Pathological Laughing and Crying Associated with a Tumor Ventral to the Pons

Case Report

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Pathological laughing and crying, defined as sudden outbursts of uncontrollable, spontaneous, inappropriate laughing and/or crying, was recognized and studied in the late 19th century.1,15,17,22 Though most commonly found in diffuse cerebral vascular disease, it has also been observed in a variety of infectious, degenerative, demyelinating, epileptic, and neoplastic cerebral diseases.7,13,15 While reports of its occurrence with intracranial tumors have been appearing for over half a century,2,24 our case is unique in that so far as we can determine, it is the first reported instance in which both pathological laughter and crying were cured by the removal of such a tumor. Two additional cases20,21 are briefly mentioned in which patho-
logical laughing preceded by months or years the appearance of other neurological signs. In these two patients, as in ours, the tumor was located ventral to the pons. We suggest that pathological laughing or crying may be a useful sign in the early diagnosis of pre-pontine tumors.

Case Report

History. The patient was a 39-year-old male college student, admitted to Cushing Veterans Administration Hospital on February 17, 1950, complaining of uncontrolled spells of laughing and crying for the past 11 months. During the same time he had occasional short episodes of involuntary shallow respirations. For the last 8 months he had noted left-leg weakness, staggering to the left and clumsiness of the left arm, and, for 2 months, dysphagia with occasional regurgitation of fluids through the nose. He had no headache, dizziness, vertigo or tinnitus.

Examination. On admission the patient was alert and oriented. Positive physical findings were confined to the nervous system and included paroxysms of inappropriate and uncontrolled laughing and crying and occasional separate 1 to 2 minute episodes of rapid, shallow respirations (over 50 per min.). Cranial nerve changes included loss of corneal reflex, decreased ability to smell, and weak palatal movements and gag reflex on the left. There was deviation of the tongue to the left with bilateral perioral facial weakness and hearing loss, greater on the left. There was slight weakness of the left leg, dysmetria of the left arm and leg, and a positive Romberg with falling to the left. Reflex changes included left hyperreflexia, diminished cremasteric and absent abdominal reflexes. Bilateral extensor planter responses were present, greater on the left.

Routine blood and urine tests were normal. Spinal fluid pressure was 155 mm. of cerebrospinal fluid; protein 222 mg%. Skull x-rays showed erosion of the superior inner aspect of the left petrous pyramid and sclerosis of the left mastoid. A left carotid arteriogram was normal. The EEG was abnormal, with bilateral paroxysmal spiking suggesting a lesion of the deep mid-line structures.

Operation. On March 16, 1950, after indigo carmine had been injected into the right lateral ventricle, the left cerebellar hemisphere and midline were exposed through a left hockey-stick incision and the dura was opened. There was no obvious deformity of the cerebellum, the 4th ventricle or the medulla. The escape of cerebrospinal fluid (which contained no dye) from the basilar cisterns permitted exploration of the left poste-
rior fossa without removal of any cerebellar tissue. An encapsulated tumor was seen anterior and medial to the 7th and 8th cranial nerves, displacing them slightly posteriorly, but not attached to them. More retraction showed the tumor to be lying anterior and medial to all of the lower cranial nerves. It was attached firmly to the dura on the medial surface of the petrous part of the left temporal bone and the basilar portion of the occipi-
tal bone down to the rim of the foramen magnum. As the surrounding tissue was brushed from the posterior surface of the tumor, it appeared to cross the midline. It was clear that removal of the mass in one piece would be impossible without severe damage to the cranial nerves and the basilar and vertebral arteries. After free-
ing the tumor from surrounding structures—medulla, pons and cranial nerves—the capsule was incised. The interior portion of the tumor was gradually removed with a metal sucker and the capsule was allowed to collapse away from the adjacent structures. All but the most caudal portion of the capsule, which was attached to the anterior edge of the foramen magnum, was re-
moved. It was estimated that 80 to 90 per cent of the tumor was removed. As soon as the cerebellum was allowed to fall back into place, blue dye appeared in the spinal fluid. The tumor was diagnosed as a meningioma.

Postoperative Course. Pathological laughing and crying and paroxysmal rapid respirations ceased on the first postoperative day and have not recurred in the 15-year follow up. The patient returned to college and gradu-
ated. There was gradual return of normal symmetrical coordination, motor power, and reflexes in the left arm and leg. The only neurological deficits 15 years after operation are decreased sensation on the left side of the face and decreased muscle mass of the left masseter and temporalis muscles.

Discussion

Pathological laughing and crying has been de-
scribed with focal lesions of the central nervous
system ranging from the cortex to the medulla, and there have been many theories to explain its occurrence. Davison and Kelman said "apparently, there are pathways which originate in the cortex and which are in intimate connection with the thalamus, hypothalamic nuclei, mesencephalon and faciorespiratory nuclei. A lesion in any of these centers or along the course of the associated pathways may account for the appearance of these responses." Wilson believed that lesions of the geniculate bundle of the pyramidal tracts anywhere in its course, especially bilateral lesions, may produce pathological laughing and crying. We have reported a unique case and suggest the early localizing value of pathological laughing or crying in patients with intracranial mass lesions.

The occurrence of pathological laughing and crying with noninvasive intracranial tumors is rare. No mention is made of it in Cushing's experiences with meningiomas nor in Edwards' and Paterson's extensive review of the symptoms and signs of acoustic neurofibromata. However, its existence with invasive malignant tumors is well documented. Previously reported cases of brain tumors in which this sign was present show that it may be of localizing value when considered in reference to deficits in thought processes, changes in the state of consciousness, or obvious neurological signs. In all cases reviewed in which this syndrome was present in patients with a cortical tumor there were associated mental changes such as poor memory, inattention, impaired perception, blunting of emotional responses, or loss of social restraints.

When this syndrome was present with tumors of the diencephalon there was an associated disturbance in the patient's state of consciousness. Cairns commented that lesions of the 3rd ventricle and diencephalon may be associated with symptoms resembling psychosis or dementia, and especially states of stupor. Ironside, however, points out that while a depression in the level of consciousness is the more common finding with diencephalic lesions, occasionally one observes "episodic states of excitement with laughter, usually a prelude to stupor with dementia or death." Cox has also mentioned the association of "exhausted" states of consciousness with diencephalic lesions. Foerster and Gagel have reported 3 similar disturbances while operating on patients under local anesthesia in the region of the 3rd ventricle. Their most dramatic case was that of a patient who burst out in fits of laughter each time Foerster swabbed the floor of the 3rd ventricle following the removal of an intraventricular cyst. Fulton and Bailey have also described pathological laughing and crying occurring transiently in a patient for the first 3 weeks following the removal of a pinealoma from the 3rd ventricle. We see then that when pathological laughing and crying occurs due to a mass lesion at the diencephalic level, there usually is an alteration in the patient's state of consciousness.

Pathological laughing or crying with posterior fossa mass lesions is not commonly associated with an early alteration in state of consciousness or thought processes. In the far-advanced state they both may be present secondary to increased intracranial pressure and hydrocephalus. Most important is the fact that pathological laughter or crying may be the first sign to occur and may precede by months the development of other more orthodox neurological signs.

In our patient this syndrome preceded typical neurological deficits of the cranial nerves, pyramidal tract, and ipsilateral cerebellum by 3 months. This was also true in Stevenson's Case #1, in which the patient had outbursts of uncontrollable laughter with headaches antedating by months the subsequent posterior fossa signs. The tumor in that case was also a meningioma arising from the dura over the basilar portion of the occipital bone just behind the sella. It was compressing the pons and obstructing the aqueduct of Sylvius, causing internal hydrocephalus. The most recent report of pathological laughing associated with a microscopically documented brain tumor is the case in which Stevenson's translacial approach was used to remove a chordoma arising from the clivus where it had been producing brainstem and cranial nerve compression. This patient developed manifest bulbar and long tract signs. Two years earlier, he experienced uncontrollable laughter at his mother-in-law's funeral though he inwardly experienced the appropriate feelings of sorrow. Pathologic laughter persisted as his only neurological symptom, while his disease was considered to be functional. The pathological laughter in this patient, as in ours, ceased immediately following the operation.

In these 3 cases of early pathological laughing and crying the tumor was located ventral to the pons and medulla. Therefore, while the occurrence of this sign suggests a posterior fossa lesion, its presence alone without additional neurological abnormalities may indicate the development of a mass lesion anterior to the pons and medulla. The bizarre occurrence of episodic forced shallow respirations as well as the laughing and crying also suggests a brainstem lesion. Stevenson states that tumors at the base of the brain are "frequently accompanied by disturbances in respiration and heart rate." Edwards and Paterson observed similar respiratory disturbances in a 53-year-old man with an acoustic neurofibroma found at autopsy. They concluded that this was due to a local brainstem compression. The early recognition of these signs has recently become of even greater importance. With the devel-