Pavor nocturnus from a brainstem glioma

Pavor nocturnus or night terrors usually occur in the absence of identifiable neuropathology. This report documents pavor nocturnus associated with a brainstem lesion.

A 15 year old boy with headaches and ataxia had a brainstem tumour on CT (see fig). The patient underwent resection of a grade I cerebellar astrocytoma arising from the fourth ventricle and adherent to the brainstem. His postoperative examination disclosed dysarthria, spontaneous vertical nystagmus, bilateral sixth nerve paresis, decreased sensation on the face bilaterally, facial diplegia, sensorineural hearing loss, mild dysmetria and ataxia, and decreased reflexes with bilateral Babinski responses.

Postoperatively he developed a sleep disorder where he suddenly sat up in bed, screamed, and appeared to be staring in fright. During these episodes he was agitated and would try going over the rails of the bed or would thrash about in bed screaming. After one or two minutes, he promptly fell back to sleep. He had no incomplete recollections of these episodes. Sometimes the only evidence of an episode was injury or blood on the floor. At other times, he recalled being frightened by images of parts of people sticking out of walls or by the belief that the bedposts were his room mates restraining him. The patient subsequently became depressed and had aggressive outbursts and paranoid beliefs. Before his tumour, he did not have a history of neurological or psychiatric difficulties, and there was no family history of a sleep disorder.

At the age of 24, polysomnography documented his night terrors. Spontaneous arousals from slow wave sleep were typically sudden and included restlessness, vocalization, and looking accompanied by interspersed alpha and delta activity of electroencephalography (EEG). After starting clonazepam (0.5 mg at bedtime), the episodes of night terrors decreased, but he developed enuresis.

There are few documented cases of pavor nocturnus from neurological disturbances. Reports suggest that night terrors may occur as a consequence of a right temporal lobe seizure focus. This report documents pavor nocturnus associated with a fourth ventricular brainstem lesion.

Pavor nocturnus is usually not associated with any psychiatric or neurological disturbance when it occurs in children. Its natural course is to disappear by adulthood, suggesting a disorder of maturation of the nervous system. When night terrors begin in adolescence or adulthood, there is a likelihood of psychological difficulties. Patients with night terrors may repress their anger or aggression and show elements of obsessive-compulsive and phobic behaviour as well as depression. Our patient has no difficulty expressing anger and does not have obsessive-compulsive behaviours. Rather, he has a brainstem lesion disrupting non-rapid eye movement (REM) sleep and resulting in night terrors. Furthermore, clonazepam, which suppresses slow wave sleep has decreased his night terrors while unmasking enuresis.

Pavor nocturnus is a consequence of partial arousal during non-REM slow wave sleep. During deep sleep, the brain continues to process subcortical sensory information such as respiratory awareness. With arousal from non-REM sleep, these sensory experiences may intrude into consciousness as single, brief, frightening experiences. Patients are terrified and may even injure themselves trying to escape the frightening image or feeling. In this patient with a brainstem lesion, the mechanism of slow wave arousal may be disruption of the nucleus of the solitary tract in the dorsum of the brainstem near the wall of the fourth ventricle. Activation of the midbrain, pons, and sleep may modulate the arousal properties of the ascending reticular activating system. Polysomnographic studies of patients with comparable brainstem lesions would help elucidate the pathophysiology of pavor nocturnus and the proposed role of the nucleus of the solitary tract.

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Ooctreotide—a new treatment for diarrhoea in familial amyloidotic polyneuropathy

Familial amyloidotic polyneuropathy (FAP) is a well known hereditary polyneuropathy which was reported for the first time by Corino de Andrade in Portugal. This disease generally begins with sensory symptoms and signs (50%) and sexual impotence in man (30%). A less significant number of subjects have constipation (20%), loss of weight (10%) or diarrhoea (10%) as an initial symptom. After two to five years of other dyssynergic symptoms are very common, namely orthostatic hypotension and severe diarrhoea.

In a group of 60 patients followed at the neurology outpatients, we found that half had regular diarrhoea which was particularly refractory in six. To control the diarrhoea we used a low fibre diet, antibiotics (tetracyclines and metronidazole), metoclopramide and loperamide.
We report the therapeutic results with Octreotide, a somatostatin analogue, in two subjects who became resistant to the usual medication. The trial arose from previous experience with diarrhoea, also a dysautonomic manifestation. In FAP we accept that there is a blind loop phenomenon with bacterial overgrowth and a mild malabsorption as in diabetic dysautonomic diarrhoea.

The first patient, a 53 year old man whose disease had advanced for years previously, had diarrhoea (7 times per day) for six months, orthostatic hypotension and loss of weight (3 kg). The other patient, a 43 year old man whose symptoms started seven years ago, had less severe diarrhoea (4 times per day) for two months sufficient to cause psychological disturbance and deterioration in orthostatic hypotension. The diarrhoea in these patients was painless and watery and the stools contained no blood, pus or mucus.

Octreotide 0-05 mg was administered by subcutaneous injection every 12 hours. The first patient was treated for 15 days which resulted in an improvement in his diarrhoea (3 times per day and stools semi-formed) and orthostatic hypotension; his weight also stabilised. The second patient was also given the same therapy for seven days. The diarrhoea became less frequent (twice per day) and the stools semi-formed; orthostatic hypotension also improved as did his psychological state. This patient reported mild nausea during the treatment. Each injection improved symptoms after 30-60 minutes and relief continued for 24-48 hours. After this period, the mild deterioration experienced in both patients was controlled with the usual therapy during the three to four months follow up.

Octreotide should be considered in the treatment of refractory diarrhoea in FAP.

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1 Andrade C. A peculiar form of peripheral neuropathy. Brain 1952;7:408-27.

Angiostrongylus cantonensis

I believe that the authors of a case report1 of Angiostrongylus cantonensis occurring in a brain abscess of a patient from India have misidentified the parasite and that this does not represent a case of human angiostrongyliasis but is in fact a case of sparganosis caused by a tapeworm parasite of the genus Sparganum. I suggest this alternative diagnosis for the following reasons:

1) Although A cantonensis infection is common in south east Asia, many parts of the Pacific region and other parts of the world, I know of no published reports of the parasite occurring in India although I have observed anecdotally that rat infections have been seen there. I do not believe any human infections have been previously reported in India.

2) The clinical picture described in the report is at variance with what we generally see in eosinophilic meningoencephalitis associated with A cantonensis infection. When the authors state “this case is unique in that the patient presented with focal neurological manifestations . . .” it agrees with a diagnosis of neurocysticercosis or sparganosis but not Angiostrongylus. Thus a brain abscess picture with a single focal lesion, no reported eosinophilia in blood or CSF, is far more consistent with the diagnosis I suggest.

3) Finally, the lesion pictured in figs 2 and 3, and in particular 3 which is supposed to be the actual worm, do not illustrate a nematode. The segment of the tapeworm is clearly seen in fig 3 and its histological features are consistent with a larval tapeworm and certainly have no features of a roundworm in section. I would suggest that at a slightly higher magnification calcareous corpuscles would be clearly visible in the segment which are unique to cestode tissue. You can barely make them out in fig 3 nor is it too low in magnification to be absolutely certain. The fact that the authors state that the worm was living and motile when the lesion was opened suggests sparganosis rather than cysticercosis. In addition there is no bladder visible which you see with cysticercosis.

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2 Purohit et al reply: It is not that human and rodent infections have not been reported from India. There are enough publications to suggest that this infection does occur in India.1,2 The absence of eosinophils can occur in parasitic infestations where the pathology is localised, for example, occurrence of abscess.

However, after reading the comments in the letter by Dr Ash and a publication by Chans et al3 we have reviewed the histopathology slides and have found no calcareous corpuscles in the tegument.

We have, however, found the following important positive features to suggest that the worm could be a sparganum: 1) Pseudo segmentations; 2) longitudinal excretory channels and 3) classical muscle bundles in the parenchyma.

Considering the possibility that the calcareous corpuscles must have dissolved during fixation the worm in question is most likely a sparganum and this case report of abscess by this parasite is definitely a very rare entity.

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Angiostrongylus cantonensis is a parasite that is commonly found in South East Asia, the Pacific region, and other parts of the world. However, there have been no published reports of this parasite occurring in India. The clinical picture described in the report is at variance with what we generally see in eosinophilic meningoencephalitis associated with Angiostrongylus cantonensis infection. The authors state that this case is unique because the patient presented with focal neurological manifestations. However, further studies are needed to determine the cause of this phenomenon and the precise localization of the responsible lesion.

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Hypergraphia associated with a brain tumour of the right cerebral hemisphere

We read with great interest the recent paper by Imamura et al on hypergraphia, namely the inextricably handicapped. We found thirty three patients with hypergraphia, of which 18 were male and 15 female. The ages ranged from 14 to 70 years (mean age 39.5) and the IQs ranged from 13 to 70 (mean IQ 29).

Five patients had temporal lobe epilepsy and thirteen others had a history of epilepsy. Six had superimposed psychiatric disorder (four with epilepsy) and other patients showed considerable behaviour disorder with aggressive outbursts as a prominent feature. There were also four patients with Down’s syndrome.

Further studies are needed to determine the cause of this phenomenon and the precise localization of the responsible lesion.

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Imamura et al reply: We thank Drs Jancar and Cooke for their comments. We did not cite several mass studies in our article, because we could not judge their hypergraphia as Waxman or Yamasdori type. It is possible that these studies included a third type of hypergraphia.

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