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Choroid Plexus Papilloma Presenting With Cerebrospinal Fluid Rhinorrhea and Otorrhea

—Case Report—

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Abstract

A 52-year-old woman presented with right rhinorrhea and right otorrhea manifesting as aural fullness for 2 years caused by a choroid plexus papilloma in the right cerebellomedullary cistern. Computed tomography and magnetic resonance imaging revealed a well defined lobulated mass at the foramen of Luschka, which extended towards the right cerebellomedullary cistern with slight dilation of the ventricular systems. The tumor was totally resected via a right lateral suboccipital approach. Histological examination revealed a choroid plexus papilloma. Postoperative course was uneventful, just after the operation rhinorrhea ceased completely, and hearing of the right ear dramatically improved. Choroid plexus papillomas rarely cause cerebrospinal fluid (CSF) rhinorrhea. Total removal of the tumor resulted in the cessation of CSF leaks.

Key words: cerebellomedullary cistern, choroid plexus papilloma, cerebrospinal fluid rhinorrhea

Introduction

Cerebrospinal fluid (CSF) rhinorrhea due to trauma and cranial surgery is common. Conversely, non-traumatic CSF rhinorrhea is uncommon. Infection of the paranasal sinuses with osteomyelitis of the adjacent bone, congenital anomaly, and direct invasion of the skull base and nasal cavity by tumors are causes of non-traumatic CSF rhinorrhea.[5,10] CSF rhinorrhea indirectly caused by remote brain tumors is rare.[5] Choroid plexus papilloma (CPP) can increase production of CSF and also can cause an obstruction of CSF flow that leads to hydrocephalus. The resultant chronic increase of intracranial pressure can cause dural and bony defects that lead to CSF rhinorrhea.[5,11] CPPs rarely cause indirect CSF rhinorrhea.[5] CPP comprise 0.3–1% of all intracranial tumors.[9–12]

We report a rare case of non-traumatic CSF rhinorrhea caused by a CPP in the right cerebellomedullary cistern (CMC), and rhinorrhea was stopped after tumor removal. The tumor originated in the fourth ventricle near the fora-

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men of Luschka, which grew extraventricularly into the right CMC.

Case Report

A 52-year-old woman with a 2-year history of right rhinorrhea and right otorrhea with aural fullness was referred to our department from another hospital. Her rhinorrhea had been treated as an allergy for 2 years without brain computed tomography (CT) or magnetic resonance (MR) imaging. Radiological examination at the previous hospital then revealed a lesion near the fourth ventricle that extended towards the right CMC.

Neurological examination on admission found a clear liquid discharge from the right nostril, slight cerebellar signs on her right extremities, and right hearing disturbances. Audiometry showed 42.5 dB of the right ear compared with 13.8 dB of the left ear. Her right otorrhea was not discovered. There were no other neurological deficits.

CT revealed a well demarcated, lobulated isodense mass with patchy calcification in the right CMC with erosion on the clivus and the petrous bone, and an isodense area in the sphenoidal sinus, suggesting liquid accumulation (Fig. 1A). CT with contrast medium showed a heterogeneously enhanced mass (Fig. 1B). Bone window CT showed erosion on the right clivus and the petrous bone (Fig. 1C). T_1-weighted MR imaging showed a well defined, lobulated isointense mass at the foramen of Luschka, which extended towards the right CMC (Fig. 1D). T_2-weighted MR imaging showed a mixed iso-/hyperintense mass and retention of CSF in the sphenoid sinus and right mastoid air cells (Fig. 1E). MR imaging with gadolinium showed a 30 $\times$ 35 mm homogeneously enhanced mass (Fig. 1F). CT and MR imaging showed slight dilation of the ventricular systems.

We performed surgery using a right lateral suboccipital approach. The dura mater was found to be very thin with a trickle of CSF. The brown, soft tumor was found under the right tonsil. The feeding artery around the tumor edge from the posterior inferior cerebellar artery was coagulated and cut. Total removal of the tumor was achieved with care taken to preserve the lower cranial nerves including the hypoglossal nerve, using stimulation and monitoring of each nerve. The postoperative course was uneventful. Just after the operation, rhinorrhea ceased completely, and hearing in the right ear improved dramatically. Audiometry showed 12.5 dB of the right ear compared with 42.5 dB from the preoperative examination. Postoperative CT and MR imaging confirmed total removal of the tumor, disappearance of the fluid accumulation in the right petrous bone, and improvement of the hydrocephalus.

Histological examination revealed a papillary structure of cuboidal epithelium with fibrovascular core and scattered foci of calcification either in the fibrovascular core or in the stroma. There were no malignant signs, suggesting the diagnosis of CPP (Fig. 2).

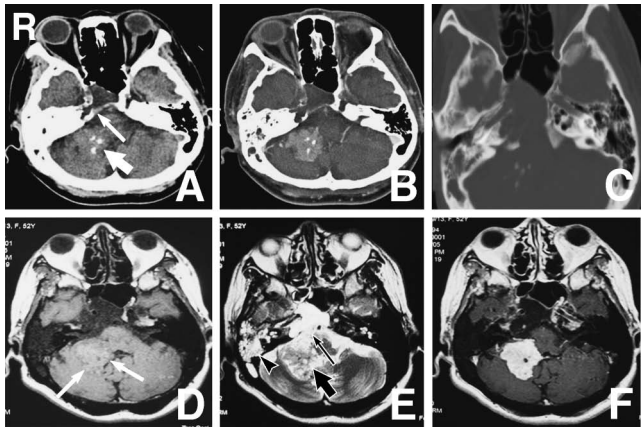

Fig. 1 Neuroimaging findings on admission. A: Computed tomography (CT) scan showing an isodense mass (*thick arrow*) with patchy calcification in the right cerebellomedullary cistern, with erosion on the clivus and the petrous bone (*arrow*). B: CT scan with contrast medium showing a heterogeneously enhanced mass. C: Bone window CT scan showing erosion on the right clivus and the petrous bone. D: T_1-weighted magnetic resonance (MR) image showing a well defined, lobulated isointense mass (*arrows*) in the edge of the fourth ventricle, near the foramen of Luschka, which extended extraventricularly towards the right cerebellomedullary cistern. E: T_2-weighted MR image showing a mixed iso-/hyperintense mass (*thick arrow*) with retention of cerebrospinal fluid in the clivus and petrosal bone area (*arrow*) and in the right mastoid air cells (*arrowhead*). F: T_1-weighted MR image with gadolinium showing a homogeneously enhanced mass.

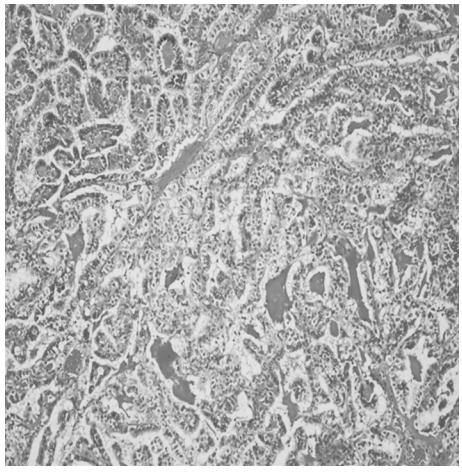

Fig. 2 Photomicrograph showing a characteristic pattern of choroid plexus papilloma with scattered foci of calcification. Hematoxylin and eosin stain, original magnification $\times$100.

Discussion

CPPs are rare benign intracranial tumors that arise from the epithelium of the choroid plexus. The lateral ventricle, fourth ventricle, and third ventricle are common sites for CPP, in that order.[1] CPP in the cerebellopontine angle (CPA) is quite rare, accounting for only 9% of all CPPs and almost exclusively occurs in adults.[7,12] CPP of the CPA could arise from direct extension of a fourth ventricle

Table 1 Summary of choroid plexus papilloma (CPP) cases causing cerebrospinal fluid (CSF) rhinorrhea

Author (Year)	Age (yrs)/ Sex	CPP location	Pathway of CSF leakage	Treatment	Outcome of CSF rhinorrhea
Vigouroux (1908)*	27/M	fourth ventricle	ethmoid sinus	unknown	unknown
Rovit et al. (1969)[8]	48/M	third ventricle	not mentioned	total removal	stopped
Lamberts (1984)[4]	34/M	fourth ventricle	ethmoid sinus	total removal and fistula repair	stopped
Symss et al. (2009)[11]	61/M	fourth ventricle and major cistern	cribriform plate	fistula repair and total removal	stopped
Present case	52/F	fourth ventricle and cerebellomedullary cistern	petrous bone to eustachian tube	total removal	stopped

*Cited from ref. 4.

tumor through the foramen of Luschka or as a primary CPA tumor.[1,12] In our case, the CPP arose from the choroid plexus near the foramen of Luschka, which commonly grow extraventricularly. This extraventricular extension could be explained as a direct extension from each recess at the foramen of Luschka.[2,10]

The incidence of CPP resulting in CSF rhinorrhea is very low. Our review found four additional cases of CPP resulting in CSF rhinorrhea (Table 1). Most CPPs were located in the fourth ventricle, with only one case located in the third ventricle. CSF rhinorrhea caused by neoplastic lesions can be differentiated into two groups according to the mechanism of fistula formation.[2,6] In the direct type, tumors caused erosion of the meninges and bone. In the indirect type, tumor expansion caused severe chronic obstructive hydrocephalus. Raised intracranial pressure may cause progressive erosion of the most anatomically fragile area at the base of the skull[2,6,8] or a lesion of the arachnoid sheaths accompanying the olfactory nerves, at the level of the cribriform plate.[2]

In our case, preoperative CT and MR imaging revealed mild hydrocephalus, and also erosion of the clivus bone and fluid accumulation in the right mastoid air cells with erosion of the right petrosal bone. At the beginning of the tumor growth, CPP caused long-standing mild intracranial hypertension as could be seen from the erosion of meninges and bone. Actually, the dura mater was found to be very thin with a trickle of CSF during the operation. Later the development of CSF leak in the form of otorrhea and rhinorrhea might have reduced the intracranial pressure. These factors could explain why our patient showed CSF rhinorrhea and otorrhea as the initial symptoms, with a lack of more serious symptoms due to intracranial hypertension. The CSF leaks might have reduced intracranial pressure and minimized the symptoms.

Many pathways of CSF rhinorrhea are possible, such as via the frontal sinus, the cribriform plate of the ethmoid bone, the sphenoid sinus, the sella, or the petrous bone to the middle ear and then the eustachian tube.[2] The CSF rhinorrhea pathway in our case seemed to be via the petrous bone. Long-standing hydrocephalus might cause indirect erosion of the posterior aspect of the petrosal bone and the mastoid, forming a fistula into the middle ear and then via the eustachian tube to the nasal cavity. This pathway was supported by the following evidence: Fluid accumulation in the mastoid cells on the CT and MR imaging; symptoms included right aural fullness and otorrhea; and hearing deterioration of the right ear due to air and bone conduction, but not sensorineural, since there was no attenuation of the acoustic nerve by the tumor. Air and bone conduction may have resulted from CSF leaks to the mastoid cells and middle ear.

Regarding CSF rhinorrhea treatment, surgical removal of the tumor usually results in immediate cessation of CSF leaks. However, if CSF rhinorrhea does not stop after tumor removal, secondary dural repair would be mandatory to close the fistula. The fistula poses a danger of meningitis, which has been reported in 40% of patients with nontraumatic CSF rhinorrhea.[3] Our patient ceased to show CSF rhinorrhea just after total removal of the tumor. Hence, secondary dural repair was unnecessary.

CPPs rarely cause CSF rhinorrhea. Overproduction of CSF and attenuation of the CSF pathway by the tumor led to intracranial hypertension. Long-standing intracranial hypertension caused progressive erosion of the meninges and bone and resulted in the development of a fistula connecting the skull base and the nasal cavity. Total removal of the tumor usually results in the elimination of CSF leaks.

References

1) Aksoy FG, Gomori JM: Choroid plexus papilloma of foramen of Luschka with multiple recurrences and cystic features. *Neuroradiology* 41: 654–656, 1999
2) Benvenuti D, Lavano A, Corriero G, Irace C, de Divitiis E: Cerebrospinal rhinorrhea in a patient with a posterior cranial fossa tumor. *Neurosurgery* 21: 742–744, 1987
3) Hubbard JL, McDonald TJ, Pearson BW, Laws ER Jr: Spontaneous cerebrospinal fluid rhinorrhea: evolving concepts in diagnosis and surgical management based on the Mayo Clinic experience from 1970 through 1981. *Neurosurgery* 16: 314–321, 1985
4) Lamberts AE: Choroid plexus papilloma with cerebrospinal fluid rhinorrhea. *Surg Neurol* 22: 576–578, 1984
5) Liang CL, Lu K, Lee TC, Su TM, Chen HJ: Non-traumatic cerebrospinal fluid rhinorrhea indirectly caused by remote brain tumor: a case report and review of the literature. *Clin Neurol Neurosurg* 103: 83–86, 2001
6) Ommaya AK, Di Chiro G, Baldwin M, Pennybacker JB: Nontraumatic cerebrospinal fluid rhinorrhoea. *J Neurol Neurosurg Psychiatry* 3: 214–225, 1968
7) Piguet V, de Tribolet N: Choroid plexus papilloma of the

cerebellopontine angle presenting as a subarachnoid hemorrhage: case report. *Neurosurgery* 15: 114–116, 1984
8) Rovit RL, Schechter MM, Nelson K: Spontaneous ''high-pressure cerebrospinal rhinorrhea'' due to lesions obstructing flow of cerebrospinal fluid. *J Neurosurg* 30: 406–412, 1969
9) Sarkar C, Sharma MC, Gaikwad S, Sharma C, Singh VP: Choroid plexus papilloma: a clinicopathological study of 23 cases. *Surg Neurol* 52: 37–39, 1999
10) Shin JH, Lee HK, Jeong AK, Park SH, Choi CG, Suh DC: Choroid plexus papilloma in the posterior cranial fossa: MR, CT, and angiographic findings. *Clin Imaging* 25: 154–162, 2001
11) Symss NP, Prasad AN, Vasudevan MC, Ramamurthi R: Exophytic choroid plexus papilloma of the fourth ventricle presenting with cerebrospinal fluid rhinorrhea: a case report. *Surg Neurol* 71: 705–708, 2009
12) Talacchi A, De Micheli E, Lombardo C, Turazzi S, Bricolo A: Choroid plexus papilloma of the cerebellopontine angle: a twelve patient series. *Surg Neurol* 51: 621–629, 1999

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