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 association with violent muscle 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 several injuries including one apparent 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. Two to three years later, the patient progressively developed a full picture of multiple system atrophy of striatonigral degeneration type with predominantly right sided akinetic rigid syndrome, unresponsive to levodopa, and subsequent impairment of postural reflexes, pyramidal signs, dysarthria and hoarseness, dysphagia, urinary incontinence and retention, impotence, postural hypotension with syncope, and a peripheral sensorimotor neuropathy. The violent behavioural episodes improved, but the nocturnal speech production persisted and snoring and episodes of stridor appeared. Electromyography showed denervation of the external urethral sphincter typically found in multiple system atrophy.⁶ Polysomnography with video showed stage 1 and 2 sleep with little deep non-REM sleep and one episode of REM sleep. During light sleep, there were episodes of bilateral fragmentary myoclonic twitches of arms, hands, and thumbs followed by widespread alpha activity for about 30 seconds. During REM sleep, there was atonia on segmental EMG apart from one episode of a sudden flinging movement of both arms with a related increase in EMG activity. No epileptic features were recorded during the episode. None of the other nocturnal behaviours described at the beginning of the disease were recorded on this occasion. There were no apnoeic episodes and no dips in oxygen saturation.

Patient 2 was a man who developed restless and disrupted sleep at the age of 42. He often shouted intelligibly, and sometimes lectured or reprimanded people. The patient was aware of this and said that it was in the context of dreaming. Periodically he would jump out of bed, shake or growl in a stereotyped fashion, or flail his arms. Once he dived out of bed in a rugby tackle, believing he was actually tackling someone. Such episodes occurred at around 5 00 am rather than soon after going to bed, from once a fortnight to twice a week. Episodes of snoring were also noticed. Two to three years later he progressively developed impaired coordination, slurred speech, impotence, and intermittent postural faintness. A full picture of multiple system atrophy of the olivopontocerebellar atrophy type then developed with prominent cerebellar, pyramidal, and dysautonomic signs and symptoms, and mild parkinsonian features. His nights were then predominantly affected by snoring and episodes of stridor. At that time, five years after the onset of nocturnal problems, a sleep study recorded a normal sleep architecture with no excessive movements. No epileptic features or apnoeic periods were recorded, but the patient made "squeaking" noises on inspiration. The baseline saturation was 93% with minor dips throughout the study. Urethral

sphincter EMG was again abnormal,⁶ and brain MRI showed moderate cerebellar atrophy.

The two patients described developed clinically probable multiple system atrophy of striatonigral degeneration (case 1) and olivopontocerebellar atrophy (case 2) types.⁷ In both cases, pronounced sleep disturbances were the first recorded complaint, preceding the onset of the first motor or autonomic symptoms by two to three years. Such sleep disorders, with a history of violent and potentially harmful behaviour and reported vivid dream mentation appropriate to the observed action, are typical of REM sleep behaviour disorder.⁵ The disorder was subsequently documented by the recording of a less severe episode in patient 1 during a sleep telemetry study.

Chronic REM sleep behaviour disorder is often reported to be idiopathic⁵ and could coincidentally have appeared in these two cases. Beyond the temporal association, however, there are other reasons to believe that REM sleep behaviour disorder was the first manifestation of multiple system atrophy in those patients. Thus REM sleep behaviour disorder has also previously been described during the course of,^{1,2} or before the onset of, multiple system atrophy⁴ and in familial olivopontocerebellar atrophy, the pathology of which bears some similarity to that of multiple system atrophy.⁸ It has also been described in idiopathic Parkinson's disease, occurring late in the disease, but occasionally preceding other symptoms.⁹ REM sleep behaviour disorder is thought to originate from a dysfunction in pontine structures generating REM sleep muscle atonia.⁵ Dream acting behaviour and disappearance of REM sleep muscle atonia have been documented after experimental pontine lesions,^{5,8} and pontine lesions are almost always found in both striatonigral degeneration and olivopontocerebellar atrophy variants of multiple system atrophy, with a more restricted involvement of the locus ceruleus in idiopathic Parkinson's disease.¹⁰ It is therefore likely that REM sleep behaviour disorder was the first manifestation of pontine involvement due to multiple system atrophy in our two patients, and has to be added to the growing list of unusual clinical presentations of multiple system atrophy of which physicians should be aware.⁷ These two cases also underline the fact that REM sleep behaviour disorder is not always idiopathic, but can instead herald the onset of a major neurodegenerative disorder.^{4,5}

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Crying seizures after cerebral infarction

Ictal crying is a relatively rare epileptic condition.^{1,3} There have been 11 such cases reported before 1993. Recently seven more cases were reported by Luciano *et al.*¹ The aetiology of the crying seizures in these patients was attributed to tumour, vascular malformations, or mesial temporal sclerosis. There have been no previous case 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.

Three weeks after an inferior wall myocardial infarction, a 66 year old right handed man developed dizziness and a left hemiparesis that resolved over a five day period. He was placed on aspirin (325 mg three times daily). The patient did well until two years later when he 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 cerebral atrophy. The patient was placed on ticlopidine (250 mg twice daily).

The patient did well until seven months later when he began exhibiting unusual behaviour including inappropriate speech, episodes in which he would suddenly stop talking and stare, and episodes of eye deviation to the left. On admission to hospital he was seen to exhibit repeated episodes of deviation of the head and eyes to the left during which time he would begin crying. During these crying episodes he remained alert, answered questions appropriately, and denied any subjective experience of sadness or depression. Electroencephalography performed during these episodes showed ictal discharges occurring repeatedly over the

Electroencephalography during an episode of ictal crying shows a repetitive sharp wave discharge, maximal over the right temporal area.

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 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.^{3,4}

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.

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Neurotropic 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 be the presenting 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 sensory 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 paranasal 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 showed a dome 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 dermal 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 mass superficial to the right maxillary antrum extending through to the infraorbital foramen. This was thought to be expanding tumour along the course of the infraorbital nerve and a palliative course of radiotherapy to the right side of the face was commenced. The patient died 11 months after presentation.