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PATHOLOGICAL LAUGHTER IN A PATIENT WITH TRIGEMINAL NEURINOMA

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ABSTRACT - We present a 47-year-old woman with a long history of anxiety and a more recent history of shock-like facial pain and episodes of laughter without any motivation. She could not explain the laughing bursts and did not have a sense of mirth preceding it. On neurological examination she presented a VI nerve palsy and trigeminal hypoesthesia (V2 and V3) on the right side. Magnetic resonance imaging exhibited a large cystic lesion on the right middle fossa causing significant compression on the brain stem. A frontoorbitozygomatic and pretemporal combined approach was performed. During intra and extradural exploration a large tumor was found on the trigeminal nerve. The whole lesion was resected, revealing to be a neurinoma on pathological exhamination. She maintained a VI nerve palsy but had complete remission of the unmotivated laughing episodes during the one year follow up.

KEY WORDS: pathological laughter, neurinoma, trigeminal neuralgia.

Riso patológico em uma paciente com neurinoma de trigêmeo

RESUMO - Relatamos o caso de uma paciente de 47 anos com história de longa data de ansiedade que apresentou início de dor facial em choques do lado direito e episódios de riso sem motivação. Ela não podia explicar os episódios de riso e não percebia uma sensação de graça que os precedia. Ao exame neurológico apresentava paresia do VI nervo e hipoestesia no trajeto dos ramos oftálmico e maxilar do trigêmeo. A ressonância magnética de encéfalo apresentava uma lesão cística na fossa média direita causando significativo efeito de massa sobre o tronco encefálico. Um acesso combinado fronto-orbito-zigomático e pré-temporal foi realizado e a exploração intra e extra-dural revelou um grande tumor no nervo trigêmeo. Toda a lesão foi ressecada, revelando ser um neurinoma no exame patológico. A paciente manteve a paresia de VI nervo mas apresentou remissão completa dos episódios de riso imotivado durante o seguimento de um ano.

PALAVRAS-CHAVE: riso patológico, neurinoma, neuralgia trigeminal.

Pathological laughter is a condition associated with neurological lesions at various levels. Several structures have been involved as parts of the pathway responsible for the expression of laughter. There seems to be no cortical center for the control of laughter, but the motor, premotor and sensory areas are involved in the voluntary control. Subcortical structures such as the hippoccampus and the denteate girus have been implicated as well[1]. The hypothalamus plays a central role in the control and expression of emotions, one of which is laughter. It receives afferences from cortical and subcortical structures which work as modulators of emotional responses. Diffuse lesions that interrupt the corticohypothalamic tracts produce neurological syndromes marked by loss of control over the emotional expressions, such as pseudobulbar palsy[2]. In these cases marked changes in emotional expressions occur without appropriate stimulus, one of the conditions that characterize pathological laughter. A lower pathway may be responsible for the motor control of laughter. A faciorespiratory mechanism has been pointed out to coordinate the facial nucleus with the nucleus ambiguous and the phrenic nuclei in the upper cervical spinal cord. This lower mechanism is also supposed to be under upper and cortical control, producing pathological laughter as well when liberated by lesions on the descending pathways. The tegmental area at the mesencephalon, as well as the central gray matter may also be part of this large network, also controlling the facial, vocal and respiratory movements. Lesions at this level have also produced stereotyped and purposeless laughter in patients[2].

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Many pathological situations have been associated with pathological laughter. Diffuse lesions such as produced by multiple sclerosis and atherosclerosis are associated with interruption of large descending tracts. Various tumors, infarcts and degenerative lesions have been associated with pathological laughter secondary to larger focal lesions in any of the mentioned structures. Nevertheless, no previous report had linked trigeminal neurinoma to pathological laughter as a symptom, as in the case we report.

CASE

A 47-year-old woman was referred from a psychiatrist with a long history of anxiety and paranoid symptoms. She had a more recent history of headache and facial pain. The pain was characterized by shocks on her right facial side, which were frequent and would not improve with common analgesics. On further questioning she complained of purposeless laughter, which would occur randomly, without any motivation at any situation. There was no sense of mirth preceding laughter and the patient could not give any explanation to the bursts. There was no association to anxiety preceding laughter, which occurred spontaneously during rest as well. During the first and subsequent consultations, laughter was observed either when the patient was reporting her illness or sometimes while she awaited (Fig 1). On neurological examination she seemed orientated and no neuropsychological alterations could be detected. Although she pointed the pain to occur in the territory of all the three branches of trigeminal nerve, hypoesthesia was found only in V2 and V3. A right VI nerve palsy was detected as well as a vestibular syndrome characterized by disturbance in gait and balance towards the right side.

She was submitted to a MRI (Fig 2) which reveled a lesion with cystic characteristics in the middle fossa, causing brain stem compression. There was no signal alteration in the brain stem which would suggest anatomical lesion.

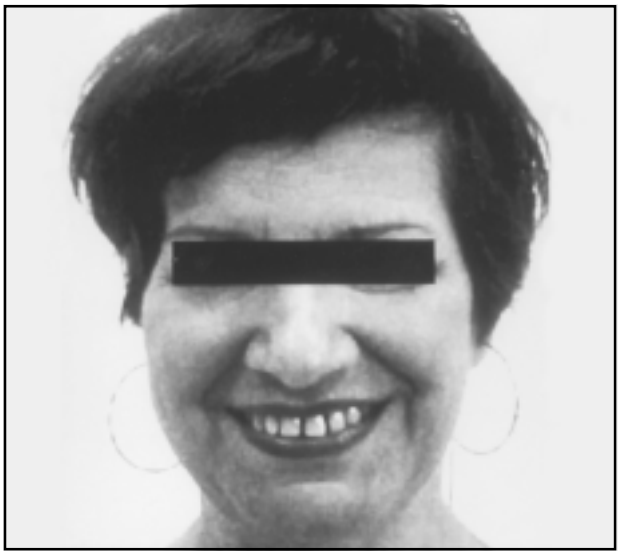

Fig 1. The patient presenting laughter without intent nor mirth.

She was submitted to a frontoorbitozygomatic craniotomy, with a pretemporal extension. An extradural and intradural approach was performed, allowing identification of a tumor of the trigeminal nerve. Complete resection of the lesion was achieved, as confirmed by MRI (Fig 3) and the pathological examination concluded it was a neurinoma.

The patient developed a worsening of the VI nerve palsy and started to receive 200 mg of carbamazepine daily. She was released from the hospital on the 10th post operative day, presenting significant improvement of the vestibular symptoms. At one year after the operation she had no more trigeminal pain. Purposeless laughter had resumed as well.

DISCUSSION

In understanding the nosology of pathological laughter one must first consider its definition. As a wider one, it is safer to consider as pathological any production of laughter without the appropriate stimulus, either external (humorous) or internal (thoughts), at inappropriate situations and without voluntary control[1,2]. Differentiation must be made from emotional liability or emotional incontinence, in which laughter and crying alternate as an expression of the patient's true (and pathological) affective state. Dementia may also be associated with inappropriate laughter which is usually not included in the series reporting pathological laughter, since it is believed to occur from the diffuse loss of higher function control over the subcortical circuitry responsible for the expression of emotion and laughter. Finally, pathological laughter is associated with a structural lesions or functional disorders of the central nervous system.

Propedeutically, it is important to obtain from the patient if there is a sense of mirth preceding laughter or if it comes merely as a motor phenomenon[2,3]. Patients sensing mirth probably present with a lesion affecting higher levels of function, including the circuit of Papez. Without appropriate stimulus, spontaneous mirth may occur from direct stimulation of subcortical structures such as the hypothalamus or the hippoccampus and denteate girus, allowing the patient to have a sensation compatible to the facial and respiratory responses unleashed[3]. Concomitantly, the motor response observed is far more realistic (as a true laughter) than in cases presenting pure motor responses, without the sense of mirth.

Lower lesions observed both in patients and in experimental models produce movements of the facial muscles coordinated with respiratory movements that only resemble laughter, including the sound produced. The facial expression may not be coherent with movements of the mouth and sounds from respiratory movements[4,5].

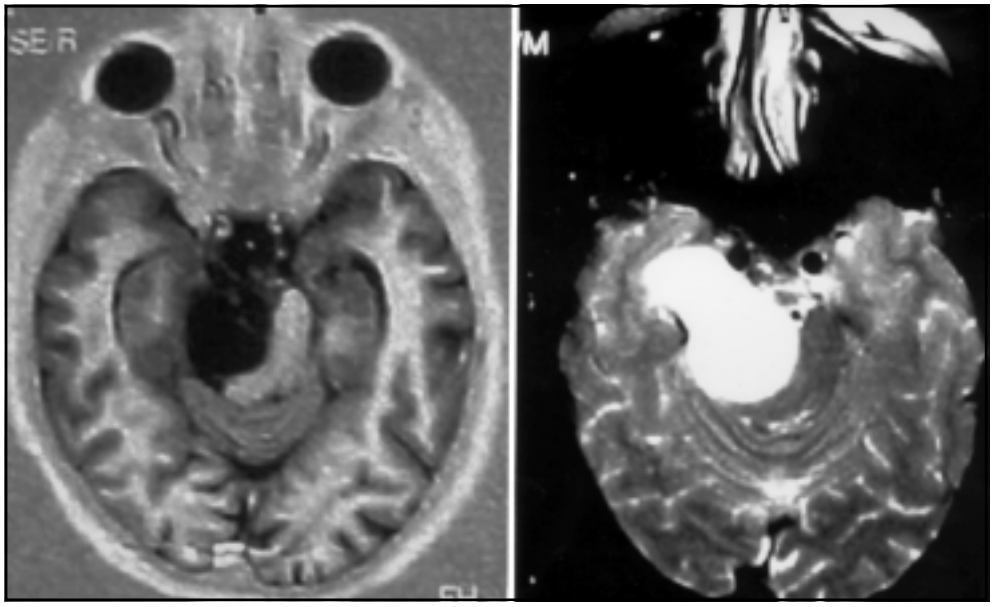

Fig 2. A: T1-weighed MR image showing a large lesion with hyposignal content and no contrast enhancement. B: At T2-weighed image, the hypersignal core suggests the content to be cystic.

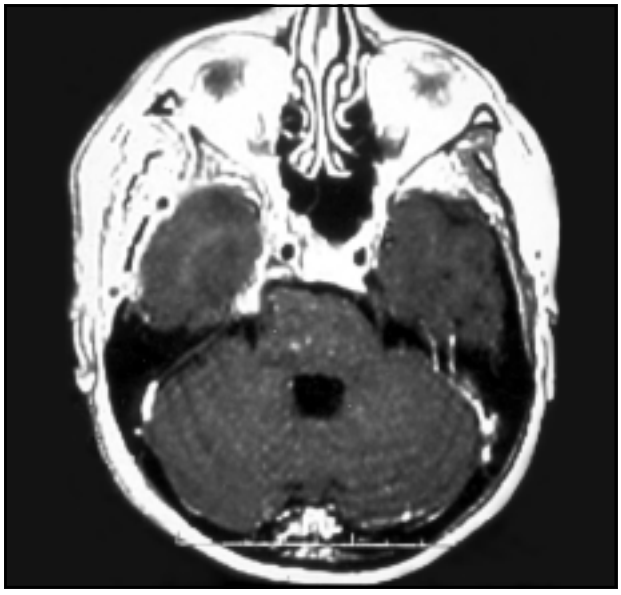

Fig 3. FLAIR weighed MR image showing complete resection of the lesion.

The pathological substrates for focal lesions associated with pathological laughter in the literature include frontal (midline) and temporal gliomas[1,3]; free edge tentorial meningiomas compressing the hypothalamus, thalamus and cerebral peduncles[1]; other tentorial edge tumors[6], clival chordoma and petroclival meningioma with marked compression of the pons and mesencephalon[7,8]; infarcts at the diencephalic structures; multiple cerebrovascular lesions from embolic disease and demyelinating lesions affecting the central white matter and diencephalon. Removal of such lesions have been reported to resume pathological laughter[1]. Several epileptic disorders have been implicated with pathological laughter as well, which can be summarized as patients with complex partial seizures of suspected temporal origin and patients with hypothalamic hamartomas and gelastic seizures[9].

We presented a case of pathological laughter in a patient with a large trigeminal tumor. Besides non motivated laughter, the patient also presented sings of cerebelopontine angle compression. There was no sense of mirth suggesting not a disorder of higher neurological structures but a direct lesion on motor effectors of laughter. There was no sign of hypoccampal compression, while the mass effect was mainly upon the brain stem. Compression at this site would explain why laughter was not comprehensible even to the patient as well as why it seemed not convincing to the examiner. The case illustrates that the mass effect relief in this topography may improve the patient's symptoms of pathological laughter.

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