Choroid Plexus Carcinoma in an Adult Patient: Case Report

Erişkin Bir Hastada Koroid Pleksus Karsinomu: Olgu Sunumu

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ABSTRACT

Although choroid plexus carcinoma is a rare and malignant intraventricular neoplasm that is usually seen in the pediatric age group, it may also present in older individuals. Radiotherapy following radical surgery is the main treatment strategy.

A 54-year-old male patient with choroid plexus carcinoma is reported. Preoperative and postoperative radiological examinations were performed. The patient underwent radiotherapy following gross total excision of the tumor but died at the end of the first year despite treatment.

KEY WORDS: Adult, Carcinoma, Choroid plexus

ÖZ

Koroid pleksus karsinomu nadir ve genellikle çocukluk çağının malign bir intraventriküler neoplazisi olmakla birlikte, yaşlı bireylerde de görülebilir. Radikal cerrahiyi müteakip radyoterapi temel tedavi stratejisidir.

Koroid pleksus karsinomu olan 54 yaşında bir hasta rapor edildi. Preoperatif ve postoperatif dönemde radyolojik incelemeler yapıldı. Gros total tumor eksizyonunu müteakip radyoterapi uygulandı. Tüm tedavi prosedürlerine rağmen hasta birinci yılın sonunda kaybedildi.

ANAHTAR SÖZCÜKLER: Erişkin, Karsinoma, Koroid pleksus

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INTRODUCTION

Choroid plexus carcinoma is rare central nervous system tumor derived from the choroid plexus epithelium, affecting mainly children under 3 years of age (1). Even though the occurrence of this neoplasm is exceptional beyond childhood, pathologists should consider a malignant choroid plexus tumor when making the differential diagnosis of an intraventricular papillary neoplasm in adults (5).

We present a 54-year-old patient with choroid plexus carcinoma. He was treated surgically by gross total resection and radiation of the whole cranium and spinal cord following surgery. According to our knowledge, this is one of the oldest patients reported in the literature.

CASE REPORT

A 54-year-old male patient was admitted to our department with complaints of headache and visual and gait disturbance. Bilateral papilledema was detected during the neurological examination. The cranial computed tomography (CT) revealed hydrocephalus and a contrast-enhancing mass lesion of millimetric size in the right lateral ventricle. Magnetic resonance imaging (MRI) was performed to evaluate the nature of this small mass lesion. A small, well-circumscribed and round tumor causing hydrocephalus was detected and interpreted as choroid plexus papilloma on MRI (Figure 1A, 1B, 1C). Lumbar puncture was performed to examine the biochemical and cytopathological features of the cerebrospinal fluid (CSF) and an increased protein level of 266 mg/dl was found. No malignant cell was observed in the CSF. Whole spinal MRI was also normal.

The mass lesion was grossly removed totally by an interhemispheric transcallosal approach to the lateral ventricles. During the operation, the surface of both ventricles was observed to be green-colored and cheese-like. This was probably due to local invasion by the tumor.

Pathological examination revealed a complex glandular and papillary architecture in some areas but a mostly solid tumor. The tumor was highly cellular and exhibited significant nuclear atypia and pleomorphism (Figure 2). Bizarre multinucleated neoplastic cells and brisk mitotic activity were present. Most neoplastic cells had a large and deeply eosinophilic cytoplasm giving them an "oncocytic"

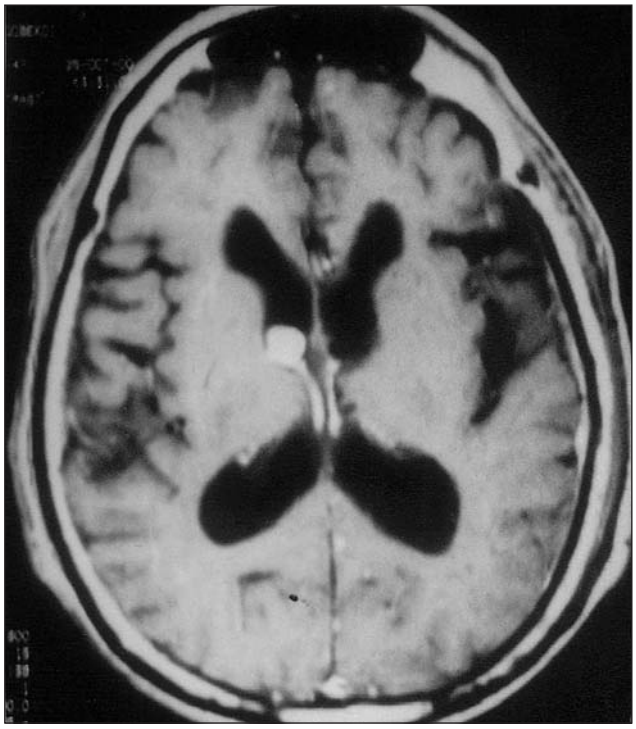

Figure 1A: The axial MRI scan of the patient showing a contrast-enhancing mass lesion in the right lateral ventricle.

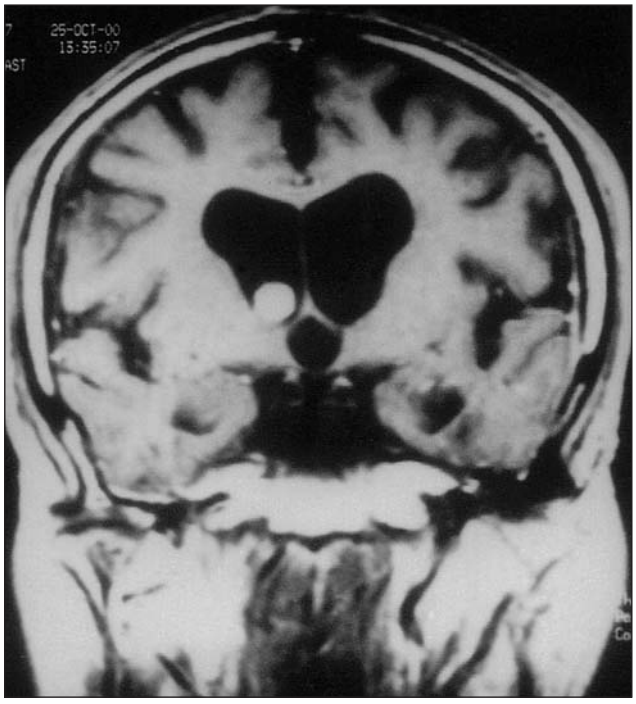

Figure 1B: The coronal MRI scan of the patient showing a small, round, well-circumscribed mass lesion in the right lateral ventricle.

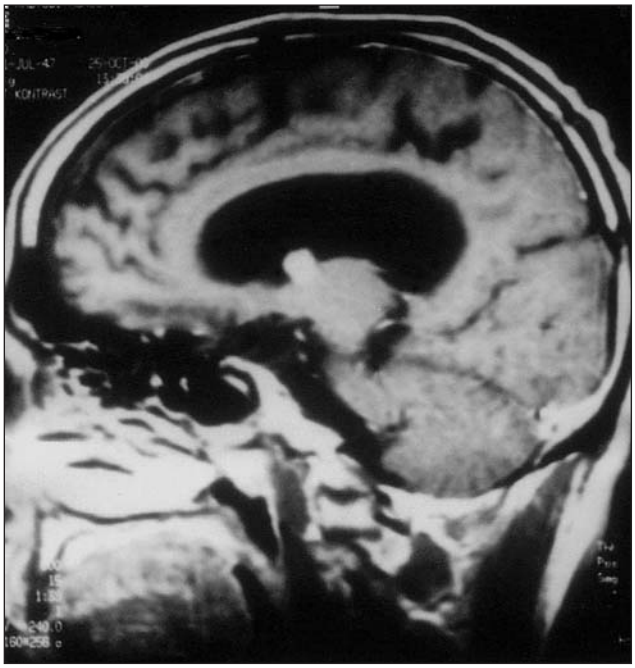

Figure 1C: The sagittal MRI scan of the patient showing the mass lesion and hydrocephalus.

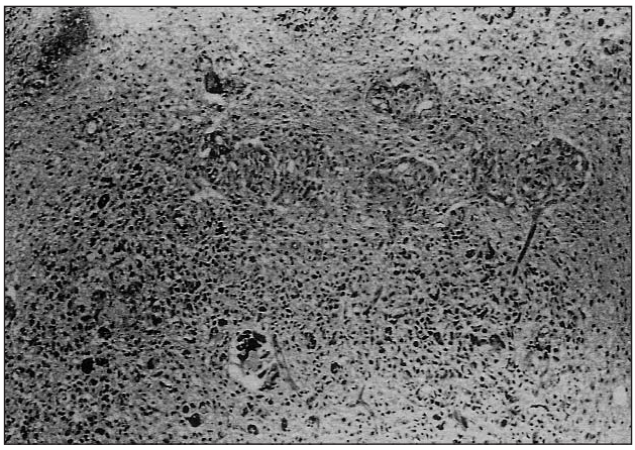

Figure 2: Complex gland-like structures are present in some areas of this tumor. Note the significant nuclear atypia and large eosinophilic cytoplasm of the neoplastic cells.

appearance. Immunohistochemically, most neoplastic cells were reactive for cytokeratin and vimentin. Immunoreactivity for glial fibrillary acidic protein (GFAP) was also present in a few tumor cells. S-100 staining was not present in the neoplastic cells. Malignant ependymoma and metastatic carcinoma were considered in the histopathological differential diagnosis. A metastatic malignancy was ruled out on the basis of clinical and neuro-radiological findings. Malignant ependymomas are generally more monomorphic tumors with perivascular pseudorosettes and usually show vascular proliferation and necrosis. Immunohistochemically, ependymomas, in distinction to papillomas, are widely positive for GFAP and negative for cytokeratin.

Following surgical treatment, the patient underwent radiotherapy of the cranium and all the spinal axis. Postoperative cranial MRI five months after the surgery confirmed the absence of recurrence (Figure 3). The patient died at the end of the first year following the operation due to deep venous thrombosis and pulmonary edema.

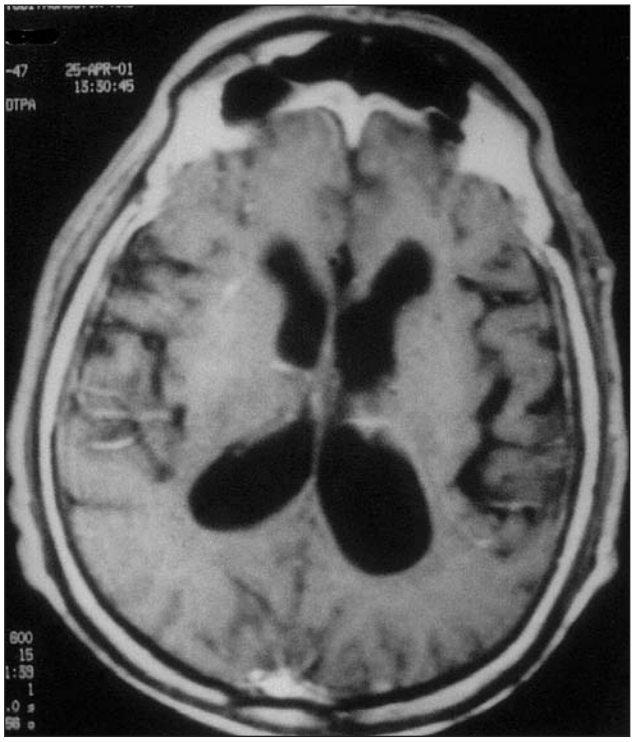

Figure 3: The postoperative axial MRI scan of the patient showing absence of the tumor.

DISCUSSION

Choroid plexus tumors are rare intraventricular neoplasms derived from choroid plexus epithelium, and account for only between 0.4-0.6% of all intracranial and 2-3% of pediatric neoplasms. Plexus papillomas outnumber choroid plexus carcinomas by a ratio of 5:1 and around 80% of choroid plexus carcinomas arise in children (6). Most reported cases of lateral ventricle choroid plexus carcinomas are under 40 years old, and the majority of these are children. Our case is one of the oldest patients diagnosed with choroid plexus carcinoma in the lateral ventricles.

Clinically, this group of tumors tend to cause hydrocephalus and increased intracranial pressure (3). Bleggi-Torres et al (1) reported 15 cases of choroid plexus carcinoma in 2000 and pointed out that the main symptoms of this tumor are hydrocephalus (62.5%), intracranial hypertension (25%) and convulsion (12.5%). In their series, the oldest patient was 21 years old and the results supported the poor prognosis and high mortality rate of choroid plexus carcinoma (1). In our patient, hydrocephalus was present at the time of admission and papilledema was the main neurological finding.

Mainprize et al (5) presented another case of choroid plexus carcinoma in a 22-year-old female patient. They stated that pathologists should consider this malignant tumor in the differential diagnosis of intraventricular neoplasms in adults. The MRI features of our patient supported the diagnosis of choroid plexus papilloma but choroid plexus carcinoma was identified by histopathological examination following surgery.

Choroid plexus carcinoma is a rare and frequently lethal tumor. Its cure depends on the achievement of gross total resection. Greenberg (3) suggested that the contribution of adjuvant therapies, both irradiation and chemotherapy, in the context of gross total resection is unclear, but where such resection is not possible there may be a role for adjuvant therapy to permit more nearly complete resection. He also pointed out that the use of chemotherapy following an initial biopsy may influence the prognosis of this malignant tumor. Souweidane et al (7) also supported the use of preoperative chemotherapy to achieve volumetric reduction of choroid plexus carcinoma in a 15-month-old girl. They achieved a reduction in tumor volume of 29.5% with preoperative chemotherapy. We prefered irradiation following gross total resection of the tumor in our patient and obtained a good result in the short term.

Inamura et al (4) reported an autopsy case of primary choroid plexus adenocarcinoma arising in a 40-year-old female, associated with a high serum level of carbohydrate antigen 19-9 (CA 19-9). The immunohistochemical examination of tumor tissue specimens revealed intense reactivity for CA 19-9. In our patient, the preoperative serum level of CA 19-9 was normal, and the tumor specimens did not show reactivity for CA 19-9.

Connor et al (2) published the most recent article on choroid plexus carcinoma cases in 2002 and emphasized the MRI features of such carcinomas in the preoperative and postoperative period. They suggest that total surgical resection is a significant prognostic factor in the postoperative period. We also reported on the preoperative and postoperative MRI features of our patient. Our patient is the oldest reported patient with intraventricular choroid plexus carcinoma. Another interesting characteristic of our case is oncocytic transformation.

In conclusion, choroid plexus carcinoma should be kept in mind in the differential diagnosis of all intraventricular mass lesions during the radiological examination, especially in adult patients. Early surgical resection and irradiation is an effective protocol to achieve good results.

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