Recurrence of Choroid Plexus Papilloma with Malignant Transformation

—Case Report and Lectin Histochemistry Study—

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Abstract

A very rare primary choroid plexus carcinoma occurred in a 44-year-old male presenting with occipitalgia, nausea, and blurred vision. The tumor had progressed from a choroid plexus papilloma in the fourth ventricle which was totally removed 6 years previously. Lectin histochemistry might be useful for the differential diagnosis of primary choroid plexus neoplasms and other brain tumors such as secondary carcinoma.

Key words: choroid plexus neoplasm, malignant transformation, lectin histochemistry

Introduction

Primary choroid plexus neoplasms are rare, comprising 0.4–0.6% of all intracranial tumors,^{2,10,14} while choroid plexus carcinomas are even more infrequent.^{3,5,8,10,12,14} Diagnosis of primary choroid plexus carcinoma is difficult, despite certain morphological characteristics visible by conventional light microscopy.^{8,14} Infiltration, implantation, or seeding into the subarachnoid space occurs in both benign and malignant choroid plexus neoplasms,^{1,5,6,8,14} and cannot therefore be considered evidence of malignancy. Choroid plexus carcinoma may be confused with gliomas such as ependymoma, and with metastatic carcinoma.^{1,3,5,8,12,14} Lectin histochemistry using peanut agglutinin may be useful in the differential diagnosis of primary and secondary choroid plexus neoplasms.^{11,15}

Here, we present a patient with choroid plexus carcinoma which progressed from choroid plexus papilloma, and discuss the usefulness of lectin histochemistry in the diagnosis.

Case Report

A 38-year-old male, previously in good health, was

admitted to the Gifu University Hospital in December, 1984 with pulsatile occipitalgia and nausea persisting for several days. A general physical examination revealed no abnormalities, and no apparent neurological deficit was observed. Precontrast computed tomographic (CT) scans showed a mass in the fourth ventricle which was homogeneously enhanced postcontrast (Fig. 1 *left*). The neoplastic lesion was totally removed *via* a suboccipital craniectomy on January 29, 1985, after emplacement of a ventriculoperitoneal shunt (Fig. 1 *right*). Histological examination of the surgical specimen confirmed papillary structures composed of a single layer of columnar epithelium supported by a stroma of vascularized connective tissues (Fig. 2 *left*). The diagnosis was a choroid plexus papilloma.

He remained well until November, 1990 when occipitalgia and nausea developed again, as well as blurred vision. On the second admission, neurological examination revealed mild disorientation and choked disk. Cytological examination of the cerebrospinal fluid obtained from the shunt system discovered no malignant cells. A plain chest x-ray film was unremarkable. CT scans and magnetic resonance (MR) images showed two separate mass lesions with homogeneous enhancement: one in the suprasellar region and the other in the left cerebellopontine angle (Fig. 3). Tumor invasion into the surrounding brain was suspected.

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Fig. 1 Pre- (upper) and postcontrast (lower) CT scans before (left) and after (right) surgery, showing a homogeneously enhanced lesion in the fourth ventricle which was totally removed.

Fig. 2 left: Photomicrograph of the original tumor specimen, revealing papillary growth of single or partly pseudostratified layers of columnar epithelium, indicating a choroid plexus papilloma. HE stain, $\times 112$. right: Photomicrograph of the recurrent tumor specimen, revealing the transition from more normal-appearing choroidal epithelial cells to densely packed and atypical epithelial cells with occasional mitotic figures (arrowhead). HE stain, $\times 170$.

The suprasellar tumor was removed subtotally through a frontotemporal craniotomy on December 11, 1990. Six weeks later, the cerebellopontine angle tumor was partially removed, as the lower cranial nerves were totally enclosed. Histological examination of the specimen revealed papillary architecture of choroid plexus tissue with areas consisting of sheets of densely packed and atypical epithelial cells. The tumor cells varied considerably in size and appearance, with hyperchromatic nuclei occasionally containing mitotic figures (Fig. 2 right).

A lectin histochemistry study was performed using tissue from the original and the recurrent tumors.

Biotin-labeled peanut agglutinin (EY Lab., San Mateo, Cal., U.S.A.) was used in the avidin-biotin peroxidase complex method.¹³⁾ Another two samples of choroid plexus neoplasm (1 malignant and 1 benign) from adults were also studied for comparison. Peanut agglutinin staining was negative or weak in all four samples, but reactivity was intense after treatment with neuraminidase (Sigma, St. Louis, Mo., U.S.A.) (Fig. 4).

The postoperative course was uneventful, but MR images revealed a lumbar spinal lesion (Fig. 5). He received 54 Gy Lineac irradiation to the whole brain

Fig. 3 Axial (left) and coronal (center, right) MR images of the head, demonstrating two separate recurrent lesions: one in the suprasellar region and the other in the cerebellopontine angle.

Fig. 4 Peanut agglutinin histochemical study of the recurrent tumor. *left*: Before neuraminidase treatment, only the vascular endothelia and erythrocytes were stained, but not neoplastic cells. $\times 225$. *right*: After neuraminidase pretreatment, the neoplastic cells also showed intense reactivity. $\times 170$.

Fig. 5 Spinal MR image, demonstrating a lumbar lesion (arrow).

and 30 Gy locally to the lumbar spine. He was discharged 3 months after the last operation with a mild limb ataxia. He is doing well, although follow-up MR images revealed that the residual tumor in the cerebellopontine angle had not reduced in size 8 months after discharge.

Discussion

Malignant transformation of choroid plexus papilloma^{9,14} is very rare. Russell and Rubinstein¹⁴ reported two pediatric cases which recurred with increased cytological malignancy 8 and 11 months

after subtotal removal of the original papilloma. Masuzawa *et al.*⁹ reported an 11-year-old child, in whom a fourth ventricle papilloma became malignant 4 years after subtotal removal of the original papilloma. Although the mechanism of malignant transformation is unclear, choroid plexus papillomas do become malignant. Our case was exceptional in that the original papilloma had been totally removed, and recurrence was very delayed compared with other reported cases.

Differential diagnosis of choroid plexus carcinoma in adult cases requires great care, as it may be confused with metastasis from adenocarcinoma.¹⁴ Electron microscope and immunohistochemical methods are helpful for the diagnosis of choroid plexus neoplasms,^{4,7,9,10,12} and lectin histochemistry is useful for the differential diagnosis of primary and secondary choroid plexus neoplasms. Several authors^{11,15} have studied the binding sites of peanut agglutinin on human brain tumors. Primary choroid plexus neoplasms become positive with peanut agglutinin after neuraminidase pretreatment, in contrast with brain tumors such as gliomas or secondary brain lesions. Ours and two other choroid plexus neoplasms showed markedly increased reactivity to peanut agglutinin after neuraminidase treatment. This result is characteristic of primary choroid plexus neoplasms and is helpful in differential diagnosis.

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