Ocular flutter, postural body tremulousness and CSF pleocytosis: a rare post-infectious syndrome

Sir: The neurological sequelae of non-specific febrile illnesses include meningoencephalitis, myelitis and polyradiculitis. A rare, distinctive post-infectious syndrome comprising ocular flutter, body tremulousness and cerebrospinal fluid (CSF) pleocytosis has been described.1-3 Outcome is favourable but the infective agent and pathophysiology remain uncertain.

Three weeks prior to presentation a 31 year old man, accustomed to jogging 6 km per day, found himself unusually short of breath whilst running and experienced generalised myalgia, arthralgia and headache. One week later he developed an unsteady gait and became nauseated. When sitting or standing his trunk and limbs felt jittery and shaky. Sudden loud noises precipitated brief shock-like jerks of the limbs and trunk. His symptoms improved over the following week but subsequently recurred, prompting his admission.

On examination he was afibrile and abnormal findings were confined to the nervous system. Initially he was noted to have horizontal gaze-evoked nystagmus but after a few days ocular examination revealed sudden bursts of conjugate horizontal saccadic oscillations without intersaccadic interval (ocular flutter) occurring both spontaneously and with fixation, with eyes open and closed, during pursuit and saccades, and in all direction of gaze (fig). Down gaze was particularly provocative. The ocular excursion was full and optokinetic responses were normal bilaterally. When lying in bed limb ataxia was minimal and there were no involuntary movements. However, of standing, irregular jerks of the head, trunk and limbs were present. Standing on either leg accentuated the truncal tremulousness which was difficult to characterise. Anxiety, chorea, myoclonus and truncal ataxia were all considered by different examiners. The absence of involuntary movements when lying contrasted with the disorder seen when assuming the upright posture. Palatal myoclonus was not observed.

Cerebrospinal fluid (CSF) examination revealed 8 lymphocytes per μl, protein 0.28 g/l (normal 0.15-0.45), glucose 3.9 mmol/l (normal 2.8-4.4) and IgG albumin ratio 0.24 (normal 0.04-0.24). Electrocardiograph showed first degree heart block and anterolateral T wave inversion. Electroencephalograph was normal but irregular eye movement artefact was present throughout the recording, more obvious with the eyes closed, and suggestive of ocular flutter or opsoclonus. Attempts

Fig Electro-oculography, looking down. Burst of conjugate horizontal saccadic oscillations without intersaccadic interval (ocular flutter).
isolate virus from blood, CSF and urine failed. Full blood count, erythrocyte sedimentation rate, chest radiograph, visual evoked potentials, brain stem auditory evoked potentials, and cranial CT scan were normal.

Gradual resolution of symptoms and abnormal findings occurred over the following 6 weeks. In the 1960’s Smith and Walsh and later Cogan, described a benign encephalitis with ocular oscillation and truncal ataxia.1-3 Following a prodrome of malaise and mild fever, such patients developed ocular flutter or opsonolus and shivering movements of the head and body. Cerebellar and long tract signs also occurred in some patients but the sensorium usually remained clear. Spinal fluid protein and cell count were often elevated. The illness resolved, usually within a few weeks or months, although the course was occasionally protracted, especially in children (Kinsbourne’s myoclonic encephalopathy with “dancing eyes and dancing feet”).4

The movement disorder of the trunk was precipitated by sitting or standing, particularly on one leg, and by loud noises, but did not occur whilst lying. The initial suggestion by some observers that the patient was either very anxious or hysterical has been noted in other case reports.1 3

The prodromal systemic symptoms, CSF pleocytosis and temporal profile, suggested that the neurological disorder was a sequel to infection, although no viral or bacterial agent was isolated. If so, it probably represented a late or delayed phenomenon occurring after the acute stage of infection at a time when the organism could not be isolated.

The possible pathophysiological substrate for opsonolus and ocular flutter is probably related to a disorder of the inhibitory control of saccadic burst neurons by pontine pause cells.5 Baringer postulated that the postural tremulousness was a result of a post-infectious lesion affecting the cerebellar vermis.5

The self limited course in all reported cases has precluded any clinicopathological correlation. Awareness of this easily recognisable syndrome may provide the opportunity to further study its possible pathogenesis. Recognition, in any case, provides reassurance for the patient and attending clinician.

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References

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Two cases of influenza B encephalitis
Sir: It is a common belief that “influenza” is often associated with neurological sequelae. Osler1 has been quoted as saying “Almost every form of disease of the central nervous system may follow influenza”. He made this assertion before respiratory viruses had been classified, and before virological proof was possible. We report two cases of severe encephalopathy associated with influenza B infection. We are not aware of any other well documented cases of encephalitis associated with influenza B.

Five days after admission to a psychiatric unit for treatment of an anxiety state a 37 year old housewife developed a fever of 39°C, mild neck stiffness and became disorientated. Two days later her condition deteriorated, and she was transferred to a neurological ward. She could not speak, did not respond to commands, reacted in a semi-purposeful manner to painful stimuli and had marked neck stiffness, with Kernig’s sign positive. The pupillary reflexes were normal. There was no papilloedema. Oculocephalic responses were normal. Muscle tone and tendon reflexes were generally increased, with extensor plantar responses. There was no obvious sensory loss in any limb. EEG showed loss of alpha rhythm with widespread delta activity with a tendency to periodicity. Computed tomography (CT) and isotope brain scans were normal. Chest radiograph showed patchy pneumatic changes. Lumbar puncture was traumatic. The CSF contained 6,700 red blood cells (rbc) and 43 lymphocytes per µl; sugar 2.7 mmol/l with a corresponding blood sugar of 7.6 mmol/l protein g/l. CSF repeated 7 days later contained 584 rbc, 75 lymphocytes/µl; protein was 0.32 g/l and electrophoresis showed oligoclonal bands. Eight days after her initial fever she required assisted ventilation for six days. She developed status epilepticus and frequent myoclonic jerks. She was treated initially with dexamethasone and anticonvulsants. She also required antibiotics for repeated chest infections due to Staphylococcus aureus.

After about four weeks she became more alert, but remained rigid, with periods of myoclonus. She was treated with a levodopa preparation without benefit. She was allowed home after four months. Able to walk with the aid of one person but slightly confused. One year after discharge she was free of fits and myoclonus. She now lives an independent life. There is no evidence of residual intellectual deficit. Five days after the onset of the fever the complement fixation titre of antibody to influenza B was <1:10; 16 days later it was 1:320. There was no rise in complement fixation antibody titre to mumps, measles, herpes simplex, varicella zoster influenza A virus or Mycoplasma pneumoniae. Using influenza B/Singapore 222/79 as antigen in the haemagglutination inhibition test, the patient’s sera showed a rise of antibody from <1:10 to 1:80. Viral CSF was not isolated from acute phase faeces of CSF in baboon kidney or HEp2 cells.

A 38 year old housewife had a four day prodromal illness of headache and generalised muscle aches, then became pyrexic, confused and dysphasic with mild weakness in her right arm. Two days later she had a solitary generalised convulsion, developed mild neck stiffness and became stuporous. Her temperature was 38.5°C. She responded to painful stimuli only. (There was no verbal response). There was no papilloedema. The pupillary responses were normal. The eyes were deviated to the left. The right arm was flaccid with diminished reflexes. The plantar responses were equivocal. The CSF was normal in all respects. CT brain scan was normal and the EEG showed extensive low frequency activity, maximal in the left posterior temporal region. A provisional diagnosis was made of herpes simplex encephalitis. Treatment was started with acyclovir 500 mg tds IV and dexamethasone. The next day she was responding to questions, two days later she was orientated in time and place. There was a slight dysphasia and she was able to move her limbs nor-