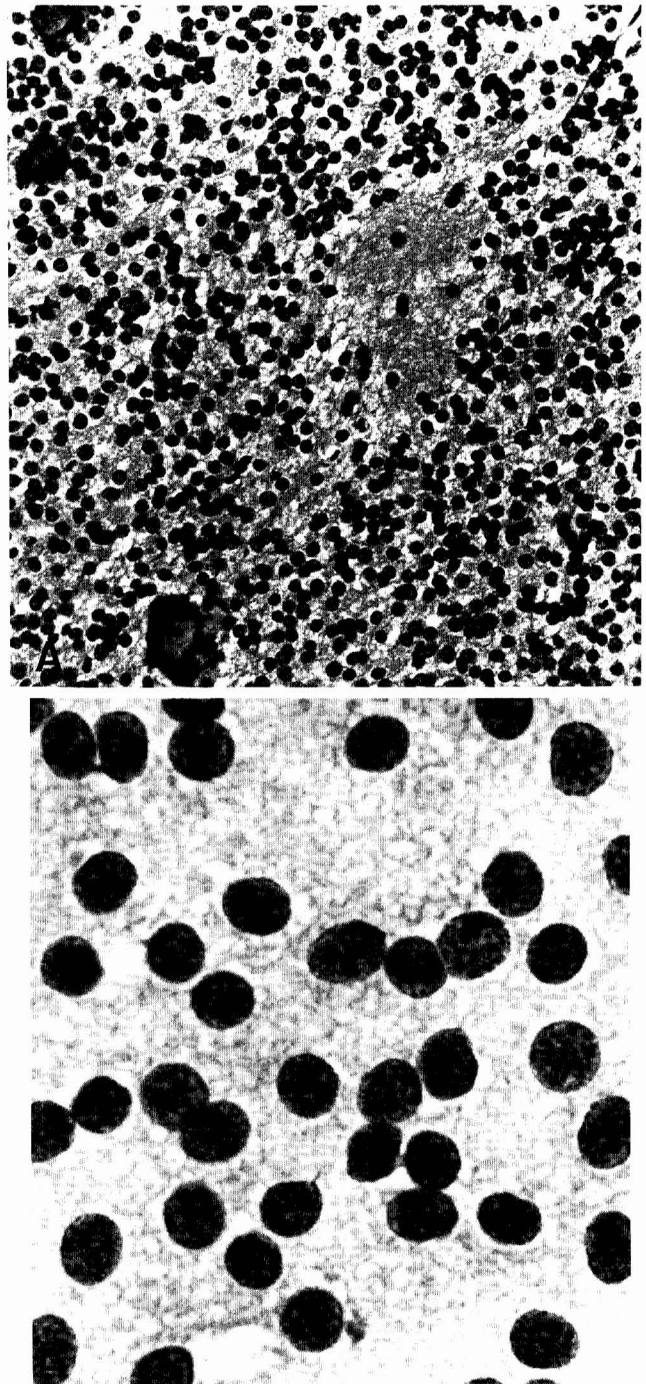


FIG. 4. *A*, smear preparation of the tumor showing a uniform population of cells with rounded nuclei, a fine fibrillar background occasionally forming acellular areas, and focal calcification (hematoxylin and eosin smear, $\times 250$). *B*, smear preparation showing cytological detail of cells with rounded nuclei, indistinct cytoplasm, and no delineation from the fibrillar background. The nuclei have finely dispersed chromatin with several small indistinct nucleoli (hematoxylin and eosin smear, $\times 1350$).

suggestive of staining of the included astrocytes but did not stain the tumor cells or processes. In one tumor, the Ki-67 index indicated that less than 1% of tumor cells were proliferating (Case 3, Fig. 8).

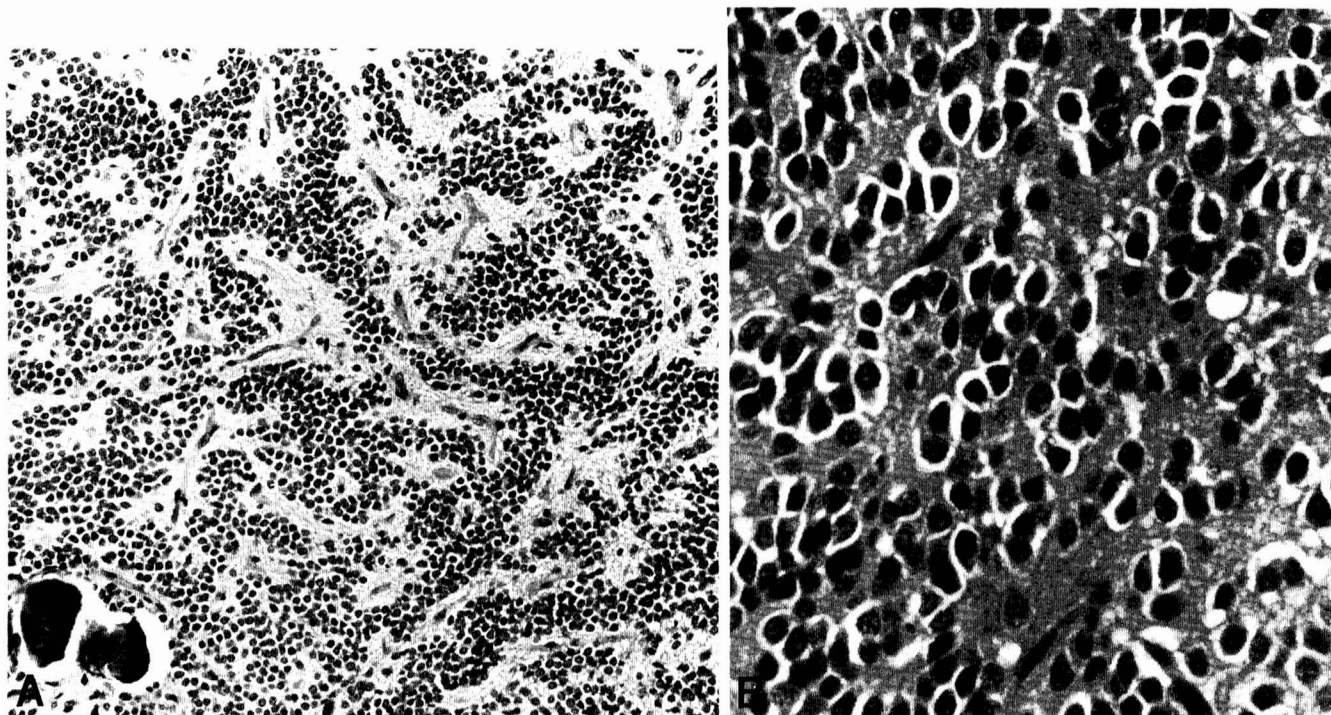


FIG. 5. *A*, paraffin section showing perivascular pseudorosette formation, linear arborizing fine vascular pattern, and focal calcification (bottom left) (hematoxylin and eosin, $\times 185$). *B*, higher power detail shows that true rosette formation is subtle but just evident. Note perinuclear vacuolation reminiscent of that seen in the oligodendroglioma (hematoxylin and eosin, $\times 920$).

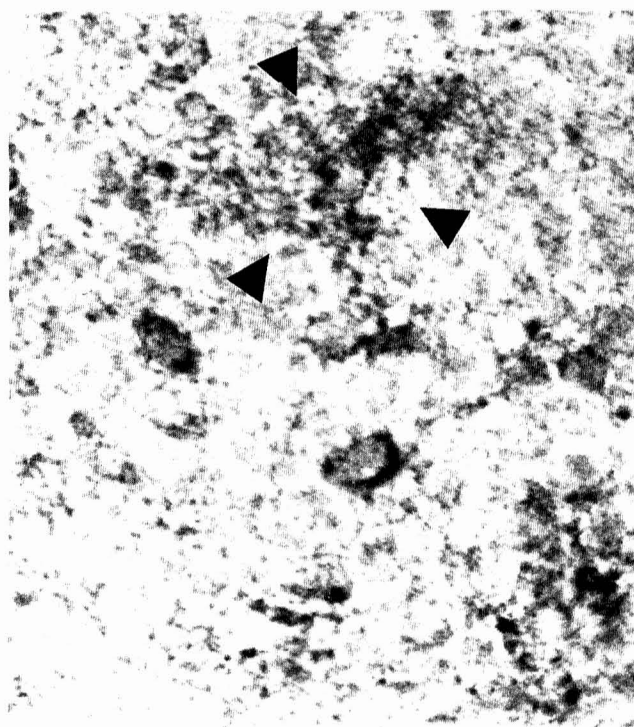


FIG. 6. Synaptophysin is seen as a fine granular intercellular punctate pattern particularly in the ill-defined true rosette areas (arrowheads) with occasional cells exhibiting perinuclear cytoplasmic positivity ($\times 774$).

Electron microscopy

Electron microscopic examination confirmed that the tumors were composed of a monomorphic cell type. Neuronal differentiation was strongly evident; the cytoplasm contained numerous neurosecretory granules, mainly dense core vesicles but also clear vesicles. Numerous dendritic cytoplasmic processes were also present, containing vesicles as well as parallel bundles of microtubules. Occasional junctional complexes were noted, but no definite synapses were identified (Fig. 9).

DISCUSSION

The intraventricular neurocytoma is a recently described tumor of neuroepithelial origin which on light microscopic examination may mimic oligodendroglioma or ependymoma. Other recognized central nervous system tumors of neuronal origin are infrequent, and those described usually occur in children in the cerebral parenchyma (6, 7, 9, 11, 14, 17, 18). It has been suggested that this tumor may represent a differentiated form of neuroblastoma; however, in contrast to neuroblastoma, the neurocytoma is composed of monotonous small cells with little pleomorphism, no necrosis, and no mitoses; they also are associated with a good clinical prognosis (11, 14, 19, 20). Our findings are in keeping with previously reported tumors, variously named, with the common feature of being mature neuronal tumors of the ventricular region (6, 8, 11, 13-15, 19, 20). These tumors not only pose problems for nomenclature but also in differential diagnosis, both clinically and pathologically.

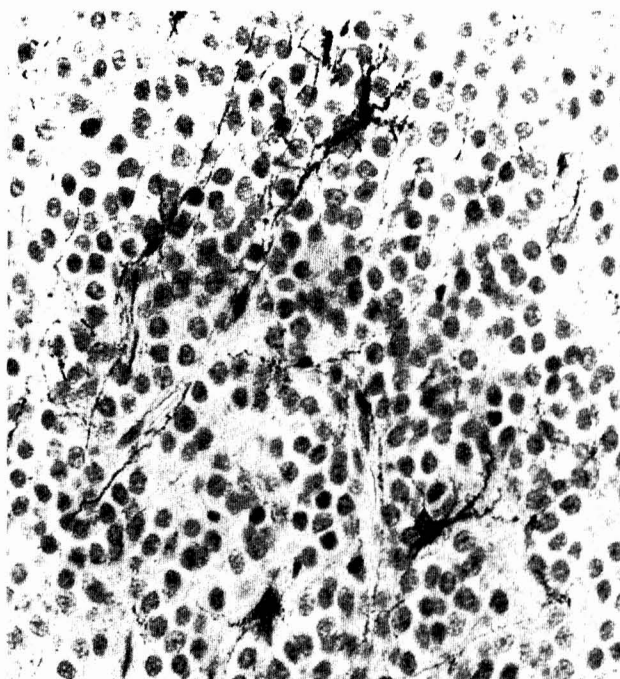


FIG. 7. GFAP-positive staining of occasional astrocytes within the tumor. The tumor cells are negative ($\times 470$).

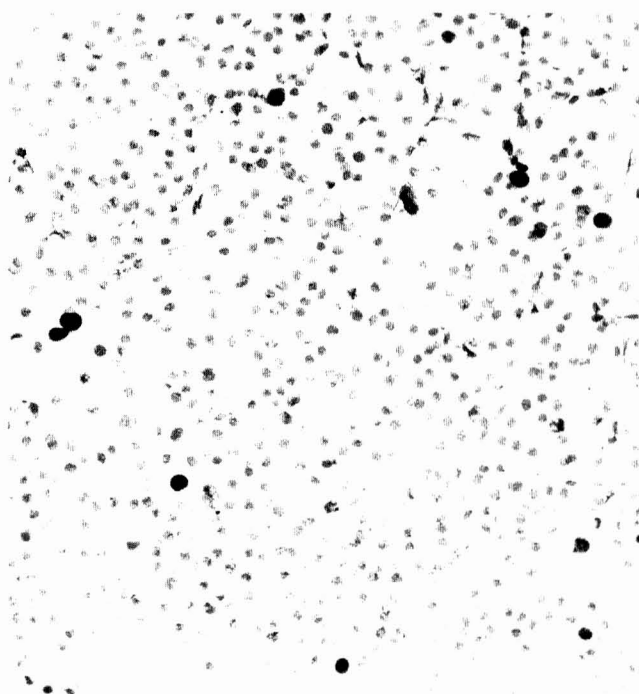


FIG. 8. Less than 1% of tumor cells show positive for Ki-67, seen as densely stained nuclei in this di-aminobezine/hematoxylin preparation ($\times 250$).

Nomenclature

Previous authors who have reported tumors with the features we have described above have used various terms, thus adding to the difficulty of accurately determining the number of cases in the literature. These terms include *intraventricular*

neurocytoma, *intraventricular neuroblastoma*, and *differentiated neuroblastoma* (6, 8, 11, 13, 19, 20). Hassoun et al. (6), who first described these tumors, introduced the term *neurocytoma*, in view of the striking feature of well-defined synapses on electron microscopy. Nishio et al. (11) maintained the term *intraventricular neurocytoma* for their tumors, despite the lack of synapses. Jerdan et al. (8) and Wilson et al. (20) described tumors with similar features except for the lack of synapses and they called their tumors *differentiated neuroblastomas*. Poon et al. (15) did identify synapses but describe their tumor as an *intraventricular neuroblastoma*. Pearl et al. (13) also use this term. They failed to find synapses, but in one case they describe the presence of asymmetrical junctions suggestive of abortive synapses. We believe that in order to avoid nosological confusion and to stress the benign biological behavior of these tumors, which have features characterizing a differentiated neuronal lesion, they should be termed *intraventricular neurocytoma*. This is in keeping with the proposals of Hassoun et al. (6) and Nishio et al. (11). In our opinion the presence or absence of synapses in these lesions seems academic, since, provided that the tumors correspond to a small cell but mitotically inactive neuronal lesion, they are associated with a good prognosis.

Accepting differences in terminology, we have reviewed 17 cases of this type of tumor that have been reported since 1982 (Table 1). Despite reported variability in finding synapses, these tumors have a similar clinical and pathological pattern, which justifies grouping as a distinct entity. The cases reported are as follows: Hassoun et al. (6), Cases 1 and 2; Jerdan et al. (8), Cases 3 and 4; Wilson et al. (20), Case 5; Pearl et al. (13, 14), Cases 6, 7, and 8; Townsend and Seaman (19), Cases 9 and 10; Nishio et al. (11), Cases 11 to 16; Poon et al. (15), Case 17. We compare these with our three cases (Cases 18 to 20).

Clinical findings of reported cases

Neurocytomas affect a relatively young population with an age range of 15 to 52 years (average, 28 years). The sex ratio is 11:9 (male:female). The presenting symptoms vary but most common are those related to raised intracranial pressure with headache (77%), nausea, and vomiting being the most frequent complaints. Symptoms ascribed to hydrocephalus may also be present, i.e., loss of memory, apathy, and disorientation. Visual symptoms were found in a small number of patients and consisted of diplopia, blurring of vision, and decreased visual acuity. The duration of symptoms is variable, but they have usually been present for several months at the time the patient is seen (range, 2 to 6 months). The variation in the symptomatology resulting from these lesions is illustrated by two patients in whom the tumor was asymptomatic and discovered as an incidental finding on the CT scan after head injury (Cases 11 and 16), and by one patient in whom the sudden onset of coma was the presenting feature (Case 10). Neurological examination demonstrates no specific features apart from those attributable to space occupancy and the tumor location.

Radiological findings in reported cases

The radiological studies are based on CT scans and, in one of our patients, MRI scans, which typically revealed an intraventricular mass, occupying one or both lateral ventricles with or without third ventricle involvement. One tumor was restricted to the third ventricle. CT scanning demonstrated normal or slightly increased attenuation, and there was slight to moderate enhancement after injection of contrast material.

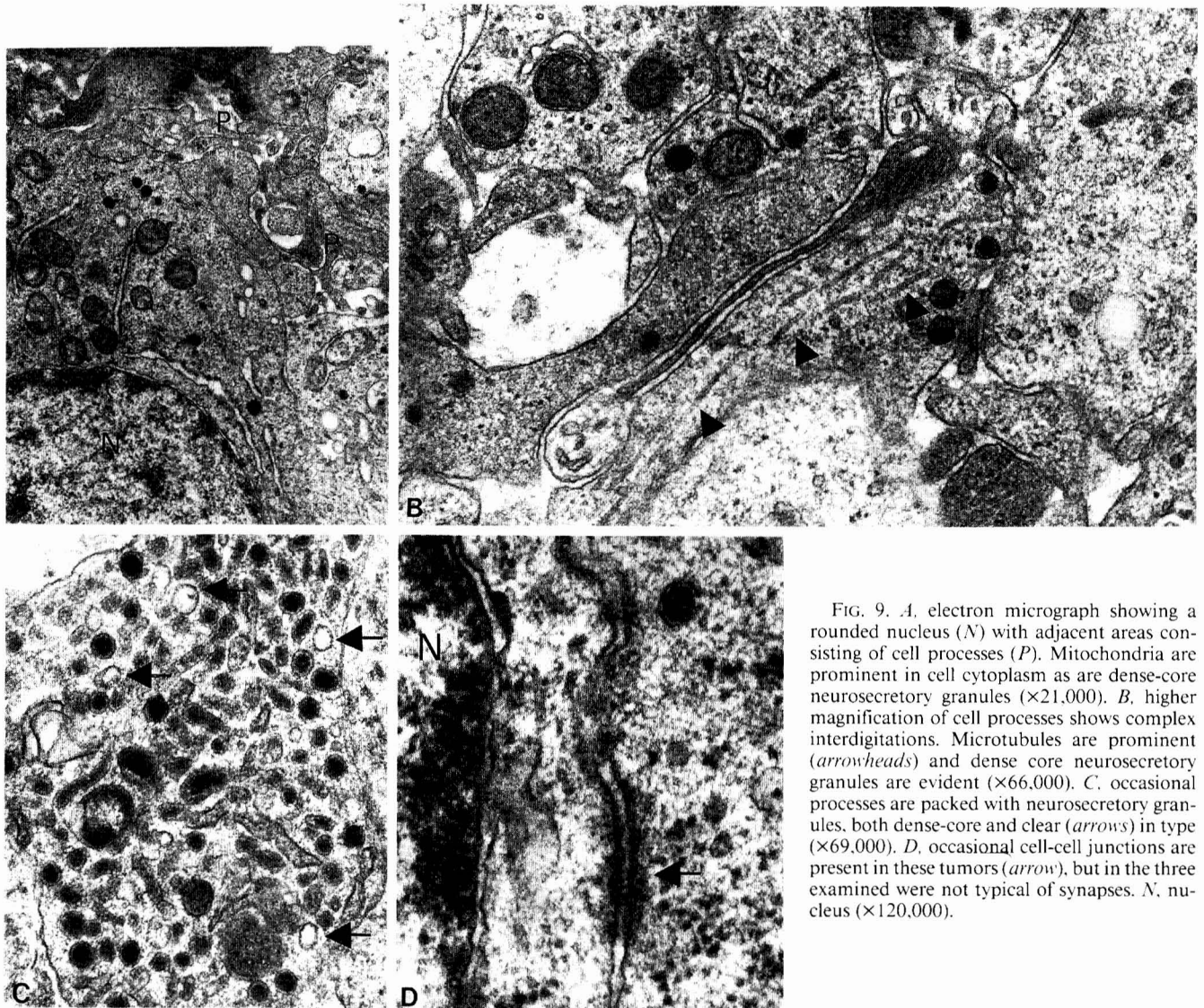


FIG. 9. *A*, electron micrograph showing a rounded nucleus (*N*) with adjacent areas consisting of cell processes (*P*). Mitochondria are prominent in cell cytoplasm as are dense-core neurosecretory granules ($\times 21,000$). *B*, higher magnification of cell processes shows complex interdigitations. Microtubules are prominent (arrowheads) and dense core neurosecretory granules are evident ($\times 66,000$). *C*, occasional processes are packed with neurosecretory granules, both dense-core and clear (arrows) in type ($\times 69,000$). *D*, occasional cell-cell junctions are present in these tumors (arrow), but in the three examined were not typical of synapses. *N*, nucleus ($\times 120,000$).

Areas of calcification and of cystic change within the tumor are frequently found, and associated hydrocephalus is an almost constant feature. The presence of an intraventricular mass lesion with these characteristics in a patient in the described age range should raise the possibility of the lesion being a neurocytoma.

Pathological findings in reported cases

1. Light microscopy. These tumors typically have a uniform monotonous appearance on light microscopic examination, being composed of cells with regular small nuclei. Cellular pleomorphism and mitoses are not features of these tumors although rare mitoses have been described. While most of the cell nuclei are small and rounded, occasional nuclei are slightly larger and have a single large nucleolus reminiscent of ganglion cell maturation. For the most part cells are evenly dispersed in a finely fibrillar stroma; however, there may be ill-defined rosettes and there may be conspicuous perivascular pseudorosette formation with nuclear free zones of fibrillarity around small vessels. The vessels within these tumors are small, of even caliber, and tend to be arranged in

a linear arborizing pattern. Microcalcification is commonly present. In formalin-fixed tissue, perinuclear cytoplasmic vacuolation may occur in tumor cells.

In considering differential diagnosis, two other tumors occur in the region of the ventricles and also have a uniform cell population that may mimic neurocytoma. These are oligodendroglioma and ependymoma.

Oligodendrogliomas, like neurocytomas, are composed of cells with evenly spaced, rounded nuclei. Both tumors have a vascular pattern in which small straight arborizing vessels with a nuclear free perivascular zone are a conspicuous feature. In addition, both tumors may exhibit artefactual vacuolation of cytoplasm seen after formalin fixation, which gives rise to the "poached egg" appearance of cells. The fact that both tumors commonly exhibit calcification means that this feature is not helpful in differential diagnosis. On light microscopic examination and especially on smear and cryostat sections, we believe that the differentiation of neurocytoma from oligodendroglioma is very difficult. The finding of true neuroblastic rosettes, nuclei with a slightly ganglionic appearance, and ill-defined cell boundaries in the presence of a lesion related to the ventricles should strongly suggest the diagnosis

TABLE 1
Clinical Features of Patients with Intraventricular Neurocytoma

Case Number	Age (yr)/Sex	Symptoms ^a	Site ^b	Radiotherapy	Follow-up ^c
1	32/M	Frontal syndrome	R, III	No	Died 14 months after surgery, no evidence of recurrence
2	39/M	Frontal syndrome, RICP	R, L, III	?	Well at 2 yr
3	23/M	RICP	R, L	?	?
4	48/M	RICP	R, L	?	?
5	25/M	Frontal syndrome, headaches	R, L	No	Died postoperative day 2 (intraventricular hemorrhage, hydrocephalus)
6	23/M	Visual haziness (L), headaches	L	Yes	No increase in tumor size after 4 yr, 6 mo
7	52/F	Neck pain, fatiguability	L	Yes	No increase in tumor size after 6 mo
8	19/M	Nausea, headaches, diplopia	?L	Yes	?
9	25/F	Headaches	R, III	No	Autopsy diagnosis
10	42/F	Sudden coma	R	?	Well at 2 yr
11	15/M	Asymptomatic	?L	Yes	Well at 2 yr, 5 mo (K 100)
12	22/F	Headaches	?L, III	Yes	Reasonably well at 18 yr, 11 mo (K 80)
13	22/F	Headaches	?L	No	Moderately disabled at 4 yr, 8 mo (K 50)
14	24/M	Headaches	R, L, III	Yes	Reasonably well at 1 yr, 3 mo (K 80)
15	30/M	Headaches	R, L, III	Yes	Well at 1 yr, 3 mo (K 90)
16	39/F	Asymptomatic	R, L	Yes	Moderately disabled at 2 yr, 1 mo (K 50)
17	17/F	Headaches, amenorrhea	III	No	No residual tumor at 7 mo
18	17/F	Diplopia, headaches	R, L	Yes	Well at 4 yr
19	25/F	Headaches	R, L	Yes	Well at 2 yr
20	26/M	Hallucinations, decreased visual acuity	R, L	No	Well at 1 yr, 5 mo, but decreased right visual acuity

^a RICP, raised intracranial pressure; L, left.

^b R, L, and III refer to involvement of right lateral, left lateral, and third ventricles, respectively.

^c K, Karnofsky score.

of intraventricular neurocytoma, which must be confirmed by immunohistochemistry and electron microscopy.

Ependymoma also commonly occurs in an intraventricular position, usually the fourth ventricle. Again, ependymomas have features in common with neurocytomas, being composed of small cells with nuclear free fibrillar zones around vessels. Calcification is present in 15% of ependymomas and in a lesion without characteristic ependymal tubules there may be a striking similarity with neurocytoma.

Immunohistochemistry may provide useful information with which to make a diagnosis and distinguish these superficially similar tumors occurring around the ventricles. Previous authors have used various immunohistochemical markers, but no helpful positive staining pattern has been described. Two authors describe neuron-specific enolase positivity as a characteristic of neurocytomas, but, contrary to earlier beliefs, it has been found that this enzyme is not specific for tumors of neuronal origin and is of limited value unless interpreted in conjunction with other immunohistochemical reagents (2). GFAP has been used by almost all authors and a consistently negative staining pattern has been found, demonstrating that there is no glial differentiation in these tumors, although scattered non-neoplastic astrocytes were present in three cases (11, 15). This feature was also seen in our three patients. Widespread tumor cell positivity with GFAP staining would mitigate against a diagnosis of neurocytoma but may be seen in ependymomas or occasionally in oligodendroglioma. Nishio et al. (11) describe NFP positivity, but we have not demonstrated that. The fact that neurofilament expression may be one of three molecular weight species and furthermore be subject to post-translation modification by phosphorylation means that any single anti-NFP antibody may not be detecting all forms of expression. Synaptophysin staining was positive in these tumors in two cases examined.

This is a recently described membrane glycoprotein of presynaptic vesicles and its presence is specific for neuroendocrine tumors of low- and high-grade malignancy, unlike NFP which is present in only a proportion of these lesions (3-5, 10). Synaptophysin has not previously been assessed in neurocytomas, but the striking staining pattern we found indicates its usefulness in establishing a definite neuronal phenotype. We attribute the lack of staining in one case to poor fixation as immunoreactivity is lost with long formalin fixation. Previous authors have commented on improved immunostaining of paraffin-processed material if Bouin's fluid rather than formalin is used as a fixative (3, 10). Leu-7 staining was seen in all three neurocytomas, and we did not find the pattern helpful in differential diagnosis.

The proliferation index of the intraventricular neurocytoma was studied using the monoclonal antibody Ki-67, which is directed against a nuclear antigen present only in proliferating cells in the G₁, S, G₂, and M phases of the cell cycle (1, 12, 16, 21). The one tumor examined had a very low proliferation rate (<1%), comparable to benign tumors such as meningioma, craniopharyngioma, pituitary adenoma, and neurinoma and in contrast to high proliferation rates seen in primitive neuroectodermal tumors and glioblastoma multiforme (1, 12, 21).

2. Ultrastructure. In our hands, the most reliable and useful diagnostic procedure is electron microscopy, which confirms that these tumors have a uniform neuronal cell population. The nuclei are rounded and contain finely dispersed chromatin and a conspicuous nucleolus (not always visualized). The cytoplasm is sparsely populated and contains rough endoplasmic reticulum, polysomes, mitochondria, and Golgi apparatus. Large numbers of microtubules, rarely present in tumors other than of neuronal origin, are a feature. Membrane-bound dense core vesicles are abundant. Numerous

cytoplasmic processes are present between cells and these contain parallel bundles of microtubules, dense core vesicles, and some clear vesicles. In other cases abortive or well-defined synapses have been described (6, 11, 14, 15, 19). It has been suggested that the presence of synapses could represent a more mature form of the tumor and therefore indicate a better prognosis (11, 15, 19). In those cases where they have been identified, only a small number of synapses are present; therefore, the possibility of sampling error must be considered in the cases where they are not found.

Management and prognosis in reported cases

Details of the surgical procedure are not given for all of the previously reported patients. The most usual finding at operation is that of a relatively well-circumscribed intraventricular mass without obvious invasion of cerebral parenchyma and most frequently attached to the septum pellucidum. In one patient (Case 9), in whom diagnosis was made at postmortem examination, a soft friable mass was found in the right lateral and third ventricles, attached to the septum pellucidum and the corpus callosum; indeed it has been proposed that the septum pellucidum is the site of origin of these tumors (6, 19). The surgical removal was total in 7 patients, subtotal in 10, and not reported in 2 patients. The completeness of tumor removal appears unrelated to the site of ventricular involvement.

Patient follow-up is summarized in Table 1, which shows that 50% of patients received postoperative radiotherapy. In one of our patients (Case 19), there has been evident shrinkage in the residual tumor bulk after radiotherapy, documented by scans taken before and after radiotherapy scans. The majority of patients have an excellent prognosis, including one patient (Case 12) who has been followed for 18 years and 11 months. Only one recurrence has been reported so far (11).

CONCLUSIONS

Intraventricular neurocytomas are uncommon tumors, consisting of mature neuronal cells and occurring in a young population. The usual mode of presentation is with symptoms of raised intracranial pressure; however, as more cases are recognized, it is probable that a wider range of presenting signs and symptoms will become apparent. The appearance on light microscopy closely mimics oligodendroglioma or occasionally ependymoma and a definitive diagnosis should be based on immunohistochemistry and electron microscopy. As illustrated by the retrospective diagnosis in two of our patients, awareness of the diagnostic possibility coupled with electron microscopy are the two most important adjuncts to identifying such tumors. It is likely that many tumors diagnosed as intraventricular oligodendrogliomas are in fact neurocytomas. Although only a small number of cases have been reported and the follow-up periods are short, the available data suggest that these tumors follow a benign course and have a good prognosis after surgical treatment.

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COMMENT

The authors describe and review a recently reported new tumor entity, the intraventricular neurocytoma, which characteristically occurs in young adults and for which the prog-

nosis appears favorable. These tumors have usually been diagnosed as oligodendrogliomas, but their neuronal nature is documented by electron microscopy. Because of the characteristic differential diagnosis, age of onset, and location of this new tumor type, it is important that it be recognized by neurosurgeons and neuropathologists, and that the necessity of doing electron microscopy in such cases to establish the neuronal features be emphasized. The authors rightly believe that the tumor is probably a well-differentiated cerebral neoplasm and that separate designation as an intraventricular central neurocytoma is important.

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Carcinoma Metastatic to a Cerebellar Vascular Malformation: Case Report

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A 73-year-old woman with a spontaneous intracerebellar hemorrhage was seen and was found to have metastatic adenocarcinoma within a vascular malformation. There was no evidence of other metastatic disease. The association of these two lesions is uncommon and has rarely been reported. The rich vascularity of the malformation may explain the hematogenous deposition of metastatic neoplasm at this site. (*Neurosurgery* 26:1054-1057, 1990)

Key words: Arteriovenous malformation, Metastatic carcinoma

INTRODUCTION

Spontaneous intracerebellar hemorrhage is most commonly associated with hypertension, although intracerebellar hemorrhage may also be seen with primary or metastatic neoplasms, vascular malformations, or blood dyscrasias (1, 5). We report here the case of a patient who was found to have a spontaneous cerebellar hemorrhage within a suspected metastatic neoplasm. Pathological evaluation of the operative specimen, however, revealed a vascular malformation with a focus of neoplastic cells. The association of a metastatic neoplasm located within a vascular malformation has rarely been reported.

CASE REPORT

This 73-year-old, right-handed woman sought treatment for a 2-week history of intermittent headache that had worsened in the 3 days before admission. On the day before admission she experienced vomiting and ataxia. A computed tomographic (CT) scan of her head, performed at her local hospital, revealed a 3-cm, hyperdense, right cerebellar lesion with surrounding hypodensity, anterior and left displacement of the fourth ventricle, and obstructive hydrocephalus (Fig. 1). A CT scan of the chest obtained during the same evaluation demonstrated a right paratracheal, mediastinal soft tissue mass extending from the carina to a point approximately 7 cm superior to it. The patient's past medical history was

significant for asthma of 10 years' duration and for a smoking habit of 1 pack of cigarettes per day for 50 years; she had quit smoking 8 years previously. She had no history of hypertension.

Examination

At admission, the patient was slightly somnolent, but awoke in response to voice and was oriented. Neurological examination was remarkable for mild horizontal nystagmus on left lateral gaze; bilateral dysmetria that was worse on the right than on the left; unsteady stance; gait ataxia; and bilateral extensor responses to plantar stimulation. The remaining examination revealed mild bilateral papilledema, scattered rales throughout the lung fields, and normal breast tissue; the findings of rectal examination were normal, and the stool was guaiac negative. A blood coagulation profile, including platelet count, was normal. The findings of the chest x-ray were consistent with the changes of chronic obstructive pulmonary disease, but without evidence of the paratracheal mass demonstrated on the CT scan of the chest.

Admission

A right frontal ventriculostomy was placed on the day of admission for ventricular decompression. Laboratory examination of the cerebrospinal fluid was unremarkable, and no atypical cells were seen. Magnetic resonance imaging performed postoperatively confirmed the right cerebellar mass and did not demonstrate additional lesions.