CASE REPORT

Pseudobulbar Palsy Caused by a Large Petroclival Meningioma: Report of Two Cases

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ABSTRACT

Two patients sought treatment for symptoms of bulbar motor dysfunction and the marked emotional lability that characterizes pseudobulbar palsy (PBP). Magnetic resonance (MR) imaging showed large petroclival masses with severe compression and displacement of the brainstem. A suboccipital craniectomy with a transpetrosal, transtentorial approach to the tumor was performed in both patients. One patient required a second-stage surgery to resect an extension of the tumor into the cavernous sinus. In both patients pathological evaluation confirmed the diagnosis of syncytial meningioma. After surgery, PBP resolved in both patients. Large posterior fossa tumors should be included in the differential diagnosis of patients with a clinical picture of bulbar motor dysfunction coupled with emotional lability.

KEYWORDS: Meningioma, pathological laughter, pseudobulbar palsy, tumor

In 1837 Magnus reported the first case of pseudobulbar palsy (PBP) in a patient who had suffered multiple infarcts.¹ The term pseudobulbar palsy, however, was not coined until 1877 when Lepine used it to differentiate lower cranial nerve palsies caused by supranuclear lesions from cranial nerve palsies caused by lesions of the bulbar nuclei.² PBP is characterized by dysarthria, dysphagia, facial and tongue weakness, and emotional lability. It is caused by bilateral dysfunction of the

corticobulbar tracts, which exert supranuclear control over the brainstem motor nuclei that, in turn, control mastication, speech, and deglutition.³ An important difference from primary disease of the bulbar motor nuclei is the spontaneous, involuntary laughing or crying associated with PBP. In addition, the jaw jerk reflex typically becomes exaggerated in PBP. Unlike lower motor neuron bulbar paralysis, there is no atrophy or fasciculations of the affected muscles.

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PBP is commonly described in conditions causing bilateral corticobulbar lesions such as amyotrophic lateral sclerosis, lacunar infarcts, or demyelinating conditions such as multiple sclerosis.⁴ One of the abnormal expressions of emotion sometimes described in patients with PBP is pathological laughter, which is characterized by involuntary, uncontrollable, inappropriate laughter. Pathological laughter alone has been reported in association with multiple tumors of the posterior fossa, particularly those involving the pons and midbrain. Both extra-axial tumors⁵⁻¹² and intra-axial tumors¹³⁻¹⁶ of the posterior fossa have been associated with this condition. Unlike pathological laughter, PBP in association with posterior fossa neoplasms is extremely rare. We present two patients with reversible PBP associated with large petroclival meningiomas.

ILLUSTRATIVE CASES

Case 1

A 37-year-old woman had several months' history of spontaneous laughing spells and episodes of uncontrollable crying. These emotional outbursts were embarrassing and distressing to her. She also reported progressive dysphagia as well as difficulties with balance and gait. On neurological examination, she exhibited mild dysmetria in the left upper and lower extremities. Her cranial nerve examination was unremarkable and a gag reflex was present. Her extremity reflexes were brisk bilaterally and plantar responses were extensor. She also had a moderately ataxic gait. Magnetic resonance imaging (MRI) revealed a large, left-sided mass in the petroclival region that extended into the cavernous sinus (Fig. 1). The brainstem was severely compressed and displaced.

Because the tumor extended into the cavernous sinus from the petroclival region, the patient underwent two staged operations. First, she underwent a suboccipital craniectomy. Through a

transpetrosal, transtentorial approach about 60% of the tumor was resected, leaving the extension in the cavernous sinus and petrous apex. Three weeks later, she underwent a left frontotemporal craniotomy to resect the residual tumor (Fig. 1). Postoperatively, the patient developed delayed onset cerebrospinal fluid rhinorrhea, which was treated by repacking the temporal bone with a muscle and fat graft. Histological examination identified a benign syncytial meningioma.

The patient was in excellent condition after surgery. Her preoperative subjective swallowing difficulties, emotional outbursts, and cerebellar ataxia resolved completely. She exhibited mild hypesthesia in the left V1 and V2 distribution and left-sided hearing loss. Imaging studies revealed no evidence of tumor until 5 years after surgery. At that time MRI showed a small recurrence in the petroclival region. She has no new complaints and is scheduled to undergo Gamma Knife™ (Elekta, Stockholm, Sweden) treatment.

Case 2

A 57-year-old woman sought treatment for a 10-month history of progressive dysarthria, dysphagia, and frequent episodes of emotional lability. Her emotional lability was characterized by frequent outbursts of inappropriate laughing and crying spells. She reported right-sided weakness, gait difficulties, and the recent onset of urinary incontinence. On admission, she exhibited a sensory deficit in the distribution of the left 5th cranial nerve and subtle left-sided weakness. Her gag reflex was decreased and her speech was dysarthric. Mild right hemiparesis and right hemisensory deficit were noted and her plantar responses were extensor bilaterally. MRI of the brain revealed a 4- × 3- × 2.5-cm mass in the left petroclival region with marked mass effect on the pons and midbrain. Mild hydrocephalus was also present. Audiometry revealed mild left sensorineural hearing loss at 30 decibels.

Figure 1 Upper: Preoperative T1-weighted, gadolinium-enhanced MRIs of Case 1, showing a large, left-sided petroclival meningioma extending into the cavernous sinus and compressing the brainstem severely. Lower: Immediate post-operative, gadolinium-enhanced MRIs showing a small residual tumor. The brainstem is completely decompressed.

A retrolabyrinthine transpetrosal, transtentorial approach was performed. Intraoperatively, a large vascular tumor with an indistinct interface with the brainstem was encountered. A thin rim of tumor was left adherent to the midbrain and lateral petrous wall. Histological examination identified a benign syncytial meningioma.

Postoperatively, the patient made a good recovery. Her emotional lability resolved immediately and her dysarthria, dysphagia, and gait difficulty improved gradually. One year after surgery, she exhibited mild residual left-sided V_2 and V_3 sensory loss as well as mild right hemisensory loss. Follow-up imaging has revealed no evidence of progression of the residual tumor.

DISCUSSION

Multiple posterior fossa neoplasms have been reported in association with disordered emotional expression, typically in the form of pathological laughter. Bhatjwale et al.⁵ described four patients with massive trigeminal neuromas whose principal symptom was pathological laughter. Pathological laughter also has been associated with posterior fossa meningiomas,^{6,7,10,11} epidermoid tumors,¹³ trochlear nerve neurinomas,² and clival chordomas.^{8,12} Intrinsic brain tumors associated with pathological laughter include pontine glioblastomas multiforme,⁴ hypothalamic glioblastomas multiforme,¹⁵ and metastatic melanomas of the pons.¹⁶ Like PBP, pathological

laughter is most often associated with degenerative, vascular, and demyelinating processes.⁴ Poeck¹⁷ reviewed the published pathological anatomy in three cases of pathological laughter and found that the supranuclear motor pathways are involved bilaterally, always in the brainstem between the thalamus and medulla. In PBP, which more aptly describes the clinical presentation of our two cases, the bilateral lesions can involve not only the pontine and/or mesencephalic levels but also the thalamus at the capsular and cortical levels.

It is extremely rare for tumors of the posterior fossa to manifest with PBP. Tsuzaka et al.¹⁸ reported one patient with a primary lymphoma of the central nervous system who presented with PBP. This patient had a lesion on the right cerebellum as well as multiple lesions in the left frontal lobe and basal ganglia. PBP also has been reported in children who underwent posterior fossa surgery for large midline vermian or fourth ventricular tumors.¹⁹ In these cases, PBP was attributed to retraction pressure that caused edema to track along fiber pathways. Likewise, Hattori et al.²⁰ reported a patient with PBP who had an arteriovenous malformation involving the thalamus and midbrain. This patient, however, also had Moyamoya disease with areas of diffuse cerebral infarcts and atrophy.

There are at least three possible causes for the PBP in our two patients with posterior fossa tumors, namely compression, ischemia, and demyelination. The relatively large size of the neoplasms in our two patients may have caused compressive dysfunction of the pontomesencephalic areas that contain the corticobulbar tracts bilaterally. Compression and ischemia are likely interrelated in that compressive mechanical injury leads to compromise of blood flow through small vessels. The portions of the brainstem made ischemic by compression may remain in an ischemic penumbra and may have significant potential for recovery after decompression. The acute resolution of symptoms in our patients after surgery supports such a theory. Furthermore, the absence of abnormality on pre- and postoperative MRI argues against infarction or demyelination as responsible for the

PBP in these two patients. We suggest that large compressive posterior fossa tumors should be included in the differential diagnosis of patients with a clinical picture of bulbar motor dysfunction coupled with emotional lability.

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Commentary

Certainly pathological laughter has previously been reported in association with different types of tumors in the posterior fossa. However, pseudobulbar palsy (PBP) associated with posterior fossa tumors as the initial manifestation is very rare. PBP tends to be associated with neurodegenerative and demyelinating disorders, which are mostly irreversible pathological processes.

If diagnosed properly, PBP associated with resectable tumors of the posterior fossa is reversible. This potential emphasizes the importance of imaging studies in patients with PBP. However, new posterior fossa lesions in patients already diag-

nosed with neurodegenerative or demyelinating disorders cannot be excluded. In such cases, the appearance of new PBP symptoms warrants reimaging. This caveat is especially important for neurologists, who usually follow these patients.

Typically, petroclival meningiomas manifest with symptoms related to the trigeminal, vestibulocochlear, and glossopharyngeal nerves and with gait ataxia and dementia due to hydrocephalus. PBP and hemiparesis are late symptoms, associated with large or giant tumors. If a patient has no cranial nerve symptoms, PBP may be the initial symptom, especially in patients in rural areas or otherwise stoic individuals.

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Commentary

Well-articulated and referenced, this case report presents a rare and seldom recognized system complex associated with petroclival meningiomas. The authors are to be commended for their excellent presentation.

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