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 previous procedure, in the hope that this might help the radiouclide 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 persisted the meningeal enhancement. However, its intensity was smaller than that seen on the MRI at admission.

Intracranial hypotenion associated with postural headache is a common clinical phenomenon after lumbar puncture, but it may also occur spontaneously, 1 a rarely reported syndrome. 2 Our patient fits well with the typical description of SIH—namely, spontaneous frontal headache exacerbated by erect posture and relieved by supine position. It may be associated with nausea and vomiting; less commonly, patients experience vertigo, diplopia, photophobia, or neck stiffness. 3 In this patient, examination was unremarkable, with no alteration of the patient’s headache may include lumbar puncture that may allow measurement of the CSF pressure. This was not performed in our patient but it is usually low, less than 10 cm H2O. Because the CSF in SIH sometimes contains an abnormal number of white blood cells, or high protein content, or both, 4 suspicion of meningeal infection or tumour may lead to further evaluation, including contrast enhanced MRI. Several recent reports 5-10 have shown subdural hygromas and diffuse meningeal enhancement on MRI with gadolinium associated with SIH.

In all reported cases, the enhancement disappeared as clinical symptoms resolved. This may be related to infiltration of the pachymeninges secondary to reversible disturbance of the choroid plexus, 11 a dural venous dilatation in response to low CSF pressure, 12 or to a fibrocollagenous proliferation of the leptomeninges. 13 The last is consistent with our finding because of the persistence of symptoms for months after disappearance of clinical symptoms. Such an enhancement can lead to diagnostic confusion. However, the clinical presentation should be sufficient to make the distinction in most cases.

Spontaneous spinal CSF leaks are now increasingly recognised as a cause of postural headache associated with intracranial hypotenion. 14 However, in most of the cases, the site of the CSF leak may be difficult to determine and will remain unknown. 15 It has been reported that small dural tears can occur at the nerve roots from even minor activities, and are often a history of a trivial fall, vigorous exercise, or violent coughing preceding the onset of the headache in many cases of SIH. 16 The sneezing and the altitude with possible degussactivities may have been a promoting circumstance for our patient. Radionuclide cisternography has been used to visualise CSF leaks in SIH, 17 showing a layer of radiotracer in the bladder and less activity than expected over the cerebral convexities, suggestive of unusually rapid uptake of the tracer into the circulation. 18 This characteristic finding is the diffusion of tracer into the extra-arachnoid space, outlining the spinal roots.

The headache in SIH usually resolves spontaneously with strict bed rest, some-times with analgesics and antiemetics. More aggressive treatment may be necessary when the headache persists or is incapacitating. According to some authors, epidural puncture headache, an epidural pseudocyst in the epidural space, is a frequent finding. It is not always recommended 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. This 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 al 1 described a disorder characterised by histiocytic hyperplasia, preponderant hemophagocytosis, and proliferation of macrophage cells. The term macrophage activation syndrome was proposed. Others terms are paraneoplastic 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, intrabuccal dysaesthesia associated with hypoesthesia in the second branch of the trigeminal nerve occurred. Eight days later he developed left hypoaesthesia and a right T8 band of hypoesthesia. At this point, laboratory tests, CT, and cerebral MRI were normal; a CSF examination showed 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, ceftriaxone (2g/day) was given for three weeks. Two months after the onset of the symptoms, 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 haemoglobin was normal. Some systemic lupus erythematosus, rheumatoid arthritis serologies, and biological variables of sarcoidosis were all negative. A second CSF examination and another cerebral MRI with gadolinium injection were normal. A few days later an erythematous skin rash occurred on his legs; biopsy showed leukocyto-tlastic vasculitis. His fever increased and corticosteroid therapy was started. Temperature returned to normal. However, in the third month fever reappeared, with weight loss and liver failure. Pancypentria appeared, platelets decreased to 30 × 109/l, white blood cells to 3.5 × 109/l, haemoglobin to 8.1 g/dl, and triglyceridaemia increased to 7 mmol/l. Serum tumour necrosis factor (TNFα) concentrations were six times higher than normal. Finally, bone marrow aspiration established the diagnosis of macrophage activation syndrome. Macrophage cytoplasmi contained erythroblasts, neutrophils, and platelets (fig 1). Atypical lymphocytes were also present, (fig 2), suggesting myeloma, and lymphoma. This was confirmed by marrow biopsy. Subsequently, hepatomegaly and splenomegaly occurred. Despite aggressive therapy with bolus steroids, the patient died from myocardial incompetence and bilateral lung infiltration. Postmortem examination was not performed.

Diagnosis of macrophage activation syndrome may be evoked in the presence of heterogenous non-specific clinical symptoms and biological anomalies. Asthenia, weight loss, sweating, and fever can be the first signs of the disease. 3 But what clearly indicates macrophage activation is the combination of patients with unexplained postural headache. This cranial onset is a rapidly deteriorating clinical status. Pancypentria, arthrogenic anemia, severe thrombocytopaenia, hypertriglyceridaemia and hypoaesthesia usually occur. 4 Diagnosis is established by bone marrow examination showing the most characteristic signs—that is, clearly differentiated macrophages presenting abnormal signs of active haemophagocytosis. Of note, all clear aetiopathogenic explanation exists for the occurrence of macrophage activation syndrome but it can occur after infections (Epstein-Barr virus, HIV, bacterial and parasitic pathogens), 5 in patients with an underlying disease: immune deficiency, solid tumours, inflammatory diseases, haemopathies, 6 lymphomas. 7 Macrophage activation syndrome was not seen in this patient. In the absence of a clear explanation, further examination was performed. 

Figure 1 Macrophages containing numerous erythroblasts and platelets. Myelogram (originally 400).
Figure 2  Lymphoid cells with azurophil granules. Myelogram (originally × 100).

Paraneoplastic opsonoclonus associated with cancer of the gall bladder

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

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 angiomata 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 transferase (100 IU/L), and CA 19-9 (111 kU/L). Abdominal ultrasound showed a heterogenous, polypoid tumour structure in the gall bladder associated with hyperchoic lesions in the liver and a thrombus 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 florid, mononuclear infiltrate of undifferentiated adenocarcinoma most suggestive of a pancreato-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. It is probably the result of a diencephalic or mesencephalic lesion with production of the abnormal movement by removal of normal saccadic generator inhibition. Dysfunction of the pause neurons, which play a part in inhibiting the phasic neurons responsible for the appearance of jerks is likely. The frontal lobe angiomata does not explain the opsonoclonus.

The opsonoclonus was considered paraneoplastic because it was not associated with an infectious or tumoral cholinergic lesion. Other possible causes (toxic, metabolic, degenerative, and vascular) 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, 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 adults and are sometimes discovered during necropsy.

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 investigation of a paraneoplastic opsonoclonus.

Lethal hyperoral behaviour from the Klüver-BucY syndrome

Clinicians have not sufficiently appreciated the danger of hyperoral behaviour in neurological disorders. A man aged 40, who exhibited this hyperoral behaviour is the Klüver-BucY syndrome. Originally described in monkeys after anterior bitemporal lobectomies, this syndrome includes indiscriminate dietary behaviour and a tendency to examine objects by mouth. The complete syndrome also results in placidity, hypersexuality, hypermetamorphosis or a tendency to attend to any visual stimulus, and visual agnosia. Both reports refer to patients with the Klüver-BucY syndrome who died as a consequence of their hyperoral behaviour.

Patient No 1 was a 40 year old man with epilepsy who developed persistent hyperoral behaviour after prolonged status epilepticus lasting several hours. On resolution of the seizures and recovery of consciousness, he

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