Choroid Plexus Papilloma Originating in the Sella Turcica
—Case Report—

Tetsuro SAMESHIMA, Rokuya TANIKAWA, Toshihide SUGIMURA, Naoto IZUMI, Toshitaka SEKI, Takahiro MAEDA, Toshiyuki TSUPO, Masaaki HASHIMOTO, Teruo KIMURA*, and Kazuki NABESHIMA**

Abashiri Neurosurgical Hospital, Abashiri, Hokkaido; *Dohtoh Neurosurgical Hospital, Kitami, Hokkaido; **Department of Pathology, Fukuoka University Hospital, Fukuoka

Abstract

A 51-year-old female presented with a rare case of choroid plexus papilloma originating in the sella turcica manifesting as headaches that was not readily distinguishable preoperatively from pituitary adenoma. Head magnetic resonance imaging revealed a tumor extending from the sella turcica to the suprasellar cistern. The tumor was removed via an endonasal transsphenoidal approach. Histological examination indicated a papillary structure covered with a layer of columnar epithelial cells that resembled normal choroid plexus. These findings, together with immunohistochemistry, led to a diagnosis of choroid plexus papilloma.

Key words: choroid plexus papilloma, sellar region, pathology, magnetic resonance imaging, differential diagnosis

Introduction

Choroid plexus papilloma is a rare benign tumor that normally arises from the ventricular choroid plexus, and accounts for 1% or less of all primary intracranial tumors. Pediatric cases tend to occur in the lateral ventricles, whereas adult cases tend to occur in the fourth ventricle. More unusual locations include the posterior fossa, cerebellopontine angle, brain stem, third ventricle, sacral canal, and suprasellar region. Only one case has been reported in the sellar region. We describe another case of sellar choroid plexus papilloma.

Case Report

A 51-year-old female complained of headaches. Head magnetic resonance (MR) imaging at another hospital indicated a neoplastic lesion in the sella turcica, so the woman was referred to this hospital. Neurological examination detected no abnormalities and no visual field or vision impairment was found. Endocrinological results also detected no abnormalities.

Head radiography revealed slight enlargement of the sella turcica, and MR imaging revealed a tumor extending from the sella turcica to the suprasellar cistern. T1-weighted MR imaging revealed an isointense lesion with hypointense areas, with heterogeneous enhancement after administration of gadolinium-diethylene-triamine-penta-acetic acid (Gd-DTPA). T2-weighted MR imaging showed the lesion as hyperintense and heterogeneous isointense. Dynamic MR imaging showed the tumor as initially heterogeneously enhanced in the early phase and heterogeneously enhanced in the late phase. No extension into the third ventricle or contact with the normal choroid plexus were noted (Fig. 1). In addition, portions thought to be the pituitary stalk and normal gland were displaced superiorly.

Based on these findings, the preoperative diagnosis was pituitary adenoma and surgery was performed via an endonasal transsphenoidal approach. The tumor was friable and light yellow, somewhat cauliflower-like in shape, and contained small cysts and a fluid component thought to be necrotic tissue and patchy calcification. The tumor was heterogeneous in some areas. Cystic fluid contained old blood components, and cysts were thought to have formed as a result of bleeding in the tumor. Pinkish tissue thought to be normal pituitary gland was noted above the tumor. The tumor was almost completely removed.

Histological examination indicated a papillary structure covered with a layer of columnar epithelial cells. Immunohistochemistry showed that the tumor was positive for epithelial membrane antigen (EMA), cytokeratin AE1/AE3, vimentin, and transthyretin, and negative for glial fibrillary acidic protein (GFAP), S-100 protein, synap-
Fig. 1 Magnetic resonance images showing a tumor within the sella turcica and extending into the suprasellar cistern. A: T₁-weighted image showing the isointense and slightly hyperintense tumor. B: T₂-weighted image showing the hyperintense and heterogeneously isointense tumor. C: T₁-weighted image following gadolinium-diethylenetriaminepenta-acetic acid administration showing the heterogeneously enhanced tumor. D: Dynamic early phase image showing the tumor as initially heterogeneously enhanced.

tophysin, and chromogranin A (Fig. 2). Based on these findings, the histological diagnosis was choroid plexus papilloma.

Discussion

Choroid plexus papillomas tend to occur in the lateral ventricle and fourth ventricle, or more rarely in the third ventricle (70–80%, 14%, and 7%, respectively). In addition, the lateral segment of the choroid plexus in the fourth ventricle is divided into a peduncular part located in the lateral recess and a floccular part protruding from the ventricle into the cerebellopontine angle. Choroid plexus papilloma developing from the floccular part appears as a cerebellopontine angle tumor that is continuous with or close to the foramen of Luschka. Choroid plexus papilloma developing between the cerebellar hemispheres is presumed to have developed from the medial segment of the fourth ventricle or the peduncular part of the lateral segment and then extended in the direction of the cisterna magna.

Choroid plexus papilloma may also develop at a primary intraparenchymal location completely unrelated to the ventricular system, such as the intrinsic brainstem region, cerebellar parenchyma, frontotemporal cortex, and spinal intramedullary region, as well as the suprasellar region on the surface of the brain but not intraparenchymally, and in the sacral canal. These papillomas arise from ectopic development or metaplasia of an ependymal rest. The only previous case of choroid plexus papilloma of the sellar region was associated with partial hemorrhage. The present case also involved a papilloma that was completely separate from the ventricular system and is thought to have developed from ectopic choroid plexus tissue.
Histological examination indicated that the tumor had a morphology mimicking the structure of the choroid plexus, and the tumor cells had a morphology of simple cuboidal epithelium or pseudostratified columnar epithelium. Cells were arranged in papillary fashion along a narrow interstitium with abundant vessels, and a basement membrane was observed beneath the cells.

Choroid plexus papillomas are reported to be positive for EMA, cytokeratin AE1/AE3, vimentin, transthyretin, S-100 protein, and GFAP, but there appear to be many variations. In the present case, S-100 protein and GFAP were negative, and synaptophysin and chromogranin A, which are specific for neuroendocrine tumors, were also negative. These findings are similar to those in the previous case. 13)

Neuroimaging findings for choroid plexus papillomas are nonspecific. Typically, the tumors appear as isointense on T1-weighted MR imaging and heterogeneously hyperintense on T2-weighted MR imaging, with marked enhancement following administration of Gd-DTPA. In the present case, head radiography revealed slight enlargement of the sella turcica. The tumor appeared as isointense with hypo- and hyperintense areas on T1-weighted MR imaging, heterogeneously hyperintense on T2-weighted MR imaging, and with heterogeneous enhancement following administration of Gd-DTPA. However, no extension into the third ventricle or continuity with the normal choroid plexus were noted. Dynamic MR imaging showed the tumor as initially heterogeneously enhanced in the early phase and heterogeneously enhanced in the late phase. These findings resembled those for pituitary adenoma in involving a relatively large tumor. 10) Therefore, the preoperative diagnosis was pituitary adenoma.

Choroid plexus papillomas are rare tumors and do not follow a specific clinical course or result in specific imaging findings. Therefore, the preoperative differential diagnosis is difficult for such papillomas in unusual locations or with extraventricular extension completely separate from the ventricular system.

References


Address reprint requests to: Tetsuro Sameshima, M.D., Abashiri Neurosurgical Hospital, 4–1–7 Katsura-machi, Abashiri, Hokkaido 093–0041, Japan.

e-mail: tetsurosameshima@gmail.com

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