This unusual presentation of multiple system atrophy.

Patient 1 was a man who initially developed a sleep disorder at the age of 57. During sleep, he began to talk or even shout in an unusual, recorded motor jerks such as flinging his arms or lifting himself off the pillow. On several occasions he abruptly got out of bed and injured himself by colliding with furniture. His spouse had sustained severe head injuries following one particular attempt at strangulation. These violent attacks lasted for a few minutes and occurred between midnight and 3:00 am. They could be aborted by forceful wakening. However, a few years later, the patient progressively developed a full picture of multiple system atrophy of striatogniral degeneration type with predominantly right sided akinetic rigid syndrome, unresponsive to levodopa, and predominant right side postural akinetic rigidity, pyramidal signs, dysarthria and hoarseness, dysphagia, urinary incontinence and retention, impotence, postural hypotension with syncope, and autonomic dysfunction.

The violent behavioural episodes improved, but the nocturnal speech production persisted and snoring and episodes of stridor appeared. Electroemography showed desynchronization of the subcortical and cerebral sphincters, a pattern not typically found in multiple system atrophy. Polygraphic sleep showed stage 1 and 2 sleep with little deep non-REM sleep and no episode of REM sleep. During light sleep, there were episodes of bilateral focal myoclonic twitches of arms, hands, and thumb followed by widespread alpha activity for about 30 seconds. During REM sleep, there was atonia in the lower extremity and also in the lower part of the chest. During all episodes of REM sleep, there were episodes of tonic atonia in the lower part of the body. The EMG of the diaphragm showed a high percentage of tonic activity, with a consequent increase in diaphragm tone. There were no episodes of atonia with EMG activity, and no episodes of REM sleep with tonic atonia in the lower part of the body. The EMG of the diaphragm showed a high percentage of tonic activity, with a consequent increase in diaphragm tone. There were no episodes of atonia with EMG activity, and no episodes of REM sleep with tonic atonia in the lower part of the body.

Crying seizures after cerebral infarction.

Ictal crying is a relatively rare epileptic condition. There have been 11 such cases reported before 1993. Recently seven more cases were reported by Tison and colleagues. The aetiology of the crying seizures in these patients was attributed to tumour, vascular malformations, or mesial temporal sclerosis. There have been no previous reports of ictal crying after cerebral infarction. We report a case of crying seizures in a patient after cerebral infarction with a seizure focus in the right temporal region. The patient, a 66-year-old woman with a left posterior wall myocardial infarction, a 66-year-old woman with a left hemiparesis that resolved over a five-day period. She was placed on aspirin (325 mg three times daily). The patient did well until two years later when she awoke with a left hemiparesis. There was no sensory loss or ataxia. Carotid doppler imaging, trans-thoracic echocardiography, and cardiac monitoring were unremarkable. Brain MRI showed multiple focal ischaemic changes in the white matter, most notably in the posterior limb of the right internal capsule, and mild diffuse cerebellar atrophy. The patient was treated on ticlopidine (250 mg twice daily).

Correspondence to: Dr F Tison, Département de Neurologie, Hôpital Pellegrin, Place Amélie Raba- Léon, 33076 Bordeaux Cedex, France.


Neuropathic malignant melanoma presenting as a trigeminal sensory neuropathy

Cranial neuropathies may occur as a complication of malignant melanoma of the head and neck and may not be a fleeting feature. We have not been able to find an account of this in the neurological literature.

A 63-year-old woman presented with a seven-month history of altered sensation over the right cheek and nostril; for five months the cheek and the roof of the mouth on the right side had been numb and a continuous dull pain had developed over the right face. There was no relevant medical history. There was no family history of neurological problems and the patient had not received cytotoxic medication in the past.

On examination there was mild infraorbital oedema on the right and severe loss to light touch and pin prick over the cheek, the side of the nose, the hard palate and upper gum on the right side. Movements of the mandible, face, tongue and palate were symmetric and sensation over the soft palate and posterior pharyngeal wall was normal. The rest of the clinical examination was normal.

Routine blood tests, a chest radiograph, and examination of the CSF were normal. Plain radiographs of the maxillary sinuses, fine section axial CT through the skull base and parasagittal sinuses (which included images of the nasopharynx) and MRI of the brain stem and middle cranial fossa were all normal. Examination under anaesthesia of the postnasal space and biopsies of the right lateral nasopharynx and the right fossa of Rosenmuller were normal.

Two months later the patient noticed enlargement of a lesion on the tip of her nose. This lesion had been present for as long as the patient could remember and until this time it had not changed or attracted her attention; medical staff had noticed the lesion when the patient first presented and had considered it to be benign in appearance. Examination revealed a 13-mm, pink, shaped nodule on the tip of the nose measuring 13 mm by 10 mm with a purple red surface; no brown pigmentation was present to suggest a melanocytic lesion. Histopathological examination of a punch biopsy of the lesion revealed an amelanotic malignant spindle cell tumour. A biopsy of the skin of the right cheek was then performed and histopathological examination revealed thickened inflamed nerves with infiltration of the perineurium by cytologically malignant cells (figure). Immunocytochemical studies showed that both the spindle cells in the first biopsy and the larger cells in the perineurium of the dorsal nerves in the second biopsy were positive with polyclonal anti S-100 antibodies (figure, insert) and monoclonal HMB-45 antibodies. The pathological diagnosis was of a spindle cell neurotropic malignant melanoma.

The swelling of the right cheek worsened and repeat CT of the head two months later showed a 20 mm by 15 mm soft tissue mass in the right temporal area (figure), confirming the occurrence of partial seizures with ictal crying. He was loaded intravenously with phenytoin with resolution of the crying seizures. Brain CT showed periventricular leukomalacia and mild diffuse cerebral atrophy. Neurological examination showed a left hemiparesis, unchanged from previous examinations.

Crying can represent an affective behavioural manifestation of sadness or depression, can occur as an ictal event or postictal phenomenon, or can occur in association with a non-epileptic seizure. Outbursts of involuntary and uncontrollable laughing or crying may also accompany the pathological emotion seen in patients with pseudobulbar palsy, which may occur in vascular, degenerative, or demyelinating diseases of the brain.

The neuroanatomical localisation of crying remains unknown. In most patients with crying seizures, the EEG has provided evidence of a frontotemporal seizure focus in the non-dominant cerebral hemisphere. Evaluation of our patient also showed the presence of a seizure focus in the temporal region of the non-dominant cerebral hemisphere, presumably caused by ischaemic cerebrovascular disease as there was no evidence of an intracranial tumour, vascular malformation, or infection. This is in support of the concept that the limbic areas in the non-dominant hemisphere are related more to the experiencing of negative emotions, such as fear and anxiety, than positive emotions such as happiness. 1

To our knowledge, this is the first reported case of crying seizures after cerebral infarction. As sudden outbursts of uncontrollable crying may also accompany the pathological emotion seen in patients with pseudobulbar palsy, the differentiation of these two possibilities with EEG recordings should be considered.

DAVID Z WANG
Creighton-Nebraska Neurology Programme
ROBERT E STEG
NANCY FUTRELL
Creighton University School of Medicine

Correspondence to: Dr Robert E Steg,
Department of Neurology, Creighton University School of Medicine, 601 North 30th Street, Omaha, Nebraska, 68111, USA.

Crying seizures after cerebral infarction.

D Z Wang, R E Steg and N Futrell

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