Ependymomas and Ventricular Tumors

9

Manfred Westphal

Contents

9.1 Definition

Ependymomas arise from the ventricular surface, which is made up of ependymal cells. They occur throughout the whole central nervous system, including the filum terminale. They may occur outside the ventricular structures, representing the rare ectopic ependymoma.

Ventricular tumors include the ependymomas, but also all tumors that arise from the matrix of the choroid plexus, mainly plexus papillomas. Also meningiomas can occur in the choroid plexus as well as hemangioblastomas, which are either sporadic or associated with von Hippel-Lindau disease.

In addition, all glial tumors can extend into and compromise the ventricles, but are rather to be considered exophytic and can have any glial histology from pilocytic astrocytoma to glioblastoma. The only specific astrocytoma of the ventricular system is the subependymal giant cell astrocytoma, which is associated with tuberous sclerosis and arises from the subventricular zone. A neuroglial tumor arising preferentially in the ventricles is the neurocytoma.

Secondary tumors to the intraventricular choroid plexus are metastases. The most frequent types are renal cell carcinoma and melanoma because they seem to have a certain tropism for this kind of highly vascularized tissue matrix [45].

9.2 Epidemiology

A comprehensive source for the epidemiology of tumors is the Central Brain Tumor Registry of the United States, CBTRUS (2007–2008). Adjusted to the

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US 2,000 standard population ICDO code C71.5 (ventricular tumors) came out with an adjusted rate of 0.21 (per 100,000 person years). In the most recent survey, a total of 978 cases were reported among 73,583 cases of primary CNS tumors, translating into 1.3%.

Ependymomas. Given the relative rarity of these tumors, and possibility for error with the histological diagnosis, especially for ependymoma, numbers have to be viewed with caution. Also, the epidemiological numbers for ependymomas frequently do not contain subdivisions among intraparenchymal, intraventricular, intramedullary, and cauda equina. In the spinal cord they are the most frequent neuroepithelial neoplasm [27]. The numbers vary between 3% and 9% of all neuroepithelial tumors in different reports–most of which have another primary focus. In children, ependymomas are the third most common tumor and account for 10% of all posterior fossa tumors in this population [43].

Choroid Plexus Tumors. In a recent survey, the prevalence of choroid plexus tumors was given as 0.3 cases per 1 million people [47]. This translates into 0.4–0.6% of all reported intracranial tumors. The tumors are more frequent in children [1], with a mean age at diagnosis of 3.5 years and no clear gender preference either in children or adults. Choroid plexus carcinomas are only a small subgroup of these tumors, 80% of which occur in children [33] with a median age of 3 years at presentation.

Neurocytomas make up 0.25–0.5% of intracranial tumors [8]. They occur in all ages, but mainly in young adulthood, so that 75% of the cases are diagnosed between the ages of 20–40 years [20]. They affect women and men equally. The most common site of occurrence is the anterior part of the lateral ventricle.

Ventricular Meningiomas. In larger series of unselected meningiomas, a ventricular location was seen in only 1.3–1.5%, but in a much higher incidence of 9.4% in the pediatric age group (reviewed in [26]). They are mostly located in the lateral ventricles, with locations in the third or fourth ventricle being very rare. They originate from the choroid plexus.

Metastases become more frequent with the extended life span of patients with cancer. Ventricular metastases are still rare and make up 6% of all ventricular tumors and less than 1% of intracranial metastases [15].

9.3 Molecular Genetics

Ependymomas appear to have a distinct genetic phenotype that is different from other glial tumors. The otherwise characteristic mutations of p53, deletions or mutations of CDKN2A and CDKN2B, PTEN, or amplification of the EGF-R are not found [6]. The only association is with NF-2 correlating to the epidemiological finding that in these patients there is an increased frequency of intramedullary ependymomas. There seems to be a different genetic pattern for intramedullary ependymomas and supratentorial lesions [19], and recent use of gene expression analysis techniques has provided some differentially expressed genes between the different histological subtypes, but without clues to specific pathogenesis [23]. No specific gene alterations that lend themselves to the development of targeted therapeutics have been detected for intracranial ependymomas dealt with in this chapter.

Plexus papillomas have provided even fewer cytogenetic or molecular genetic findings. Some anomalies were found in the 9p region, but no further specific characterization has been done. Like with meningiomas, comparative genomic hybridization provides a long list of sporadically altered chomosomal regions, but of pathophysiological distinction is possibly only an alteration in the Notch pathway [33].

There are no extensive investigations on the molecular genetics of neurocytomas. The limited available data indicate only a gain of chromosome 7 in some cases, but no specific regions or genes have been implied [1, 8].

No specific genetic alterations have been reported for ventricular meningiomas that would distinguish them genetically from the majority of the other meningiomas.

9.4 Etiology and Prevention

No specific causes are known for ependymomas or other ventricular tumors. Thus, there is no specific strategy for prevention. Except for the association with NF-2 [6], there are not even environmental or epidemiological predictors for these tumors. In known NF-2 cases careful observation knowing about the possibility to develop such tumors may lead to earlier detection and an optimal timing for therapy. Likewise, there is

one other genetic syndrome associated with a pediatric ventricular tumor, the subependymal giant cell astrocytoma, which is associated with tuberous sclerosis [51].

Neurocytomas are considered neuro-glial progenitor tumors [49, 50] and as such may be attributable to a deficit in definitive differentiation. There is no consistent co-incidence with genetic syndromes or any kind of environmental exposure or ethnic preference.

9.5 Signs and Symptoms

Intracranial ependymomas occur mainly in children and there especially in the posterior fossa. When confined to the fourth ventricle, they may cause obstructive hydrocephalus with sudden headache of undulating intensity, nausea, vomiting (projectile), diplopia, and papiledema (Fig. 9.1). When they invade the floor of the fourth ventricle, they may also cause cranial nerve disorders, mostly again diplopia, but also facial weakness. A preferred site is the foramen of Luschka where the tumors extend partly into the fourth ventricle and partly into the cerebellopontine angle. This causes similar symptoms, but frequently also unilateral problems with the caudal cranial nerves and in cases of major compression of the medulla also hemiparesis or hemidystaxia because of loss of sensation. Spinal intramedullary ependymomas have different symptoms and are discussed in 54.1.

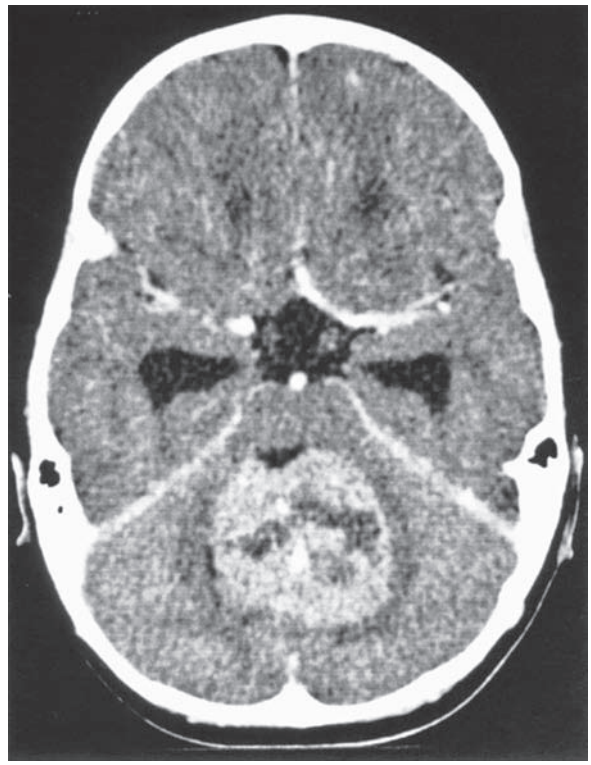


Fig. 9.1 Ependymoma in the fourth ventricle causing acute obstructive hydrocephalus. An immediate direct approach is desirable to avoid any complications from shunting-associated relief of supratentorial pressure

The less frequent supratentorial intraventricular/periventricular ependymomas occur preferably in adults and will cause some kind of hydrocephalus. In a location within the lateral ventricle, it could be a trapped compartment or a univentricular hydrocephalus. When located around the foramen of Monro, hydrocephalus can be biventricular (Fig. 9.2) with intermittent crises of severe headache like in the case of colloid cysts of the third ventricle. Ependymomas in the third ventricle can cause severe endocrinological problems when involving the hypothalamus or memory disorders when involving the fornices. The more dorsal locations lead to compression of the Sylvian aqueduct and cause triventricular hydrocephalus.

Ependymomas in adults have a tendency also to occur ectopically intraparenchymally somewhere in the hemispheres (Fig. 9.3). There they will lead to local symptoms identical to any other glioma, including paresis, speech disorders, visual field impairment as well as seizures. The histology is usually unexpected, and there are no specific neuroradiological clues despite the high incidence of anaplastic tumors in this group. A dysembryogenic component of faulted migration pattern possibly in association with inadequate apoptotic potential must be suspected as part of their development.

In contrast to the ependymomas, plexus papillomas usually do not infiltrate their surroundings and respect the ependymal layer. Therefore, the symptoms are those of local compression and of hydrocephalus that ranges between partial hydrocephalus from a trapped compartment to complete internal, non-communicating, obstructive hydrocephalus due to a lesion in the caudal part of the fourth ventricle. The same symptomatology pattern is true for the ventricular meningiomas and metastases. Meningiomas by virtue of their capacity to secrete large amounts of vascular endothelial growth factor may cause edema, and metastases even more often do so as they may originate from the subependymal layer or, when originating from the plexus, have less respect for the ependyma than meningiomas or plexus papillomas.

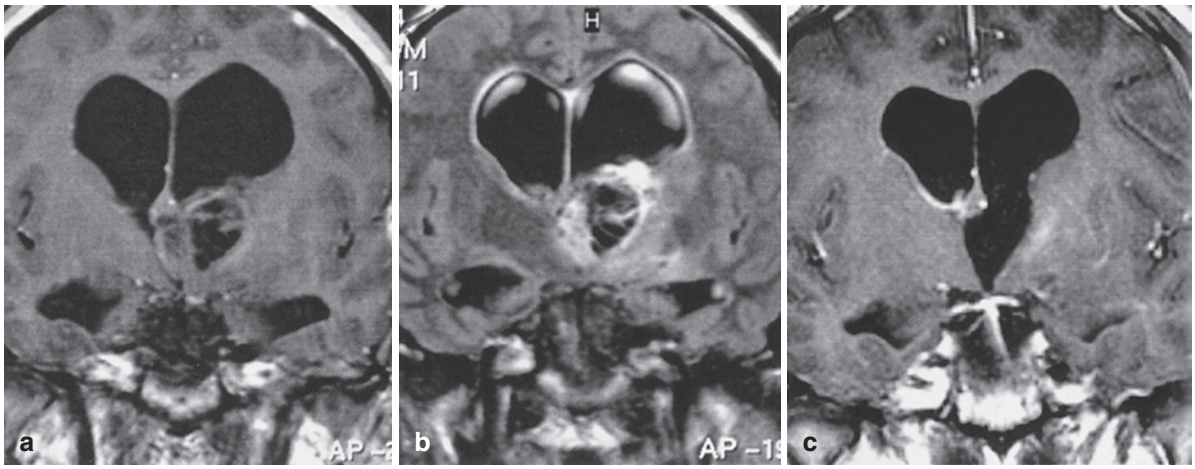


Fig. 9.2 Ependymoma arising from the ventricular wall around the area of the foramen of Monroe (**a, b**). The lesion is inhomogeneously enhancing and has a cystic component and causes biventricular hydrocephalus. Via a frontal transcortical approach, this tumor can be removed (**c**), but because of the broad base there is an extreme likelihood for recurrence warranting regular follow intervals. Because of the immediate vicinity of the hypothalamus and the as yet undefined role of radiation for ependymoma, radiation is delayed until the activity of this tumor has revealed itself

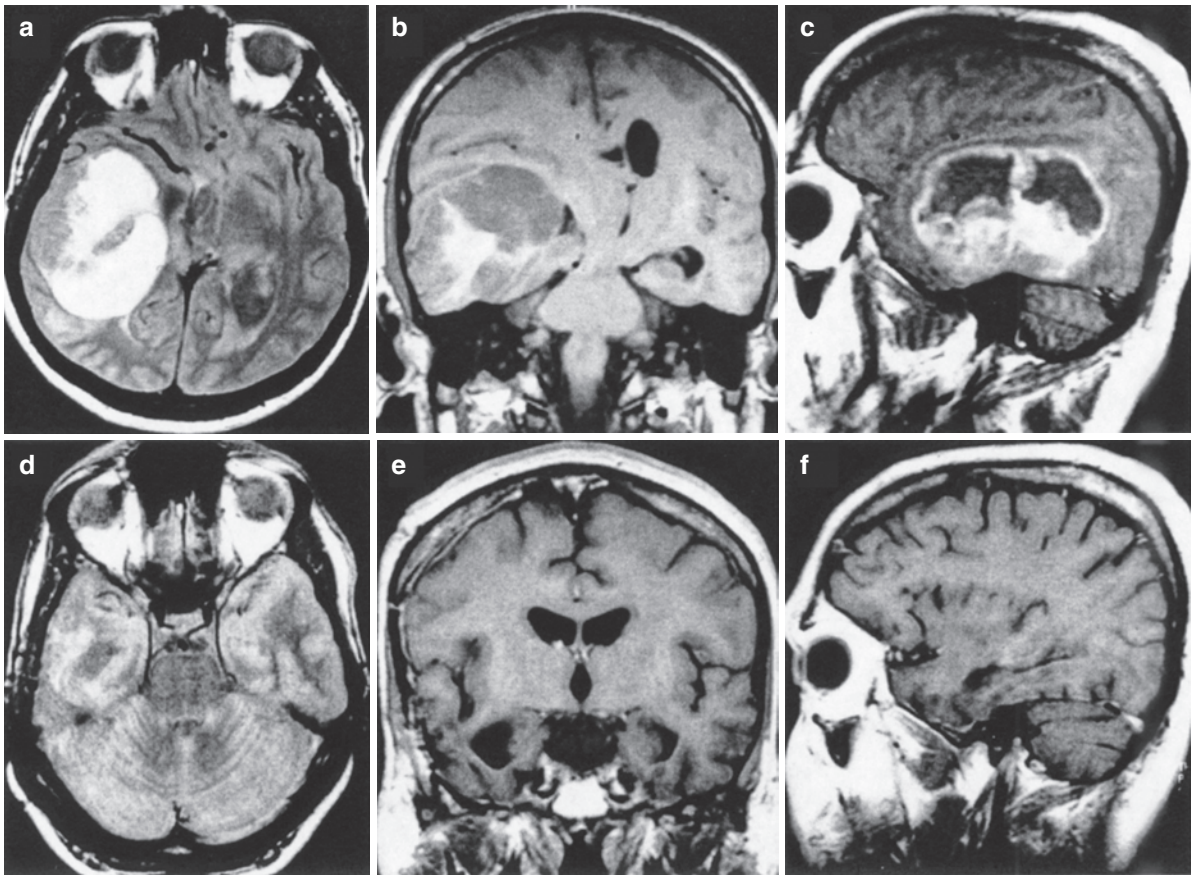


Fig. 9.3 Intraparenchymal ependymoma in a 46-year-old adult male patient (**a–c**) that was not suspected when the lesion was approached. (**d–f**) The patient was treated afterwards with external beam radiation and has had a stable follow-up of 4 years

9.6 Staging and Classification

According to the WHO classification, subependymomas and the myxopapillary ependymomas of the filum terminale are considered to be WHO grade I tumors. There are four variants of grade II ependymoma: cellular, papillary, clear cell, and tanycytic. Anaplastic ependymoma is considered to be grade III [27].

Tumors of the choroid plexus are either classified as grade I, corresponding to choroid plexus papilloma, or grade II when mitotic activity is present or histopathological signs of atypia, which is to be expected in 15% of the cases. Grade III corresponds to choroid plexus carcinoma.

Astrocytomas, oligodendrogliomas, or oligoastrocytomas that extend exophytically into the ventricular system are graded according to the WHO grading system.

Neurocytoma is considered to be grade II in the latest edition of the WHO classification [1, 8]. There is much discussion about anaplasia in neurocytomas [17, 24], but at present it is preferred to speak about neurocytomas with atypical features when necrosis or a high mitotic labeling index is present, and an intensified clinical follow-up is recommended. The concept of malignancy has been used for the extra-ventricular location [44], but even in this case has not been introduced into the WHO classification [8].

9.7 Diagnostic Procedures

Clinical Signs. As most of these tumors occur in children (see that section), the experienced neuropediatrician will suspect a posterior fossa tumor or another intracranial lesion causing hydrocephalus because of obstruction of the CSF pathways from some of the typical symptoms described above. Drowsiness, nausea, vomiting, papiledema, and singultus are grave warning signs. The "stiff neck," when children try to avoid neck pain from descended tonsils by holding their head upright and almost fixed, is a pathognomonic sign that must not be missed.

Ventricular tumors cause obstruction of the CSF pathways and thus create a pressure gradient that results in a vector of forces that can be activated by selectively decompressing one compartment by puncture, i.e., lumbar. This can create downward or in rare cases upward herniation (when supratentorial relief by external ventricular drainage is associated with posterior fossa pressure), and therefore lumbar puncture can only be performed when this possibility has been excluded. If obstruction of the CSF pathways warrants relief, a ventriculocisternostomy (third ventriculostomy) is preferable to a shunting procedure. In any case, CSF must be ascertained to look for markers and cytology.

Neuroradiology. Overall, imaging is the most important diagnostic modality. CT allows the detection of calcifications in tumors and the assessment of the type and severity of hydrocephalus. Much more information is obtained in the MRI, which allows detecting or at least speculating about the area of origin of the tumor. Furthermore, the three planes of representation are crucial for the planning of the surgical approach.

Ependymomas will most often be located infratentorially. They show extension into the cerebellopontine angle (Fig. 9.4) or are located exclusively in the fourth ventricle. They can have cystic components, show signs of prior hemorrhage, and vary between minimal to intensive enhancement after contrast, sometimes within the same tumor. When located in the lateral ventricles, they can have a broad base of ependymal attachment and appear like an exophytic lesion of the brain. However, they may arise from the septum pellucidum or any other area including the roof of the ventricles and then are sometimes undistinguishable from neurocytoma by imaging (Fig. 9.5).

Choroid plexus papillomas tend to show a heterogeneous internal structure, frequent calcifications, and very little reaction of the surrounding brain. They show intense staining due to their vascularization from the choroid plexus. They are usually located centrally in the ventricles and occur anywhere where choroid plexus can be found. In children they are more frequent than in adults and are found usually in the lateral ventricles (see that section). In adults they tend to be more frequent in the fourth ventricle (Fig. 9.6).

Ventricular meningiomas show little calcification in CT, are homogeneously enhancing, and show very little reaction of the surrounding brain when only moderately distending the ventricles. As they slowly grow expansively in their "empty" compartment, they may grow to considerable size before becoming detected (Fig. 9.7). When they are large and lead to compression of the veins in the ventricles, they can cause congestive edema. The best imaging is obtained by MRI, which is also needed for the planning of the surgical approach. In the case of a suspected meningioma, angiography is

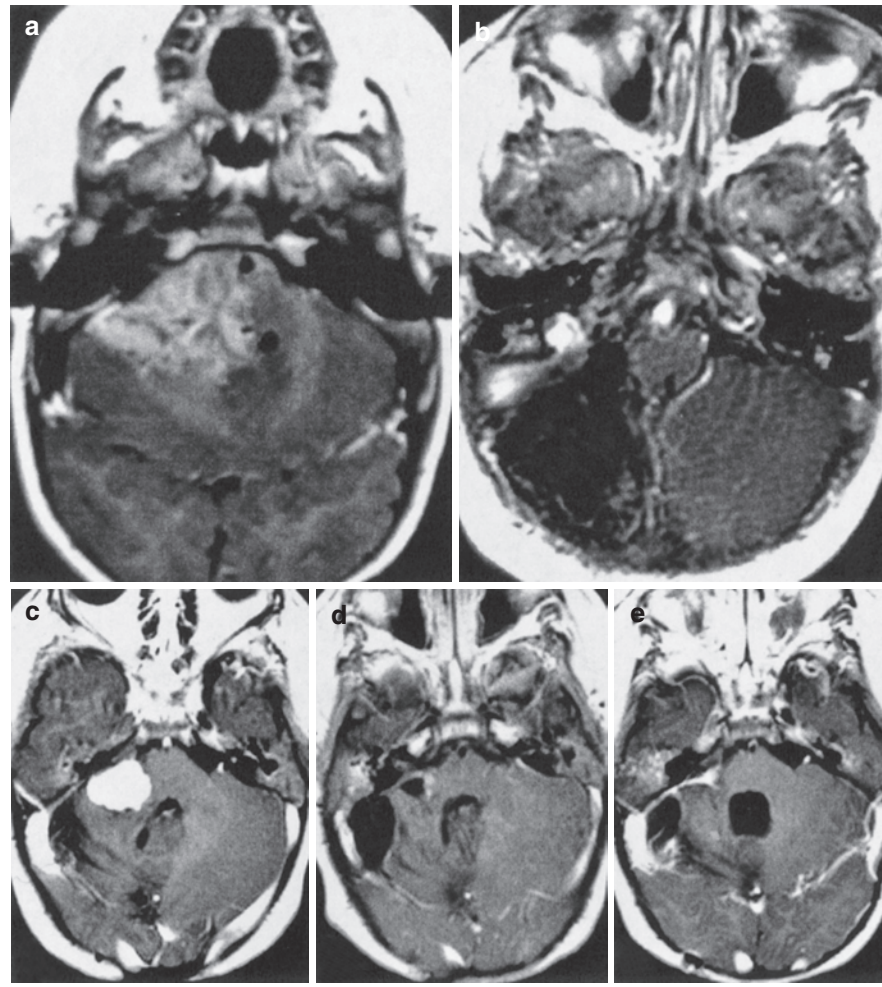


Fig. 9.4 Ependymoma of the foramen of Luschka of a 9-month-old child with extension around the brain stem and into the fourth ventricle (**a**). After near total resection and radiation, the condition was stable for 2.5 years (**b**), and then a recurrence developed that was again resected with radiation of residuals (**c**) that were tightly adherent to the nerve root exits of the caudal nerves and the facial nerve(**d, e**)

still sometimes performed, but it does not provide much useful information and is an unnecessarily added procedural risk to the patient. The blood supply is usually from the vessels of the choroid plexus, and nothing more than a faint blush can be seen. Because of this very peripheral vascularization, no high capacity feeding vessels can be distinguished, and as there is no option for embolization, it is not necessary.

Neuroctomas are usually located in the lateral ventricles, have a broad attachment to the ependyma, are homogeneous in texture even when large, and show moderate enhancement after application of contrast media (Fig. 9.8). They tend to occur more frequently in the area around the foramen of Monro.

The major differential diagnosis for all intraventricular tumors is metastasis. The diagnosis of a metastasis is usually made in the context of a known primary. For intracranial metastases in general one finds 20% in the absence of a known primary, but this does not apply to the types that preferentially metastasize into the choroid plexus, which are renal cell carcinoma and melanoma. They may have any of the appearances mentioned above. Interestingly, metastases seem to have an increased angiogenic activity, which is known to be associated with an increased production of VEGF, and therefore edema is much more frequent and intense than with plexus papilloma, meningioma, or ependymoma.

Hemangioblastoma of the choroid plexus is another rare entity and may be very similar to a renal cell carcinoma, but usually has more prominent (venous) blood vessels, which can be seen as flow voids in its surroundings. As in other locations of hemangioblastomas, there may be cysts or signs of prior hemorrhage. In many cases a von Hippel-Lindau disease is known.

Another rare differential diagnosis can be cavernous hemangioma, which is rare [4], but can be a surprise during surgery or when obtaining the final histological report (Fig. 9.9).

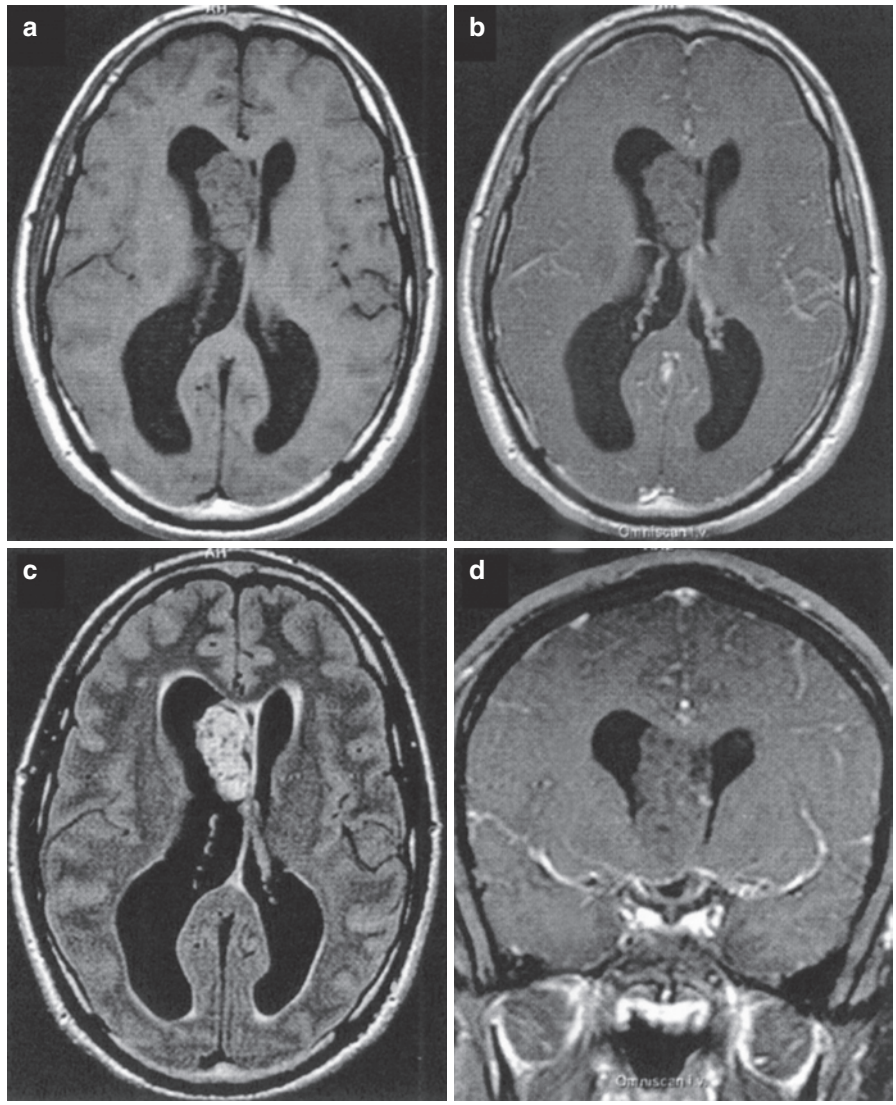


Fig. 9.5 Ependymoma of the region of the foramen of Monroe that was suspected to be neurocytoma but clearly does not have that histology and has a different immunohistochemical marker profile typical of a differentiated ependymoma (WHO II)

9.8 Treatment

Nearly all patients with ventricular tumors require a surgical intervention. Only rarely a tumor in the anterior part of the third ventricle (infundibulum) or posterior part (pineal region) will turn out to be a pure germinoma (see that section), will be diagnosed purely by CSF cytology and marker analysis in CSF and serum, and will be treated by radiation as the main component with optional chemotherapy [32].

Hydrocephalus is frequently a presenting sign and needs special consideration in respect to the timing of surgery. During the removal of a ventricular tumor, it may be very helpful to have distended ventricles to work in, and therefore definitive surgery is usually scheduled within a short period after diagnosis or even as an emergency. It is undesirable to have a shunt placed as a first measure in a center that cannot definitively deal with the lesion and then see the patient referred weeks later when the ventricular system has become normal or even slim. Also, placing a shunt prematurely before the definitive diagnosis can be dangerous. There is a danger for peritoneal seeding, although in one of the larger series that analyzed this phenomenon, this was more frequent for germinomas and medulloblastoma than ependymomas or plexus papillomas [39]. In a series of patients with extraneural metastasis of ependymoma, two patients with peritoneal seeding had shunts [31]. If surgery and relief of hydrocephalus cannot be in close timely association, a third ventriculostomy (IIIVS) is indicated

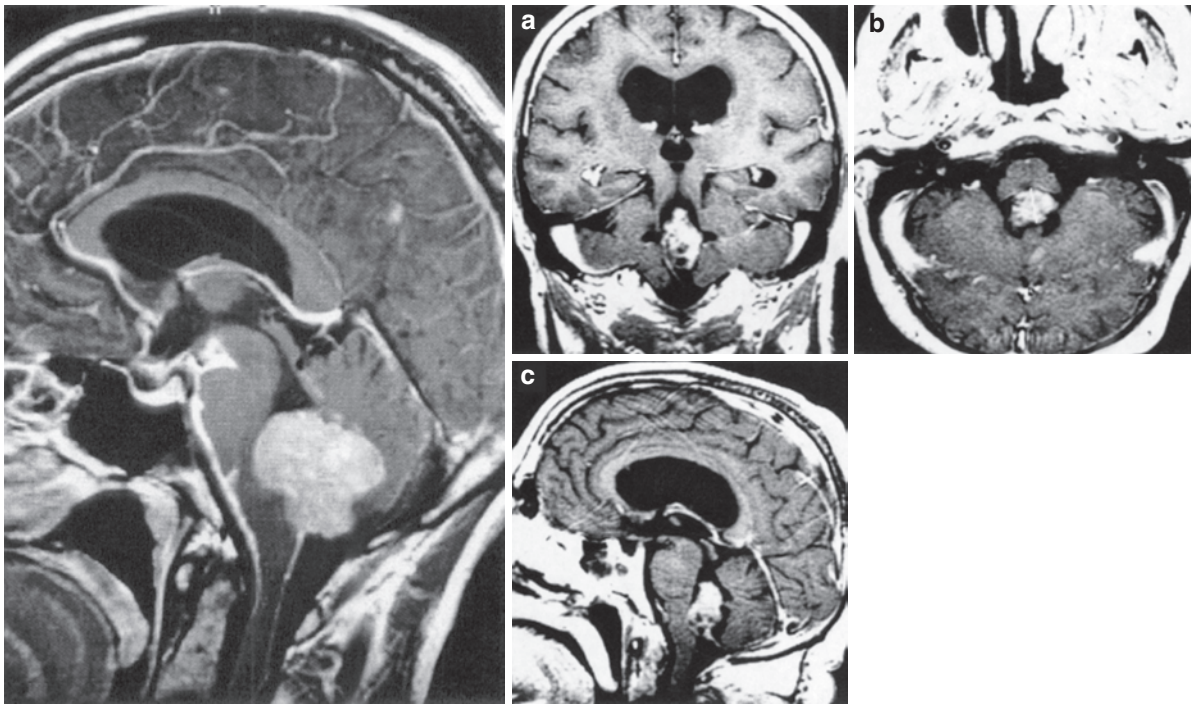


Fig. 9.6 Two examples of adult patients with plexus papillomas of the fourth ventricle. (*left panel*) MRI with contrast of a large tumor in the fourth ventricle that was suspected to be adult medulloblastoma in a 38-year-old male patient but turned out to be a plexus papilloma, which has been removed [11]. (**a–c,** *right panel*) Plexus papilloma at the exit of the fourth ventricle in a 60-year-old man

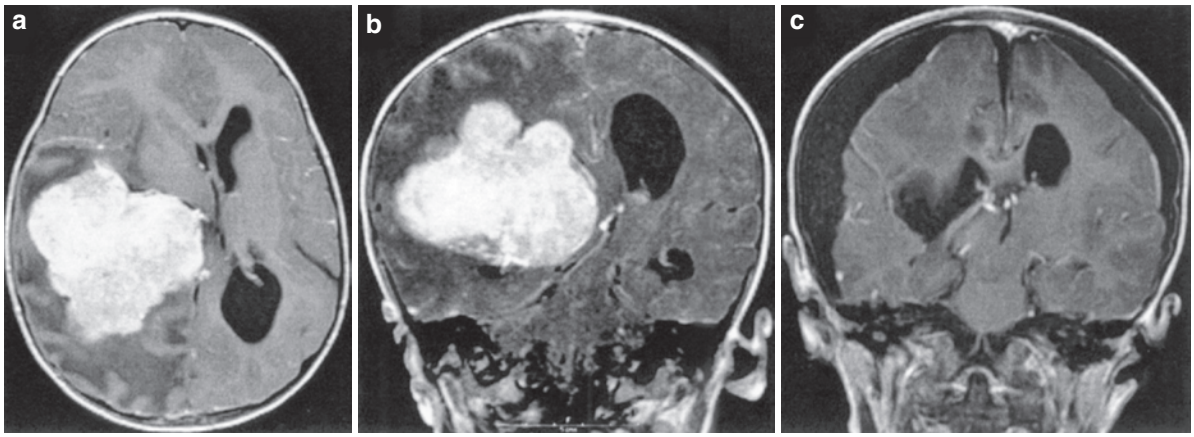


Fig. 9.7 Homogeneously enhancing tumor in the right trigonal area in a 4-year-old boy that turned out to be a ventricular meningioma (**a, b**) that could be removed without sequelae (**c**). Ventricular meningiomas tend to be more frequent in boys, which is in contrast to that disease elsewhere and later in life

because it is a small procedure, will leave no external drains that potentially could become infected, and also provides a biopsy opportunity so that a histological workup can be obtained [18]. The ventricular collapse after IIIVS is also less compared to ventriculoperitoneal shunting or external drainage so that the situation for surgery does not deteriorate as much.

The approaches to the ventricular system are standardized for every location and are plentiful [2]. The approaches to the lateral ventricle depend on the

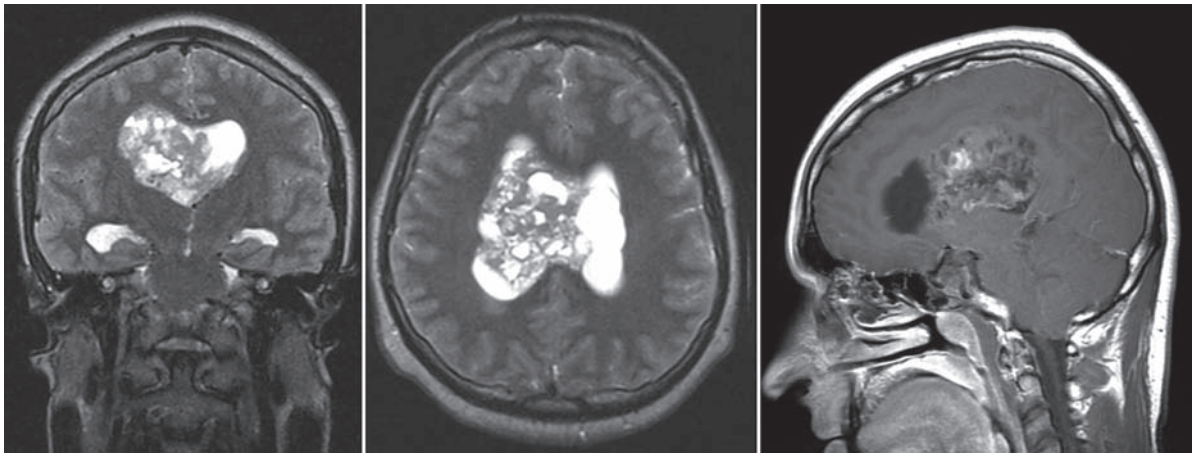


Fig. 9.8 Preoperative images histologically confirmed neurocytoma arising from a large area of the wall of the lateral ventricle, the roof, and the septum pellucidum allowing for extensive decompression but no complete resection. This patient was referred to radiotherapy without any presence of atypical features but for the sake of saving morbidity by trying to expose almost all of the lateral ventricle

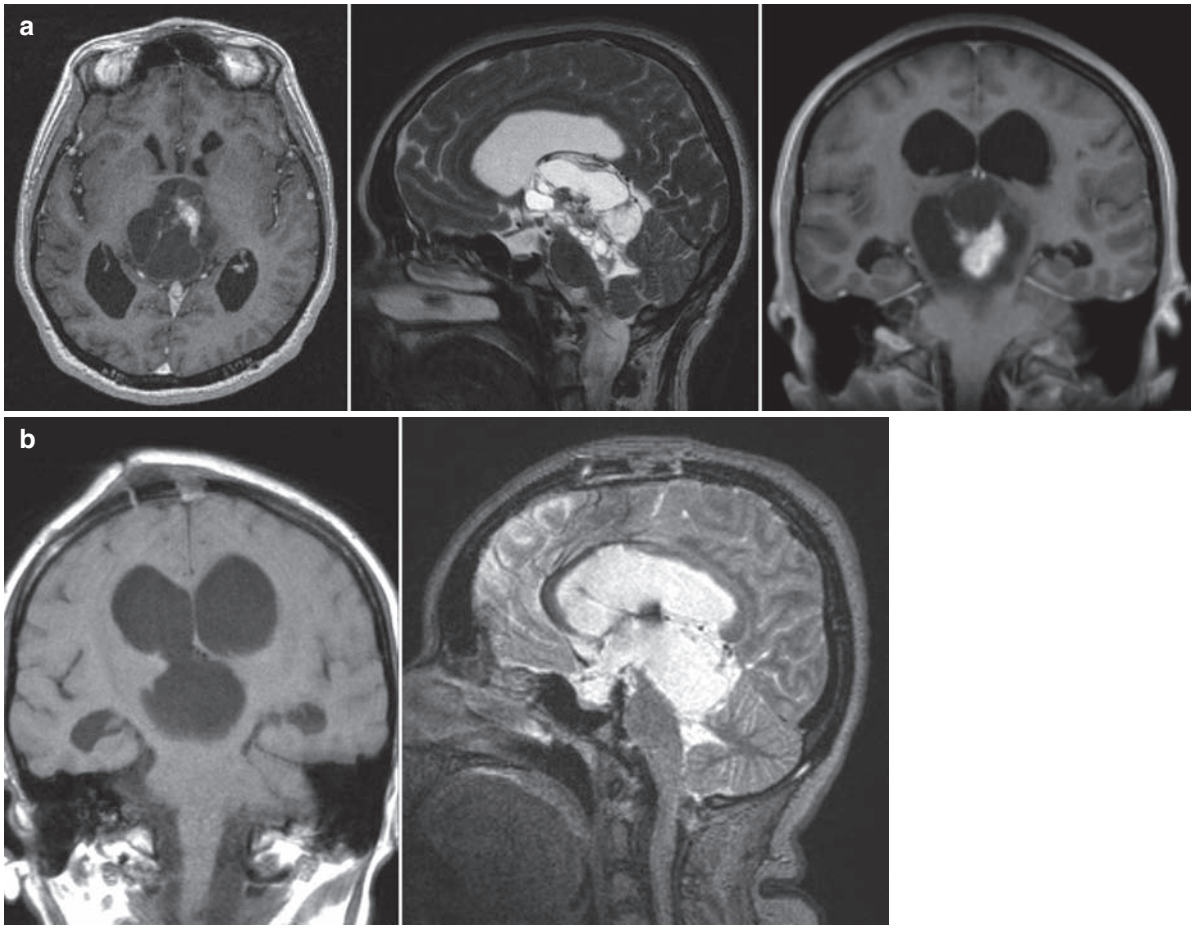


Fig. 9.9 (**a**) Large lesion in the third ventricle causing complex oculomotor disturbances with many heterogeneous cysts and signs of prior hemorrhage that was found to be consistent with a slowly developing pilocytic astrocytoma but turned out to be cavernous hemangioma. (**b**) This lesion was approached by the subchoroidal approach allowing a good overview of the whole third ventricle way down to the fourth ventricle where a perforated gliotic velum with no impairment to the CSF passage was left behind

location of the tumor and the hemispheric dominance. The approaches are always transcortical and are symmetrical for the hemispheres except for the trigonal area, where special consideration has to be taken on the dominant side and a more basal or dorsal approach taken, which leaves the angular gyrus area undisturbed. The third ventricle can be approached from the subfrontal region by the translaminar route or the transcortical, transforaminal route in cases of a distended foramen of Monro. The interhemispheric, interforniceal approach is used for all processes in the central part of the third ventricle, but here a subchoroidal approach is a valid alternative [48, 52] (Fig. 9.9). The posterior part can easily be reached via a supracerebellar, infratentorial approach [13, 42], which gives easy access to all processes originating around the pineal region. Should the venous system be placed inferiorly or is there only limited extension into the pineal region, transcallosal or transtentorial approaches may be more opportune [22]. Most infratentorial processes are approached by suboccipital craniotomy and a direct midline approach from the foramen of Magendie into the fourth ventricle. The further the head is anteflected, the more it is possible to work underneath the vermis without splitting it, thus avoiding the posterior fossa syndrome mainly associated with prolonged mutism [7, 35].

9.8.1 Ependymomas

Few therapies for intracranial tumors are discussed as controversially and inconclusively as those for ependymoma. There is almost no dissent about usually resecting them as radically as possible, and a wealth of information exists, predominantly in the pediatric literature, showing that the extent of resection is the most important prognostic variable, correlating positively with survival (reviewed in [11]). There is, however, no agreement on the use of adjuvant therapies at the present time [43].

The role of surgery is firmly established. In the microsurgical age, mortality has dropped dramatically, and morbidity is related mainly to involvement of the brain stem and cranial nerves. The numbers of patients surviving 5 years after complete (microsurgical) resection versus subtotal/partial resection show a statistically highly significant difference (reviewed in [43]). In addition to extending survival, complete resection apparently leads to a significant reduction of spinal seeding. As spread of the tumor beyond the site of origin can already be seen on preoperative imaging, it is not surprising that the neuroradiological evidence of spread is a negative prognostic sign [5]. The pattern for adults is somewhat different, because they arise more often in the supratentorial compartment and thus have a different surgical morbidity pattern when arising in the fourth ventricle [40]. Another poor sign seen in adults is the more frequent intraparenchymal localization because these tumors are nearly always anaplastic and correlated with poor survival [10].

The role of radiation is controversial in many aspects. It has been stated and seems to be accepted for adult patients that in cases of MR-confirmed complete resection of grade II ependymoma, radiation can be deferred until recurrence on an individual basis [38]. It has also been found that it may be beneficial for incompletely resected low grade tumors, but counterintuitively not for anaplastic lesions that have been completely removed [28]. As for the indication for craniospinal irradiation versus local radiation to the posterior fossa where most cases occur, there is apparently no difference in the rate of distant failures between groups receiving local radiation only compared to craniospinal regimens. When recurrences occur, they are almost always earlier in the posterior fossa before going to the spinal compartment. The doses delivered to the tumor area are between 45 and 50 Gy, and areas of macroscopic disease are given an extra 10 Gy. In the pediatric age group, there is now sufficient evidence that the intensity of radiation, involved field, and age at the time of treatment will cause significant neuropsychological sequelae [29]. On the other hand, these are experiences from the past, and with the modern radiation techniques allowing for extreme limitation of the target volumes and minimalization of collateral damage, the outcomes may be much better when present day treatment series are evaluated. There is already a small study showing that radiosurgery for focal residual or recurrent disease may extend survival time [25].

The role of adjuvant chemotherapy is defined in the pediatric population and extrapolated to adults. It is still not firmly established, and no highly efficient regimen evaluated by a large study has been reported to date. The efficacy of chemotherapy is about that of radiation, sparing the children the radiation-induced sequelae that are a well-researched complication [46]. There is promise from a study alternating between a

combination of carboplatin with vincristine and ifosphamide with etoposide [30]. The main goal of all chemotherapy in children is the delay of radiation, which can be successfully achieved [9].

Plexus papillomas are resected and then followed at regular intervals. As they can be easily resected, there is no use for adjuvant therapies [16, 53], and this is an accepted regimen for children as well as for adults. Even local recurrences of well-differentiated plexus papillomas in the adult are rather reoperated [12]. Carcinomas of the plexus are much more difficult, occur mostly in children, and carry a poor prognosis. They are treated with a combination of surgery, radiation, and chemotherapy. Prognosis is significantly worse than for papilloma [53].

Plexus meningiomas are resected like papillomas and then followed [26]. Radiosurgery may be considered in selected cases, but because of the high incidence of increased radiation toxicity and unsatisfactory tumor control in the long term, this modality is reserved for patients who for some reason cannot be given surgical treatment [26].

Neurocytomas, although a potentially benign and well-differentiated tumor entity, are difficult to treat. They can be completely resected in the anterior part of the lateral ventricles when not encasing the fornices. When they extend too far into the third ventricle and the fornices cannot be distinguished, a subradical approach must be taken. The same is true when the tumors arise with a broad ventricular base in the middle part of the ventricles or the trigonal area (Fig. 9.8). The most controversial issue for neurocytomas is the indication for and the timing of radiation. There is clearly a benefit for incomplete resection and for recurrent well-differentiated lesions and a strong recommendation for giving radiation to patients with tumors with anaplastic features [21, 37]. Because of the efficacy of radiation in manifestly present tumor, radiation after gross total resection is not beneficial because while prolonging the event-free survival, it does not extend overall survival [21]. When during follow-up rapid regrowth is seen, the patients should undergo radiation, which will lead to long-term control or complete cures, respectively [36]. There is also a report about chemotherapy that was successfully given to patients with recurrence despite resection and radiation [3].

Neuro-endoscopy has an important role in the treatment of ventricular tumors, mostly for the relief of hydrocephalus and biopsy [18]. In particular lesions in the posterior part of the third ventricle that are related to the complex pathology of the pineal region (see that section) need a ventriculostomy rather than external ventricular drainage or shunt (Fig. 9.10) unless the patient has a combination of obstructive and malresorptive hydrocephalus. During ventriculostomy it is possible to take an endoscopic biopsy so that chemotherapy or radiation may be started before a surgical option is considered.

There are tumors that do not arise from the proper ventricular tissue matrix but nevertheless have exophytic growth into the ventricles, leading to partial hydrocephalus. These are frequently best approached by the direct transventricular route to get to the exophytic part, which in some cases may be the only part that can be resected (Figs. 9.11–9.13).

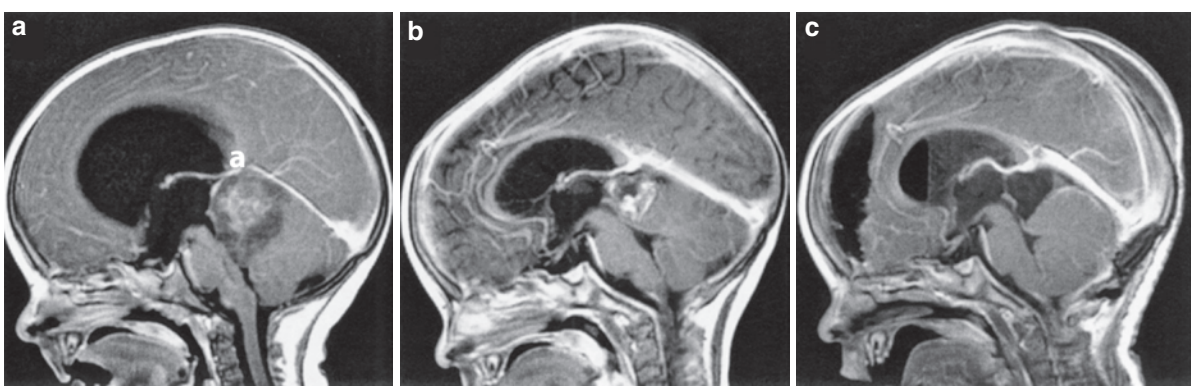


Fig. 9.10 Histologically confirmed atypical teratoid/rhabdoid tumor (ATRT) in an 8-month-old boy that was biopsied on the occasion of a ventriculostomy (**a**). The diagnosis led to chemotherapy after which the tumor shrank significantly (**b**) and was then removed completely (**c**). A malresorptive component of the hydrocephalus required shunting after the ventriculostomy

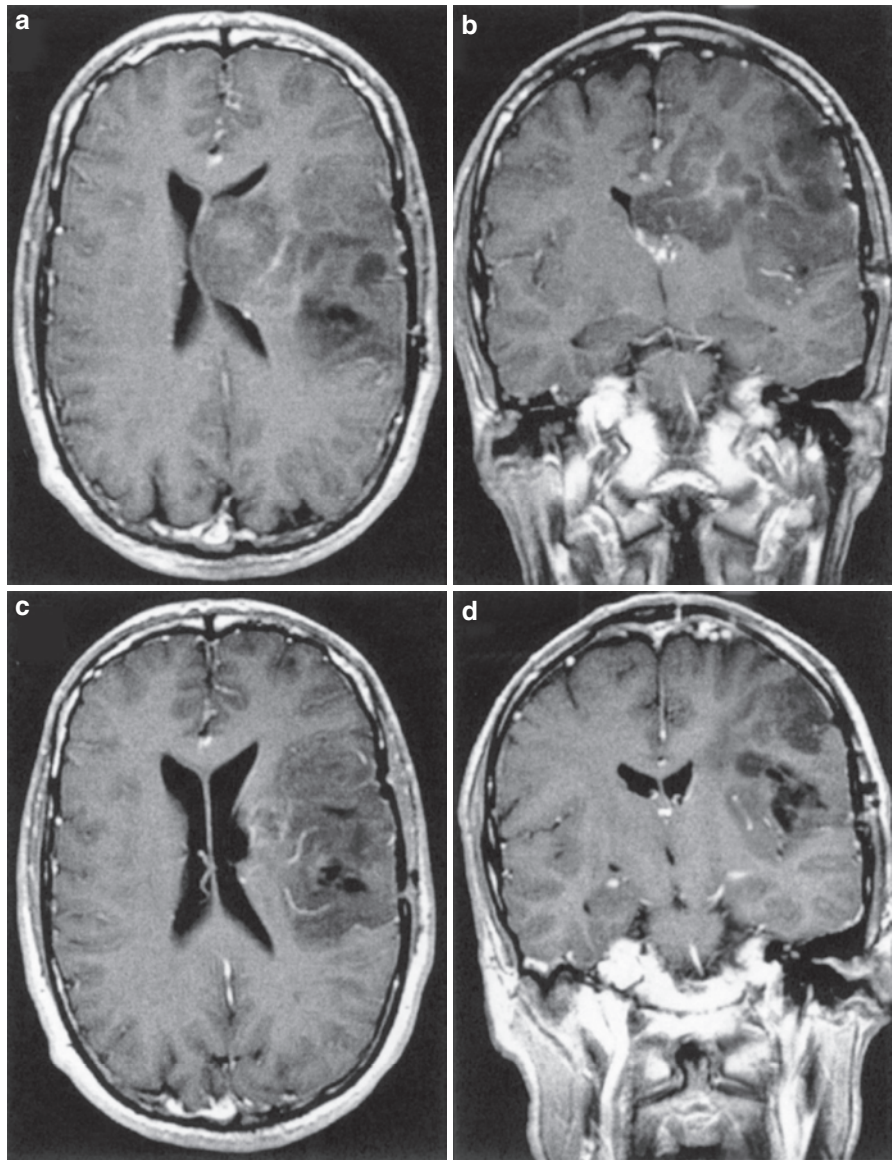


Fig. 9.11 A case of a hemispheric glioma in a 40-year-old male patient that extended into the lateral ventricles with a large exophytic component. In this case this was a histologically proven oligodendroglioma of the left insular region that had remained stable over several years without treatment but then developed a nodular satellite obliterating parts of the lateral ventricle (**a, b**). This was removed by a direct transcortical approach (**c, d**)

Craniopharyngiomas and optic nerve gliomas, which also frequently affect the third ventricle, can sometimes easily be decompressed via the translaminar approach (Fig. 9.14), but may not be approachable when recurrent and when no dissection plane is present to the hypothalamic boundaries (Fig. 9.15). Aggressive surgical approaches to the third ventricle and its surroundings have unwanted sequelae, in particular eating and weight disorders [34, 41]. Therefore, radiation is indicated when tumors are infiltrative or recurrent [14].

9.9 Follow-Up and Prognosis

In all patients with ependymomas, the extent of resection is documented by postoperative MRI, preferably within 2 days after surgery. In the more aggressive tumors (WHO III), a spinal MRI is always performed to obtain staging information. Depending on the treatment protocol chosen, patients are observed without any further treatment and followed at regular intervals or treated with radiation and/or chemotherapy, after which they are also followed. The intervals for imaging are initially

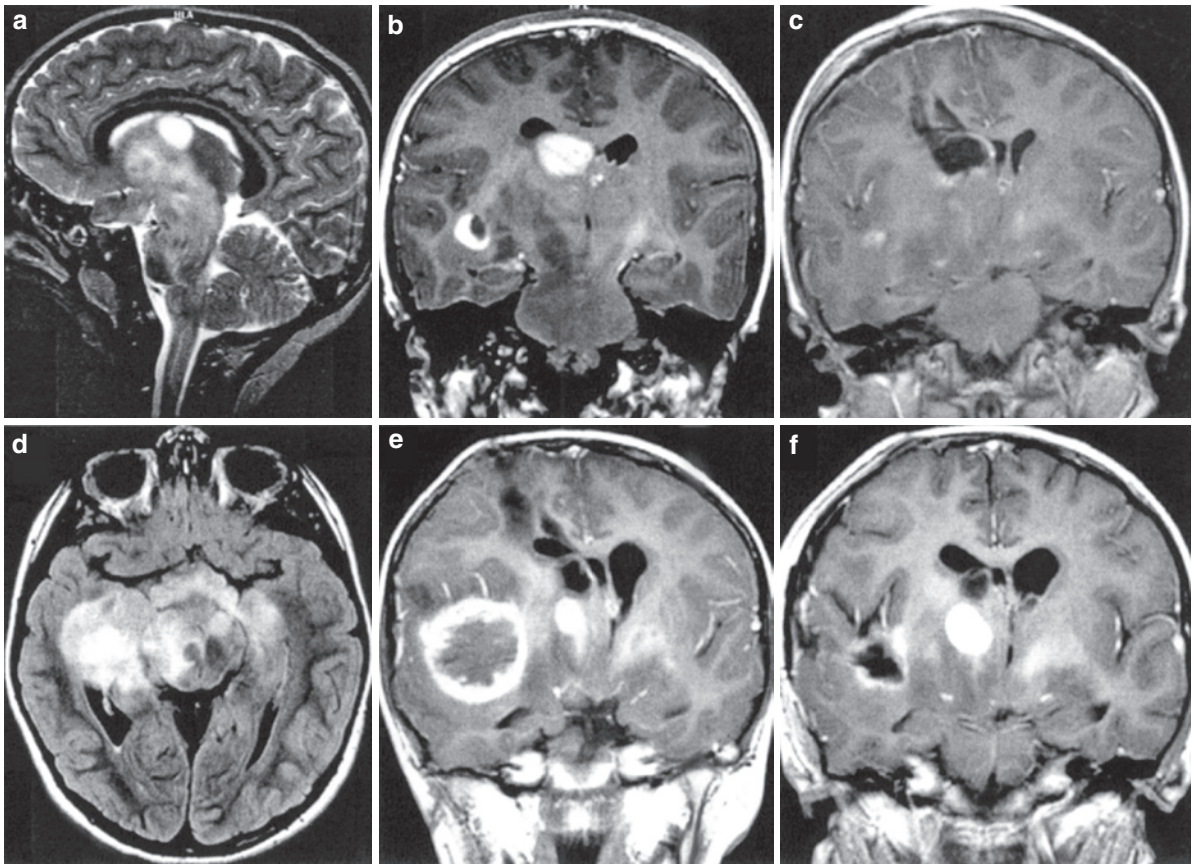


Fig. 9.12 Very complex pediatric (5 years, female) pilocytic astrocytoma of the optic nerve involving the basal ganglia and brain stem, which intermittently forms contrast-enhancing, rapidly growing foci, one of which protruded into the ventricle (**a, b**), was well-delineated, and could be removed completely by a direct transcortical approach (**c**) like other foci that were removed at later stages (**d, e, f**)

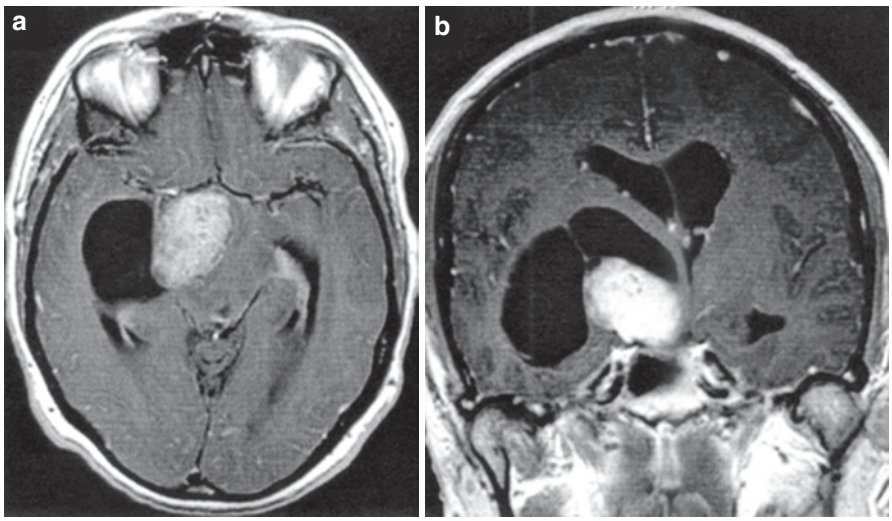
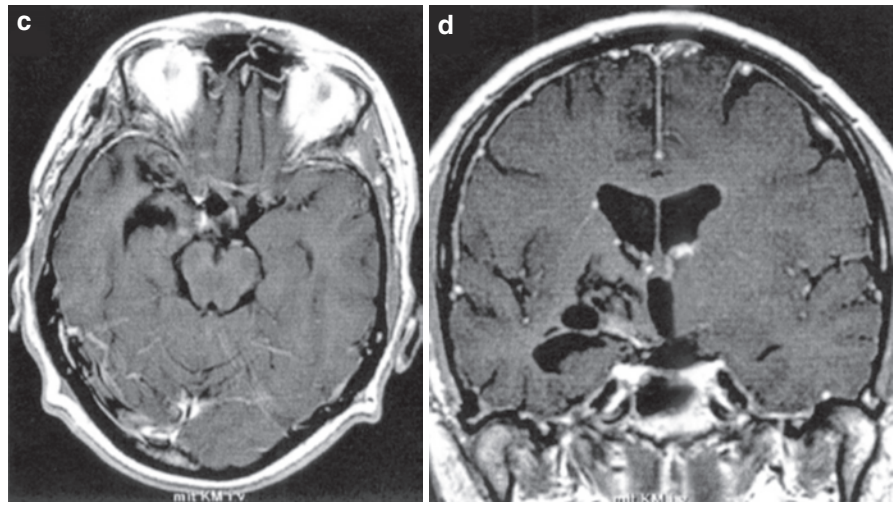


Fig. 9.13 Large tumor of the temporal horn that after near-complete microsurgical removal (**c, d**) turned out to be an exophytically grown pilocytic astrocytoma of the optic tract in a 40-year-old woman

Fig. 9.13 (continued)



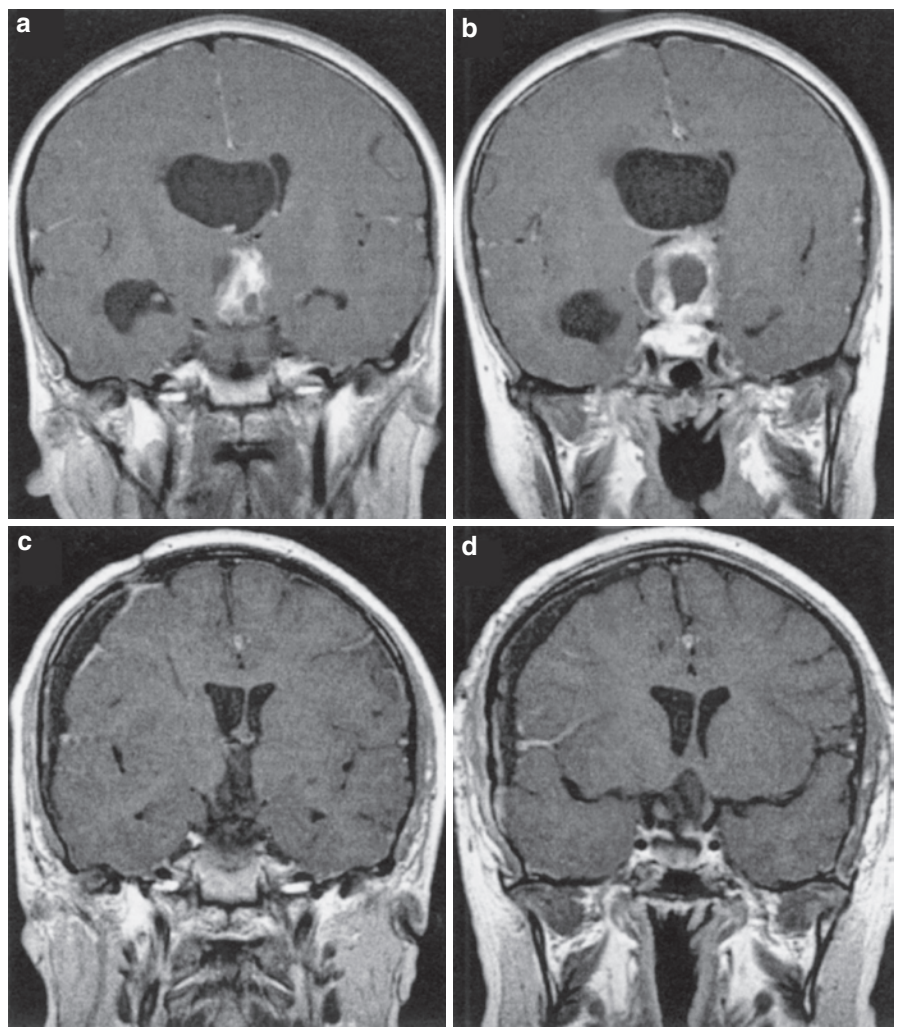


Fig. 9.14 Gross total microsurgical resection (**c, d**) of a mostly cystic, recurrent craniopharyngioma of the third ventricle, (**a, b**) which has presently been stable for 3 years after adjuvant radiotherapy

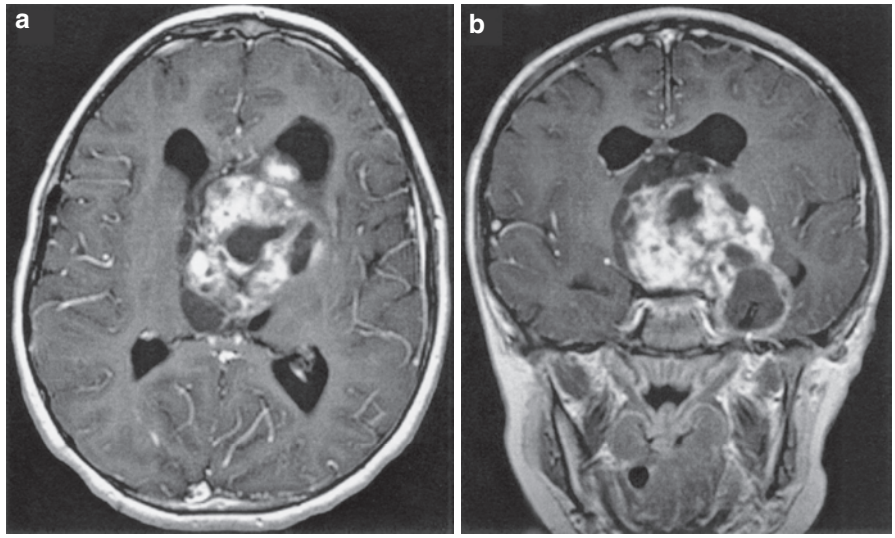


Fig. 9.15 Progressive development of an optic nerve glioma (pilocytic) that had been subradically approached already a few years earlier. As the cysts turned out to be part of the tumor all around, a dissection plane could not be expected so that in the absence of major neurological deficits, shunting and adjuvant therapies were preferred to a resection attempt

every 6 months but can be shorter when the tumors are WHO III and the treatment protocols call for shorter intervals. When the situation is stable, the periods between the follow-up visits can be extended to yearly to eventually biannual visits. There is much controversy as to what the prognostic parameters are because histology is not predictive of outcome, and even the occurrence of extraneural metastases occurs to the same degree with grade II and grade III lesions [31].

Plexus papillomas, meningiomas, and hemangioblastomas are followed with initially 6-month intervals, then yearly visits after 2 years, and then biannual follow-up after 5 years. Neurocytomas may require radiation already after the first operation when they are grade III. Thereafter, they are followed with 6-month intervals. Also for the grade II lesions, close observation for regrowth is required for 3 years with 6-month intervals.

Metastases are treated and followed according to the histology of the primary tumor and the development of the disease outside the brain. With neurological deterioration in the absence of solid tumor and no local recurrence, the development of meningeal carcinomatosis has to be considered.

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