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Choroid plexus tumors in children

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The choroid plexus is anatomically located within each of the four ventricles at the parenchymal/ventricular junction and is derived from the specialization of ventricular ependymal cells along certain segments of the neural tube. The roof of the neural tube in areas of the forebrain and hindbrain expands into thin sheets of epithelium with an underlying loose mesenchymal stroma. The stroma consists of leptomeningeal cells, blood vessels, and connective tissue. The epithelial layer consists of cuboidal cells linked by tight junctions, thereby creating a blood–cerebrospinal fluid (CSF) barrier. Although the evolutionary origin of the choroid plexus is unknown, it is the source of most CSF. In lower mammals, there is a well-developed lymphatic system and the volume of CSF produced is small. The loss of a lymphatic system in the brain may have led to the corresponding increase in the volume of CSF produced in primates.

As with other tissues of the central nervous system, benign and malignant tumors can arise from the choroid plexus. It is assumed that malignant tumors arise de novo, with conversion from a benign to malignant phenotype occurring rarely [1]. Guerard was the first to describe a choroid plexus tumor in 1833. The first surgical resection was reported by Bielschowsky and Unger in 1906. Thereafter, both Davis and Cushing [2] and Dandy [3] reported their experiences with this unusual tumor.

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Epidemiology

Choroid plexus papillomas (CPPs) and their malignant counterpart, choroid plexus carcinomas (CPCs), are uncommon, comprising only 0.5% to 0.6% of all brain tumors. They are encountered in all age groups but are mainly seen in childhood. There is a particular preponderance in infants and extremely young children. In his review of all published cases before 1974, Laurence [4] reported that 45% presented in the first year of life, whereas 74% occurred in the first decade. Wolff and colleagues [5] studied all available cases reported in the literature until 1998 and performed a detailed meta-analysis. This report noted a male-to-female ratio of 1.2:1, a median age at diagnosis of 3.5. years, and a striking difference in tumor location and age. Supratentorial tumors (lateral and third ventricles) occurred mostly in infants, with a median age of 1.5 years at diagnosis in this group. By comparison, the median ages at diagnosis of tumors in the fourth ventricle and cerebellopontine angle (CPA) were 22.5 and 35.5 years, respectively. Laurence [4] did note that 50% of cases reviewed were situated in the lateral ventricles, 37% in the fourth ventricle, 9% in the third ventricle, and the remainder in other locations.

As expected, reviews from pediatric centers report that a higher percentage (1.8%–2.9%) of their cases are choroid plexus tumors [6–8]. Two reviews of tumors in the first year of life by Galassi et al [9] and Haddad et al [10] found that choroid plexus tumors comprised 14% and 12.8% of all cases, respectively. Most series noted previously have not reported any predilection for right or left ventricle or sex. CPCs comprise 29% to 39% of all choroid neoplasms [8,11,12].

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Pathology

Gross appearance and histopathology

Typically, most astrocytic tumors are soft and poorly cohesive masses. In contrast, CPPs recapitulate the normal appearance of the choroid plexus with well-differentiated secondary structures, resulting in a "cauliflower-like" appearance (Fig. 1). The surface of the tumor is similar to the soft fronds of normal choroid, with the overall shape being roughly globular. The stroma can be fibrous and tough in consistency. Evidence of previous hemorrhage is sometimes apparent. Because papillomas are benign, they tend to expand the ventricle rather than invade adjacent brain. Nevertheless, the proximity of these tumors to deep-seated structures, such as the internal cerebral veins and limbic structures, can make their removal difficult.

The microscopic appearance of papilloma is also similar to that of the normal choroid plexus. There are many papillae covered with a simple cuboidal or columnar epithelia. The stroma of these fibrovascular structures is composed of connective tissue and small blood vessels. The presence of the connective tissue stroma is notable mainly because it allows one to distinguish between CPP and papillary forms of ependymoma (whose stroma is composed of fibrillary neuroglia).

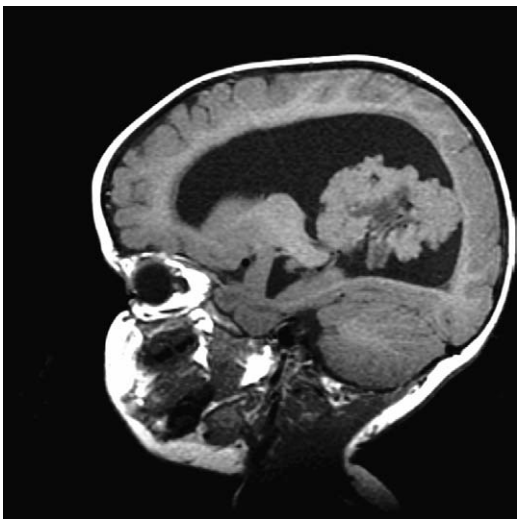

Fig. 1. A large choroid plexus papilloma located within the lateral ventricle. A sagittal, precontrast, T1-weighted image illustrates the gross architecture of the tumor. The mass consists of many small fronds of tissue surrounding a central mesenchymal stroma.

In addition, choroid epithelial cells do not contain cilia or blepharoplasts as do ependymal cells. Mitotic figures are rare. Unusual pathologic features, such as mucinous degeneration, tubular differentiation, and formation of structures like bone, are sometimes observed [13].

Grossly, CPCs tend to be softer and more friable than papillomas, but it can be difficult to differentiate the grade of the neoplasm based only on the gross appearance. Although carcinomas rarely metastasize from the intracranial or intraspinal compartment, they can disseminate throughout the CSF pathways [14]. The definitive diagnosis is usually based on routine histopathology. There are three major features that have been associated with malignancy. First are cytologic criteria of malignancy, such as nuclear atypia, increased nuclear-to-cytoplasmic ratio, and prominent mitotic figures. Second is the loss of the normal papillary architecture. Third is the presence of brain invasion by the tumor, which usually involves extension through the ependymal lining. The significance of stromal invasion has been questioned in a recent report, where three of four patients with CPP and invasion after gross total resection (GTR) were cured without the need for adjunctive therapy [15].

Occasionally, diagnostic confusion can arise in the setting of a tumor with epithelial features. The epithelial nature of the frank malignancy can create confusion, because other tumors, such as metastatic adenocarcinoma and medulloepithelioma, are histologically similar. If the tumor arises in a young patient, the chance of the tumor being metastatic is extremely low. Electron microscopy can reveal details, such as cilia, that are not normally present in choroid plexus tumors. Rarely, if a tumor demonstrates some atypical features without evidence of invasion, it can be classified as an atypical papilloma.

Villous hypertrophy of the choroid plexus is a poorly defined entity. Characteristically, the choroid plexus of both lateral ventricles is enlarged and is associated with hydrocephalus from birth. The hydrocephalus is related to hyperactivity of the choroid, whereas the cytologic appearance of the tissue is normal. Villous hypertrophy has been used synonymously with bilateral CPP, but this is not accurate in the strictest sense if histologic evidence of neoplastic growth is not present and expansion of the choroid plexus is occurring diffusely [16]. Finally, a recent report highlighted the presence of rhabdoid cells within CPCs [17]. These cells are

more typically encountered in atypical teratoid rhabdoid tumors (ATRTs), with electron microscopy required to distinguish between the two diagnoses.

Immunohistochemistry

Usually, the diagnosis is clear from standard hematoxylin-eosin stains, whereas immunohistochemistry is often inconclusive. The calcium-binding protein S-100 is positive in most choroid tumors [18,19]. This is of limited diagnostic value, because glial tissues and normal choroid express S-100 in a parallel fashion with glial fibrillary acid protein (GFAP). Other markers, such as vimentin, GFAP, and cytokeratins, can be positive but also lack specificity [20,21]. Prealbumin or transthyretin (TTR) was initially believed to be a specific marker, but another report noted that 20% of choroid tumors were TTR-negative [22,23]. These investigators did find that prognostic information could be gleaned from immunohistochemical data. A poor prognosis was found in those tumors when less than 50% of the cells in a given tumor were heavily stained for S-100. In addition, the absence of TTR-positive cells also correlated with a poor prognosis. Cellular proliferation as measured by Ki67/MIB1 labeling is low in papillomas and significantly higher for carcinomas [17,24].

Genetics and molecular biology

Choroid plexus tumors arise sporadically without a known cause. Aside from one report that described two cases occurring in one family, a genetic predisposition is not present for most cases [25]. Some experimental evidence links SV40, a primate DNA virus, with choroid plexus tumor formation. As with most viruses, a portion of the virus genome expresses proteins used to subvert normal cell functions and to interfere with growth regulatory pathways. When expressed in mice, large T antigen, the major regulator of late viral gene products of the SV40 virus, induces the formation of choroid plexus neoplasms [26]. The large T antigen is expressed only in the choroid plexus and seems to interact with the product of the p53 gene [27]. T antigen interferes with the function of both p53 and retinoblastoma (RB) tumor suppressor proteins. This function is likely required for viral replication and growth but may also have the effect of increasing tumor formation. Using polymerase chain reaction (PCR), SV40 DNA sequences were demonstrated in 50% of choroid plexus tumors and most ependymomas [28]. Active T-antigen and p53 complexes have also been demonstrated in brain tumors [29]. More recently, the expression of transgenes of the viral oncoproteins E6 and E7 from human papilloma virus has also been shown to produce tumors in 71% of offspring, of which 26% of the tumors were choroid plexus tumors [30].

A subset of central primitive neuroectodermal tumors (PNETs), CPCs, and medulloblastoma were recently shown to have frequent mutations in the *hSNF2/INI1* gene, which encodes for a component of the adenosine triphosphate (ATP)–dependent chromatin remodeling complex [31]. The same authors proposed that constitutional mutations in this gene lead to a greater incidence of renal and extrarenal malignant rhabdoid tumors, CPCs, central PNETs, and medulloblastoma, a complex they have coined as the "rhabdoid predisposition syndrome" [32]. The penetrance of the disease is high, with many probands developing malignant tumors before 3 years of age. Some pathologic data also suggest a connection between malignant rhabdoid tumors and CPCs [17].

A number of chromosomal abnormalities have been identified in CPP and CPC [33]. Surprisingly, even benign CPP (32 of 34 cases) demonstrated chromosomal aberrations. The patterns of aberrations in CPP differ from those observed in CPC. Wyatt-Ashmead and colleagues [17] reported the presence of deletions involving either an entire copy (3 of 4 cases) or a portion of chromosome 22 (1 of 4 cases) among a group of 5 cases of CPC studied.

Clinical features

For most patients with a choroid plexus tumor, hydrocephalus is responsible for the presenting symptoms. It is caused by overproduction of CSF and, in certain cases, the obstruction of CSF pathways, although it seems that overproduction is the major contributing factor (Fig. 2) [34]. Resolution of hydrocephalus has been reported after complete tumor removal, suggesting that CSF hypersecretion was responsible for ventriculomegaly [35–37]. Variations are likely to exist, because a normal rate of CSF production has been reported in a patient harboring a papilloma [38].

The most common presentation of choroid plexus neoplasms is related to increased intracranial hypertension secondary to obstructive hydrocephalus or CSF overproduction [4,8,39]. Because most cases occur in infants and young children, there are characteristic features of raised

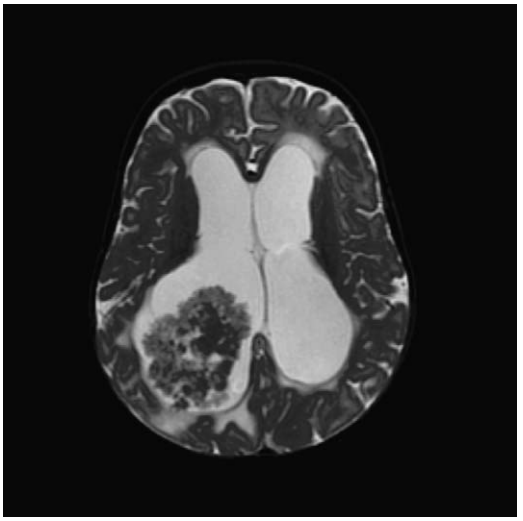

Fig. 2. An axial T2-weighted image demonstrates severe ventriculomegaly in a patient with a lateral ventricle choroid plexus papilloma. The entire ventricular system is enlarged, suggesting that hydrocephalus is primarily caused by cerebrospinal fluid (CSF) overproduction. Transependymal absorption of CSF is visible at the margins of the ventricles.

intracranial pressure (ICP). Ellenbogen et al [8] described the usual presenting signs and symptoms. The most common symptoms were nausea/vomiting, irritability, headache, visual difficulty, and seizure. As expected, the most common signs were craniomegaly, papilledema, and decreased level of consciousness. The duration of symptoms reported in this series varied from 2 months in those patients younger than 2 years of age to 6 months on average in those patients older than 2 years. Although choroid plexus neoplasms are viewed as slow-growing tumors, the presence of stupor or coma as the presenting sign in 25% of children suggests a more acute clinical course in some patients. Rapid decompensation can occur either from massive hydrocephalus or from tumoral hemorrhage. Of 21 patients who had CSF examined, 2 were found to have grossly bloody fluid. Lateralizing signs are found in a few patients and are usually related to asymmetric ventricular dilatation. Hydrocephalus was present in 78% of cases at the Hospital for Sick Children and in 95% of cases at the Children's Hospital in Boston [8,39].

Neuroimaging

The CT scan appearance of CPP is often characteristic for this tumor. The mass is well demarcated from the brain, lobulated, and often has punctate calcification. These tumors enhance homogeneously and intensely after contrast, reflecting a luxuriant blood supply [40]. An enlarged choroidal artery leading into the tumor mass can sometimes be seen in postcontrast images. At times, the massive size of these lesions may obscure the exact site of origin. Some carcinomas display a diffuse border between tumor and normal brain, which may reflect areas of brain invasion. On the basis of CT, certain features distinguish a suspected choroid plexus tumor from other possibilities. Cerebellar astrocytomas tend to be less homogeneously staining and often have cystic areas. Medulloblastomas are characterized by a more heterogeneous appearance, although they also stain vividly with contrast and may cause confusion with a fourth ventricular choroid papilloma. Ependymomas arise physically in similar locations but tend to enhance inhomogeneously. Finally, meningiomas can occur in an intraventricular location, but these tumors are rarely encountered in children.

Papillomas are isodense to brain on T1-weighted images (see Fig. 1). Areas of high signal indicate hemorrhage necrosis. After gadolinium administration, the tumor enhances brightly (Fig. 3A), although this can be patchy in nature, reflecting areas of high flow. T2-weighted images demonstrate an intermediate to high signal intensity with areas of heterogeneous internal signal (see Fig. 3B) [41]. MR spectroscopy of CPP and CPC is characterized by a prominent choline peak and absence of *N*-acetyl aspartate [42]. As with CT, an enlarged choroidal artery is often noted, especially with larger tumors. The vascularity of these tumors is easily demonstrated with specific perfusion sequences. MR angiography can sometimes help with defining the location of feeding vessels (see Fig. 3C). With CPC, the boundary between the tumor and surrounding brain can be indistinct in areas, but this is not a universal finding.

Treatment

Overall objectives

GTR is associated with the most favorable outcome in patients with choroid plexus tumors [5,43]. Therefore, any treatment plan should be tailored to reach this objective. The following steps are likely to be encountered as part of the surgical plan: (1) temporary or permanent

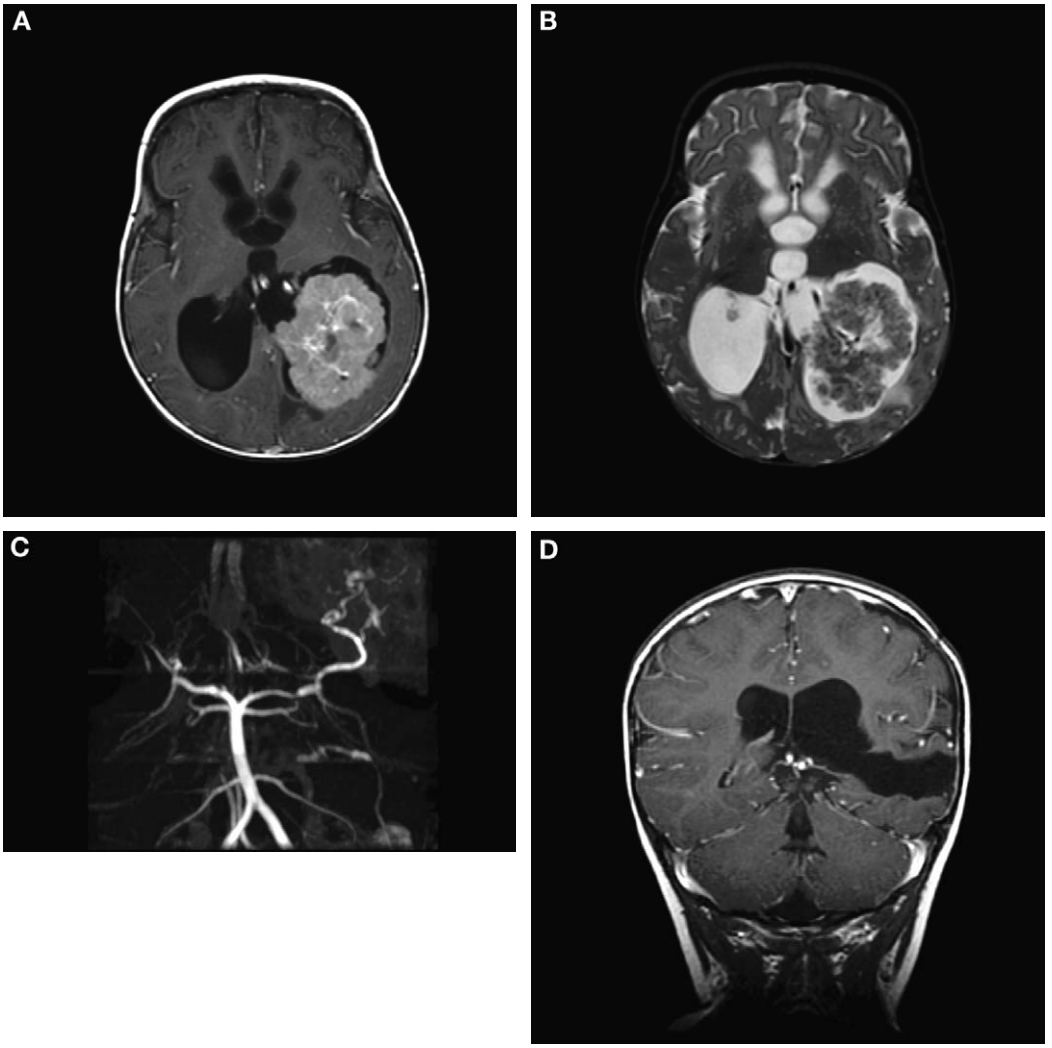

Fig. 3. An 18-month-old girl with a large left ventricular choroid plexus papilloma. (*A*) T1-weighted image after injection of gadolinium. (*B*) T2-weighted axial image. (*C*) MR angiogram demonstrating a choroidal artery arising from the posterior cerebral artery leading to the tumor. (*D*) A postoperative, coronal, T1-weighted image demonstrating the surgical route clearly shows an enlarged choroidal artery leading into the tumor. Postoperative coronal MRI demonstrates the route through the temporal lobe used to access the feeding artery initially and then the actual tumor itself.

resolution of hydrocephalus, (2) definition of the anatomic location and blood supply, and (3) planning of the surgical approach to allow maximal exposure and resection.

Management of hydrocephalus

Because most patients present with symptoms of intracranial hypertension secondary to hydrocephalus, the initial step is to decide whether CSF drainage is required. In infants less than 8 to 10 months of age, the presence of open sutures allows compensation by slow enlargement of the head, and treatment of hydrocephalus can often wait until the planned surgical date. If rapid neurologic deterioration is occurring, an external ventricular drain should be inserted immediately, regardless of the age of the patient. Placement of a ventriculo-peritoneal (VP) shunt, although an acceptable alternative, does not allow external CSF drainage and ICP monitoring in the intraoperative

and postoperative settings. Furthermore, the presence of intraventricular blood and debris after resection may lead to blockage of the shunt. If it is necessary to delay definitive surgical treatment beyond a few days, a VP shunt may allow time for the procedure to be performed electively.

A substantial number of patients have resolution of hydrocephalus after tumor resection. Matson and Crofton [36] and others have reported that the successful removal of a tumor obviates the need for shunting. It is likely that other factors, such as ventricular blood, postoperative changes, or meningitis, can also render a patient shunt dependent, however. Ellenbogen et al's series [8] noted that 37% of surviving patients required shunting. Two other series reported much higher rates of shunt dependency, ranging from 57% to 78% [39,44]. Raimondi and Gutierrez [45] have recommended that third and fourth ventricular tumors require immediate shunt placement, followed by a delay of 7 to 14 days before surgery. At our institution, a VP shunt is only placed in the postoperative period if there is clear evidence of hydrocephalus.

Preoperative imaging

The primary preoperative imaging study is a high-quality MRI scan with postgadolinium sequences performed in all three planes. Choroid plexus tumors enhance brightly and can be distinguished from normal brain tissue easily. Obtaining multiple planes assists with surgical planning and determining the relation of the tumor to various structures within the ventricles. Conventional catheter angiography is not required for diagnosis. Rather, its primary role is as a preoperative adjunct to reduce tumor vascularity [46]. Angiography has demonstrated that the vascular supply of papillomas is from normal choroidal vessels, which often enlarge as the tumor grows. Tumors of the lateral ventricle or third ventricle are generally supplied by branches of the anterior or posterior choroidal arteries. Mass effect tends to displace the internal occipital artery and the basal vein of Rosenthal in an inferior direction. A fourth ventricular tumor receives its blood supply from medullary or vermian branches of the posterior inferior cerebellar artery.

Operative treatment

As with most intracranial tumors, the exact approach is determined by avoiding eloquent tissue (eg, primary motor or sensory cortex, speech centers, visual cortex). The two features of choroid plexus tumors that can make resection exceedingly difficult are profuse vascularity and large size. Within the tumor, arterial vessels arborize rapidly; thus, control of hemorrhage within the tumor requires slow and tedious dissection. The most effective strategy focuses on initial exposure of the feeding artery and its ligation. In certain cases, approaching a tumor from the preferred angle to facilitate tumor resection does not allow early visualization of the feeding artery. In this case, a separate initial approach to allow coagulation of the feeding artery before tumor resection should be considered. In general, en bloc excision is recommended [45], although this may not be feasible with large tumors. Detailed descriptions of surgical approaches to the lateral and third ventricles are provided elsewhere in this issue.

For lateral ventricle papillomas, a direct cerebrotomy posterior to the angular gyrus allows access to the entire trigone and permits the pedicle of the tumor to be identified and coagulated. For more anteriorly located tumors, an incision can be made in the frontal convolutions and the lateral ventricle approached from an anterolateral direction. Lateral ventricular tumors can also be approached through a cerebrotomy through the superior or middle temporal gyri. Tumors located within the temporal horn are easily approached through the middle or inferior temporal gyri (see Fig. 3D). Extension of the cortical incision anteriorly or posteriorly allows access to the anterior or posterior choroidal vessels. Obviously, this approach must be tailored to avoid injury to eloquent areas in the dominant hemisphere.

Third ventricular tumors are rare and are approached via a midline transcallosal route. The anterior aspect of the ventricle is entered through a generous opening in the corpus callosum extending from the rostrum to the supraoptic recess. In this way, the tumor can be separated from the choroid of the tela choroidea, where it is usually attached, and the accompanying bridging vessels can be identified and divided.

Fourth ventricular tumors almost always produce triventricular obstructive hydrocephalus and may require preoperative CSF drainage and stabilization as noted previously. Tumors in this location arise from the caudal part of the roof of the fourth ventricle (Fig. 4). The tumor may extend into the lateral recesses or through the foramen of Magendie. The approach is via a standard midline posterior fossa craniectomy

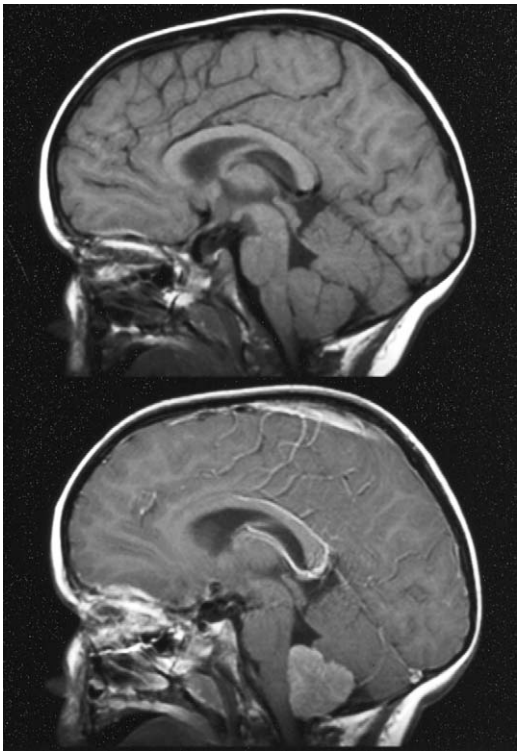

Fig. 4. A typical choroid plexus papilloma arising in the fourth ventricle. T1-weighted images before (*top*) and after (*bottom*) gadolinium enhancement are shown.

or craniotomy exposing the vermis and tonsils. The blood supply from branches of the posterior inferior cerebellar artery (PICA) are visualized from a medial vantage point. If vessels arise from the region of the superior vermis, it may be necessary to split the vermis to reach these vessels. In certain situations, the tumor may extend through the foramina of Luschka; in such cases, the tonsil can be reflected laterally to allow access to this area. Alternatively, at that sitting or during a staged procedure, a retrosigmoid approach can be used to reach the space anterolateral to the brain stem.

Special considerations in the pediatric population

The primary perioperative concerns are those related to blood loss, resuscitation, and anesthesia. It cannot be emphasized enough that the anesthesia team should be completely familiar with managing fluid replacement and blood loss in a pediatric patient. Although self-evident, it cannot be assumed that experience with adult perioperative problems guarantees expertise with similar problems in a child.

A 10-kg child has a circulating blood volume of 800 to 1000 mL (70–80 mL/kg). The entire blood volume can easily be lost during the resection of a particularly vascular tumor. Therefore, preoperative planning in the small child must anticipate the need for intraoperative transfusion, potentially several blood volumes, and the accompanying coagulopathy. Specifically, several units of blood and blood products should be available before starting the procedure. Adequate intravascular access is crucial, and a central line should be considered if peripheral access is limited. During surgery, continual communication with the anesthesiologist allows transfusion to be started before the development of adverse events, such as hypotension and hypoxia. Infants from 2 to 6 months of age reach a physiologic hemoglobin and hematocrit nadir, which may need to be considered if a significant degree of blood loss occurs. Prevention of a coagulopathy is critical, because hemostasis is exceedingly difficult once a coagulation defect is present. Finally, if several blood volumes have been replaced and tumor resection is not complete, serious consideration should be given to staging the procedure (Fig. 5).

Treatment of choroid plexus carcinomas

Overall, reported results in the literature confirm that GTR has a favorable impact on survival for carcinomas (see section on outcome). For this reason, as with papillomas, aggressive surgical treatment with GTR should be the primary objective. Nevertheless, GTR with carcinoma is achieved in less than 50% of cases. Combined with adjunctive therapy, radiation, or chemotherapy, survival after GTR ranges from 67% to 91% [47]. Technical considerations with CPC include the expected increased tumor vascularity, lack of a well-developed plane between the brain and tumor, and excessive friability of the tumor tissue. The rate of recurrence associated with GTR alone suggests that adjunctive therapy is useful, although definitive guidelines are not available [47].

Most chemotherapy regimens rely on cyclophosphamide, etoposide, vincristine, and a platinum agent [12,43,48]. Wolff and colleagues [5] noted that only 8 of 22 carcinomas responded to chemotherapy, a disappointing observation. Use of combination chemotherapy (ifosfamide, carboplatin, and etoposide) after an initial surgical procedure was found to reduce tumor volume and

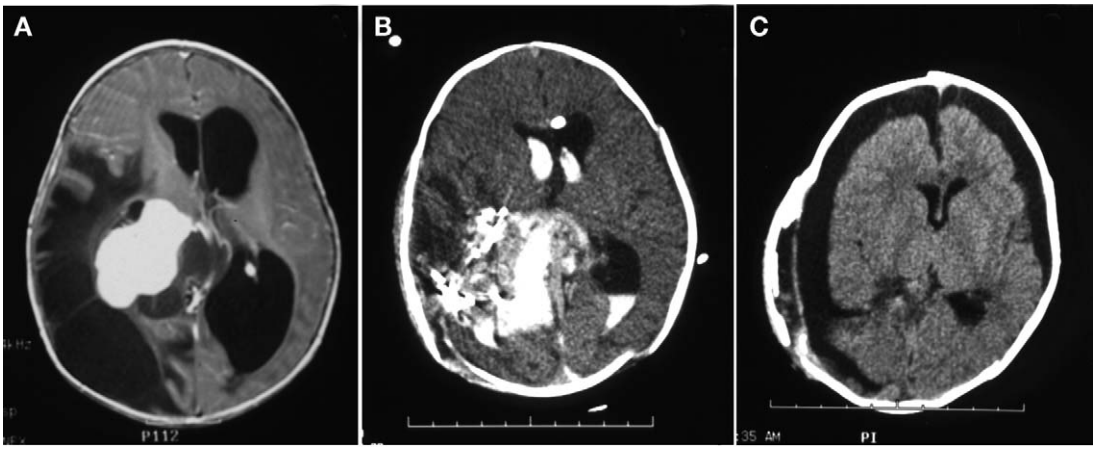

Fig. 5. A 2-month-old child with a right atrial choroid plexus papilloma is shown. (*A*) The preoperative T1-weighted image demonstrates bright enhancement after injection of gadolinium. (*B*) During the initial attempted resection, the deeper vascular pedicle could not be secured; after two blood volumes had been lost, the procedure was stopped. The postoperative CT scan demonstrates a large intraventricular and intraparenchymal hematoma. (*C*) After 2 weeks, the patient was taken back to the operating room, where the residual tumor and hematoma were resected. Postoperative CT shows resolution of the hydrocephalus with large extra-axial cerebrospinal fluid collections bilaterally. The patient did not require placement of ventriculo-peritoneal (VP) shunt.

allow a more complete resection during the second-stage operation [12,50]. Importantly, the vascularity of the tumor seemed to be greatly reduced, because measured blood loss during the second procedure was on average 15% of blood volume compared with an average of 64% of blood volume during the first procedure.

Postoperative radiation is usually recommended if the child is older than 3 years of age, although this therapy has not been subjected to a clinical trial. Radiation is also used in the presence of leptomeningeal dissemination, subtotal resection, and drop metastases. In one series, 10 patients with CPC were treated with either chemotherapy or craniospinal radiation [51]. Some of these patients demonstrated no evidence of disease after chemotherapy alone, but some required radiation to achieve disease control. The authors do suggest that radiation can be used as salvage therapy, but whether radiation for all patients with carcinoma would reduce relapse remains unclear. Fitzpatrick and colleagues [47] noted that after subtotal resection, radiation therapy, alone or in combination with chemotherapy, offered a survival advantage. The question of whether to use adjunctive therapy after GTR remains unclear, although the presence of relapse despite chemotherapy and radiation in a small group of patients suggests that surgery alone is not sufficient for CPC. Wolff and colleagues [49] support this view and state that GTR alone is insufficient for carcinoma and should be supplemented with radiation. The role of conformal radiation and radiosurgery is unknown; neither is the role of intrathecal chemotherapy. The experience reported by Packer et al [48] suggests that disease relapse confers a poor prognosis.

Outcome

Most patients with CPPs can expect excellent long-term survival. The survival for CPC, however, is much worse. In a recent meta-analysis, the 1-, 5-, and 10-year survival rates for papilloma were 90%, 81%, and 77% compared with only 71%, 41%, and 35% for carcinoma [5]. The extent of surgery is the most important treatment variable influencing long-term survival for papilloma and carcinoma [5,8,48]. The degree of surgical resection varied from 96% for papillomas to one of six cases in a series of carcinomas [8,12]. The overall crude survival rate in Ellenbogen et al's series [8] was 88% for patients with papillomas and 50% for those with carcinomas.

Packer et al [48] reported that GTR for carcinoma without adjunctive therapy offers the highest likelihood of success. Four of five patients with GTR remained disease-free at a median of 45 months after diagnosis. Five of six patients who

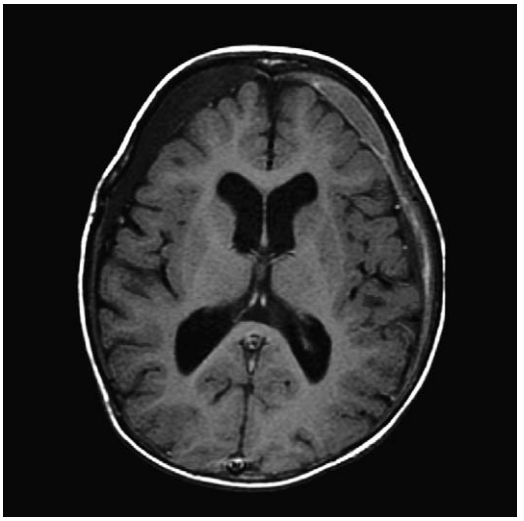

Fig. 6. Persistent subdural collections in a child several months after removal of a lateral ventricle choroid plexus papilloma. The patient remained asymptomatic with a gradual reduction in the size of the collections; thus, surgical drainage was not required.

had a subtotal resection relapsed. Although the authors of this study were pessimistic regarding the value of adjunctive therapy, the timing of this therapy may prove to be important with regard to survival after multimodality treatment. Two other reports noted that 5-year survival after GTR of carcinomas ranged from 26% to 40% [43,52]. Berger et al [43] also noted that surgery was the most important prognostic factor for CPC. The role of radiation also remains unclear. A brief report recently noted that the 5-year survival for patients with carcinoma and GTR followed by radiation was 68% compared with 16% for those not irradiated [49]. The two groups were not exactly comparable, but the clear suggestion is that surgery alone is insufficient to prevent recurrence of carcinomas.

Although papillomas are histologically benign and potentially curable, morbidity and mortality are significant concerns. With respect to operative mortality, modern series provide figures of 8% to 9.5% [39,44]. In the series from the Hospital for Sick Children, the cumulative mortality was 36%, most of which (six of eight cases) occurred in patients less than 12 months of age. Morbidity remains an important problem. In one series, 33% of patients with papillomas had persisting motor sequelae and psychomotor retardation [44]. In another series, 26% of patients were classified as having a fair or poor recovery [8].

As noted previously, the treatment of hydrocephalus goes hand in hand with the treatment of choroid plexus tumors. One treatment-related complication is the development of large subdural collections. These can result from a transcortical approach or by craniocerebral disproportion. In the former, the cause is believed to be a persistent ventriculosubdural fistula. Boyd and Steinbok [53] report that placement of pial sutures at the conclusion of the procedure can prevent the development of subdural collections. In the latter, reduction in the size of the ventricles and removal of a large mass can lead to an enlarged calvarium relative to the size of the brain (see Fig. 5C; Fig. 6). At times, these subdural collections may require treatment by the placement of a subdural-peritoneal shunt.

Summary

Choroid plexus tumors represent a well-defined subset of brain tumors that occur mainly in young children. Surgical resection for papilloma is usually curative, although careful surgical planning is required to minimize the potential risks. Although adjunctive therapy for carcinoma includes chemotherapy or radiation, the long-term survival for carcinoma remains poor.

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