Neurosurg. Rev.
14 (1991) 127–133

Tumors of the lateral ventricles

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Abstract

Tumors are only rarely found in the lateral ventricles. Although various oncotypes of these tumors differ in growth rate and invasiveness they present the same clinical pattern with the same diagnostic and surgical problems. Thus we can consider them as a group. This series comprises 51 primary tumors arising strictly from the structures of the lateral ventricles, the majority from the trigone, operated on between 1952 and 1988: 20 meningiomas, 19 ependymomas, 9 papillomas of the choroid plexuses, and 3 subependymomas. As most of these tumors were benign, the response to surgical treatment was, as other authors have found, good with permanent cure or long survival in the majority of cases. Advances in neuroradiological techniques have greatly facilitated the work-up and differential diagnosis of these tumors. Of the various surgical approaches, the parieto-occipital is preferred by our department, even for tumors of the dominant hemisphere. Our operative mortality of 10.6% tallies with that of other workers.

Keywords: Clinical features, CT, lateral ventricle tumors, MNR, surgical approaches.

1 Introduction

Tumors of the lateral ventricles are a catch-all group of tumors, each having particular gross macro- and microscopical histological characteristics and differing in growth rate and in tendency to infiltrate the adjacent parenchyma. Nonetheless, it makes sense to consider them together because of their common site, frequent similarities of clinical development, common procedures of preparation and common problems of surgical approach implicit in the site. Since tumors of the lateral ventricles grow in a non-functional space, they may attain considerable size before they are diagnosed. Most of these tumors are benign or of low malignancy, and their removal may, therefore, result in permanent cure or long survival. Further, their low malignancy generally renders them resistant to treatment other than surgical excision. This treatment has to be planned and performed with extreme accuracy, for otherwise morbidity and mortality can be high. It is important to bear in mind that these tumors are relatively rare and, therefore, outside the routine experience of the majority of neurosurgeons. We report the case series of our department, emphasizing the salient clinical features of these tumors, the principal specific characteristics revealed by modern neuroradiological imaging, the criteria of choice for surgical approach, and the longterm results.

2 Material and method

Of the 6300 tumors of the cerebral hemispheres operated on in our department between January 1952 and December 1988, 51 were located solely or mainly in the lateral ventricles. All the tumors included in this series were primary tumors arising from the structures belonging to the lateral ventricles (choroid plexus, tela choroidea, ependyma). The series included 20 meningiomas, 19 ependymomas, 9 papillomas of the choroid plexuses, and 3 subependymomas.

2.1 Meningiomas

Of 1388 intracranial meningiomas operated on in this period 20 were of the lateral ventricles (1.4%). The age range of the patients, 13 females and 7 males, was 14 to 64 years with a mean of 32 years. The length of the clinical history ranged from 6 months to 4 years (mean 27 months). The first symptoms were headache in 12 cases (60%), epilepsy in 5 (25%), psychic disorders in 2 (10%) and paresis of the contralateral lower limb in 1 (5%).

Neurological examination at admission showed: motor deficits in 13 cases (65%), intracranial hypertension in 10 (50%), homonymous lateral hemianopia in 9 (45%), sensory deficit and aphasia in 6 (30%), dysmetria in 4 (20%), nystagmus in 3 (15%), and trigeminal hypoesthesia in 2 (10%).

CT scanning, performed in eight patients, imaged a hyperdense lesion with enhancement after contrast medium in all. Hydrocephalus was present in three cases and calcifications in four. All patients underwent angiography, which supplied indirect signs of the tumor in every case. The blood supply via the anterior and/or posterior choroidal arteries was evident in all and a tumor blush was clearly visible in 18 cases (90%).

The lesion was located on the left side in 12 cases, on the right in 8, and occupied the region of the trigone in all. 18 meningiomas were fibromatous, 1 psammomatous, and 1 endotheliomatous.

One patient died of cardiac arrest on day 6; 2 patients were lost to follow-up. For the other cases the follow-up ranges from 12 months to 31 years. Two patients with moderate hemiparesis and 6 with mild hemianopoia lead normal lives. The neurological status of the others is absolutely normal.

2.2 Ependymomas

Of 140 intracranial ependymomas operated on in the period, 19 were located in the lateral ventricles (13.6%). 13 patients were female and 6 male (F/M: 2.2/1) and their age range was 7 to 59 years (mean 28.8). The mean length of the clinical history was 14.6 months; the longest was 9 years and the shortest was in a stroke case with neoplastic apoplexia (hemorrhage) not preceded by other symptoms. The first symptoms were headache in 10 cases (52.6%), epilepsy and psychic disorders in 4 (21.7%), and clouding of consciousness in 1 case (5.2%).

Neurological examination at admission showed: signs of intracranial hypertension in 12 cases (63.6%), psychic disorders in 5 (26.3%), phase disorders and homonymous lateral anopias in 4 (21%), hemiparesis in 3 (15.7%) and ataxia in 1 (5.2%). The examination was negative in 2 cases (10.5%).

Before CT was available, the great majority of patients underwent plain X-ray examination, pneumoencephalography, and/or ventriculography and brain scintigraphy. CT scanning was done in 10 patients, revealing increased density in 8 (80%) and patchy density in 2 (20%); hydrocephalus was present in 5 cases (50%) and intratumoral calcifications in 2 (20%). The tumor was enhanced after injection of contrast medium in 9 cases. Angiography was done in 12 patients and supplied indirect cases. Angiography was done in 12 patients and supplied indirect evidence of an intraventricular tumor in all of these; a tumor blush was present in only 4 cases (33.3%).

In 10 patients the tumor occupied the right lateral ventricle (52.6%), in 8 the left (42.1%), and in 1 (5.2%) both ventricles. The lesion was located in the ventricular trigone in 9 cases (47.4%), in the frontal horn and midventricular body in 4 (21%), and in the occipital horn and temporal horn in 1 (5.2%).

Removal was macroscopically total in all cases except in one in which the tumor infiltrated the caudate nucleus. In the cases considered, surgical inspection showed that four fifths or more of the tumor projected into the ventricle. In 10 patients the ependymoma was grade I, in 5 grade II, in 3 grade III and in 1 grade IV. All patients received radiotherapy at a mean dose of 5000 rads.

There were 4 perioperative deaths, all in the early years of the series and all classifiable histologically as grade I. The grade IV ependymoma recurred after about a year and the patient died. Of the 3 grade III ependymomas, one recurred 2 years later and one recurred 3 years later. These patients underwent reoperation and have had no recurrence one year after the second operation. The third patient with the grade III tumor is well 1.5 years after operation. Of the 5 grade II ependymomas 1 recurred 5 years later. The patient underwent reoperation but died 2 years later (the recurrence was more malignant: grade III). The other 4 patients with grade II ependymomas are well 6, 5, 3 and 2 years after operation. Of the 6 patients with grade I ependymoma, 1 has been followed up for 20 years and is well, 1 developed a frontoparietal primary neuroectodermal tumor after 9 years, during which time he was free from neurological deficits, and the other 4 patients are well 10, 7, 4 and 3 years after operation, although one presented with epileptic seizures at 4 years, controlled by drugs.

2.3 Papillomas

Of 27 intracranial papillomas operated on in the period, 9 (33.3%) were located in the lateral ven-

tricles. 6 patients were male and 3 female (M/F: 2/1) and their ages ranged from 50 to 1.5 years with a mean of 26.3 years. The mean length of clinical history was 8.2 months (range 15 days to 4 years). The first symptoms were headache in 6 cases (66.6%) and epilepsy in 3 (33.3%).

Neurological examination on admission showed: intracranial hypertension in 6 cases, hemiparesis in 4, homonymous lateral hemianopia in 3 cases, and normal results in 2 cases.

Only in the 3 most recent cases of the series was CT scanning done: the lesion was hyperdense in 2 cases and isodense in 1; it was enhanced after contrast medium in all cases. Hydrocephalus was present in 2 patients and intratumoral calcifications in 1. Angiography was done in 8 cases and showed: indirect signs of an intraventricular tumor in 6 cases, tumor blush in 4, and vascular hypertrophy in 1. Before CT was available, diagnosis was made on the basis of the same examinations as for ependymomas.

In 5 patients the tumor was located on the right and in 4 on the left. In 8 it occupied the trigone and in 1 the temporal horn. Removal was complete in all cases. On histological examination the tumor proved to be a papilloma in 8 cases and a carcinoma in one.

The mean follow-up was 6.5 years (range 3 months to 22 years). 6 patients are in perfect neurological health; the patient operated on for carcinoma of the choroid plexus died 1.5 years later (after radiotherapy with 6000 rads to the entire brain); 1 patient has a mild hemiparesis but leads a practically normal life; 1 patient, 1.5 years after operation, has occasional epileptic seizures.

2.3 Subependymomas

Of 8 cases of subependymoma treated at our hospital, 3 were located in the lateral ventricles. 2 were women and 1 a man, aged 21, 20, and 49 years, respectively. The tumor was in the right ventricle in 1 case, in the left in 1, and in both in 1. It arose from the septum pellucidum in all cases. The length of clinical history was 18 years in the 21 year old woman with subependymoma of the right ventricle, who presented epilepsy and mental retardation; 1 year in the 20 year old woman with subependymoma of the left lateral ventricle, who presented a frontal lobe syndrome and intracranial hypertension; and acute in onset with subarachnoid hemorrhage in 40 year old man. CT scans were taken in 2 cases, showing a space-occupying lesion of patchy density with enhancement after contrast medium. Angiography, done in all 3 cases, showed no tumor blush and only indirect signs of intraventricular tumor. The lesion was totally removed in the 3 cases. At follow-up after 12, 8, and 2 years all 3 patients are in good general and neurological health.

3 Discussion

Tumors of the lateral ventricles are rare. CUSHING [8] in his series of 2023 verified intracranial tumors found 3 papillomas of the choroid plexus and 6 (possible) ependymomas. A more recent series of 330 meningiomas included 3 in the lateral ventricles [9]. ZÜLCH [38] in a series of 6000 brain tumors reported only 41 in the lateral ventricles. LAPRAS et al [27] in a series of 692 intracranial tumors operated on between 1967 and 1972 reported 26 cases (3.7%). BARONE [4] reporting on 47 cases of intracranial ependymoma found 9 in the lateral ventricles. NAMER [30] reporting on 81 cases of intracranial ependymoma operated on between 1973 and 1982, stated that 24 were supratentorial and only one was in a lateral ventricle.

Tumors arising from the tela and from the choroid plexus clearly fall under the heading of intraventricular, whereas tumors arising from the wall (ependymoma and adjacent neuroglia), which not only invade the ventrice but may also grow in the surrounding parenchyma, are harder to classify topographically. We, therefore, think it correct to consider ependymoma, but not glioma originating from the neuroglia of the wall of the lateral ventricle, as ventricular. Although the ependymoma may grow into the brain substance, it tends to expand rather than to infiltrate the nervous tissue, as the glioma does [5]. The symptoms reflect an obstruction of the cerebrospinal fluid pathways and resulting expansion and thus compression of the surrounding substance of the brain. This gives rise to few other signs, while tumors that grow partly within the periventricular tissue give rise to a more pronounced clinical pattern.

DANDY [10] attempted, in 1934, to define the clinical pattern of lateral ventricle tumors in the light of 15 personal cases and another 25 from the literature. The most common symptoms were headache and vomiting. In the majority of cases other signs were also present: epilepsy, hemianesthesia, hemiparesis, and hemianopia. He concluded that "no clinical syndrome allows accurate

localization". In other papers [9, 17] focal signs are often absent.

DELANDSHEER [13] at the end of his review of 175 meningiomas of the lateral ventricle culled from the literature said that two associated patterns are characteristic: the presence of a clinical syndrome consistent with a posterior cranial fossa lesion or a cerebral hemisphere lesion, and the intermittent and paroxysmal nature of certain symptoms ("ventricular attacks"). For ependymomas of the lateral ventricles, too, pseudocerebellar syndromes [18] and paroxysmal phenomena [4], especially bouts of headache depending on changes in the position of the head, have been reported. The latter must be due to an intermittent obstruction to the CSF circulation.

The type and severity of the signs and symptoms depend, of course, on tumor size and direction of growth. Another factor is age; at least two age-related clinical patterns emerge: one dominated by hydrocephalus in neonates and a pattern of intracranial hypertension associated with focal signs of various types in children and adults.

Until 1950, the radiological diagnosis of tumors of the lateral ventricle was based on air or iodized contrast ventriculography. The first angiographic observations of these tumors were made in 1952–53 [1, 19, 29]. The anomalies found with carotid angiography in cases of intraventricular meningioma have been fully discussed in the literature (hypertrophy of the anterior and posterior choroidal arteries, "sunburst appearance" and homogenous tumor blush in the capillary phase, indirect signs of intraventricular tumor, especially in the venous phase [36]). However, papillomas may present a similar angiographic pattern. Further, as reported by several authors [20, 22], the angiographic image may, in some cases, be similar to that of a deep parietal tumor, especially if the blood supply comes from the anterior choroidal artery. In these cases vertebral angiography may be useful because it reveals: a) anomalous vascularization of the posterior choroidal vessels, which are often stretched, b) outlining of the tumor both by encircling vessels and by a residual haze. In the case of intraventricular ependymomas the angiogram usually indicates the existence of a space-occupying lesion, through displacement of the anterior and middle cerebral arteries or stretching of the anterior choroidal artery, but only rarely a tumor blush. In any case, in all tumors of the lateral ventricles, carotid angiography displays in the arterial and venous phases vascular changes caused by hydrocephalus, where present, and in the venous phase significant displacements of the internal cerebral vein, of the subependymal veins, of the basilar vein, of the the septum pellucidum vein, and of the thalamostriate vein.

Scintigraphy proved very useful in our patients at first but it was later supplanted by the CT scan, which supplies a better definition of site, size, and nature of the tumor. In the pre-CT era it was difficult to radiologically distinguish between a primarily intraventricular and a pseudoventricular tumor [6–25], but now CT scanning makes definition of the origin easier [22]. CT scanning was done in 23 of our cases (10 ependymomas, 8 meningiomas, 3 papillomas of the choroid plexus, and 2 subependymomas); it clearly showed the primarily intraventricular origin of the tumor in all. The diagnosis may be more laborious if the intraventricular tumor is accompanied by hypodensity of the periventricular white substance due to edema, peritumoral ischemia, or transependymal reabsorption in the presence of active hydrocephalus.

The density pattern of these tumors, naturally in connection with the clinical pattern, may also supply a reasonably sure diagnosis of nature. This was the case in all 8 of our meningiomas studied with CT: both plain and enhanced CT patterns were characteristic of this disease (uniformly hyperdense lesion before, and clearcut outline with marked enhancement after, injection of contrast medium [16]). Hydrocephalus was present in 3 cases of meningioma and was confirmed to the ventricle occupied by the tumor.

In the case of ependymoma and papilloma of the choroid plexuses, differential diagnosis may be more complex, although certain features intrinsic and extrinsic to the tumor may point to the correct diagnosis. Among the intrinsic features, a homogeneous isodensity with the cerebral parenchyma on the plain scan is more frequent in papilloma and a patchy hyper- and hypo-density in ependymoma [2–22]; fine calcifications are more frequent in papilloma [2–12–34]. One of the distinguishing extrinsic features is hydrocephalus, which is more often tetraventricular in papilloma of the choroid plexus [22].

As to the differential diagnosis of subependymomas with CT, these tumors seem to present a lower attenuation value than do ependymomas [3–28–35] and less enhancement [3–28].

NMR, with its high spatial resolution, supplies a beautifully precise diagnosis of site and definition of the relations between tumor and surrounding structures in intraventricular tumors [14–37]. A diagnosis of nature is not yet possible. As ELSTER [14] points out in connexion with ependymomas, "no specific NMR characteristics are known that may distinguish supratentorial ependymomas from the more common glial tumors" and in connexion with papillomas of the choroid plexuses "the relaxation times are not specific".

The commonest site of tumors of the lateral ventricle is the trigonal region. In our series 37 tumors were at this site: 20 meningiomas, 9 ependymomas and 8 papillomas of the choroid plexus. This site allows a surgical approach that allows control of the vascular pedicle of the tumor and obviates the risk of onset or worsening of motor, visual, and speech deficits (the last-named if the dominant hemisphere is affected). Several approaches to the trigone have been reported: occipital, temporoparietal, posterior middle temporal, posterior parieto-occipital, transcallosal. The occipital route has been abandoned because it increases the risk of visual deficits. A temporoparietal incision involves the shortest cut through the brain substance. However, this is its only advantage, given the frequency of postoperative neurological deficits (homonymous hemianopia and speech deficits in the dominant hemisphere) and the difficulty of controlling the anterior and oosterior choroidal arteries hidden by the tumor. The posterior middle-temporal and posterior parietal-occipital routes theoretically avoid damage to the visual and speech areas, and, in fact, the majority of authors report a low frequency of postoperative complications, which are usually transient anyway, for both routes. Some surgeons prefer the posterior middle temporal route because affords easy access to the vascular pedicle of the tumor and to the tumor itself, which develops ventrally to occupy the temporal horn. The transcallosal approach offers early control of the posterior choroidal arteries but only limited exposure. It is thus not suitable for tumors that attain some size, as intraventricular tumors often do. In addition, visual-verbal disconnexion syndrome has been reported when the splenium of the corpus callosum is resected in patients with homonymous hemianopia. To overcome this drawback, JUN and HUTIK [22] have proposed a modification of the traditional transcallosal approach of KEMPE and BLAYLOCK [23]: partial resection of the splenium from the right side to facilitate sparing of the interhemispheric visual association fibers located ventrally.

Since 1952 we have used the parieto-occipital approach for tumors arising from the trigonal region in 35 of the cases reported; in one case we performed a standard occipital craniotomy because the diagnosis was meningioma of the tentorium and in one other case we used the middle temporal approach because angiography had shown a substantial blood supply to the tumor from the anterior choroidal artery. As to the risk of visual deficits with this approach, it should be remembered that the optic radiations run inferolaterally to the ventricle. The risk of hemianopia is due to the fact that tumors of the trigone (not only ependymomas but sometimes also meningiomas and dysembryogenetic tumors) often go beyond the ventricular wall so that the optic radiations may be damaged on dissection. The use of the parieto-occipital approach has been criticized in cases in which the dominant hemisphere is affected. In our experience, however, if the skin incision is made very dorsally and low down, the aphasia and dyslexia that may arise in the post-operative period disappear more rapidly.

Tumors arising from the midventricular body tend to invade both lateral ventricles. The midventricular body was the site of 4 ependymomas and of 3 subependymomas of the present series, and in 2 cases the tumor extended to both ventricles. A lateral transcortical approach is contraindicated because tumors of the midventricular body lie under the motor and sensory cortex. In these cases the middle frontal gyrus incision is the approach of choice. This was used in the 7 cases in which the tumor arose from the cella media. Problems may arise with this route if the tumor receives its blood supply from the posterior choroid plexuses and lies on the mid-dorsal aspect of the body. In such cases the transcallosal route may be advantageous, even though it may result in cognitive and memory disturbances. The midfrontal gyrus incision is also indicated for tumors located in the frontal horn. We used this approach in four cases of ependymoma. In two cases in which the tumor was in the temporal horn (one ependymoma and one papilloma), we performed a midtemporal corticotomy and in the ependymoma of the occipital horn an occipital corticotomy.

The role of radiotherapy in preventing recurrence of ependymoma is well established [7, 30, 31, 32].

Doubts remain about the systematic irradiation of the spinal cord because of the risks involved [7, 30, 31, 32]. In agreement with NAMER et al [30] we have performed radiotherapy only for subtentorial ependymomas, which have the greatest risk of seeding. For the ependymomas included in this series, we irradiated the brain alone using 5000–6000 rads. In the latest cases we gave stereotactic radiotherapy using the metal clips left during the operation at the site of the insertion of the tumor on the ventricular wall as targets.

We did not use chemotherapy in any of the patients.

Perioperative mortality in the series as a whole was 10.6% (five patients: four with ependymoma and one with meningioma), that which compared well with of other series [30, 4]. These deaths occurred early in the series, in cases treated with the surgical and anesthetic procedures of those days.

The longterm outcome is good in meningiomas and in papillomas of the choroid plexuses, given the possibility of radical surgery and the tumor biology, and, in fact, we have had no recurrences. Less happy is the prognosis of ependymoma. In the experience of other workers as well as our own, much depends on the histological grade [4–15–24–30]. In fact, with total removal followed by radiotherapy we had a higher frequency of recurrence in ependymomas of grades III and IV. Of the 4 cases of these grades (3 grade III and 1 grade IV) 3 had recurrences; one each after 1 year (grade IV), 2 years, and 3 years. One patient with a grade III ependymoma and no recurrence has been followed up for only 1.5 years. Of the 5 patients with grade II spendymoma only 1 had a recurrence, which was fairly late (5 years). Unlike other authors [18], we found a higher histological grade in this recurrence. The recurrence rate in the present series seems to be unrelated to patient age or tumor site. Long survival of patients with subependymoma is well known [28–33], especially for tumors of the septum pellucidum [33]. The mean follow-up for these tumors in our series is 7.3 years with a range of 2 to 12 years: the patients seem to be in good neurological and general health. Furthermore, the histological pattern of the tumors ("pure" subependymomas [33]) promises well for the next follow-up. Even in ependymomas there have been reports, though not many, of very long survival [11–18]. We have one such case in this series: a man with a grade I ependymoma, who enjoys good general and neurological health 20 years after his operation.

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Submitted November 28, 1989. Accepted January 4, 1990.

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