

# Tumors of the Choroid Plexus

## Koroid Pleksüs Tümörleri

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**Abstract: Objective:** Choroid plexus neoplasms originate from choroid plexus epithelium. They are classified as papillomas and carcinomas histopathologically. The aim of this study is to discuss the signs, symptoms, treatment modalities and surgical results of this entities.

**Method:** Ten cases of choroid plexus tumors operated on in Neurosurgery Department of Marmara University Medical School - İstanbul between 1992 and 2000, are presented with the review of literature. Radiological investigations of the patients are reevaluated. Patients operative, histopathological and postoperative follow up records are detected.

**Findings:** One of the tumors was choroid plexus carcinoma and the others were choroid plexus papillomas. The youngest patient was two months old, the oldest one was 39 years old and the average age was 12,4. Female to male ratio was (5:5) one. Three patients were adults and the others were children. Major symptoms were headache, alteration in conscious level and enlargement of head. Most encountered neurological signs were papilla edema, alterations in conscious level, increased head circumference and sixth nerve palsy. Five of the tumors (50%) were localized in the trigon of the lateral ventricle, three were in the fourth ventricle(30%) and one each in the third ventricle(10%) and in cerebellopontine angle (CPA) (10%). All of the tumors were excised totally except two. One of the residual mass was excised with reoperation and the gamma knife surgery was applied to the other one. Two patient died in early postoperative period. The others are still alive. Average postoperative follow up is 25 months. **Conclusion:** Total surgical excision is the most important prognostic factor for choroid plexus tumors.

**Key words:** Choroid plexus, papilloma, carcinoma

**Özet: Amaç:** Koroid pleksüs tümörleri koroid pleksüs epitelinin köken alan neoplazmlardır. Histopatolojik olarak papillomlar ve karsinomlar olarak sınıflandırılırlar. Bu çalışmanın amacı bu tümörlerin belirti, bulgu, tedavi yöntemleri ve cerrahi sonuçlarını tartışmaktır.

**Metot:** 1992-2000 yılları arasında Marmara Üniversitesi Tıp Fakültesi Nöroşirürji Bölümü'nde opere edilen 10 koroid pleksüs tümörü olgusuna ait sonuçlar literatürle beraber tartışılmıştır. Olguların radyolojik incelemeleri tekrar gözden geçirilmiştir. Ameliyat bulguları, histopatolojik ve ameliyat sonrası izlem kayıtları yeniden değerlendirilmiştir.

**Bulgular:** Bir olgu koroid pleksüs karsinomu diğerleri koroid pleksüs papillomu olarak rapor edilmiştir. Olguların en küçüğü 2 aylık, en büyüğü 39 yaşında, ortalama yaş 12,4 dır. Kadın/erkek oranı (5:5) birdir. Üç olgu erişkin, diğerleri çocuktur. Baş ağrısı, bilinç bulanıklığı ve başda büyüme en sık rastlanılan şikayetlerdir. Nörolojik muayenelerinde, staz papiller, bilinç bulanıklığı, baş çevresinde büyüme ve 6. sinir parezisi en sık görülen bulgulardır. Beş olguda lateral ventrikül trigonunda (%50), 3 olguda 4. ventrikül içinde(%30), 1 olguda 3. ventrikül içinde (%10) ve bir olguda serebellopontin açıda (%10) tümör tespit edilmiştir. İki olgu haricinde tüm lezyonlarda total tümör eksizyonu uygulanmıştır. Subtotal eksizyon yapılan tümörlerin biri daha sonra tekrar opere edilerek rezidü tümör eksize edilmiş diğer olguda ise gamaknife cerrahisi uygulanmıştır. İki olgu erken postoperatif dönemde eksitus olmuş, diğer hastalar yaşamaktadır. Ortalama takip süresi 25 aydır.

**Sonuç:** Koroid pleksüs tümörlerinde total cerrahi eksizyon en önemli prognostik faktördür.

**Anahtar kelimeler:** Koroid pleksüs, papilloma, karsinoma

## INTRODUCTION

Tumors of the choroid plexus originate from choroid plexus epithelium. (9,21,26,36). Papillomas constitute 0,3-0,5% of all intracranial tumors. Carcinomas are less encountered than papillomas (15-30% of papillomas) (9,21,26,33,36). In childhood, overall ratio of choroid plexus tumors is higher than adults and it makes the 3% of all intracranial tumors of children (22,27). They are usually localized in lateral ventricles but by the increasing age, fourth ventricular tumors are seen more frequently (4,21,33,46). Increased intracranial pressure signs and symptoms are prominent features of these tumors (7,21,26,27,46). Hydrocephalus is considered to be a primary cause of this clinical picture (5,17,21,26,27, 29,34,46). Total surgical excision leads to cure for papillomas. Eventhough the carcinomas are highly malignant tumors, gross total resection of the tumor is the most important part of therapy (7,20,26,32,36,40,46).

## PATIENTS AND METHOD

Ten cases of choroid plexus tumors are presented here which were treated in Neurosurgery Department of University of Marmara-İstanbul between 1992-2000. Average age of the patients is 12,4. The youngest patient was 2 months old and the oldest one was 39 years old. Female to male ratio is 5:5. Seven patients were children (70%) and the others were adults (30%).

All of the patients were operated on and followed up in our clinic.

## FINDINGS

The complaints of the patients were headache (60%), alterations of conscious level (30%), diplopia (30%), increased head circumference (20%), nausea-vomiting (20%), tinnitus (%10) and weakness (10%). Neurological examinations revealed; Papilledema (50%), decreased conscious level (40%), sixth nerve palsy (30%), enlargement of head circumference (20%), sunset appearance (20%), cerebellar findings (20%), third nerve palsy (20%), visual field deficits (10%), hemiparesis (10%) and spasticity(10%).

Diagnoses were all made by computed brain tomography (CT) and magnetic resonance imaging (MR). Tumors were seen in the lateral ventricle

(Figure 1a,b) in five patients (50%), in the fourth ventricle (Figure 2a,b) in three patients (30%) and one each in the third ventricle (10%) and in CPA. In adults two of the tumors were in fourth ventricle and the other one was in CPA. While all of the adult tumors were localized in the posterior fossa only one out of seven childhood tumors was in so. Our papilloma cases except the tumor in CPA were all intraventricular and homogenously enhancing masses. In carcinoma case MR revealed a homogenously enhanced, intraventricular mass with 8 cm radius.

First imaging studies revealed hydrocephalus in six cases. Ventricles of the patient having a CPA tumor was found normal as a result of previous shunt operation had done in other institution. The preoperative hydrocephalus rate was 70 % for our cases.

Four lateral ventricle tumors were excised by transcallosal (Figure1c,d), the fifth was by posterior transcortical. While fourth ventricular tumors were operated on by standart suboccipital route (Figure 2c), tumor of CPA was excised by transpetrosal and the third ventricle tumor by pterional way. In two of the tumors, residual masses were left after surgery, one in left lateral ventricle and one in fourth ventricle. Five months after first operation 10 mm residual mass was excised from lateral ventricle. Gamma knife surgery was applied for 5mm residual tumor mass in the fourth ventricle. One of the patients that had a lateral ventricular tumor (carcinoma) and the patient with third ventricular papilloma had died in early postoperative course because of hypovolemic status due to the massive perioperative bleeding.

After tumor excision, one of the patient with lateral ventricular tumor needed a ventriculoperitoneal shunt. Because of the increased subdural cerebrospinal fluid (CSF) accumulation a subduroperitoneal shunts were applied in another two cases with lateral ventricular tumor. In the other three patients having hydrocephalus preoperatively, ventricles became normal after tumor excision. As a result two (%25) ventriculoperitoneal and two (%25) subduroperitoneal shunts were needed after operation.

The seventh nerve was injured perioperatively in CPA tumor case. Four months after primary surgery seven to twelve nerve anostomosis was done.

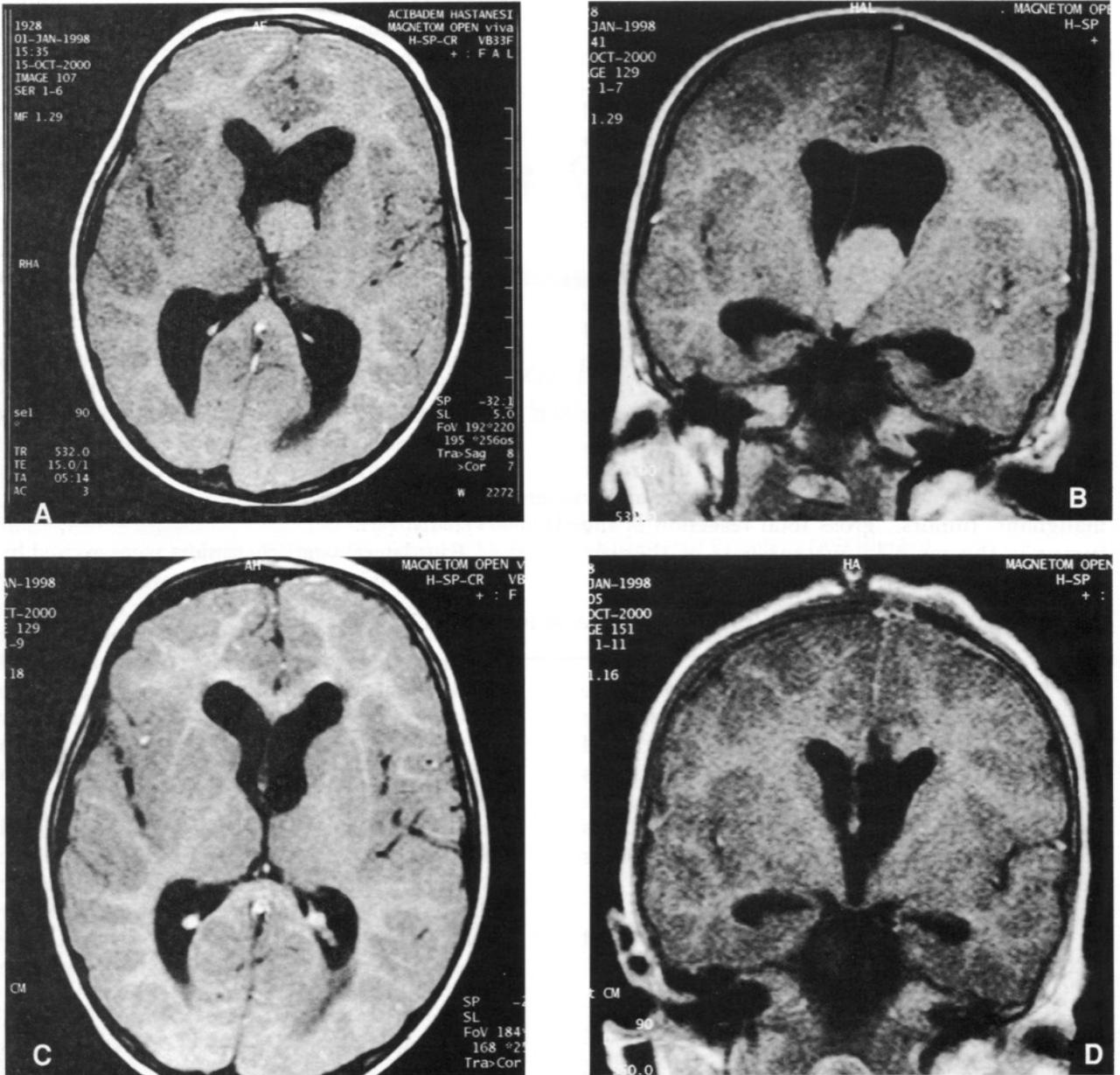


Figure 1a: Lateral ventricle located homogeneously enhanced, smooth surfaced papilloma that causes mild hydrocephalus (T1 weighted axial image with GdDTPA), 1b. Tumor protruding into the 3<sup>rd</sup> ventricle through foramen Monro (T1 weighted Coronal image with GdDTPA), 1c. Postoperative T1 weighted image shows total tumor excision and resolving of the hydrocephalus, 1d. Postoperative coronal image shows the callosotomy incision

Those patients had 15 operations and cumulative operative mortality is 13% (2/15) for this series. Average follow up period is 25 months. Any postoperative additional deficits were not seen but seventh nerve injury. Histopathological diagnosis were choroid plexus papilloma (Grade I - WHO 1993) for nine cases and carcinoma (Grade III - WHO 1993) for the tenth one.

### DISCUSSION

Choroid plexus tumors have a higher incidence in childhood and they constitute 3% of all intracranial tumors (22,26,27). Carcinomas are seen in elder age groups (4,21,26,29,33,35,46). These tumors are generally seen in the first year of life in children (7) as in three of our patients. Papillomas can be seen

congenitally like teratomas, astrocytomas, primitive neuroectodermal tumors and craniopharyngiomas (8,37). Some cases diagnosed as choroid plexus intrauterin were reported too (1,13). Choroid plexus tumors are epithelial tumors (9,21,26,36,39) and are classified as papillomas and carcinomas. Due to the classification of WHO in 1993 papillomas are classified as Grade 1 and carcinomas are grade 3-4 tumors (9,21,26,39). Nine of the tumors of this series are papillomas and the remaining one is carcinoma. Mitosis, atyp and necrosis were demonstrated in

carcinoma case but lack of vascular endothelial proliferation put this tumor into the grade 3 category.

Focal neurological findings like hemiparesis, visual field deficits, cerebellar, brain stem and cranial nerve symptoms due to the localization of the tumor can be detected with these patients, but as in our series the increased intracranial pressure signs are more prominent in diagnosis (7,21,26,27,46). Slowly growing tumor mass, accompanying hydrocephalus and the intraventricular localization of tumors are causes of this clinical picture.

In children choroid plexus tumors are usually localized in the trigonum of the lateral ventricles. By the increased age and in adults, localization of the tumor shifts to the infratentorial space and mostly into the fourth ventricle (4,9,20,21,26,46). But carcinomas are generally originate from lateral ventricle even in adults (21). The third ventricular tumors are very rare (12,21,28,29,42,43). In adults the pontocerebellar angle is another localization where tumors can be observed (3,21,44). Rarely, tumors in suprasellar space (22), in lateral cerebellomedullary sistrum (which lead to glossopharyngeal neuralgia) (19) and double sided lateral ventricles have been reported (4,21,46).

All of the cases presented here were diagnosed with both MR and CT. In CT papillomas

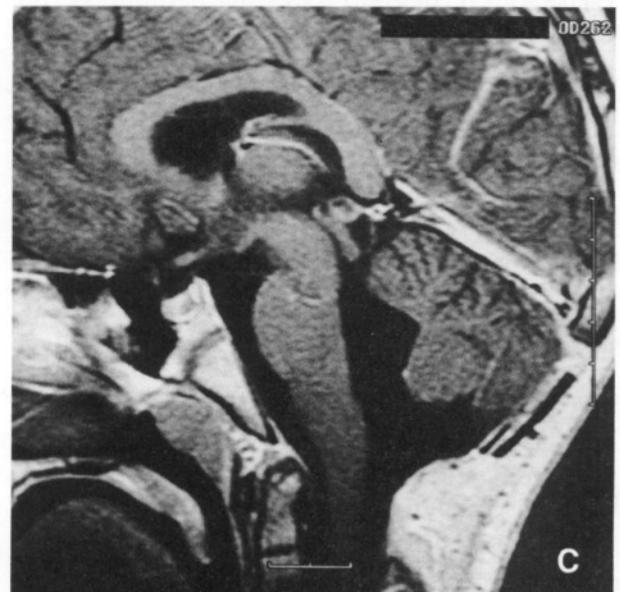
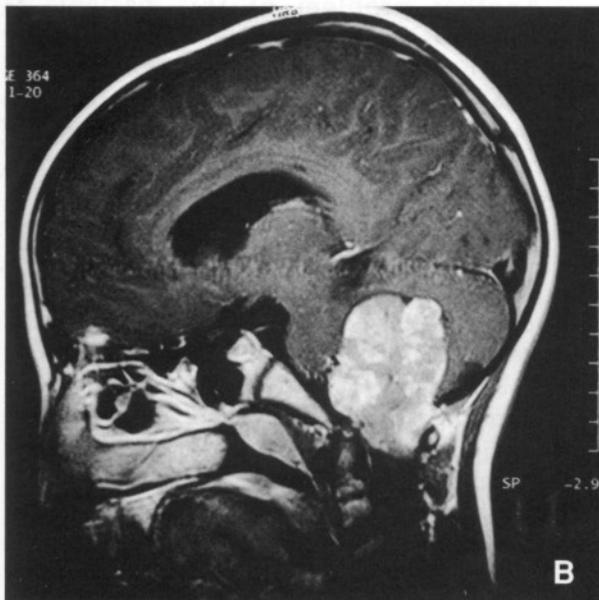
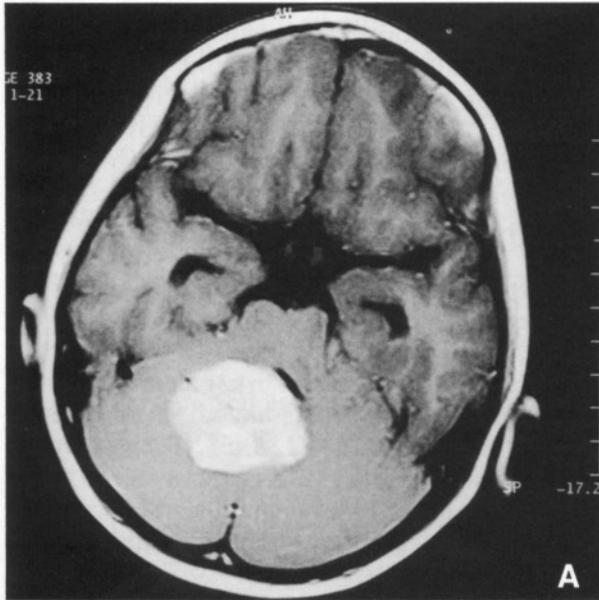


Figure 2a. Fourth ventricular, enhanced papilloma (T1 weighted axial image with GdDTPA), 2b. Sagittal image of the tumor filling all of the fourth ventricle (T1 weighted image with GdDTPA), 2c. Postoperative T1 weighted image shows total tumor excision.

are generally lobular, intraventricular, hyperdense or isodense masses and hydrocephalus usually accompanies to the tumor. Calcification can be seen and tumors enhance homogenously (4,5,21,25,26,31,46). In MR they appear as a isointense masses with brain tissue and enhancement with Gd-DTPA is very obvious (4,5,21,25,26,31,46). Carcinomas may have cystic and hemorrhagic portions. The contour of the tumor is usually irregular and the peritumoral edema secondary to brain invasion is very important. Cystic changes and hemorrhages are usually considered as malignant features (4,5). Some papillomas dissect the ependymal surface and show a malignant appearance (4,5,24) by penetrating the brain tissue. This can be observed intraoperatively and it's recommended that choroid plexus tumors with a benign cellular appearance but evidence of local parenchymal invasion and loss of the normal villus architecture at site of invasion be classified as papillomas. Patients with these tumors respond to surgical therapy alone without the need of adjuvant treatment (24).

Choroid plexus papillomas are benign tumors. Total surgical excision is the primary way of treatment (20,26,34,36,46). Partial resection causes the symptoms continue and increases the risk of malignant transformation (10,11,21). For resolving hydrocephalus, relieving compression symptoms and avoidance of the recurrence and malignant transformation, total surgical resection is necessary (11,16,21,33). In contrast to the papillomas, the carcinomas are malign lesions but total surgical excision is the first step in their treatment too (6,20,21,32,46). Prognosis is worse than papillomas. There are series reporting 100% 5 years survival rates for papillomas (35). This rate decrease to 26-40% for carcinomas (6,35). But it has been reported that if the total excision is achieved, 5 years survival rate of 85 % may be established (35). Adjuvant therapies are advocated for carcinomas specially for residual or recurrent lesions (20,30,40), but some authors suggest reoperation for carcinomas (6) too.

As a surgical approaches interhemispheric transcallosal or transcortical (transsulcal) transventricular routes can be use for lateral ventricular tumors. Decreased exposure for former and increased risk of seizure and neurological deficits for latter are disadvantages (21,36). Transcortical or transcallosal approaches give way CSF to reach subdural space easily and Nagib et al (27) offers sulcal dissection for lateral ventricular tumor. By this way after tumor resection, closed lips of sulcus

prevents compressing subdural accumulation by limiting the CSF escape to the subdural space. It's more easy to reach and coagulate feeding vessels by the transcortical (transsulcal) way. In bigger tumors, narrow operative corridor and bulk of the tumor makes this manoeuvre very hard in transcallosal approaches third ventricular tumors can be resected from transcallosal or transcortical way too (20,21,36,38,46). By the way some authors advocate combined approaches for giant third ventricular tumors (42).

Generally fourth ventricular choroid plexus papillomas adhere to the choroid plexus at the inferior of the ventricle (31). Although the pontocerebellar tumors usually grow into the subarachnoid space, some time they may invade the brain stem too (44). While the exploration of this location lower cranial nerves must be protected (19,44). In order to increase the operative view, transpetrosal approach was used for CPA tumor of this series.

Cerebrospinal seedings in papillomas and more higher rates in carcinomas have been reported in choroid plexus tumors (21,33,45). Because of this finding if new neurological signs appeared in follow up, all parts of the central nervous system must be examined again. There are no cranial or spinal spread in this series.

Another problem of this patients is hydrocephalus. It is related to the overproduction of the CSF by tumor tissue (17,20,26,34,46). Ghatak (17) et al observed secretuar organellas and histological structure in a papilloma case just like the normal choroid plexus, studied with electron microscopy. In some cases eventhough the total tumor excision is achieved, hydrocephalus persists. Inflammatory ependimitis, arachnoiditis, and CSF pathway obstruction have all been considered to be contributing factors (5,7,26,27,36,46). Sometime intraventricular localization of the tumors may lead to obstructive type hydrocephalus but it usually resolves after tumor excision (5,21,46). As mentioned before there is a risk of subdural bleeding or CSF accumulation in patients with very large ventricles (27,45). After tumor excision progressive subdural CSF accumulation is observed in two patients with lateral ventricular papillomas of this series and subduroperitoneal shunt operations were performed. In our series 50% of patients needed V/P or S/P shunts and because of this high ratio, detection of postoperative CSF

circulation problems are the most important part of follow up.

There are some alternative treatment modalities in the literature. Gaab et al (16) offers an endoscopic excision for small tumors and Duke (15) reported a tumor control in one papilloma case with stereotactic radiosurgery. After one year follow up, the tumor rest disappeared of our gamma knife applied case.

Adjuvant therapy is very dispute in carcinoma but there is consensus for total surgical excision is the primary condition for long survival (20,21,32,40,46). But chemotherapy is advocated in infants or residual or recurrent tumors and craniospinal radiotherapy for patients older than two years old (6,14,21,35,40,46). Because the carcinomas are very fragile and vascular tumors, some studies have been focused on preoperative chemotherapy that serves to tumor shrinkage and more comfortable operative status (18,41).

### CONCLUSION

1. Total surgical excision is the first condition that improves the survival rate in choroid plexus tumors.

2. It is possible to resect the tumors with minimal morbidity if the appropriate surgical approaches are used.

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*Chromosomal imbalances in choroid plexus tumors.*

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Among choroid plexus papillomas, children more often showed +8q, +14q, +12, and +20q; adults mainly presented with +5q, +6q, +15q, +18q, and -22q. Although the number of aberrations overall as well as of gains and losses on their own bore no significance on survival among choroid plexus tumors, a significantly longer survival among patients with choroid plexus carcinomas was associated with +9p and -10q.