



Sir,

Opsoclonus-myoclonus syndrome (OMS) is a rare but distinctive disorder, characterized by irregular, continual, and conjugated chaotic saccades of the eyes accompanied by appendicular myoclonus and truncal ataxia. No etiological factor can be found in 50% of adult cases. In 20% of cases it may be paraneoplastic and may also be a manifestation of brainstem stroke or infection. The most common tumors, which give rise to OMS are breast and lung cancers and infectious agents are Epstein-Barr virus and enteroviruses such as Coxsackie B.^{1,2}

A 41-year-old woman presented with severe vertigo and gait unsteadiness for ten days. There was no history of any antecedent viral infection or vaccination. Neurological examination revealed coarse eye movements with severe truncal ataxia and limb myoclonus. Laboratory examination (routine hematological tests, urine and cerebrospinal fluid examination, serological tests for infections, oncological bands work-up, magnetic resonance imaging of brain and electroencephalography) revealed no abnormality. As assays for paraneoplastic antibodies couldn't be performed due to technical limitations, the patient was considered to be a case of idiopathic OMS.

Therapeutic trial with prednisolone (1 mg/kg/orally) and clonazepam (4 mg/day) was attempted. Because of the lack of any beneficial effect, intravenous immunoglobulin (IVI g) (0.4 g/kg/day for five days) was added on the sixth day and Prednisolone and clonazepam were tapered in the following five days. On the third day of IVI g treatment, muscle jerking and eye movements stabilized. She began to sit without assistance and during the next two weeks the patient gradually recovered. She was symptom free within eight weeks. Physical symptoms and repeated laboratory examinations for occult malignancy after one-year interval was negative. She has been asymptomatic for two years.

The response to immunotherapy is unclear in paraneoplastic and idiopathic OMS, because the rarity of this disorder hinders controlled clinical studies and the possibility of spontaneous remissions makes interpretation difficult in isolated cases. In adults under 40 years of age, the clinical evolution is more benign and the effect of IVI g seems more effective.²

IVI g is an immunomodulating agent that has multiple activities, including neutralization of pathogenic autoantibodies, suppression of inflammatory mediators, complement inactivation, functional blockage of Fc receptors on macrophages and modulation of T-cell functions.³ Treatment with IVI g has been reported in a few idiopathic adult-onset OMS cases in literature. Pless et al. first reported beneficial effects of IVI g in 1996.⁴ Bataller et al analyzed 10 idiopathic and 14 paraneoplastic adult OMS patients. IVI g was used in five of the idiopathic cases. They concluded that idiopathic OMS presents an age dependent prognosis and immunotherapy seems to be associated with a faster recovery.²

Our case suggests that patients with idiopathic OMS treated with IVI g may have a faster recovery and this treatment should be recommended in cases with severe neurological dysfunction.

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Metastasis of frontal oligoastrocytoma to cerebellar vermis

Sir,

Metastasis of a low-grade glioma through the cerebrospinal fluid (CSF) is very rare. We report a case of CSF spread of a right frontal oligoastrocytoma to cerebellar vermis.

A 46-year-old woman presented with symptoms of raised intracranial pressure and difficulty in walking of two months duration. On examination, she had bilateral papilloedema and cerebellar signs on the left side. Four years earlier she had undergone excision of right frontal mixed oligoastrocytoma (WHO Grade 2) followed by 54 Gy of Cobalt teletherapy to the brain. Magnetic resonance imaging (MRI) of the brain showed a 2 cm homogeneous contrast enhancing mass in the inferior vermis with extension into the left cerebellar hemisphere and hydrocephalus. There were postoperative changes in the right frontal lobe in the form of loss of brain tissue and there was no evidence of recurrence (Figures 1a and 1b). A differential diagnosis of metastases from a systemic tumor; CSF spread of the frontal glioma or radiotherapy-induced tumor were considered. On referring to her previous radiotherapy records it was found that the posterior fossa was not included in the radiation field. Chest X-ray and an ultrasound abdomen were normal. She underwent right ventriculoperitoneal shunt followed by midline suboccipital





Letter to Editor

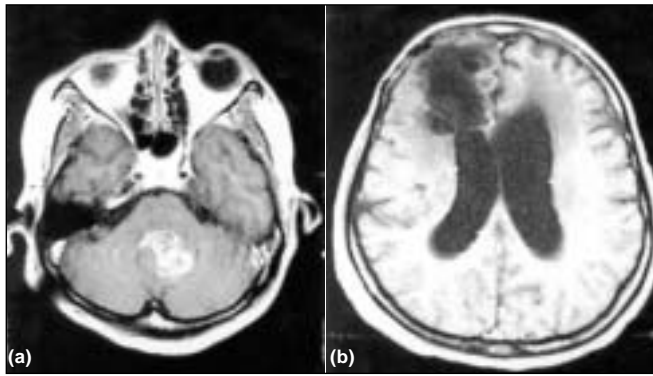


Figure 1: (a) T1W axial post gadolinium MR image showing a 2 cm enhancing mass in the vermis with mass effect on the fourth ventricle. (b) T1W axial post gadolinium MR image showing postoperative changes in the right frontal region. There is no evidence of recurrence. There is hydrocephalus

craniectomy and radical excision of the mass. The biopsy was reported as oligoastrocytoma (WHO Grade 2). She was given 56 Gy of external radiotherapy.

Choucair et al, in a large study, have reported the frequency of CSF metastases at 5-7%.¹ Compared to malignant primary tumors of the central nervous system low-grade gliomas are rarely known to metastasize through the CSF. In 1929, Cairns² reported the first case of CSF spread of an oligodendroglioma through the ventricles. In our case, similar histology of the frontal and vermian masses, rarity of primary vermian mixed oligoastrocytoma and the fact that the vermis was not included in the radiation field after the first surgery suggest that the vermian mass is likely to be due to CSF spread. The possible mechanisms of tumor dissemination include exfoliation of cells into the CSF of the ventricles or the subarachnoid spaces. Previous craniotomy procedures, shunt procedures, site of tumor, glioma type and tumor cell differentiation are known to affect CSF spread of gliomas.³⁻⁵ Oligodendrogliomas are more commonly associated with CSF dissemination. Friedberg et al⁶ have proposed CSF analysis of specific matrix metalloproteinases profile as a means to detect CSF spread of tumors.

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Fulminant subdural empyema-an unusual complication of pyogenic meningitis

Sir,

An otherwise normal 56-year-old diabetic patient presented with a one-day history of multiple generalized tonic-clonic seizures followed by altered sensorium. There was no history of fever. There was no history of trauma or any focus of infection. There was no focal neurological deficit or signs of meningitis.

Hematological investigation revealed leucocytosis with a total white blood cell count of 16,600/cu.mm and an ESR of 45 mm. Blood sugar was 351 mg%. Computed tomography (CT) scan of the brain revealed no abnormality (Figure 1). The lumbar CSF analysis revealed 1350 cells/mm with 96% neutrophils and 04% lymphocytes. Blood and CSF cultures did not reveal any growth. A diagnosis of pyogenic meningitis with diabetes was considered and the patient was placed on broad-spectrum antibiotics. Two days after admission to the hospital she developed recurrent attacks of seizures, lapsed into altered sensorium and developed a left pupillary dilatation. Repeat CT scan revealed a left fronto-parietal hypodense, extracerebral fluid collection with severe brain edema causing midline shift and obliteration of the basal cistern (Figure 2). An emergency left frontal burr hole was done and thick pus was evacuated. Gram's stain revealed pus cells and gram-nega-

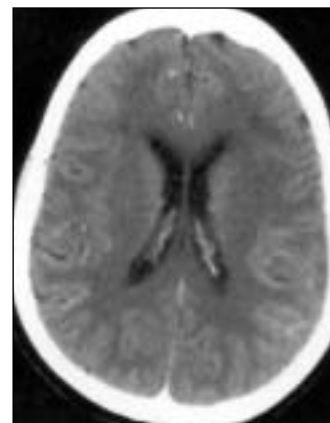


Figure 1: Plain and contrast CT scan of the brain done on 07/11/2003 shows no significant abnormality

