malities; in particular there was no arachnoid cyst. Consequently, a third blood patch (20 ml) was carried out at a higher level (L2) than the first two, guided by the radionuclide cisternography findings. The patient was headache free a few minutes later and able to ambulate the next day. Postural headache had not recurred within 12 weeks of follow up and the patient returned to work full time. Follow up brain MRI with gadolinium showed complete resolution of the subdural hygroma but persistent enhancement of the subdural punctional headache, an epidual epidural patch of cerebrospinal fluid is often used.14 Pain relief is often immediate but sometimes several blood patches are needed to be effective. It is not unusual to recommend that the precise level of the leak is identified with contrast or isotope cisternography in patients in whom blood patch therapy has failed. In conclusion, SIH features are well established. Misleading MRI findings should suggest the diagnosis, and a search for a cryptic CSF leak should be considered in patients with unexplained postural headache, even when treatment has failed. It is important to bear in mind that several blood patches are often needed.

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Transient multiple cranial nerve involvement as a first sign of macrophage activation syndrome

In 1979, Risdall et al1 described a disorder characterised by histiocytic hyperplasia, preponderant hemophagocytosis, and proliferation of macrophage cells. The term macrophage activation syndrome was proposed. Our patients are heterogenous and not specific.2 Neurological signs are rare. We report a man with macrophage activation syndrome in whom the first signs were multiple cranial nerve involvement. A 43 year old man developed left peripheral facial nerve palsy. Three days later, intrabucal dysesthesia associated with hypoesthesia in the second branch of the trigeminal nerve occurred. Eight days later he developed left hypoacusia and a right T8 band of hypoesthesia. At this point, laboratory tests, CT, and cerebral MRI were normal; a CSF examination disclosed normal protein concentration and 10 lymphocytes/μl. In the third week, the signs disappeared, but left optic neuritis occurred. Antibacterial and antiviral, including HIV, serologies were negative except for Epstein-Barr virus suggesting a latent infection. Although Borrelia serology was negative, ceftarixone (2 g/day) was given for three weeks. Two months after the onset of the symptomatology, left eye vision had improved but right optic neuritis appeared. He lost 8 kg over two months; his temperature was 38°C. Neurological examination showed only a bilateral asymmetric visual loss. Laboratory test results were white blood cells 6-4 ×109/l, haemoglobin 13-4 g/dl, platelets 200 ×109/l, fibrinogen 2-30 g/l. Erythrocyte haemolysis was normal. No other abnormalities were found. It is unusual to find significant abnormalities in patients with macrophage activation syndrome. In this case, a CSF leak was found. This CSF leak could be related to a macrophage activation syndrome.
tion syndrome has a poor prognosis, with a 50% mortality rate. The nervous system is seldom involved in the syndrome. If such involvement appears, it usually does so towards the end of the course of the disease. A patient with sensorimotor neuropathy related to axonopathy and occasional demyelination has been recently reported, but in the context of a fulminating illness.1

The distinctive feature of our finding is the occurrence of transient cranial nerve involvement as the probable first sign of macrophage activation syndrome. It could be claimed that the symptomatology is related to the lymphoma. However, very little is known about neurological complications in T cell lymphoma, and their occurrence is probably rare.2 Kaufman et al3 have reported an involvement of the nervous system in 14 patients out of 104 cases, eight being related to direct complications. In only one patient, palsy of the sixth cranial nerve was the first sign. Neurological signs occurred between 10 and 102 weeks after diagnosis of lymphoma.

If polyneuropathy occurs in T cell lymphoma it is due to infiltration and the clinical evolution is usually stereotyped with slowly evolving sensorimotor signs.4 Because there is no postmortem examination in our case, infiltration of peripheral nerves cannot be eliminated; but it is unlikely, considering the improvement in neurological signs. Meningoradiculitis could be evoked, but if that were so, there would have been a worsening of the initial signs.5 Moreover, CSF examination and cerebral MRI were normal. All these indications lead us to suggest that the neurological signs in our patient could be related to a remitting-relapsing neuropathy due to non-cureaneous T cell lymphoma infiltrating peripheral nerves, to vasculitis or, more likely, to the neurotoxic effects of cytokines. Cytokines, especially TNF, are secreted in large amounts in macrophage activation syndrome, and TNF can induce general side effects and cerebral damage.6

We are grateful to L. Rose for her help with the English version of this paper.

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Figure 2. Lymphoid cells with atrophophil granules. Myelogram (originally × 100).

Paraneoplastic opsonoclonus associated with cancer of the gall bladder

Opsonoclonus is an ocular dyskinesia consisting in ample, conjugated, arrhythmic, multi-directional ocular movements which persist even with the eyes closed.7 This syndrome has been described during the course of different cancers.8 In infancy, neuroblastoma is the cancer most often associated with opsonoclonus (in 2% to 7% of the cases).9 In adults, opsonoclonus is less common. Nevertheless, it is associated with a tumour in 20% of cases.10

Here, we report a case of opsonoclonus associated with a cancer of the gall bladder.

A 72 year old, treated hypertensive woman, experienced the sudden onset of vertigo followed by impaired consciousness. At initial examination her Glasgow score was 13. She showed opsonoclonus associated with a bilateral kinetic cerebellar syndrome. The cranial nerves were intact and there was no sensory or motor deficit. Complete physical examination only showed conjunctival icterus.

Brain MRI showed a left frontal angiomia measuring 7 mm in diameter without any impingement on cerebral parenchyma; the brain stem was normal. Two spinal taps were normal. A chest radiograph was normal. Laboratory studies showed an increase in alanine aminotransferase (43 IU), y-glutamyl transpeptidase (100 IU/L), and CA 19-9 (111 kU/L). Abdominal ultrasound showed a heterogenous, polyloid tumorous structure in the gall bladder associated with hepatocholecystic lesions in the liver and a thrombosis of the portal vein. Abdominal CT disclosed thickening of the left lateral wall of the gall bladder, liver metastases, and hilar adenopathy. Liver biopsy showed a sclerosing, necrotic-solidly replaced and necrotic carcinoma most suggestive of a pancreatico-biliary origin.

Tests for anti-Hu, anti-Ri, and anti-Yo antibodies were negative. Immunoglobulin IV (0-4 g/kg/day) and cortisone (Solumedrol, 0-5 g/day for five days) was ineffective. The patient died five weeks later. No necropsy was performed.

The diagnosis of opsonoclonus remains clinical. It usually has an abrupt onset.1 It is probably the result of a dicephalic or mesencephalic lesion with production of the abnormal movement by removal of normal saccadic generator inhibition.12 Dysfunction of the pause neurons, which play a part in inhibiting the phasic neurones responsible for the appearance of jerks is likely. The frontal angiomia does not explain the opsonoclonus.

The opsonoclonus was considered paraneoplastic because it was not associated with an infectious or tumoral aural lesion. Only possible causes (toxic, metabolic, degenerative, and vascular)13 were excluded.

The normal MRI, lumbar punctures, and the absence of anti-Ri antibodies, which have been associated with paraneoplastic opsonoclonus occurring with carcinomas of the breast,14 did not cast doubt on the diagnosis. Breast cancer and small cell cancer of the lung represent 70% of reported cases associated with opsonoclonus in adults15 and are sometimes discovered during necropsy.16 In the present case, the histological differentiation seen during liver biopsy was strongly suggestive of a primary lesion in the gall bladder. Therefore, it is not likely that the lesions discovered were metastases from one of the above cited cancer localisations.

The possibility of this association means that the gall bladder should be included in the investigations of a paraneoplastic opsonoclonus.
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*J Neurol Neurosurg Psychiatry* 1997 62: 292-293
doi: 10.1136/jnnp.62.3.292