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A 27 year old man was hospitalised because of disturbed consciousness on 11 February 1985. He had had sore throat and a fever of 38°C for a week. On admission he was confused but followed simple commands. Bilateral abductor palsies, spasms of both masseters and the left orbicularis oris, and trismus were present. The deep tendon reflexes were bilaterally brisk with extensor plantar responses. He showed marked ataxia of the extremities. Meningeal signs were absent. On the second day, his extremities were markedly rigid, and dystonic postures, such as opisthotonus, were frequently observed. In addition to the bilateral impairment of upward and lateral gaze, conjugate downward spasmodic movements of the eyes (oculogyric crises) appeared. They consisted of slow tonic downward deviation of the eyes and dilatation of the pupils, each crisis lasting for several minutes (fig). They occurred spontaneously but could also be very easily induced by applying the noxious stimuli such as pinching. They were sometimes but not always accompanied by the exacerbation of the dystonic rigidity of the trunk and extremities. Adenine arabinoside 600 mg/day was started and continued for 10 days. Over the next three days he was stuporous, though trismus, masticatory and facial spasms, and dystonic rigidity subsided gradually. The initial hyperreflexia was replaced by areflexia. Tracheostomy was performed but artificial respiration was not required. On the seventh day, as his consciousness began to improve, he showed marked bulbar palsy and flaccid tetraplegia, though he communicated by blinking (locked-in syndrome).

Although the oculogyric crises also began to subside, they still occurred even at this stage; they finally disappeared on the eighth day. Rapid recovery followed, and his condition returned to normal by mid March. One year later he is well with no signs or symptoms of Parkinsonism.

Cerebrospinal fluid (CSF) obtained on the second day showed: xanthochromia; lymphocytosis, 60/mm³; total protein, 61 mg/dl; IgG, 5.2 mg/dl; and glucose, 86 mg/dl. Electroencephalogram showed marked slowing of the background activity. Brainstem auditory evoked potentials showed normal waves 1, 2, and 3, but waves 4 and 5 were absent bilaterally, suggesting a lesion in the upper pons. Computed tomographic scan and magnetic resonance imaging of the brain were unremarkable. Herpes simplex virus type 1 IgG immunofluorescence antibody titre in the CSF and serum were elevated to 1:2 and 1:160 respectively, both of which returned to normal later. Other viruses showed no remarkable elevation of the antibody titres. No virus was isolated from the throat and CSF.

Although the attack of dystonia and downgaze in the acute phase of this case might mimic what is sometimes called a pontine fit, the downgaze attack should be more accurately called the oculogyric crisis for two reasons: (1) the clinical feature was typical,¹ and (2) it was more prominent than dystonia and frequently occurred alone, independently of dystonia. The oculogyric crisis and dystonia are closely related with each other: both are known to be seen frequently in postencephalitic Parkinsonism

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Oculogyric crisis in acute herpetic brainstem encephalitis

Sir: So far oculogyric crisis has not been reported in acute herpetic brainstem encephalitis, although recently some 10 cases have been reported with its typical clinical features fully documented. We report a patient who showed this unique neurological symptom only at the onset of the disorder.

Figure Note the extreme downward deviation of the eyes. Each crisis lasted for five to ten minutes. The oculogyric crises occurred spontaneously (left), but were induced easily by applying the noxious stimuli such as pinching (right). (Informed consent was obtained from the patient for this photograph.)

and antipsychotic drug intoxication.^{1,2} Although oculogyric crises due to encephalitis other than von Economo's encephalitis have been reported sporadically, they are usually seen in the chronic phase of the illness.³ This case is quite unusual in that they occurred only at the onset of the acute brainstem encephalitis.

It is difficult to explain such an extreme degree of downgaze attack solely as a release phenomenon due to the impairment of upgaze: an irritative lesion near the downgaze centre or its pathways seems to be the more plausible explanation in this case. Although the precise anatomical localisation of the downgaze centre is still unknown, there is evidence that thalamic structures may exert some influence on downgaze, and that its pathways may run medial-dorsally to the red nuclei, more ventrocaudally than those for upgaze, in the midbrain.⁴

In this case, herpes simplex virus type 1 is suspected as the aetiological agent from the typical clinical features, the serological investigations, and the efficacy of adenine arabinoside. Recently acute herpetic brainstem encephalitis has been reported from all parts of the world.⁵⁻⁹ Although the similarity of Bickerstaff's encephalitis and acute herpetic brainstem encephalitis has been pointed out by some workers,⁶ the latter has some unique clinical features: symptoms suggesting the stimulative lesions in the brainstem such as trismus, masticatory spasm, facial spasm, blepharospasm, palatal myoclonus, and hiccup have been described only in acute herpetic brainstem encephalitis and not in Bickerstaff's encephalitis.^{5,7,9,10} Until the causal virus of Bickerstaff's encephalitis is identified, the relation between these two conditions will remain unclear.

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Palatal myoclonus influenced by head posture

Sir: Palatal myoclonus is a segmental myoclonus syndrome which is often set apart from other movement disorders because of its persistence during sleep and throughout the life of the individual.¹⁻³ Recently, however, Jacobs *et al* have reported the case of a 19-year-old man who, following a head injury, developed palatal myoclonus that disappeared during sleep and varied with intensity during the day depending on which way he turned his head.⁴ The following case illustrates that palatal myoclonus may be triggered by head movements and again highlights the fact that palatal myoclonus may be an intermittent phenomenon.

For over 2 years a 32-year-old female had noticed repetitive clicking noises in her head. These were audible to observers over 5 metres away but were present only when she tilted her head to the left or right. The intensity of the noises varied from day to day but could not be heard at all during ear or throat infections, to which she was especially prone. There were no other central nervous system abnormalities on examination, but inspection of the throat revealed irregular, bilateral, jumping movements of the palate and pharynx with her head in 45° lateral

flexion (left or right), but not at other times. A clicking noise synchronous with the myoclonic jerks could be heard at a rate of approximately 120 per minute. After a few seconds both the jerking and the clicking noises could be started again by repeating the movement of lateral flexion. The clicking noises were assumed to be arising from the pharynx or from opening and closing movements of the mouth of the Eustachian tube.⁵ CT brain scan was normal.

The mechanism of generation of palatal myoclonus is unknown but post-mortem studies have demonstrated trans-synaptic hypertrophic degeneration of the inferior olivary nuclei due to a lesion of one of its afferent inputs, the dentato-olivary pathway.^{6,7} The switching on of palatal myoclonus in our patient by certain head movements suggests that other afferent pathways to the inferior olivary nuclei may be important in modifying the expression of this movement disorder; in this case proprioceptive input via the cuneate nuclei may be pertinent.

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