and eye movement abnormalities without evidence of myasthenia over 12 years.³

There are a few postmortem studies. The relevant findings are: in anti-GAD+ SPS, reduction of small spinal neurons and changes in larger alpha-motor neurons, as well as selective depletion of Purkinje cells (GAD-containing neurones), have taken place; in paraneoplastic SPS (anti-GAD-, amphiphysin positive), brainstem encephalitis has led to lymphocytic infiltrates, neuronal loss, gliosis and perivascular lymphocytic cuffs.

The pathogenic role of anti-GAD is controversial. Up to 65% of SPS cases have anti-GAD, but in paraneoplastic SPS, associated with antibodies to amphiphysin and gephyrin, anti-GAD are rarely present. Although uncommon, anti-GAD have been reported in other neurological diseases such as cerebellar ataxia, drug-resistant epilepsy, tremor of the mouth floor and periodic alternating nystagmus. Anti-GAD have also been found in diabetes and other autoimmune diseases in a lesser proportion.

In vitro, anti-GAD inhibit GAD activity; however, in vivo it is uncertain (antibodies circulate extracellularly, but GAD is intracellular).

In SPS, the concentration of GABA is reduced in CSF and assessed by magnetic resonance spectroscopy; GABA is decreased in the sensorimotor cortex. GABA is the major inhibitory neurotransmitter in the striatum and GAD activity was detected in spiny and non-spiny striatal neurons. Medium spiny projection neurons comprise 90–95% of the striatal neuronal population and are inhibitory via GABA. The basal ganglia inhibit undesirable or competing movements and postural synergies initiated by cerebral cortical mechanisms, with help from the cerebellum.

GABA levels are decreased in the contralateral sensorimotor cortex and lentiform nuclei in focal hand dystonia. In Huntington's disease, striatal neuronal dysfunction would produce an inability to inhibit unwanted movements, resulting in chorea. In our patient with SPS, striatal neuronal dysfunction might have produced inability to inhibit simultaneous co-contraction of agonist and antagonist muscles. Either functional or structural striatal abnormalities may play a role in SPS, but more studies are needed to confirm this hypothesis.

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Möbius syndrome in association with the REM sleep behaviour disorder

We report a 34-year-old woman with Möbius syndrome in association with lifelong sleep disturbance diagnosed as rapid eye movement (REM) sleep behaviour disorder (RBD). RBD proved treatment resistant and the possible structural reasons for this are discussed.

Case history

The patient was a 34-year-old woman who was noted at birth to have bilateral abducens nerve palsy, complete facial diplegia, club feet, malformation of the right upper limb and chest wall with an underdeveloped right hand and an absent pectoralis major. These abnormalities were non-progressive and a diagnosis of Möbius syndrome was made based on the characteristic clinical features. The Poland anomaly (pectoralis major hypoplasia and mammary hypoplasia) is associated with 13% of cases of Möbius syndrome.¹

From birth, the patient's mother noted sleep disturbance with brief crying and vocalisation as well as twitching arm and leg movements during sleep. The sleep disturbance became more marked over time and by the age of 3 years until the current day she has suffered episodes of screaming and abnormal movements occurring in the second half of the night on a nightly basis, usually between 02:00 and 04:00. Her mother described screaming with non-stereotyped, flailing movements of her arms and legs lasting several minutes. The patient's legs often moved as if running in the bed. She had injured both herself and her mother when restrained during an attack and was initially confused after being woken.

The history from the patient was of persistent unpleasant dreams, with recurring themes. Examples included being chased through rooms with a gun and being hunted by guards in a concentration camp. The dreams did not change in a different environment but increased during menstruation and stressful life events.

There was no other past medical history; in particular there was no history of snoring, restless legs syndrome or sleepwalking. There was no family history of any psychiatric or sleep disorder. She was a teetotal non-smoker.

Investigations

A diagnosis of RBD was made on the basis of her clinical description and she was investigated at Papworth Hospital. MRI of her brain with axial T1 weighted, T2 weighted and fluid attenuated inversion recovery sequences were normal, with no pontine abnormality seen. Routine 12 lead EEG while waking and drowsy was normal.

She underwent polysomnography on two consecutive nights using a standard protocol including video recording, sleep EEG (leads C3-A2, O1-A2), electrooculogram, and submental

and bilateral anterior tibialis electromyography (EMG). Respiratory effort, airflow and oxygen saturation were measured to exclude sleep apnoea syndrome. Sleep stages were scored according to standard criteria in 30 s epochs. Sleep latency, sleep efficiency and percentage of stage 1, stage 2, stage 3, stage 4 and REM to total sleep time were recorded.

There was no sleep disordered breathing or periodic leg movements of sleep. REM sleep was difficult to stage because the facial diplegia and ophthalmoplegia affected interpretation of the submental EMG and electrooculogram. Her sleep stages corresponded to 90 min cycles with clearly defined slow wave sleep and at 02:15 and 04:30 on the second night during non-slow wave sleep she had brief episodes with vocalisation and some flailing movements of her right arm (fig 1).

Treatment

The patient tried numerous medications sequentially over many years without benefit. In order, clonazepam, melatonin, carbamazepine, sertraline and temazepam were used at therapeutic doses, each for at least 3 months. None of the medications decreased the disturbing dreams. Cognitive behavioural therapy was also unsuccessful.

Discussion

Möbius syndrome was initially defined as congenital facial diplegia and bilateral abducens nerve palsies. The description has since been broadened as many patients have other cranial nerve palsies, with variable ophthalmoplegia, bulbar dysfunction and tongue atrophy all seen. Musculoskeletal abnormalities occur in one third of patients with clubfeet, the commonest deformity, but brachydactyly, arthrogryposis and the Poland anomaly are also seen.¹

The pathogenesis of Möbius syndrome has been debated but simultaneous limb malformations with cranial nerve dysfunction suggest disruption of normal morphogenesis during a critical period in the development of the embryonic structures of these regions. In a review of 37 patients with Möbius syndrome, the authors concluded that the widespread involvement of various motor nuclei and axons made it more likely to be a syndrome of rhombencephalic maldevelopment, rather than an isolated cranial nerve or nuclear developmental disorder.¹ Imaging studies showing a variety of brainstem anomalies, including absence of the facial nerve, support this theory.¹

Typical idiopathic RBD affects elderly males, often in association with neurodegenerative diseases such as Parkinson's disease and dementia with Lewy bodies. Animal studies indicate that the pedunculopontine region of the upper brainstem is involved. RBD is usually well treated with clonazepam.²

It is possibly surprising that a disorder that causes brainstem abnormalities does not produce abnormalities of REM sleep more frequently. None of a large cohort of patients enrolled into a specialist Möbius clinic complained of disturbing dreams (Padberg GW, personal communication, 2006). Sleep disturbance in Möbius syndrome has been reported in association with both central and obstructive sleep apnoea and thought to be secondary to palatal dysfunction.¹ There are no individual reports of RBD in association with this condition in the literature although there are reports

Figure 1 Sleep staging from the polysomnogram is shown, with wake (WK), movement (MVT), rapid eye movement (REM) sleep and the non-REM sleep stages, S1, S2, S3 and S4. There was some delay before the patient fell asleep but the overall sleep architecture was then normal with two episodes of abnormal movements during REM highlighted with the black arrows. The patient was in bed from 23:00 to 06:00 and the two events occurred at 02:15 and 04:30.

of other patients with REM abnormalities,^{3,4} suggesting that the variable developmental, pontine disorder in Möbius syndrome can cause REM disturbance if the critical neural networks within the pedunculopontine nucleus are affected. Secondary causes of RBD with structural pontine lesions support this theory.⁵ It is also possible that the condition is underreported or underdiagnosed and that more cases will emerge as RBD becomes more widely recognised.

The description of the stereotyped disturbing dreams with dream enactment in the absence of other causes of sleep disturbance on the polysomnography is characteristic of RBD, but atypical features include the early age of onset and treatment resistance. The young age of onset could be explained by a developmental rather than a neurodegenerative disorder. The reason for the lack of response to treatment remains unclear.

The case is unusual but highlights the need to question patients with Möbius syndrome specifically about sleep disturbance and it supports the theory that Möbius syndrome is a variable disorder affecting brainstem development.

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BOOK REVIEW

9/11 Mental health in the wake of terrorist attacks

Edited by Yuval Neria, Ezra Gross, Randall Marshall. Cambridge University Press, Cambridge, 2006, £60.00 (hardcover), pp 630. ISBN 0521831911

This book comprehensively describes the psychological response to the 9/11 terrorist attacks in New York and, to a lesser degree, Washington DC. The impact of what happened on the local and US national population is considered through various epidemiological studies, as well as personal accounts from some of those more directly involved. These complement each other well and highlight the fact that some individuals developed marked psychological difficulties as a result of the attacks but the majority of those exposed have not. The results, particularly of Galea and colleagues' excellent work, suggest marked resilience among those involved. These findings and increasing research evidence regarding the lack of effectiveness of universal interventions raise questions about some of the outreach and intervention work described in one section of the book.

There is clearly a need to provide practical and pragmatic support in an empathic manner following traumatic events and this appears to have been effectively done in many instances but questions remain unanswered regarding if, when and how more formal interventions should be provided. The absence of methodologically robust evaluation research concerning the programmes adopted suggests that an opportunity has probably been missed to further assess what best to do following traumatic events. Without doubt, many of those involved appear to have welcomed the support they received, but it is impossible to determine whether or not the interventions were truly effective in reducing levels of short term mental distress or the development of longer term psychological sequelae. These issues are discussed in more detail in the chapters of Litz, Shelev and Wesseley, which

provide more critical commentaries on the approaches taken with full acknowledgement of the difficulty in deciding how best to respond.

The interesting chapters regarding community responses and effective leadership in extreme crisis support recent guidelines that suggest a move towards a more sociological approach to encourage resilience and support recovery. There are useful examples of the adoption of a stepped/stratified care approach and the provision of training and support for local service providers to provide specific interventions that have been shown to be effective, such as prolonged exposure treatment for post-traumatic stress disorder.

The chapter authors include a good mixture of recognised experts in the field and individuals who were directly involved and have developed an expertise as a result of this particular experience. A major strength of the book is the self-critical approach that many of the authors have taken when writing their chapters. They carefully discuss methodological issues and dilemmas that are likely to face anybody trying to provide services and conduct research in this area.

This book will undoubtedly serve as a very helpful resource for anybody who wants to learn more about the psychological aftermath and response to the 9/11 terrorist attacks and should be used to inform the planning of psychosocial responses in the future.

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CORRECTION

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J Moriarty. Psychiatric disorders in neurology patients. *J Neurol Neurosurg Psychiatry* 2007;**78**:331. In the text and in the references of this paper the first author of its companion paper has been spelt incorrectly. The correct reference is Jefferies K, Owino A, Rickards H, *et al*. Psychiatric disorders in neurology inpatients: estimate of prevalence and usefulness of screening questionnaires. *J Neurol Neurosurg Psychiatry* 2007;**78**:414–16.