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CT Characteristics of Intraventricular Oligodendrogliomas

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Intraventricular oligodendrogliomas are a rare subset of oligodendrogliomas. Analysis of the clinical and CT characteristics of 11 cases in the literature and eight new examples revealed specific radiographic features, which included the presence of an anterior, midline mass within the lateral ventricles composed of clumped calcifications within a dense, enhancing matrix. Hydrocephalus is a constant feature, and these tumors present with signs of increased intracranial pressure. Eighteen of the 19 cases were benign and all were pure oligodendrogliomas, without admixture of other cellular elements. Intraventricular oligodendrogliomas grow slowly and are associated with prolonged survival but, due to their location, are difficult to remove and frequently require shunting. Subsequently developing symptoms, including herniation and death, are more frequently associated with complications related to hydrocephalus than with tumor growth.

Intraventricular oligodendrogliomas can be distinguished from other intraventricular lesions by fairly specific CT characteristics. The ability to recognize them is helpful in prognosis, and awareness of associated complications related to hydrocephalus may assist in the long-term survival of affected patients.

Oligodendrogliomas comprise 5–9% of all primary intracranial gliomas [1, 2]. Only 1.3–10% of all oligodendrogliomas arise in the ventricular system [1, 3–5]. Although rare, intraventricular oligodendrogliomas (IVOs) represent a distinct subgroup, differing from parenchymal oligodendrogliomas in presenting symptoms and CT characteristics.

Since the first descriptions of IVOs by Dickson in 1926 [6], 56 cases have been reported. In 11 of these, the CT characteristics of the tumors were described. Between 1975 and 1987, we encountered eight surgically proved IVOs, all of which were demonstrated on CT examinations. A review of these cases and those described in the literature revealed that IVOs have characteristic CT features that help to distinguish them from other tumors and that help to predict prognosis and to plan surgical therapy. Because of their deep location, complete surgical removal is frequently difficult, and follow-up CT studies demonstrate unique complications that should be anticipated. This report presents the clinical, CT, and postoperative features of IVOs that distinguish them as a separate subgroup of oligodendrogliomas.

Subjects and Methods

Of the eight patients in this study, seven had preoperative CT examinations that were available for review: five were performed with and without contrast enhancement (usually a bolus of 28.2–42.5 g/l), one only with enhancement, and one only without. The remaining patient had only a small biopsy of the mass prior to the available CT and the majority of the tumor was not disturbed by surgery.

Postoperatively, 37 CT scans were obtained, all indicated clinically as immediate postoperative studies or for follow-up of tumor progression. Twenty-six of the studies were performed with and without contrast enhancement. The CT examinations were performed on a variety of scanners.

No patient in the current series had complete resection of the tumor. Three had subtotal resections and the rest only biopsies. Six of the eight required ventricular shunting. Three patients received radiation therapy after surgery (6000 rads) and each had at least one CT examination after therapy. Angiograms were performed in five cases, and one patient had two ventriculograms.

TABLE 1: CT Characteristics of 19 Intraventricular Oligodendrogliomas

|	
Location	
Midline and right lateral ventricle	8
Midline and left lateral ventricle	7
Midline alone	1
Third ventricle	2
Fourth ventricle	1
Matrix (CT)	
Density	
Increased (relative to brain)	15
Decreased	0
Isodense	2
Unknown (CT with contrast only)	2
Homogeneity	
Homogeneous	5
Inhomogeneous (cysts)	7
Evaluation not possible	7
Enhancement	
Present	15
Absent	2
Contrast CT only	2
Calcification	
Clumped	14
None	2
Evaluation not possible	3
Vascularity (Angiography)	
Type	
Tumor blush	5
Vascular irregularity	4
Tumor blush and vascular irregularity	3
Avascular	1

The characteristics of these cases were compared with those of 11 cases in the literature [7–12] for which sufficient clinical and CT data were described to permit comparisons (Table 1).

Results

Analysis of the cases in the current study and those in the literature group revealed a slight female predominance of 11 women to 8 men. The patients ranged in age from 17–50 years (average = 28.2 years). The majority of the patients presented with headaches (14/19) frequently accompanied by nausea, vomiting, and visual disturbances. Symptoms had been present for an average of 5.1 months, but two patients presented with an acute loss of consciousness. Papilledema was the most common finding on physical examination (11/19).

On the CT examinations, all the IVOs produced local widening of the ventricle or ventricles in which they arose and conformed to the shape of the ventricle, fulfilling criteria for an intraventricular lesion [13] (Fig. 1). The IVOs most frequently involved the lateral ventricles, primarily the left in seven cases and the right in eight. One lesion was located directly in the midline of the lateral ventricles and two arose in the third ventricle but extended into a lateral ventricle. Only one tumor was located in the fourth ventricle. The largest components of the lateral ventricular tumors were to be found in the region of the septum or foramen of Monro. In no case was either the atrium of a lateral ventricle or the occipital horn involved. Lateral ventricular hydrocephalus was demonstrated in 16 cases, and in two others shunts had been inserted prior to the diagnosis. No peritumoral edema was demonstrated in any case, and periventricular lucency associated with acute hydrocephalus was present in only one case (Fig. 2).

The density of the tumor matrix was greater than that of the adjacent brain in 15 of the 19 cases (Fig. 2) and isodense


A B
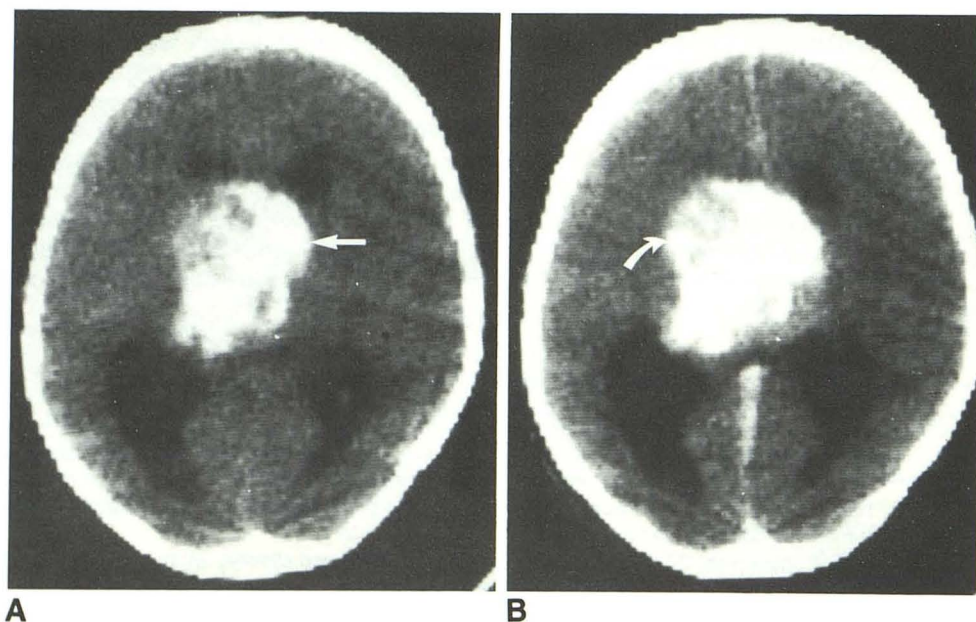
Fig. 1.—31-year-old woman with severe postpartum headaches accompanied by diplopia. Scans without (*A*) and with (*B*) contrast enhancement show intraventricular mass composed of heavily clumped calcification (*straight arrow*) and moderately enhancing tumor matrix (*curved arrow*). The mass has produced lateral ventricular hydrocephalus.

Fig. 2.—*A* and *B*, Initial unenhanced (*A*) and enhanced (*B*) scans of 26-year-old woman obtained 2 months after a small biopsy was performed. A shunt inserted into right lateral ventricle extends through tumor (*straight arrow*). Tumor matrix is slightly denser than brain and enhances moderately. Note small cysts within tumor matrix (*curved arrow*).

C and *D*, Scans at pineal level obtained 3 months (*C*) and 7 months (*D*) later reveal progressive enlargement of right frontal horn (*open arrows*) due to entrapment by tumor. Last image (*D*) was obtained after patient suddenly lost consciousness. Patient died 1 day later, presumably from acute herniation; but permission for an autopsy was denied.
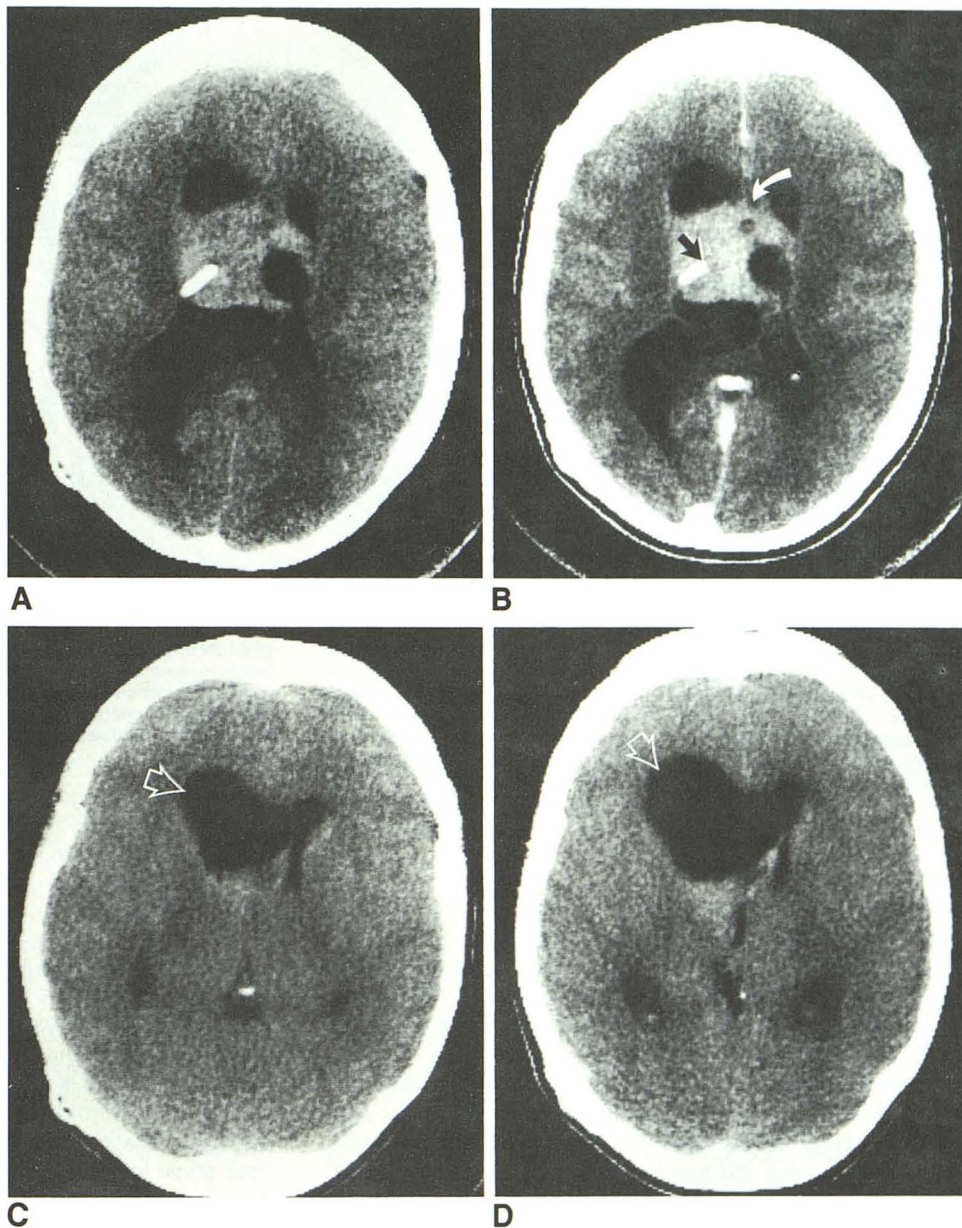
in two. In no case was it uniformly hypodense. Within the matrix, small cystic, hypodense foci were noted in seven cases. The majority of the lesions (14/19) demonstrated clumped internal calcifications. In the 17 cases in which enhancement could be defined, the tumor matrix enhanced in 15, usually slightly to moderately.

Descriptions of the angiographic findings were available in 13 cases. A tumor blush was noted in eight, and in three of these it was accompanied by abnormal vascularity. In four cases, only vascular irregularities were present; and in one, only an avascular mass was found.

During the surgical procedure, the origin of the IVOs could be defined in seven cases. In three, the tumors arose in the immediate vicinity of the foramen of Monro; in one, it arose

from the septum pellucidum. In two cases, the origin of the tumor was the lateral ventricular wall, adjacent to the right thalmus in one and to the body of the caudate nucleus in the other. In the remaining two, it arose from the lateral ventricular wall, but the precise location in the wall was not stated.

The histologic analyses revealed that 18 of the tumors were typical benign oligodendrogliomas, without admixture with other cellular elements. Only one lesion, that in the fourth ventricle, proved to be malignant, but it too was a pure oligodendroglioma [12].

Follow-up periods after surgery ranged from 3 weeks to 9 years. Four patients died: two of meningitis (one at 40 days and one at 14 months after surgery) and two of apparent complications related to hydrocephalus, although autopsies
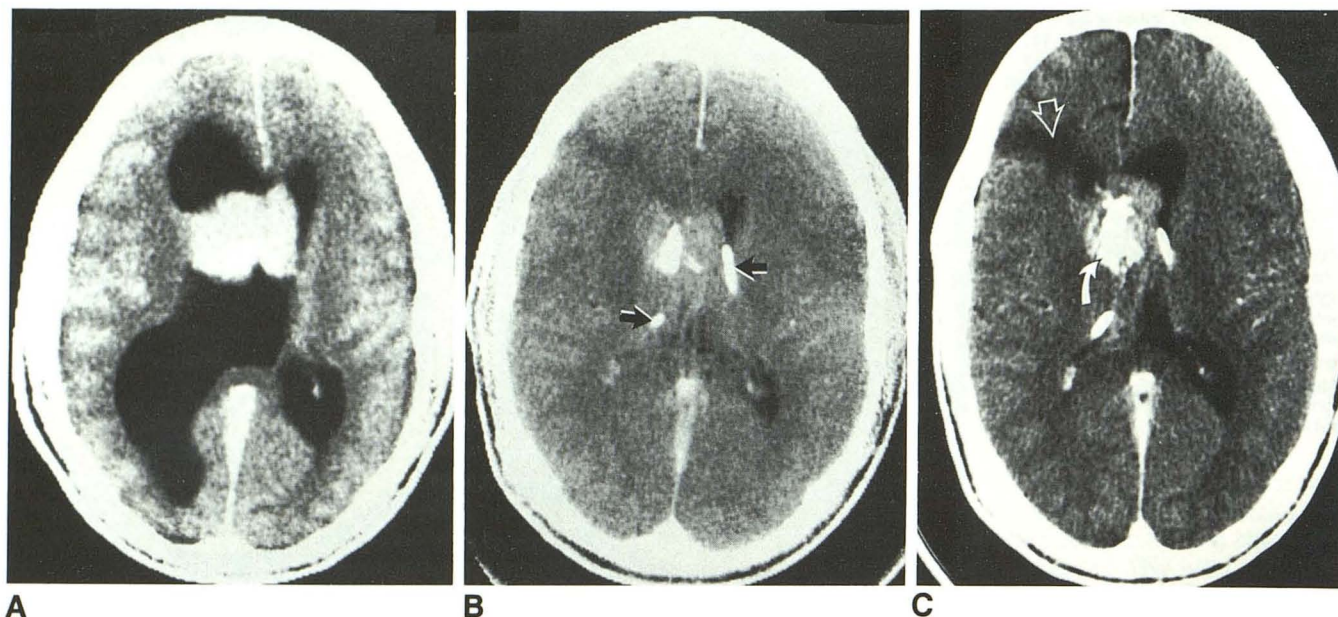
A   B   C

Fig. 3.—Preoperative, contrast-enhanced scan (*A*) of 23-year-old man demonstrates homogeneously enhancing lesion in midline of lateral ventricles obstructing the right lateral ventricle to a slightly greater degree than the left. Scan (*B*) obtained 14 months after subtotal resection of intraventricular oligodendroglioma shows bilateral shunts (*arrows*). No radiation therapy was administered, but degree of enhancement has decreased, possibly because of interference with blood supply during sur-

gery. Scan obtained 5 years after surgery (*C*) shows that calcification of tumor (*curved arrow*) has increased but the intraventricular oligodendroglioma has not changed in size and still enhances minimally. Left lateral ventricle is larger due to shunt malfunction. The band of right frontal lucency (*open arrow*) is a scar and/or fluid-containing tract related to surgery.

were not obtained (one at 9 months [Fig. 2] and one at 9 years after surgery). The patient with only a 3-week follow-up remained comatose after surgery. Of the surviving patients with benign IVOs, eight were alive and well 2 months to 6 years after surgery and four had minimal residual neurologic deficits. In one case, no follow-up was available.

In the literature, two follow-up CT studies are described. One patient had no recurrence on a study obtained 1.5 years after radical tumor resection [8]. The patient with the malignant IVO involving the fourth ventricle had a recurrence 21 months after diagnosis [12]. In the current series, tumor was present on all postoperative CT examinations. Of the six patients with more than one study after surgery, the lesions decreased in size after radiation therapy in two, one with residual contrast enhancement visible in the tumor and one with no enhancement of previously enhancing tumor matrix. In three cases, no change in the size or appearance of the tumors was demonstrated on studies obtained at 8 months, 4.5 years, and 6 years, respectively, except for an increase in calcification (Fig. 3). All these lesions persistently contrast enhanced, and none of these patients had received radiation therapy. The final follow-up study of one patient, performed 1.5 years after surgery, was not available for review but was described as showing a persistent calcified mass. The final CT scans of the two patients who had only one postoperative study (one at 1 month and one at 11.5 months after surgery) demonstrated minimally enhancing residual tumors.

Discussion

Oligodendrogliomas are highly cellular tumors composed of uniform sheets of cells with spheroidal central nuclei and clear cytoplasm. In approximately 50% of cases, oligodendrogliomas contain other glial elements, including astrocytes and spongioblasts. IVOs are therefore somewhat unusual in their lack of other intermixed cell types [2]. Although, histologically, IVOs are indistinguishable from their peripheral counterparts, their origin at a site not usually thought to contain oligodendroglia has not been fully elucidated. It is speculated that the cell of origin might be closely related to that which gives rise to ependymomas, as similarities between ependymal cells and oligodendroglia have been found. These tumors may have similar precursor cells that may evolve into either type of intraventricular neoplasm [9].

Analyses of clinical parameters demonstrate differences between peripheral oligodendrogliomas and IVOs. With reference to sex predominance of IVOs, the majority of patients in this series were female (11 women to eight men), while in the series of primarily hemispheric lesions, a five to three male predominance was encountered [1, 3–5, 14–16].

The average age at onset of symptoms of patients with IVOs in this series was found to be similar to that of patients with hemispheric lesions. In this series, the average age of the patients was 28.2 years, and in the series of IVOs [7] reported by Markwalder et al. the average age was 32.9

years. In four large series describing hemispheric oligodendrogliomas, the average ages listed were 43.3 [1], 36.3 [4], 33.0 [5], and 28.0 [3]. These findings differ from the impression of other authors who stated previously that IVOs tended to be detected at an earlier age due to early production of hydrocephalus [10, 11].

In series describing oligodendrogliomas in all locations, seizures are the most common presenting sign [1, 3, 14, 16], while with IVOs, signs of increased intracranial pressure are most common. Oligodendrogliomas are slowly growing tumors that have been known to produce symptoms ultimately related to the tumor for years prior to histologic confirmation [3]. In two large series describing hemispheric oligodendrogliomas [1, 3], slightly less than half the patients with seizures had been symptomatic for 5 or more years before diagnosis. CT evidence of slow growth of IVOs is suggested by the lack of periventricular edema usually associated with acute hydrocephalus. Despite their slow growth, however, IVOs may present with sudden loss of consciousness due to an acute elevation of intracranial pressure.

Oligodendrogliomas most frequently arise in the cerebral hemispheres, specifically in the frontal lobes [1, 4, 15–17], although they have been reported in the cerebellum and spinal cord [2] as well as within the ventricles. Only 1.3–10% of all oligodendrogliomas arise in the ventricular system [1, 3–5].

On CT, IVOs characteristically present as masses within but involving the lateral ventricles asymmetrically. They are largest anteriorly, in the region of the foramen of Monro, reflecting their frequent origin from the rim of the foramen or the septum. No IVOs in this series were found to involve the atria of the lateral ventricles, although one case of a malignant oligodendroglioma that filled the temporal and occipital horns of a lateral ventricle has been reported [13]. Owing to their characteristic midline site of origin, IVOs may originate in the third ventricle, but from there they usually extend through the foramen of Monro into a lateral ventricle. Because of their large size at presentation, however, it is frequently difficult to define the site of origin. Coronal CT scans and MR examinations may be helpful in delineating the origin of these lesions for assistance in planning a surgical approach.

The matrix of an IVO is characteristically of slightly greater density than normal brain parenchyma. In contrast, peripheral oligodendrogliomas are usually of mixed [18, 19] or low density [14, 15, 20]. Foci of hypodensity may occasionally be found within the matrix of both peripheral and IVOs [1, 18–20], and in seven of the cases described in this report, small areas of hypodensity were identified. They may represent small cysts, gelatinous areas, or old hemorrhages, as all of these features have been found histologically [2]. Hyperdense foci may occasionally be identified in the matrix of oligodendrogliomas due to acute hemorrhage [1, 20].

The matrix of an IVO usually enhances, to a minimal or moderate degree. In the periphery, only oligodendrogliomas with distinctly malignant features enhance reliably [15, 18].

Oligodendrogliomas are the most common intracranial tumors to develop calcifications [15], and in most cases, the calcifications are nodular or clumped [15, 19, 20]. They are

visible on skull films in 40% of cases [3, 4] and by CT in 70% [18] to 91% [15]. The calcifications are similar in both peripheral and IVOs.

Many peripheral oligodendrogliomas have ill-defined margins [19, 20] while those in the ventricles are well defined, probably because of their location. Edema is not usually associated with either peripheral [1, 18, 19] or IVOs unless malignant features are present histologically [19].

Peripheral oligodendrogliomas have a varying and nonspecific angiographic appearance. They may be entirely avascular [3, 14] and produce little vascular displacement relative to their size [1] or exhibit a faint capillary or venous vascular blush [1, 17]. The cases in this series demonstrated that IVOs have a greater tendency than peripheral oligodendrogliomas to have a tumor blush (11/14), and only one was entirely avascular. In the literature, abnormal vessels have been associated only with oligodendroblastomas or malignant oligodendrogliomas [1]. In this series, despite benign histology, four cases exhibited abnormal vessels angiographically.

Of the masses known to occur in the lateral ventricles, those most likely to be confused with IVOs are subependymomas, astrocytomas, ependymomas, and gangliogliomas. Subependymomas occur in the same age group as IVOs but are more likely to arise in the fourth ventricle [21] or frontal horn [13]. The matrix of a subependymoma is usually isodense and calcifications are present in only 30% of cases [21, 22]. Ependymomas may be difficult to distinguish from subependymomas, but the former tend to occur in much younger patients (77% were under age 20 in one series [23]).

Gangliogliomas are also found in young adults but they may be distinguished from IVOs in that they are usually small and tend to occur in the anterior third ventricle rather than in the lateral ventricles. While they occasionally have a hyperdense matrix, they are more frequently isodense and enhance in only two-thirds of the reported cases [24, 25].

Astrocytomas may be encountered in the ventricular system but they are usually found in a frontal horn. They calcify less frequently than IVOs (20–30%) but usually contrast enhance and frequently have a hyperdense matrix [22, 26]. However, they usually appear avascular on angiograms [22].

Choroid plexus papillomas also may have a hyperdense matrix that enhances, but they are distinguished from IVOs by their almost constant location in the trigone of a lateral ventricle. They usually occur in children (average age reported as 5.2 years in a series of 21 patients [22]), and calcifications are present in only 25% of cases. Intraventricular meningiomas also usually involve the trigone of a lateral ventricle but are more likely than choroid plexus papillomas to have a dense matrix, and all contrast enhance [22, 26].

Epidermoid tumors may occur within the ventricular system but usually in the fourth ventricle and their matrix is usually hypodense and nonenhancing. Dermoid tumors and teratomas are usually distinguishable by the presence of fat in the tumor [22].

Occasionally, especially if large, pineal and histologically related tumors may present as intraventricular masses. Ger-

minomas have a slightly dense matrix but they contrast enhance more than IVOs and rarely contain calcification. Pineoblastomas and pineocytomas frequently have calcifications and a hyperdense matrix but enhance markedly and can also be distinguished from IVOs by their location posterior to the third ventricle [27].

Because of their size and location, IVOs are frequently difficult to remove entirely. Symptoms may be alleviated by shunting, but due to blockade of the foramen of Monro by the tumor, bilateral shunts or separate shunts of both the anterior and posterior aspects of the lateral ventricles may be required. Problems related to hydrocephalus were responsible for the deaths of two patients in this series (Fig. 2) and should be anticipated, especially in those cases in which the shunt drains only one section of a lateral ventricle. Patients with shunts should also be evaluated carefully for ventriculitis. If ependymal enhancement is discovered in patients with an IVO, it is more likely to represent infection than ependymal metastatic disease, although spread of these tumors is known to occur along CSF pathways [2].

In summary, IVOs are rare neoplasms that can be distinguished from other intraventricular lesions by fairly specific CT characteristics. Recognition of these slow-growing tumors is helpful in prognosis, and awareness of associated complications related to hydrocephalus may assist in the long-term survival of affected patients.

ACKNOWLEDGMENT

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