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Case Report

Multi-Staged Endoscope-Assisted Microsurgical Resection of A Triventricular Choroid Plexus Carcinoma

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Summary

Choroid plexus carcinoma is a malignant intraventricular tumor of childhood with poor prognosis. Complete surgical resection is the most important prognostic factor. Unfortunately, complete resection of the CPC is a challenge for neurosurgeons due to extreme vascularity of the tumor, its large size, and its tendency to make invasion to the adjacent brain parenchyma. Here we present a 24-month-old boy with a triventricular CPC, to whom multi-staged endoscope-assisted microsurgical technique was performed. Complete resection was achieved. Postoperatively patient was improved neurologically and discharged home in stable condition. It is shown that multi-staged endoscope-assisted microsurgical technique definitely help neurosurgeons in dealing with this challenging tumors.

Key words: Choroid plexus carcinoma, endoscopy, hydrocephalus, surgical technique

Triventriküler Koroid Pleksus Karsinomunun Çok Aşamalı Endoskopik Asiste Mikrocerrahi Rezeksiyonu

Özet

Koroid pleksus karsinomu çocukluk çağının kötü prognozlu intraventriküler malign tümörlerindendir. Total cerrahi rezeksiyon en önemli prognostic faktörlerdür. Ne yazık ki, total cerrahi eksizyon tümörün aşırı derecede vasküler olması, çok büyük boyutlara ulaşabilmesi ve komşuluğundaki beyin parankimini invaze etmesi nedeniyle beyin cerrahları için oldukça zordur. Bu olguda, tri-ventriküler koroid pleksus karsinomu olup, çok aşamalı endoskop asiste mikrocerrahi teknik kullanılan 24 aylık erkek çocuk hastayı sunmaktayız. Tümör total olarak çıkartıldı. Postoperatif dönemde nörolojik bulguları düzelen hasta, stabil olarak taburcu edildi. Benzer derecede zorlu tümörlerin rezeksiyonun da çok aşamalı endoskop asiste mikrocerrahi tekniğin kesinlikle beyin cerrahına yardımcı olduğu gösterilmiştir.

Anahtar Kelimeler: Cerrahi teknik, Endoskop, Hidrosefali, Koroid plexus karsinomu

INTRODUCTION

Choroid plexus carcinomas (CPC) are rare malignant intraventricular tumors of childhood which derived from neuroectoderm1. Patients with CPC often manifests with a poor prognosis[1,11]. It is in agreement that the extent of surgery is the most important predictor of outcome[1,10,12]. Unfortunately, complete resection of the CPC is a challenge for neurosurgeons due to extreme vascularity of the tumor, its large size, and its tendency to make invasion to the adjacent brain parenchyma[1,5,12].

Here we present a case of a triventricular CPC of a child, which was completely resected with a multi-staged endoscope-assisted microsurgical technique.

CASE PRESENTATION

Twenty-four-month-old boy with complaints of irritability, confusion and vomiting admitted to emergency room. Initial neurological examination revealed that the patient was confused with left hemiparesis (+2/5 motor strength) and papiledema. Computed tomography (CT) of the head revealed a intraventricular mass lesion. Twenty days before this presentation right parietal craniotomy was performed for the removal of this lesion in an outside center, but due to excessive hemorrhage only biopsy can be taken (Figure 1).

External ventricular drainage system was placed for the management of the acute hydrocephalus. Through the same craniotomy the tumor was exposed in second-look surgery. Due to excessive hemorrhage, surgery was ended up with partial resection; and in the same session ventriculo-peritoneal shunt was placed. Two months after second operation patient deteriorated and magnetic resonance imaging (MRI) showed rapid growth of the tumor (Figure 2). The patient underwent tumor resection again through the same craniotomy. At this time endoscope-assisted microsurgical technique was used. Endoscope was used to provide better visualization of the tumor borders, neurovascular structures and the feeding arteries of the tumor that cannot be visualized under surgical microscope. After the coagulation of the feeding arteries of the tumor, bleeding was diminished. By dividing tentorium and the falx cerebri, resection of the posterior and the left intraventricular portion of the tumor were achieved. After this step, the rest part of the tumor in the 3rd ventricle was removed by endoscope. Immediate postoperative computed tomography of the head revealed small residual tumor located in the posterior part of the left lateral ventricle close to the midline. To achieve complete resection, next day the patient underwent his fourth operation for the resection of the residual. Falx cerebri and the tentorium was further divided to visualize the end portion of the tumor, where another vascular peduncle was seen and coagulated by the assistance of neuroendoscope; eventually radical complete resection of the tumor was succeeded (video). Histopathological examination revealed CPC with wide areas of necrosis. Neoplastic cells formed papillary structures with fibrovascular cores covered by cuboidal to columnar epithelial cells. In some areas, papillary architecture was lost and tumor cells were arranged in sheets with a prominent nuclear pleomorphism and increased cellularity. Mitotic activity was high. Immunohistochemically, neoplastic cells demonstrated focal positivity for S100 and vimentin. CK7 and glial fibrillary acidic protein were negative. Ki-67 proliferation index was about 10% (Figure 3). Postoperatively patient was improved neurologically and discharged home in stable condition. Due to aggressive nature of the tumor, the patient received chemotherapy. In the first postoperative year, patient was still doing well and MRI revealed no residual tumor (Figure 4).

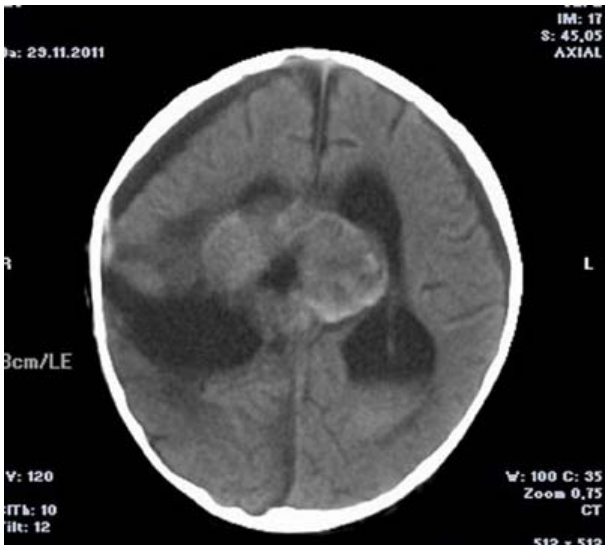

Figure 1: *Cranial CT demonstrating a lobulated intraventricular mass lesion associated with marked hydrocephalus and tip of the ventricular catheter.*

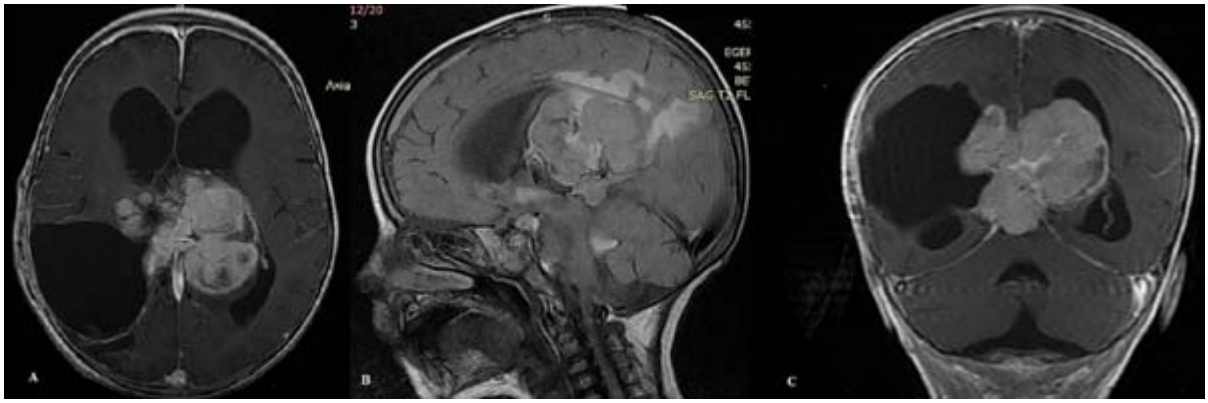

Figure 2: *Axial T1-weighted MRI with gadolinium (a) saggital T2-weighted FLAIR MRI (b) coronal T1-weighted MRI with gadolinium (c) demonstrating 10x8cm sized intraventricular mass lesion in both lateral ventricles extending through the third ventricle associated with marked hydrocephalus.*

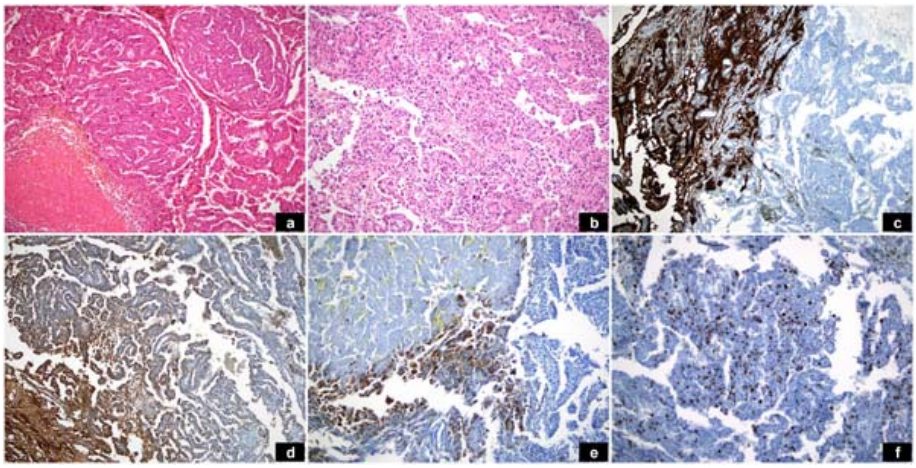

Figure 3: *(a) Histology shows tumor forming complex papillary structures with necrosis (hematoxylin-eosin x100) (b) In some areas of tumor, papillary architecture blurred and neoplastic cells demonstrated poorly formed sheets with nuclear pleomorphism and mitotic activity (hematoxylin-eosin x200) (c) While adjacent normal brain parenchyma was positive for glial fibrillary acidic protein, neoplastic cells were negative (Immunohistochemistry x 50) (d) S100 was focally positive (Immunohistochemistry x 100) (e) Some of the tumor cells were positive for vimentin (Immunohistochemistry x 50) (f) Ki-67 labeling was high (Immunohistochemistry x 50).*

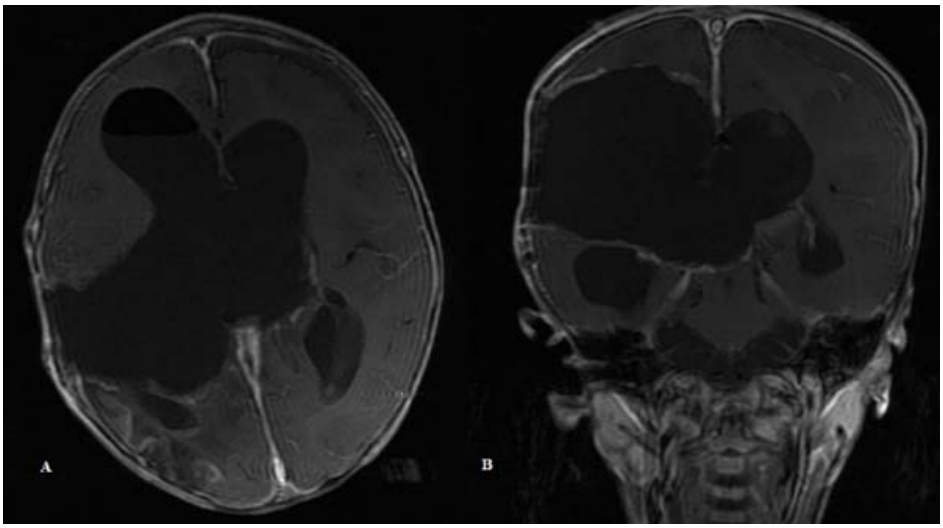

Figure 4: *Axial T1-weighted MRI with gadolinium (a) and coronal T1-weighted MRI with gadolinium (b) demonstrating no residual or recurrent tumor.*

DISCUSSION

Choroid plexus tumors are rare neoplasms derived from choroid plexus epithelium with benign and malign variants, typically classified as choroid plexus papilloma and CPC, respectively[4]. Choroid plexus carcinoma is a highly aggressive malignant tumor and classified as World Health Organization grade III, accounts for approximately 0.1% of all intracranial and 0.6% of primary pediatric central nervous system neoplasms[11].

Surgical resection is considered to be the most effective treatment for CPCs, and the surgical goal is gross total resection[1]. Gross total resection is generally considered to be the most important prognostic factor[3,4,7,12]. The five-year survival rate of 60% after complete resection was achieved where only ratio of 20% after partial resection was reported[4,7,14]. The median survival for CPC who underwent gross total resection was 58 months and those who had a subtotal resection was 36 months[13].

Unfortunately, complete resection of CPC is rarely achieved. Only 36-57% of complete resection ratios have been reported[2]. One of the most challenging points against complete resection is the extreme vascularity of the tumor[2,8,9,12]. In previous series, excessive blood loss was encountered and some patients died from intraoperative blood loss[2]. The key point of surgery is to secure the choroidal arteries at the early stages of surgery and obliterate the feeders as early as possible[9]. But as in our presented case due to size of the tumor and difficulties in navigation into the ventricles, it cannot be easily performed. At that point endoscope-assisted techniques may help surgeon to visualize and obliterate hidden feeder arteries. After the coagulation of the feeding arteries of the tumor, bleeding will be diminished and excessive hemorrhage will not be occurred, consequently. Another problematic issue in CPC surgery is the propensity of tumor to invade adjacent brain parenchyma[1,2,8].

A high-risk of hemorrhage sometimes results in tumor left behind. As complete tumor resection significantly improves the survival, second-look surgery is recommended to achieve gross total resection[6,13]. In presented case, due to hemorrhage the first two surgeries had to be completed only with biopsy and partial resection, respectively. When endoscope used to assist microsurgical technique, this giant tumor was completely resected.

Ventricles provide an anatomical corridor for endoscope causing intraventricular tumors available to endoscopic resection. Intraventricular lesions that have a non-linear axis, that extend into a cistern, or that adhere to critical neurovascular or anatomical structures can be challenging for neurosurgeons[3,5,12]. The simultaneous use of microneurosurgical technique with neuroendoscope offers several advantages. By this method multicompartmental tumors can easily be resected, neurovascular or anatomical structures can be approached by direct visualization. Blind maneuvers can also be reduced by neuroendoscope. As a result, combined approaches should maximize the relative advantages of both microsurgery and neuroendoscopy.

As conclusion, CPCs are challenging lesions that have to be completely resected for prolonged survival. Multi-staged endoscope-assisted microneurosurgical technique is shown to be effective for complete resection of the CPCs.

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REFERENCES

1. *Berger C, Thiesse P, Lellouch-Tubiana A, Kalifa C, Pierre-Kahn A, Bouffet E. Choroid plexus carcinomas in childhood: clinical features and prognostic factors. Neurosurgery 1998; 42: 470-475*
2. *Due-Tønnessen B, Helseth E, Skullerud K, Lundar T. Choroid plexus tumors in children and young adults: report of 16 consecutive cases. Childs Nerv Syst 2001; 17: 252-256*
3. *Ellenbogen RG, Winston KR, Kupsky WJ. Tumors of the choroid plexus in children. Neurosurgery 1989; 25: 327-335*
4. *Gopal P, Parker JR, Debski R, Parker JC. Choroid plexus carcinoma. Arch Pathol Lab Med 2008; 132: 1350-1354*
5. *Gore PA, Nakaji P, Deshmukh V, Rekate HL. Synchronous endoscopy and microsurgery: a novel strategy to approach complex ventricular lesions. Report of three cases. J Neurosurg 2006; 105: 485-489*
6. *Lafay-Cousin L, Keene D, Carret AS, Fryer C, Brossard J, Crooks B, Eisenstat D, Johnston D, Larouche V, Silva M, Wilson B, Zelcer S, Bartels U, Bouffet E. Choroid plexus tumors in children less than 36 months: the Canadian Pediatric Brain Tumor Consortium (CPBTC) experience. Childs Nerv Syst 2011; 27: 259-264*
7. *Menon G, Nair SN, Baldawa SS, Rao RB, Krishnakumar KP, Gopalakrishnan CV. Choroid plexus tumors: an institutional series of 25 patients. Neurol India 2010; 58: 429-435*
8. *Meyers SP, Khademian ZP, Chuang SH, Pollack IF, Korones DN, Zimmerman RA. Choroid plexus carcinomas in children: MRI features and patient outcomes. Neuroradiology 2004; 46: 770-780*
9. *Ogiwara H, Dipatri AJ Jr, Alden TD, Bowman RM, Tomita T. Choroid plexus tumors in pediatric patients. Br J Neurosurg 2012; 26(1): 32-37*
10. *Pierga JY, Kalifa C, Terrier-Lacombe MJ, Habrand JL, Lemerle J. Carcinoma of the choroid plexus: a pediatric experience. Med Pediatr Oncol 1993; 21(7): 480-487*
11. *Rickert CH, Paulus W. Epidemiology of central nervous system tumors in childhood and adolescence based on the new WHO classification. Childs Nerv Syst 2001; 17(9): 503-511*
12. *St Clair SK, Humphreys RP, Pillay PK, Hoffman HJ, Blaser SI, Becker LE. Current management of choroid plexus carcinoma in children. Pediatr Neurosurg 1991-1992; 17(5): 225-233*
13. *Wrede B, Liu P, Ater J, Wolff JE. Second surgery and the prognosis of choroid plexus carcinoma.*

Results of a meta-analysis of individual cases. Anticancer Res 2005; 25 (6): 4429-4433

14. *Wrede B, Liu P, Wolff JE. Chemotherapy improves the survival of patients with choroid plexus carcinoma: a meta-analysis of individual cases with choroid plexus tumors. J Neurooncol 2007; 85(3): 345-351*