The patient was a man who initially developed a sleep disorder at the age of 57. During sleep, he began to talk or even shout in arousals, recording 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 following 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 or by cutting him off the pillow, but the patient gradually developed a full picture of multiple system atrophy of striatogniral degeneration type with predominantly right sided akinetic rigid syndrome, unresponsive to levodopa, and subsequent impairment of postural reflexes, pyramidal signs, dystarhria and hoarseness, dysphagia, urinary incontinence and retention, impotence, postural hypotension with syncope, and autonomic instability.

The behavioural episodes improved, but the nocturnal speech production persisted and snoring and episodes of stridor appeared. Electromyography showed denervation of the diaphragmatic and phrenic nerves, a finding typically found in multiple system atrophy. Polysonomography with video showed stage 1 and 2 sleep with little deep non-REM sleep (nREM) 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 and fragmentary EMG activity. Activity per se is not abnormal, and absence of EMG activity per se is not abnormal. The patient chose to take a nap right after dinner. Periods of time that 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 type then developed with prominent cerebellar, pyramidal, and dysautonomic signs and symptoms, and mild parkinsonian features. His nights were then predominantly affected by these 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, and brain MRI showed moderate cerebellar atrophy.

The two patients described developed clinically probable multiple system atrophy of striatogniral degeneration type (case 1) and olivopontocerebellar atrophy (case 2) types. In both cases, pronounced sleep disturbances were the first recorded complaint, preceding the onset of the first motor or autonomous 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 (RBD) and are described 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 idiopathic and can 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 these patients. Thus REM sleep behaviour disorder has also previously been described during the course of, or before the onset of, multiple system atrophy and in familial olivopontocerebellar atrophy, the pathology of which bears some similarity to that of multiple system atrophy. It has also been described in idiopathic Parkinson's disease, occurring late in the disease, but occasionally preceding other symptoms.

REM sleep behaviour disorder is thought to originate from a dysfunction in pontine structures generating REM sleep muscle atonia. Atonia and the disappearance of REM sleep atonia has been documented after experimental pontine lesions, and pontine lesions are almost always found in both striatogniral degeneration and olivopontocerebellar atrophy variants of multiple system atrophy, with a more restricted involvement of the locus ceruleus in idiopathic Parkinson's disease. It is therefore likely that REM sleep behaviour disorder is 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. Its aetiology should be further elucidated.

The 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.

F TISON
G KNENNING
N P QUINN
University Department of Clinical Neurology,
Institute of Neurology,
The National Hospital for Neurology and Neurosurgery,
Queen Square, London, UK

S J M SMITH
Department of Clinical Neurophysiology,
The National Hospital for Neurology and Neurosurgery,
Queen Square, London, UK

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


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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, or 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, the differentiation of these two possibilities with EEG recordings should be considered.

David Z Wang
Creighton University School of Medicine
Omaha, Nebraska, 68131, USA.

Correspondence to: Dr Robert E Steg, Department of Neurology, Creighton University School of Medicine, 601 North 30th Street, Omaha, Nebraska, 68131, USA.


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 present as a sentinel feature. We have not been able to find an account of this in the neurological literature.

A 63-year-old man 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 sensation 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 pansinus x-rays (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 Rosenmüller 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 engendered loss 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 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 13 mm, partly supraventricular 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.