Treating Cerebrospinal Fluid Rhinorrhea without Dura Repair: A Case Report of Posterior Fossa Choroid Plexus Papilloma and Review of the Literature

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Key words
- Case report
- Cerebrospinal fluid
- Choroid plexus papilloma
- Dura mater
- Hydrocephalus
- Skull base

Abbreviations and Acronyms
CPP: Choroid plexus papilloma
CSF: Cerebrospinal fluid
CT: Computed tomography
EVD: External ventricular drainage

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BACKGROUND: **Choroid plexus papilloma revealed by nontraumatic cerebrospinal fluid (CSF) rhinorrhea has only been described 5 times, to our knowledge, in the literature. The challenges in this situation are to recognize CSF leak, to rapidly understand the pathophysiology of the leak, and to choose the best treatment strategy in emergency. We report an original case of posterior fossa choroid plexus papilloma revealed by CSF leak. We then discuss the surgical strategy and the pathophysiology of CSF leak, which is explained, in this case, by both hyperproduction of CSF and local skull base erosion.**

CASE DESCRIPTION: **We report the case of a 47-year-old man who has developed spontaneous rhinorrhea, right hearing loss, and confusion. A choroid plexus papilloma of the right cerebellomedullary cistern was diagnosed. Hydrocephalus and pneumocephalus were associated with an erosion of the homolateral skull base. The patient underwent surgical total tumor removal by a median suboccipital approach after implantation of a temporary external ventricular drainage. The patient recovered completely without any recurrence of CSF rhinorrhea. The 5-month postoperative images show total bone re-growth and resolution of hydrocephalus.**

CONCLUSIONS: **Our case shows that 1) causal reasoning is of major importance when dealing with CSF rhinorrhea and that 2) dura repair can be avoided when treating CSF leak secondary to posterior fossa choroid plexus papilloma.**

INTRODUCTION

Nontraumatic cerebrospinal fluid (CSF) rhinorrhea[1] represents only 3%–4% of rhinorrheas. They have different classifications, mostly according to CSF pressure.[2] The causes of high-pressure CSF leaks are hydrocephalus, tumors, and idiopathic intracranial hypertension.[3-7] This latter entity demonstrates that elevation of the intracranial pressure can, by itself, cause the leak. In the case of tumor, the mechanism of CSF leak could be direct, by erosion of meninges and bone, or indirect, by pressure erosion of anatomically fragile areas of the skull base.[2,8,9] The treatment of a CSF leak is an emergency because of the risk of meningitis. The 2 challenges in this situation are 1) to rapidly identify the cause of the CSF leak and 2) to treat the leak to avoid or to help antibiotic treatment of meningitis.

We report the case of a patient treated for a spontaneous CSF rhinorrhea who presented with a posterior fossa choroid plexus papilloma (CPP). Only 5 cases of CSF rhinorrhea secondary to a CPP have been previously reported, to our knowledge.[9-12] Beyond the rarity of this clinical entity, the main interest of this case is to demonstrate that treatment of the CSF leak can be achieved by treating the cause, i.e., removal of the CPP, without dura repair.

CASE REPORT

Written informed consent was obtained from the patient for publication of this case report and the accompanying images.

History

A 47-year-old man developed spontaneous rhinorrhea, right hearing loss, and sensation of fluid movement during head mobilizations. He was first treated with intranasal antibiotics and corticosteroids. His symptoms worsened for a week, with headaches, confusion, severe impairment of condition, fever, and balance disorders. He then presented to our institution. Once the rhinorrhea identified as a CSF leak, a computed tomography (CT) scan was performed (Figure 1), which showed a right anterolateral tumor of the posterior fossa, ventricular distention, ventricular and basal cisterns pneumocephalus, lysis of the right basal-occipital diploe, and erosion of the posterior wall of the clivus and dorsum sellae. These structures, the mastoid cells, and right tympanic cavity were filled with liquid. Magnetic resonance imaging (Figure 2) revealed a tumor of the right lateral recess of the fourth ventricle opening into the inferior part of the pontocerebellar angle and cerebellomedullary cistern. This lesion was hypointense on T1-weighted images, heterogeneously hyperintense on T2-weighted images, and homogeneously enhanced by gadolinium infusion.

Operation

External ventricular drainage (EVD) was implanted to treat hydrocephalus and CSF leak. A CSF examination showed

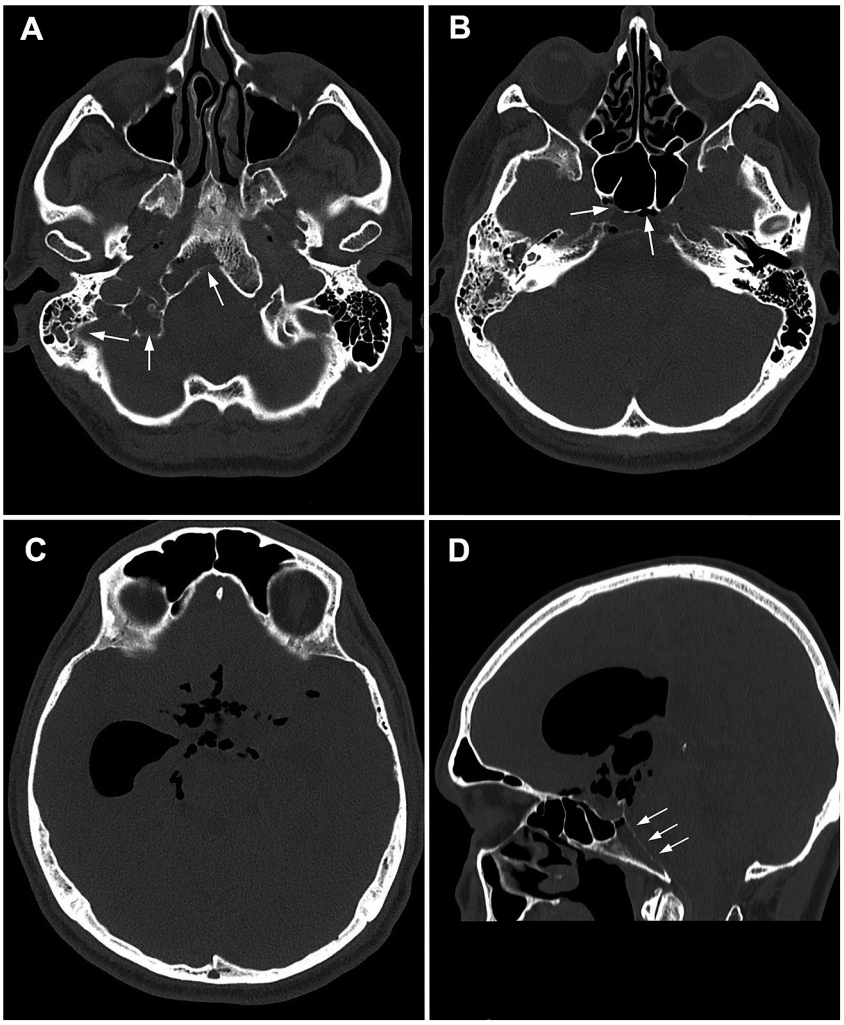

Figure 1. Cerebral computed tomography scan, bone windows. (**A**) Axial slice showing the lysis of the basal-occipital bone and the mastoid (*arrows*). (**B**) Axial slice showing the lysis of the wall of the sphenoid sinus (*arrows*). (**C**) Axial slice showing hydrocephalus and ventricular and cisternal pneumocephalus. (**D**) Sagittal slice showing the osseous lysis of the clivus (*arrows*).

biochemical meningitis without any germ, either on direct examination or on cultures. A broad-spectrum empirical double antibiotherapy was initiated. After 1 week of antibiotherapy, the patient underwent surgical tumor resection by a median suboccipital approach while in the prone position. A right occipital craniotomy with laminectomy of the posterior arch of C1 allowed immediate exposure of the tumor between the cerebellar tonsil and posterior-inferior cerebellar artery laterally and superiorly and medulla oblongata medially (Figure 3). After complete tumor resection, the vertebral artery, proximal segment of the posterior-inferior cerebellar artery, and lower cranial nerves were identifiable (Figure 4).

Postoperative Course

The EVD was removed 1 week after the procedure. CSF rhinorrhea did not recur. Hearing loss and confusion improved. Early postoperative CT scan showed a decrease in size of cerebral ventricles and drainage of the mastoid cells and middle ear. Histopathologic examination pointed to a CPP without any signs of malignancy. Postoperative CT and magnetic resonance imaging at 5 months showed a massive bone regrowth (Figure 5).

DISCUSSION

We report the case of a patient with spontaneous CSF leak treated by etiological treatment—surgical removal of a CPP of the cerebellomedullary cistern—without dura repair. Beyond the difficulty of diagnosis, the originalities of this case are 1) to discuss the pathophysiology of CSF leak associated with CPP and 2) to show that, in this case, surgery should focus on removing the CPP rather than on trying to treat at any cost both the primary cause and the dural fistula.

We found 5 cases of CSF rhinorrhea secondary to CPP reported in the literature (Table 1). Vigouroux,[13] Lamberts[10] and Symss et al.[11] described, respectively, in 1908, 1984, and 2009, fourth-ventricle CPPs with CSF leakage through the ethmoid and cribriform plate. Rovit et al.[9] described in 1969 a third ventricle CPP but did not show the location of the bone erosion. Finally, Kinoshita et al.[12] recently described a posterior fossa CPP with CSF leakage through the eustachian tube, by erosion of the petrous bone. Hence, the present case and the one of Kinoshita et al.[12] are the only ones showing a possible direct mechanism by bone erosion.

It is known that CPP leads to chronic hyperproduction of CSF, usually diagnosed by hydrocephalus.[14,15] In our case, as the fourth ventricle was not obstructed, we concluded that the cause of hydrocephalus was hyperproduction of CSF. Indeed, pneumocephaly of the lateral ventricles indirectly proves a persistent communication between basal cisterna, the ventricular system, and air cavities of the skull base (Figure 6).

Thus, we propose that the mechanism of CSF leakage was due to both a direct and an indirect cause, proven respectively by 1) the presence of CSF only inside the right side of the skull base, in direct contact with the tumor and by 2) the presence of a nonobstructive hydrocephalus. To explain the direct mechanism, we hypothesize that a direct compression of the cistern could have modified local CSF flow, causing bone erosion.

Considering surgical management, after having discussed the mechanism of CSF leak, although we can assume that removal of the tumor without EVD would probably have led to the same result, we decided to first implant an EVD to both

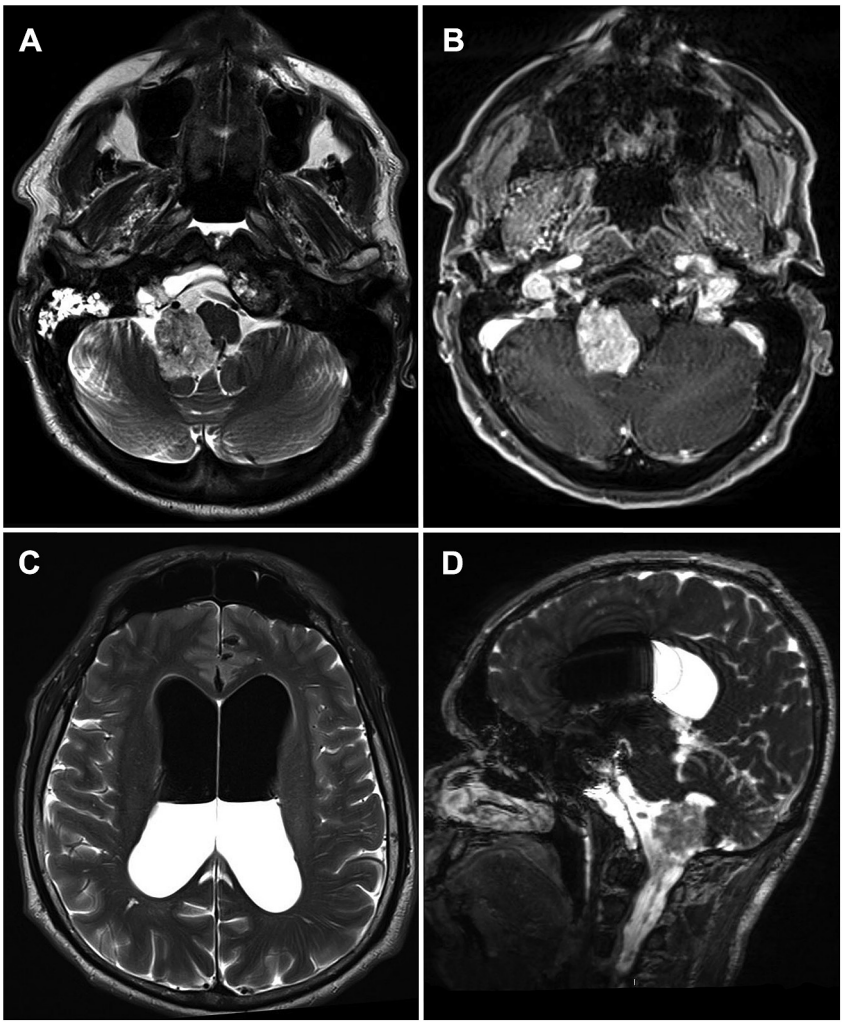

Figure 2. Cerebral magnetic resonance imaging showing a posterior fossa mass lesion. (**A**) T2-weighted axial slice showing the heterogeneous hyperintense mass into the right lateral recess of the fourth ventricle. Note the liquid filling of the right mastoid cells and clivus. (**B**) T1-weighted axial slice after gadolinium infusion showing a homogeneous enhancement of the tumor. (**C**) T2-weighted axial slice passing through the lateral ventricles showing the ventricular distension without transependymar resorption. (**D**) T2-weighted parasagittal slice showing the hydrocephalus, the ventricular pneumocephalus, and aqueous signal inside the clivus.

decrease CSF pressure and analyze the CSF before probabilistic anti-biotherapy. Once the tumor was removed, and therefore after CSF pressure had been normalized, CSF leak did not recur, allowing the removal of the EVD. It is here important to note that, during surgery, we did not intend to repair the dura mater, considering that the remaining arachnoid layer and blood covering the inferior part of the cerebellopontine angle (as seen on Figure 4), together with the decrease in CSF pressure would allow the treatment of the breach. Moreover, the absence of natural communication between the middle ear and the diploe of the basi-occipital bone allowed us to hypothesize that there were at least 2 zones of dura mater erosion, as evidenced by the fluid contamination of both the mastoid and the basal-occipital on the midline. Hence, a dura repair would have required an intraoperative exploration of these regions, which could have damaged the brainstem or cranial nerves.

Our case is the third, together with those of Rovit et al.[9] and Kinoshita et al.,[12] to show that CSF leak can be treated without dura repair. In addition, in the cases of Lamberts[10] and Symss et al.,[11] in which the CSF leakage originated from the cribriform plate, the anterior fossa dura mater had been reconstructed. It is therefore not possible to conclude about the role of dura repair in CSF leakage treatment in cases of CPP associated with cribriform plate fistula. Cribriform plate may indeed be, in these particular cases, an area of greater risk of persistence of fistula than in posterior fossa.

CSF leak due to CPP enters into the balance of several mechanisms. CSF hypersecretion causes communicating hydrocephalus, which can favor a local osteodural erosion close to the tumor, both mechanisms potentially leading to CSF leak. Our case illustrates the importance of causal reasoning to deal with nontraumatic CSF rhinorrhea. When involving the posterior fossa, surgical removal of a CPP allows excellent clinical and radiologic outcome considering both CSF leak and hydrocephalus.

REFERENCES

1. Domengie F, Cottier JP, Lescanne E, Aesch B, Vinikoff-Sonier C, Gallas S, et al. Management of cerebrospinal fluid fistulae: physiopathology, imaging and treatment. *J Neuroradiol*. 2004;31:47-59 [in French].

2. Ommaya AK, Di Chiro G, Baldwin M, Pennybacker JB. Non-traumatic cerebrospinal fluid rhinorrhoea. *J Neurol Neurosurg Psychiatry*. 1968;31:214-225.

3. Clark D, Bullock P, Hui T, Firth J. Benign intracranial hypertension: a cause of CSF rhinorrhoea. *J Neurol Neurosurg Psychiatry*. 1994;57:847-849.

4. Ransom ER, Komotar RJ, Mocco J, Connolly ES, Mullins KJ. Shunt failure in idiopathic intracranial hypertension presenting with spontaneous cerebrospinal fluid leak. *J Clin Neurosci*. 2006;13:598-602.

5. Schlosser RJ, Wilensky EM, Grady MS, Palmer JN, Kennedy DW, Bolger WE. Cerebrospinal fluid pressure monitoring after repair of cerebrospinal fluid leaks. *Otolaryngol Head Neck Surg*. 2004;130:443-448.

6. Suryadevara AC, Fattal M, Woods CI. Non-traumatic cerebrospinal fluid rhinorrhea as a result of pseudotumor cerebri. *Am J Otolaryngol*. 2007;28:242-246.

7. Yang Z, Wang B, Wang C, Liu P. Primary spontaneous cerebrospinal fluid rhinorrhea: a symptom of idiopathic intracranial hypertension? Clinical article. *J Neurosurg*. 2011;115:165-170.

8. Benvenuti D, Lavano A, Corriero G, Irace C, de Divitiis E. Cerebrospinal rhinorrhea in a patient

with a posterior cranial fossa tumor. *Neurosurgery*. 1987;21:742-744.

9. Rovit RL, Schechter MM, Nelson K. Spontaneous "high-pressure cerebrospinal rhinorrhea" due to lesions obstructing flow of cerebrospinal fluid. *J Neurosurg*. 1969;30:406-412.

10. Lamberts AE. Choroid plexus papilloma with cerebrospinal fluid rhinorrhea. *Surg Neurol*. 1984;22:576-578.

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*. 2009;71:705-708.

12. Kinoshita Y, Wasita B, Akatsuka K, Kambe A, Kurosaki M, Watanabe T. Choroid plexus papilloma presenting with cerebrospinal fluid rhinorrhea and otorrhea: case report. *Neurol Med Chir (Tokyo)*. 2010;50:930-933.

13. Vigouroux A. Ecoulement de liquide céphalorachidien. Hydrocéphalie. Papillome des plexus choroïdes du IVe ventricule [Cerebrospinal fluid leakage. Hydrocephalus. Choroid plexus papilloma of the 4th ventricle]. *Rev Neurol (Paris)*. 1908;7:281-285.

14. Ito H, Nakahara Y, Kawashima M, Masuoka J, Abe T, Matsushima T. Typical symptoms of normal-pressure hydrocephalus caused by choroid plexus papilloma in the cerebellopontine angle. *World Neurosurg*. 2017;98:875.e13-875.e17.

15. Karimy JK, Duran D, Hu JK, Gavankar C, Gaillard JR, Bayri Y, et al. Cerebrospinal fluid hypersecretion in pediatric hydrocephalus. *Neurosurg Focus*. 2016;41:E10.

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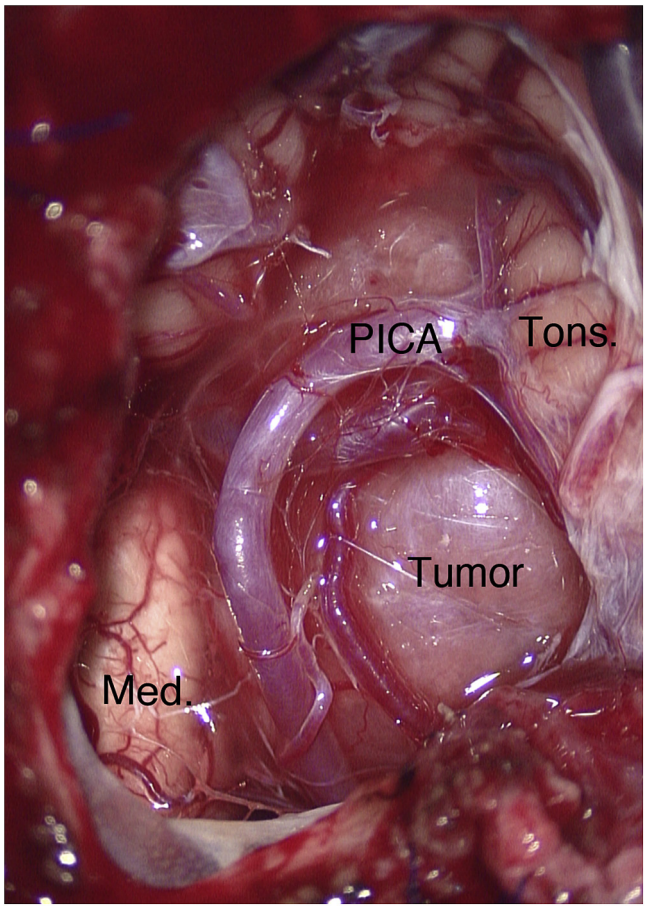

Figure 3. Intraoperative photograph. Posterior view of the tumor surrounded by the loop of the posterior-inferior cerebellar artery (PICA), between the medulla oblongata (Med.) medially, and the right tonsil (Tons.) laterally.

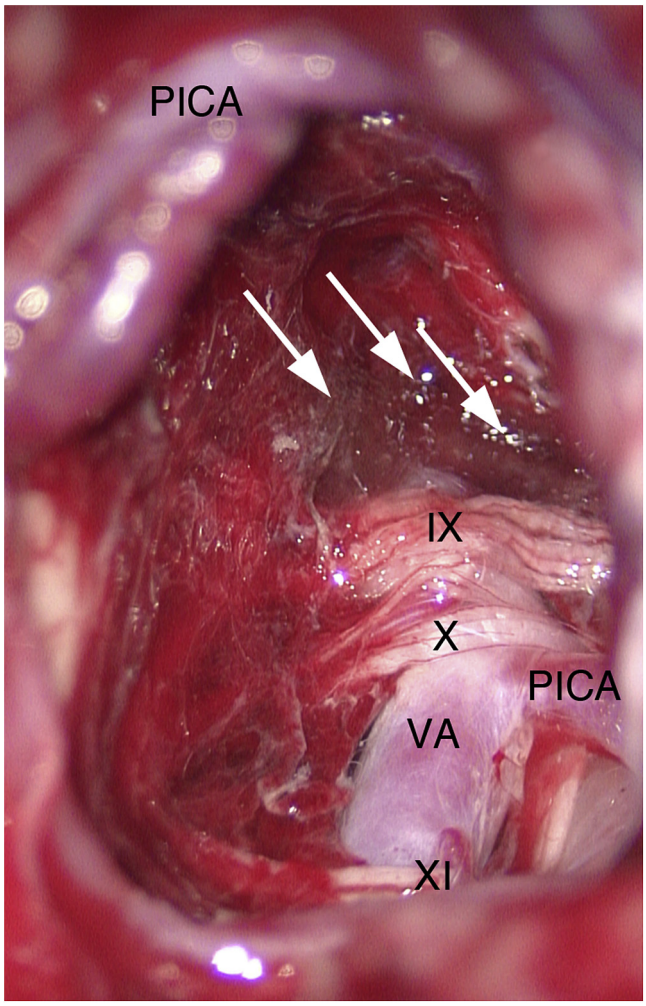

Figure 4. Intraoperative photograph. Posterior view showing the operative field after total tumor removal, the lower cranial nerves (IX, X and XI), the right vertebral artery (VA), and the posterior-inferior cerebellar artery (PICA). The tissue, including arachnoid and blood (*white arrows*), in contact with the basal-occipital bone, deliberately is not dissected to avoid the recurrence of cerebrospinal fluid rhinorrhea.

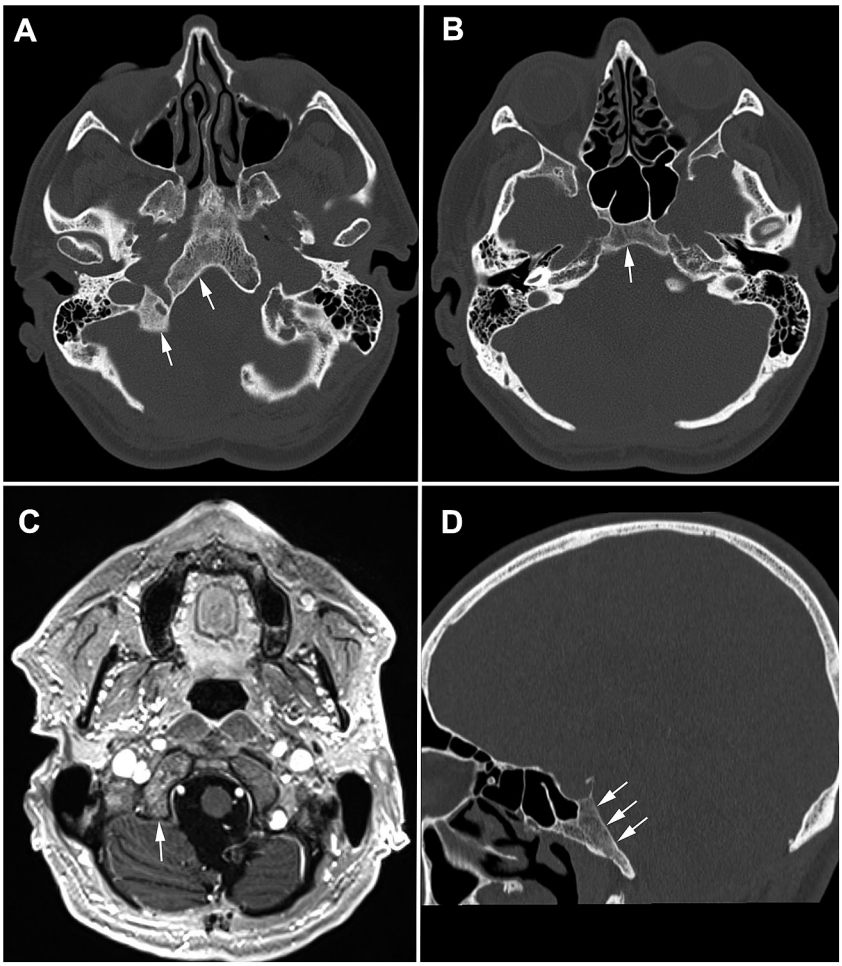

Figure 5. Five-month postoperative cerebral images. (**A** and **B**) Computed tomography (CT) axial slices in bone windows showing complete bone regrowth of the right basal occipital bone and clivus (*white arrows*). (**C**) T1-gadolinium magnetic resonance imaging axial slice showing enhancement of the diploe of the right occipital condyle, evoking bone recolonization (*white arrow*). (**D**) CT sagittal slice, in bone windows showing complete bone regrowth of the clivus (*white arrows*).

Table 1. Summary of a Literature Review of Choroid Plexus Papilloma Cases Causing Cerebrospinal Fluid Rhinorrhea

Study	Age, years/Sex	Tumor Location	Pathway of CSF Leakage	Treatment	Outcome of CSF Rhinorrhea
Vigouroux, 1908[13]	27/M	Fourth ventricle	Ethmoid sinus	NA (postmortem diagnosis)	
Rovit et al., 1969[9]	48/M	Third ventricle	Unknown	Total removal	Stopped
Lamberts, 1984[10]	34/M	Fourth ventricle	Ethmoid sinus	Fistula repair and total removal	Stopped
Symss et al., 2009[11]	61/M	Fourth ventricle and cisterna magna	Cribriform plate	Fistula repair and total removal	Stopped
Kinoshita et al., 2010[12]	52/F	Fourth ventricle and cerebellomedullary cistern	Petrous bone to Eustachian tube	Total removal	Stopped
Present case	47/M	Fourth ventricle and cerebellomedullary cistern	Petrous bone to Eustachian tube	Total removal	Stopped

CSF, cerebrospinal fluid; M, male; NA, not available; F, female.

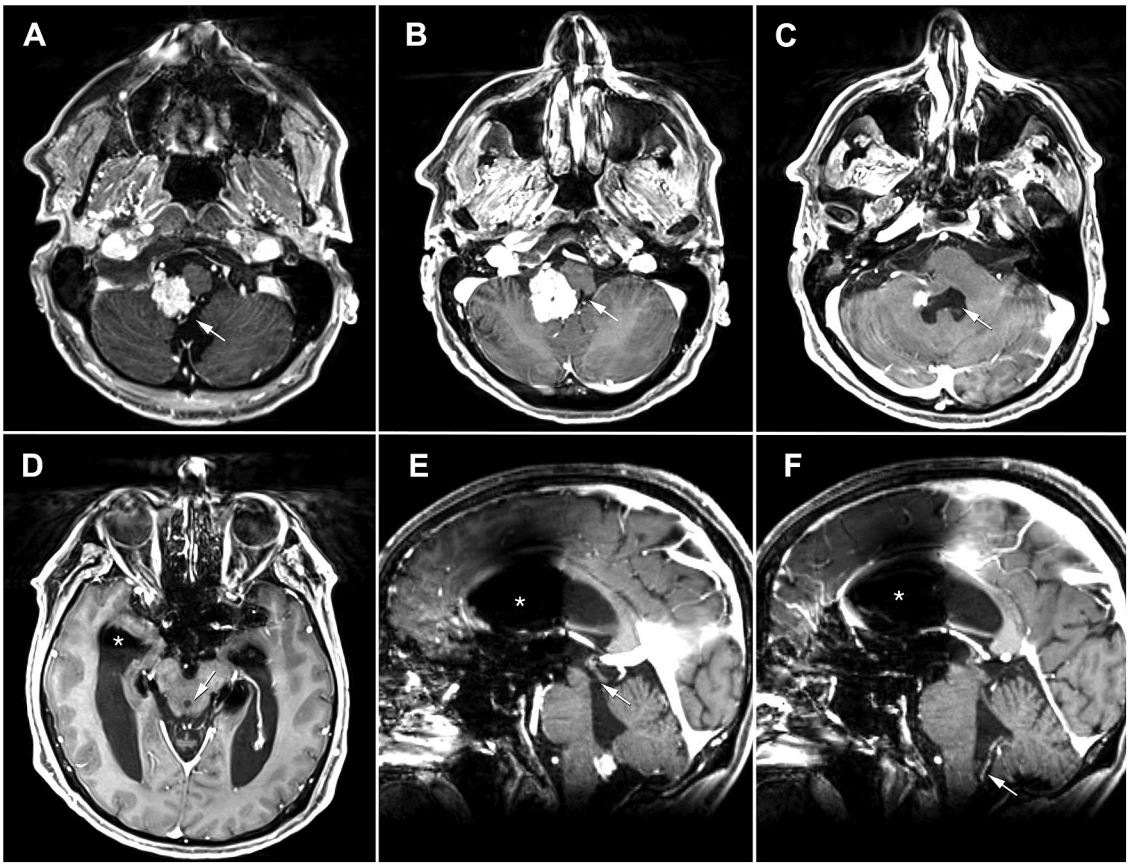

Figure 6. Cerebral magnetic resonance imaging demonstrating the absence of obstruction of the fourth ventricle. (**A**–**C**) T1-weighted axial slices after gadolinium infusion, showing the enhanced tumor compressing without obstructing the cisterna magna and the fourth ventricle (*white arrows*). (**D**–**E**) T1-weighted axial and sagittal slices after gadolinium infusion, showing the distended mesencephalic aqueduct (*white arrows*). (**F**) T1-weighted parasagittal slice after gadolinium infusion showing the nonobstructed communication between the fourth ventricle and cisterna magna (*white arrow*). *Asterisks* show pneumocephaly inside the lateral ventricles.