

DYSPHONIA AS AN UNCOMMON PRESENTATION OF PONTOCEREBELLAR CHOROID PLEXUS PAPILOMA

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SUMMARY – A case is presented of a patient with dysphonia, hearing loss and ataxia due to vestibulocochlear and vagal nerve compression by choroid plexus papilloma in the cerebellopontine angle. Choroid plexus papillomas are rare tumors usually arising in the lateral and fourth ventricle, and rarely found in the cerebellopontine angle, making the neuroimaging characteristics usually not sufficient for diagnosis. Patients usually present with headache and hydrocephalus but tumors in the cerebellopontine angle can cause vestibulocochlear dysfunction and cerebellar symptoms. Dysphonia along with hearing loss was a dominant symptom in the case presented. After complete surgical removal of the tumor, deterioration of dysphonia was noticed; it could be explained as peripheral vagal nerve neuropathy due to tumor compression and intraoperative manipulation. In this case report, we describe dysphonia as an uncommon presentation of a rare posterior fossa tumor. To our knowledge, a case of choroid plexus papilloma presenting with dysphonia has not been described before. Our case extends the differential diagnosis of dysphonia from the otorhinolaryngological to the neurosurgical field.

Key words: *Papilloma, choroid plexus – surgery; Dysphonia; Cerebellopontine angle; Case reports*

Introduction

Choroid plexus papillomas (CPPs) are rare neuroepithelial tumors with the incidence ranging between 0.4% and 0.6% of all intracranial tumors¹⁻³. CPPs are commonly found in the lateral ventricles, followed by the fourth ventricle. CPPs in the third ventricle and cerebellopontine angle (CPA) are rare^{4,5}, and usually present with headache and hydrocephalus.

We describe a rare case of a patient presenting with dysphonia as an unusual symptom, hearing loss and ataxia due to vestibulocochlear and vagal nerve compression by CPP in the CPA.

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Received August 6, 2014, accepted January 13, 2015

Case Report

A 54-year-old Caucasian man, former factory worker, complained of a several month history of intermittent episodes of dysphonia, hearing loss in the right ear, unsteady gait and headache. There were no swallowing difficulties.

Laryngoscopy showed no pathology in vocal cord movement due to intermittent dysphonia. Audiometry confirmed severe sensorineural hearing loss in the right ear with moderate sensorineural hearing loss on the left side, which could be attributed to professional noise exposure. Examination revealed an infranuclear palsy of the right facial nerve (House-Brackmann grade II) and cochlear nerve (Weber lateralized to the left), mild palsy of the right glossopharyngeal, vagal and hypoglossal nerves, and sensory ataxia.

Magnetic resonance imaging (MRI) of the brain showed a homogeneous mass in the right CPA, ex-

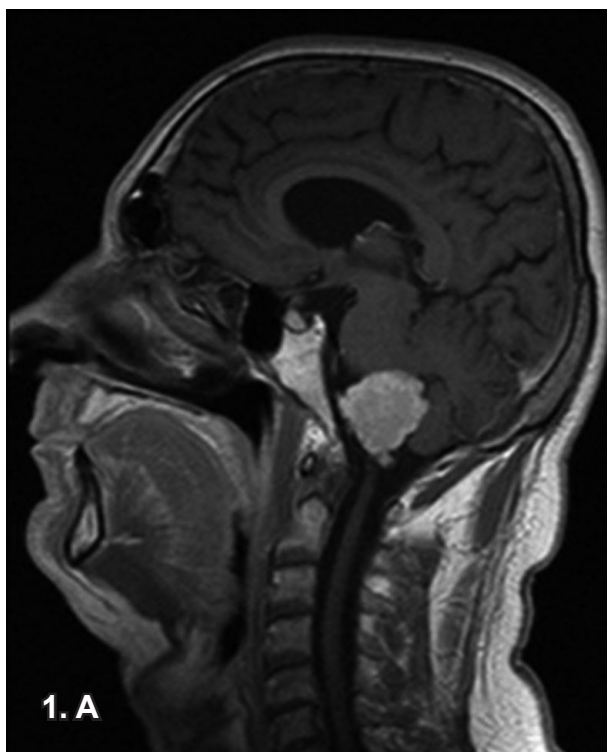
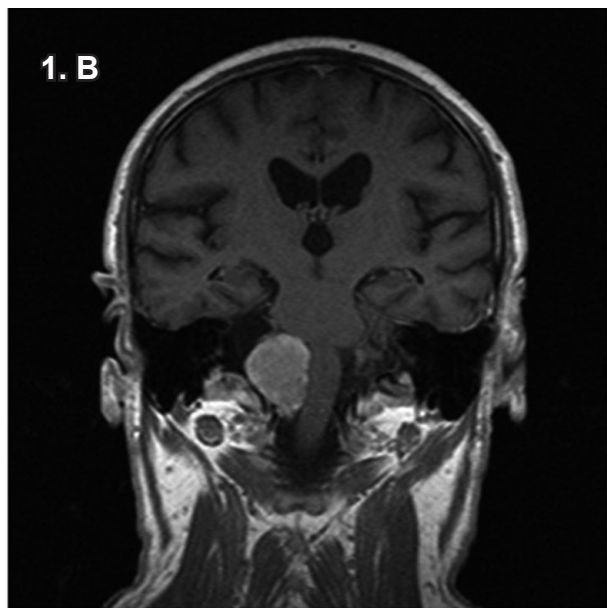


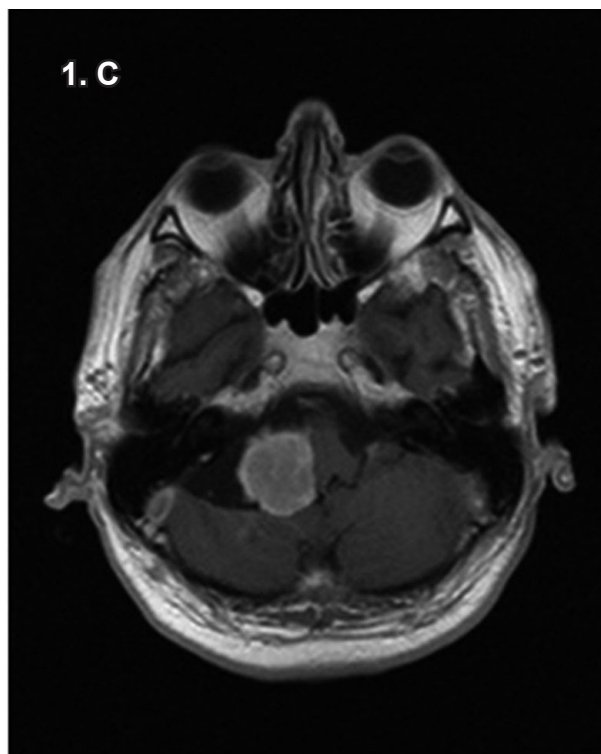
Fig. 1. Preoperative sagittal (A), coronal (B) and axial (C) contrast-enhanced T1-weighted MRIs showing a well-circumscribed and homogeneously enhancing mass extending from the right cerebellopontine angle to the cerebellomedullary cistern with compression of the brain stem and the fourth ventricle.



tending caudally to the foramen magnum level. A lesion measuring 30x26x34 mm was hypointense on T1- and hyperintense on T2-weighted images, with a central zone of marked hypointensity, which may have represented calcification. Brain stem and fourth ventricle were compressed by the lesion. Gadolinium T1-weighted images showed an intense contrast enhancement (Fig. 1 A, B, C). There were no signs of hydrocephalus.

Surgical procedure was performed in general anesthesia with intraoperative neurophysiologic monitoring. The patient was placed in a semi-sitting position and right-sided suboccipital craniectomy was performed. Upon dural opening, cerebrospinal fluid (CSF) was released from the cerebellomedullary cistern in order to relax the right cerebellar hemisphere. After retraction of the right cerebellar hemisphere, the VII/VIII complex and antero-inferior cerebellar artery (AICA) were visualized. A tumorous lesion was visualized at the level of the right-sided foramen of Luschka. The tumor had macroscopic characteristics of choroid plexus and was extending laterally and caudally.

A clear arachnoid border separated it from the adjacent cerebellum and brain stem. More adherent to



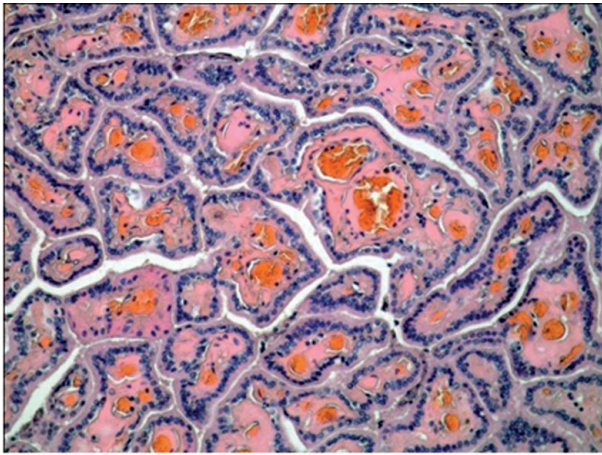


Fig. 2. Microscopic view: papillary architecture with hyalinized stroma lined by a single layer of columnar cells with isomorphic nuclei and a moderate amount of cytoplasm. (H&E, X200)

the tumor capsule were the glossopharyngeal, vagal and hypoglossal nerves. Total microsurgical excision of the tumor was performed with preservation of the lower cranial nerves. The wound was closed in a standardized fashion.

Motor evoked potentials (MEP) were used for monitoring the right facial, glossopharyngeal and hypoglossal nerves, and for bilateral monitoring of the

cochlear nerves. Final-to-baseline facial MEP ratio showed increased response in all monitored muscles innervated from the right cranial nerves. MEP of the vagal nerve was not strictly monitored.

Histopathologic examination showed papillary forms lined by a single layer of columnar cells (Fig. 2). Mitoses were very rare and there was no sign of necrosis. Immunohistochemical analysis revealed a strong positive reaction to cytokeratin (CK), S-100 protein and vimentin. Ki-67 immunoreaction was less than 5%. A diagnosis of CPP was made.

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

Outcome

On postoperative examination, deterioration of dysphonia was noticed. Postoperative electromyography of the cricothyroid muscles showed reduced interference patterns with the increase of polyphasic potentials and increase in the length of the recruited motor unit potentials. This finding could be interpreted as a sign of reinnervation. Follow up MRI of the brain three months after the surgery revealed no signs

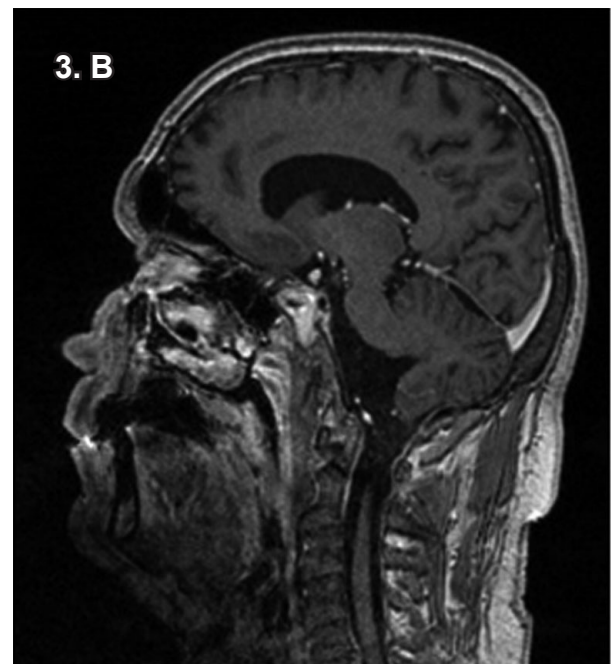
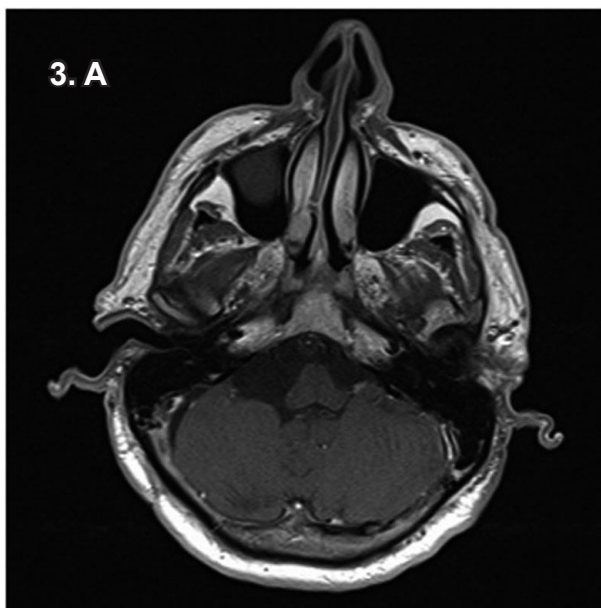


Fig. 3. Axial (A) and sagittal (B) contrast-enhanced T1-weighted MRIs three months after surgery showing no signs of residual or recurrent tumor.

of residual or recurrent tumor (Fig. 3 A, B). Clinical examination showed immobile right vocal cord, with good compensation of the contralateral fold, which resulted in normal voice. According to the electromyography finding, further improvement of vocal cord paresis could be expected. Follow up audiometry showed sensorineural deafness on the right side and unchanged threshold curve on the left side. There was no sign of facial nerve damage on follow up examination (House-Brackmann I).

Discussion

Choroid plexus papillomas originate from the neuroepithelial cells of the choroid plexus. In children, the most common location is lateral ventricle. In adults, CPP arises most frequently in the fourth ventricle^{6,7}. Extraventricular location of CPP has been reported, e.g., in the CPA, pineal region, sacral canal, suprasellar and sellar region. Rovit *et al.* report on 234 cases of CPP, 17 of them found in the CPA⁸. Kimura *et al.* report on CPP in the suprasellar region⁹, and Bian *et al.* in the pituitary fossa¹⁰. When all age groups are considered, CPPs arising in the CPA account for 7%-20% of these tumors¹¹.

The appearance of CPP in the CPA results from herniation of the tumor through the foramen of Luschka, as in the case presented, or from *de novo* development in the choroid plexus lying out of the fourth ventricle at the CPA, referred to as Bochdalek's flower basket¹¹.

Headache is the most common symptom in both children and adults. These tumors may present with signs of raised intracranial pressure due to the hydrocephalus. Hydrocephalus can be due to CSF overproduction by the tumor or due to obstruction of the CSF pathway, especially in tumors compressing the fourth ventricle^{12,13}. Tumors located in the CPA can cause vestibulocochlear dysfunction and cerebellar symptoms such as ataxia due to cerebellar, brain stem and CPA cranial nerve compression.

The symptoms described in our case, including facial nerve palsy and sensory ataxia, were mild and neglected by the patient, as well as progressive hearing loss since the patient had already had occupational noise-induced hearing loss.

Dysphonia represented an unexpected symptom. Dysphonia in this case could be explained as a pe-

ripheral vagal nerve neuropathy due to tumor compression. Postoperative worsening of dysphonia could have resulted from intraoperative manipulation. Similar episodes of dysphonia are described in patients with implanted vagal nerve stimulator as a treatment modality in patients with intractable epilepsy¹⁴. Nevertheless, in his landmark study of neuropathology of spasmodic dysphonia, Schaefer states that although peripheral vagal neuropathy could be involved, the normal findings of recurrent laryngeal nerve in dysphonic patients suggest that spasmodic dysphonia is a central nervous system disease. Stabilization of clinical findings in the study by Schaefer occurred in the 3- to 5-year interval¹⁵.

Prior to surgery, a diagnosis of CPA meningioma or metastatic tumor was suspected.

The imaging characteristics are usually not sufficient to diagnose a CPP at extraventricular locations and CPPs are often diagnosed preoperatively as meningiomas of the cerebellopontine region. The diagnosis is confirmed histopathologically in all cases of CPPs outside ventricular location.

Surgical extirpation of CPP is the treatment of choice. Long term follow up for tumor recurrence is recommended.

Conclusion

We presented a patient with a CPP atypically localized in the CPA, presenting with unusual episodes of dysphonia that worsened after complete surgical removal of CPP and gradually diminished during the 6-month postoperative period. To our knowledge, no similar case has been reported in the literature to date, thus widening the differential diagnosis of dysphonia from the otorhinolaryngological to the neurosurgical field.

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Sažetak

DISFONIJA KAO RIJETKA MANIFESTACIJA PAPILOMA PONTOCEREBELARNOG KOROIDNOG PLEKSUSA

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Prikazuje se slučaj bolesnika s disfonijom, gubitkom sluha i ataksijom uslijed pritiska na osmi i deseti moždani živac papilomom koroidnog pleksusa u cerebellopontinom kutu. Papilomi koroidnog pleksusa su rijetki tumori koji se obično javljaju u postraničnoj i četvrtoj moždanoj komori te zbog rijetke pojavnosti u cerebellopontinom kutu neuroradiološke karakteristike nisu dostatne za postavljanje dijagnoze. Bolesnici se obično klinički prezentiraju glavoboljom i hidrocefalusom, no tumori u cerebellopontinom kutu mogu dovesti do poremećaja funkcije osmog moždanog živca i cerebelarnih simptoma. U prikazanom slučaju kao dominantni simptom javila se disfonija uz gubitak sluha. Nakon potpunog kirurškog odstranjenja tumora primijećeno je pogoršanje disfonije, što se moglo objasniti perifernom neuropatijom desetog moždanog živca uslijed pritiska tumora i intraoperacijske manipulacije. U ovom prikazu slučaja opisuje se disfonija kao neuobičajena prezentacija rijetkog tumora stražnje lubanjske jame. Prema našim spoznajama dosad nije opisan slučaj papiloma koroidnog pleksusa koji se prezentira disfonijom. Ovaj slučaj proširuje diferencijalnu dijagnozu disfonije iz otorinolaringološkog polja na neurokirurško polje.

Ključne riječi: Papilom, koroidni pleksus – kirurgija; Disfonija; Cerebellopontini kut; Prikazi slučaja