Surgical Removal of a Huge Atypical Choroid Plexus Papilloma

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The authors present a case of huge atypical choroid plexus papilloma in both lateral and third ventricles. A 2-year-old girl presented with lethargy and vomiting. Magnetic resonance images disclosed a huge enhancing mass, which probably arose from the left lateral ventricle and extended to the right lateral and the third ventricles. Total removal of the mass via the interhemispheric transcralosal approach was done. Histologically, an atypical choroid plexus papilloma was identified. Second operative treatment, subdural-peritoneal shunt, was performed due to postoperative subdural effusion. The patient was discharged two weeks after the second surgical procedure without a neurological deficit.

KEY WORDS: Choroid plexus papilloma - Transcallosal approach - Subdural effusion.

Introduction

Choroid plexus tumors are rare, composing 0.4 to 0.6% of all intracranial tumors in all ages. Choroid plexus neoplasms are primary tumors of childhood. They occur more than 70% in children, and at least 50% presented in less than 2 years. They are most often found in the lateral ventricles and less commonly in the fourth ventricle in children, and these tumors generally localized in the fourth ventricle and its lateral recess in adults.

Some rare cases of bilateral choroid plexus papilloma have been published. Matson and Crofton did note that bilateral choroid plexus papillomas represent 7% of their cases. Third ventricular choroid plexus neoplasms in pediatric series represent average 10% with ranging from 7% to 29%.

Choroid plexus tumors arise from the neuroepithelial lining of the ventricular choroid plexus and their histologic and biologic properties demonstrate widely from the benign papillomas to highly anaplastic and infiltrative carcinoma.

We report a 2-year-old patient with atypical choroid plexus papilloma of the lateral and third ventricle with literature review.

Case Report

A 2-year-old girl presented with vomiting and lethargy. She had a history of poor oral intake for three months. On a neurological examination, the patient was drowsy. She had long tract signs, including an increased deep tendon reflex and positive bilateral ankle clonus.

A preoperative computed tomography scan revealed a huge lobulated mass lesion in the lateral ventricles extended to the third ventricle with hydrocephalus. She underwent an extraventricular drainage due to a progressive deterioration of her mental status.

Magnetic resonance imaging disclosed a huge intraventricular tumor, which probably arose from the left lateral ventricle and extending to the right lateral and the third ventricle. The mass lesion was enhanced densely and homogeneously and linear enhancement was noted in prepontine cistern suggesting leptomeningeal seeding. A presumptive diagnosis of a choroid plexus papilloma was made.

The mass lesion was resected via anterior interhemispheric transcralosal approach. At surgery, a huge whitish to pinkish lobulated soft tissue mass lesion was exposed, encapsulated by a thin layer of fibrous tissue. A gross total removal was achieved by piecemeal.

Histologically, an atypical choroid plexus papilloma was identified. The microscopic examination of the tumor showed the histological features of a choroid plexus tumor. However, the slightly higher number of hyperchromatic nuclei and frequent mitoses were identified. There were also
psammoma bodies. Immunostaining of S-100, and cytokeratin showed diffuse positive reactions with focal immunoreactivities to vimentin.

A postoperative CT scan demonstrated intraventricular hemorrhage, which was resolved by extraventricular drainage. On the 11th postoperative day, a slight mental deterioration developed and MRI showed a subdural fluid collection in the left fronto-parietal area (Fig. 3). Therefore, a subduro-peritoneal shunt was performed. The patient was discharged 2 weeks after the second surgical procedure without a neurological deficit.

**Discussion**

I n 1832, Guerard described the first case of a choroid plexus tumor in an autopsy specimen of 3-year-old girl. Rare case of bilateral ventricles was also noted. In our patient, a huge lobulated mass was located in the lateral ventricle body and frontal horn, more on the left than right side, involving third ventricle. The lateral and third ventricles were enlarged but aqueduct and fourth ventricle appeared to be normal, which suggests the obstruction at foramen of Monro and proximal aqueduct. The tumor in the left lateral ventricle seems to be extension of tumor tissues through foramen to right lateral and third ventricle.

The hydrocephalus is cardinal manifestation of choroid plexus neoplasm. Probably it is the result of obstruction of CSF pathway or over-production of CSF. Over production of CSF is major contribution factor. The resolution of hydrocephalus has been reported after complete resection of choroid plexus tumor, supporting the basis that over secretion of CSF was responsible for the hydrocephalus. Nevertheless, hydrocephalus persists in one-third to one-half of patients with choroid plexus neoplasms and require CSF shunt procedure after complete tumor removal. Rekate et al. demonstrated that a complex combination of CSF overproduction and limited outflow may be the cause in many patients. Also in our case, a combination of CSF overproduction and outflow obstruction may be the contributational factor to the hydrocephalus.

Matson and Crofton have reported that the successful removal of a tumor obviates the need for shunt. However, other factors, such as blood in the ventricle, arachnoid scarring, and postoperative meningitis may render the patient shunt dependent. Ellenbogen and colleagues reported that 37 percent of surviving patients require shunting in his series. Lena et al. reported much higher rates of shunt placement. Plain films may show the usual evidence of increased intracranial pressure and calcification. The introduction of CT and MRI improved the safety and accuracy of diagnosis of these tumors. Especially, MRI is the diagnostic imaging study of choice because of the detailed anatomical delineation and triplanar imaging ability. Choroid plexus papilloma could have either homogeneous or heterogeneous intensity on MRI and usually enhances intensely and homogeneously.

The gross appearance of choroid plexus papilloma appears a cauliflower like surface and generally well-circumscribed mass arising from within the ventricle. Microscopically, choroid plexus papillomas typically appear as a single layer of cuboidal epithelial cells surrounding a delicate fibrovascular stalk, arranged in papillary configuration with finger-like projection of tissues. On occasion, a choroid plexus tumors may demonstrate some atypical histopathological features without frank evidence of invasion. Microscopic features include increased cellularity, mitoses, nuclear pleomorphism and poorly formed papillary structures. These intermediate tumors may poses one or two of these features and are called or termed atypical papilloma, but do not necessarily have a more aggressive natural history and are not classified as carcinoma. The histologic examination of our patient was concordant with the features of these in-

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**Fig. 2.** Photomicrograph shows the histology of the atypical choroid plexus papilloma. There are slightly higher number of hyperchromatic nuclei and frequent mitoses (arrow) (H & E, 400).
ternediate tumors. So it was diagnosed as a atypical choroid plexus papilloma.

In general, transcortical and transcallosal approaches have been used for the surgical removal of the choroid plexus tumors. The main determining factors of choice of surgical approach were preoperative condition of patient, location of tumor, age of patient and vascular supply. Velasco-Siles and Raimondi\(^\text{30}\) provided a detailed outline of surgical approaches and pitfalls in the resection of choroid plexus papilloma. In selected patients, transcallosal approach is useful, because this permits the access to the mass in the ventricle with low risk of neuropsychological sequelae and seizure. Total and en bloc tumor excision is currently performed and recommended\(^{14}\). En block excision can’t be done in large tumors because sometimes it is very difficult to mobilize a large mass\(^{15,17}\). Therefore the authors performed total resection with peacemal removal.

One of the most problematic complication of intraventricular surgery for the choroid plexus neoplasm is the development of symptomatic subdural fluid collections\(^{2,14}\). Koos et al\(^{11}\) thought that a preoperative shunt procedure allows resolution of the hydrocephalus and decreases the risk of post operative subdural fluid collection in the majority of cases. Boyd and Steinbok\(^{2}\) have suggested that these subdural collection related to persistence of a ventriculosubdural fistula. They have argued for use of pial stay-suture to close the cortical incision, reinforced with Tissel\(\text{Baxter Healthcare Corp., Deerfield, IL, USA)}\), to prevent this complication. In our patient, postoperative condition of patient, location of tumor, age of patient and vascular supply. Velasco-Siles and Raimondi\(^{30}\) provided a detailed outline of surgical approaches and pitfalls in the resection of choroid plexus papilloma. In selected patients, transcallosal approach is useful, because this permits the access to the mass in the ventricle with low risk of neuropsychological sequelae and seizure. Total and en bloc tumor excision is currently performed and recommended\(^{14}\). En block excision can’t be done in large tumors because sometimes it is very difficult to mobilize a large mass\(^{15,17}\). Therefore the authors performed total resection with peacemal removal.

Incidence of postoperative neurological sequelae of choroid plexus papilloma is between 29% and 63%, despite these tumors are benign. Despite the treatment of choroid plexus papilloma still results high morbidity rate, the successful total removal of the choroid plexus papilloma is the most important determinant of the prognosis.

**Conclusion**

The authors report a case of huge atypical choroid plexus papilloma and suggest the extent of a surgical excision of a choroid plexus papilloma may be the most important factor for preventing post-surgical complications and determining the prognosis.

**References**