

Pathological laughter in a patient with a brain-stem epidermoid cyst

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Brain-stem epidermoid cysts (ECs) are extremely rare, with only 20 cases reported in the literature.¹ The neurological symptoms depend on the location of the ECs. They may cause headaches, palsy and unsteady gait. Pathological laughter (PL) is an unusual manifestation of neurosurgical disease and had been described in rare conditions of brain-stem compression from the surrounding tumors.² However, there are limited reports of brain-stem ECs with PL to date. Herein, we report a brain-stem EC patient presenting with PL.

A 39-year-old woman presented with uncontrollable laughter, paralysis of both lower limbs, and dizziness for 4 months. The laughter was unprovoked and not associated with mood elevation. At times, severe outbursts of PL could be provoked by conversations about private matters (for example, about her medical history). Occasionally, aconuresis was noted during laughing spells. Physical and neurological examinations were also unremarkable, except for weak limbs on both sides. Electroencephalography revealed no epileptiform discharge. An MRI revealed a large mass lesion located in the pons, which markedly indented the pons. This mass had similar signal intensity to CSF, with no enhancement on T1-weighted gadolinium-enhanced images (Figure 1a), and high intensity on the T2-weighted images (Figure 1b). Therefore, a preoperative diagnosis of brain-stem EC was made. The lesion was removed via a transpetrosal approach. After opening the dura, retracting the temporal lobe and incising the tentorium posterior to the trochlear nerve, the cyst was found to contain keratinized debris, which indirectly verified the preoperative diagnosis. To avoid risking her neurological status, the tumor was subtotally removed due to some bits of the tumor that adhered densely to the brain-stem (Figure 1c). The histological diagnosis was EC (Figure 1d). The postoperative period was uneventful, and the laughing spells disappeared immediately after surgery. There was no recurrence of the lesion or of the spontaneous laughter after 6 months.

Most of the cases reported in the literature are presumed to have arisen in the prepontine or lateral cisterns and insinuated in the brain-stem.³ In our case, the EC occupied the prepontine cistern with a significant portion of the tumor insinuating itself into the superior pons. The clinical presentation of these patients was due

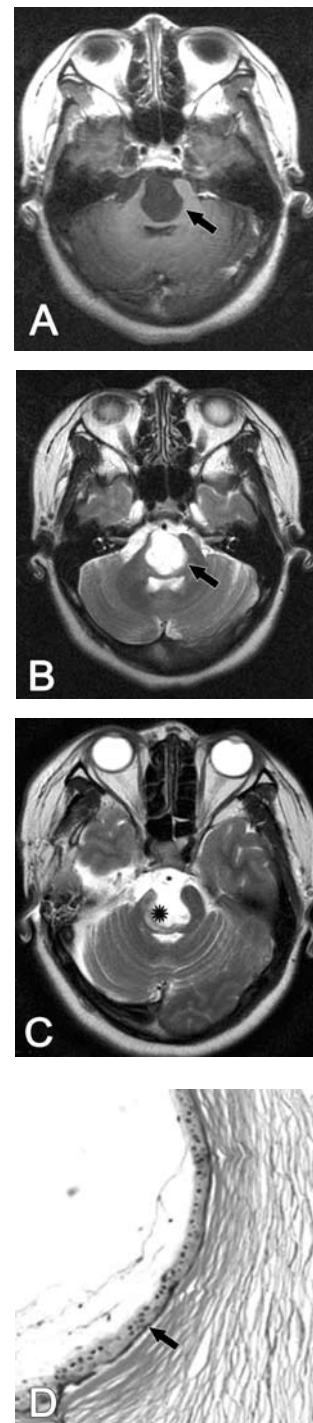


Figure 1 - Presenting MRI showing A) axial T1-weighted image demonstrating the enhancement of a mass with severe compression of the pons (arrow). B) Axial T2-weighted image showing a hyperintense mass with no surrounding edema (arrow). C) Postoperative axial T2-weighted image showing a hyperintense residual mass (asterisk). D) Histopathology slide revealing the stratified squamous epithelial lining (arrow) and laminated keratinaceous debris (hematoxylin and eosin stain, original magnification x40).

to the location of the EC. Cranial nerve palsy, headache, unsteady gait, and meningitis may be involved. Among these patients, no patient with PL has been reported.

Pathological laughter is a condition involving continuous, inappropriate, and unmotivated, relatively uncontrollable episodes of laughter without any emotional change in the patient's personality. The precise mechanism and neuroanatomy of PL remain uncertain. Parvizi et al⁴ postulated a disruption of the corticopontocerebellar pathways from higher association areas to the cerebellum, resulting in the cerebellum failing to coordinate and adjust the laughter response to the appropriate cognitive and situational context. Some researchers have speculated the existence of a brain-stem facio respiratory center, and PL was thought to ensue when this higher center were incapable of exerting their inhibitory influence on the facio respiratory center. To identify the location of facio respiratory center in the brain-stem, Elyas et al⁵ compared the images of 16 cases of PL secondary to infarction and multiple sclerosis, and found that the anterior portion of the pons might represent a zone where supranuclear inputs were modulated by both the cerebellum and other local nuclei. As in our patient, the brain-stem EC probably disrupted the cerebro-pontine-cerebellar pathway laterally and compressed the facio respiratory center in the anterior portion of the pons, resulting in PL. However, the precise mechanism underlying PL remains unexplained.

In conclusion, this paper shows a rare case of brain-stem EC presenting with PL; like most other compressive

lesions, PL is reversible and responds immediately to surgical excision of the tumor. The mechanism of PL requires further explanation.

Acknowledgments. *The authors would like to thank Shengjun Sun, Radiologist, and Junmei Wang, Histopathologist for reviewing the figures.*

Received 10th December 2011. Accepted 4th April 2012.

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ETHICAL CONSENT

All manuscripts reporting the results of experimental investigations involving human subjects should include a statement confirming that informed consent was obtained from each subject or subject's guardian, after receiving approval of the experimental protocol by a local human ethics committee, or institutional review board. When reporting experiments on animals, authors should indicate whether the institutional and national guide for the care and use of laboratory animals was followed. Research papers not involving human or animal studies should also include a statement that approval/no objection for the study protocol was obtained from the institutional review board, or research ethics committee.