Petroclival Meningioma Presenting with Pathological Laughter: Report of a Case and Review of the Literature

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Abstract- Unprovoked pathological laughter has been associated with various lesions arising from the hypothalamus, cerebral hemispheres, and brainstem. We report a 57-year-old man who presented with episodes of involuntary laughter with increasing frequency over the course of more than 4 years. A large left petroclival meningioma was found. His involuntary laughter has completely subsided after surgical removal and in the follow-up of 3 years.

Key Words: Pathological laughter, Petroclival meningioma, *Fou rire prodromique*

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INTRODUCTION

Pathological laughter is an uncontrollable laughing which is dissociated from stimulus and mood. It has been associated with bilateral or diffuse cerebral lesions[1], hypothalamic harmatomas[2], multiple sclerosis[3], cerebrovascular diseases[4,5,6], trigeminal neuroma[7,8], petroclival meningioma[9], etc. We report a case of left petroclival meningioma with this unusual symptom.

CASE REPORT

A 57-year-old right-handed male taxi driver presented with episodes of involuntary laughter. The laughter occurred with increasing frequency over 4 years and was mixed with sounds similar to inappropriate crying. He would laugh from time to time despite he was in a bad temper. He was noted to be somewhat clumsier in driving especially when making u-turns and started to have choking one year before. He gradually developed unsteady gait, urine incontinence, and left limbs weakness within the next 6 months. On examination, mentality was normal. However, there were frequent spells of unprovoked laughter status during the examination. The palmomental sign was present on right. Eye movements were full and free. The gag reflex was absent. The rapid alternate movements (RAM) were clumsier on left. He tended to fall toward his left when walking straight line and was poor walking tandem. The Romberg test was positive. Mild weakness was present in the left arm and leg. However, Babinski sign was absent bilaterally.

ACTH was low (1.04 pg/ml; reference: 9-52) but GH (0.37ng/ml; reference: < 10), prolactin, and TSH were within normal. His awake EEG revealed no abnormal findings. Computed tomography (CT) scan of brain

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showed enhanced thickened left anterior tentorium. Magnetic resournance imaging (MRI) demonstrated an extra-axial petroclival mass measuring 3.2×3.2×4.1 cm, iso-intense on T1 and T2 weighted images with homogenous enhancement, and located in left prepontine and cerebellopontine angle cisterns (Figs. A-B). The upper brainstem was distorted (Figs. C-D).

He received surgical removal of the tumor. Pathological examination of the tumor revealed as meningioma. The post-operative recovery was uneventful with complete resolution of the abnormal laughter. Follow-up CT 3 years later revealed no recurrence of the tumor. At present, he is still free of the laughter.

DISCUSSION

Normal laughing is an expression of mirth and consists of complex expression of facial movements. The precise neuroanatomy of normal laughing is still unknown, however. Pathological laughter is termed when the laughter is inappropriate, continuous, and is not associated with expected mood changes[4,5]. It may be part of a generalized affective or cognitive disorder[3]. Poeck speculated that a tonic and a phasic innervation are involved in the movements of facial expression[1]. In spasmodic pathological laughter, there is a loss of phasic control as (1) a release phenomenon following destructive lesion such as pseudobulbar palsy and in epileptic phenomenon (e.g. gelastic seizure from temporal lobe lesions[10]); (2) a manifestation of psychiatric disorders such as schizophrenia and hysteria[5,6,11]; and (3) an exposure to mood-altering chemicals such as alcohol or hallucinogens[9]. Jacob and Chand reported a case of pathological laughter related to infusion of sodium valproate[12]. On the other hand, *fou rire prodromique*, a rare syndrome with pathological laughter after an apoplectic event, is a loss of tonic control of laughing and is frequently accompanied with pathological crying[5].

Parvizi and colleagues have proposed an alternative hypothesis by considering the coordination and modulation roles of the cerebellum[13]. They have argued that

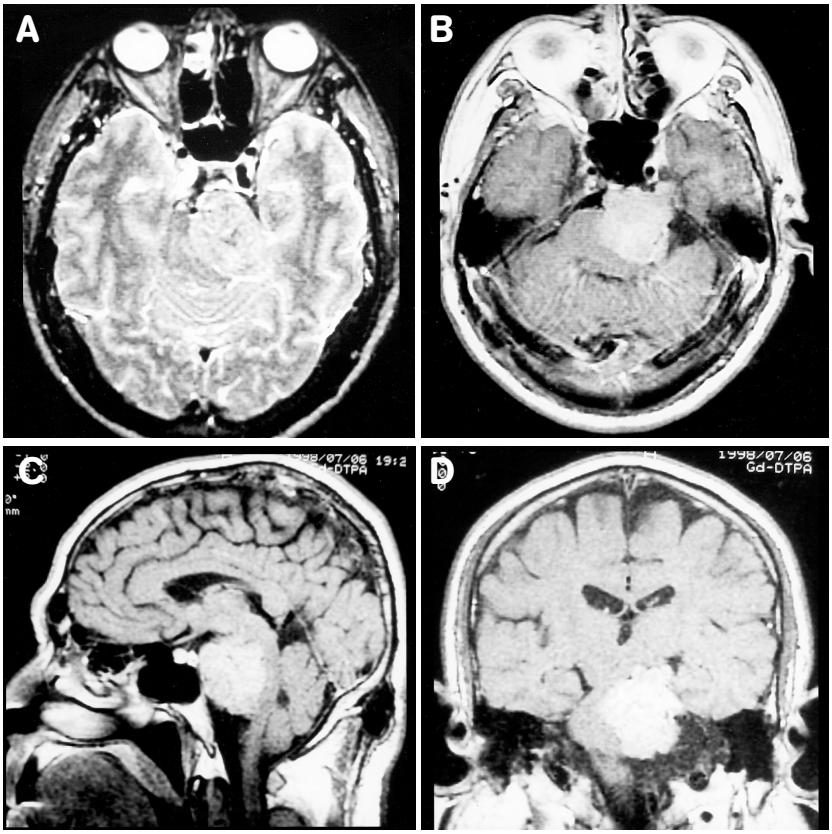

Figure. Axial T2-weighted SE (A) and axial (B), sagittal (C), coronal (D), and post-gadolinium T1-weighted SE showed a homogenous isointense and high-enhanced well-circumscribed mass lesion with extension of left Meckel's cave in widened left prepontine cistern.

projections from different cortical regions, including prefrontal and cingulate cortices, are relayed to the cerebellum by the nuclei in basis pontis. The pathological laughter and crying may be caused by lesions in the medial basis pontis resulting in deafferentation and poor communication between cerebellum, prefrontal, and cingulate inputs. Besides, cerebellum also receives projections from subcortical structures related to the processing of emotions such as hypothalamic and brainstem serotonergic raphe nuclei. Therefore, deafferentation of the cerebellum from cortical and subcortical inputs related to cognitive/affective processing, or a specific lesion in cerebellum itself, will alter the inter-communication for generating emotional responses. Anderson et al. have proposed that the serotonergic neurotransmission is impaired in patients with pathological laughter by lesions of the raphe nuclei in brainstem or their ascending projections[14]. This argument is supported by several clinical trials treating pathological laughter with selective serotonin reuptake inhibitors (SSRIs), like Sertraline. SSRIs are also effective in treating patients with pathological crying[15-18]. Furthermore, there are reports that pathological laughter and pathological crying can occur together[5,13,19,20]. Poeck has pointed out that laughter can abruptly change to crying[1]. Therefore, pathological laughter and crying may result from the same anatomical lesion, for example, raphe nuclei, and/or dysfunction of the serotonergic neural circuits. Further neuroanatomical, neurophysiological and neuroimaging studies may clarify this issue.

Pathological laughter has been reported in cases with diffuse cerebral lesions[1] or multiple sclerosis[3]. Focal lesions, that cause pathological laughter are usually located in the posterior fossa, including (1) upper brainstem tumors (intrinsic or extrinsic), i.e., pontine glioma[21,22], clival chordoma[10], trigeminal neuroma[6,7], tentorial meningioma[23], and petroclival meningioma[9]; (2) vascular events such as unilateral cerebral infarct[17], brainstem infarct[4]; and (3) cerebellar lesion[13]. Cairns first reported in 1950 night terrors in a patient with a brainstem tumor[21]. Since then there are several other reports of posterior fossa tumor presented with pathological laughter[7-9,21,23-24]. Shafqat and colleagues reported a case caused by a petroclival meningioma similar to our case[9].

A total of 15 cases have been reported (including our case)[7-9,23,24]. Among them, thirteen are male and two female. Among fourteen pathology proved tumors: 11 are meningiomas, 2 chordomas, and 1 epidermoid cyst. The ages ranged from 20 to 50 years and thirteen were below age 40 years.

The importance of afferent inputs into the thalamus in emotional expression has been emphasized by Alusi et al[6]. We would argue that brainstem lesions might disinhibit the control from the cortical level, therefore, leading to the expression of laughter without the influences from higher cortical centers. The petroclival meningiomas in our case as well as the case of Shafqat et al.[9] showed a distorted brainstem on MR imaging, and might cause pathological laughter by (1) disrupting the connection between the lowest and the intermediate levels leading to a disinhibition from the higher center in controlling laughting from both the cortical and the limbic influences[9]; (2) deafferentation of coordination or modulation of the cerebellum from the prefrontal or subcortical inputs[13]; and (3) interference with serotonergic transmission originating from pontine raphe nuclei[13,14].

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