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 fulminant 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 al.3 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 polynephropathy occurs in T cell lymphoma, it is due to infiltration and the clinical evolution is usually stereotyped with slowly evolving sensorimotor signs.2 Because there is no sign of polynephropathy 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.3 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-cutaneous 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.3

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Paraneoplastic opsonoclonus associated with cancer of the gall bladder

Opsonoclonus is an ocular dyskinesia consisting in amine, conjugated, arthritides, multi- directional ocular movements which persist even with the eyes closed.1 This syndrome has been described during the course of different cancers.2 In infancy, neuroblastoma is the cancer most often associated with opsonoclonus (in 2% to 7% of the cases).3 In adults, opsonoclonus is less common. Nevertheless, it is associated with a tumour in 20% of cases.4

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 angioma measuring 7 mm in diameter without any impingement on the 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/L), y-glu- tamyl transpeptidase (100 IU/L) and CA 19-9 (111 kU/L). Abdominal ultrasound showed a heterogenous, polyploid tumorous structure in the gall bladder associated with hypochogenic 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 severe phagocytosis, monocytic differentiation and scattered adenocarcinoma most suggestive of a pancreaticobiliary origin.

Tests for anti-Hu, anti-Ri, and anti-Yo antibodies were negative. Immunoglobulin IV (0-4 g/kg/day) and cortison (Solumedrol, 0-5 g/day for five days) was ineffective. The patient died five days 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 diencephalic or mesencephalic lesion with production of the abnormal movement by removal of normal saccadic generator inhibition.2 Dysfunction of the pause neurons, which play a part in inhibiting the phasic neurons responsible for the appearance of the jerks is likely. The true face angina does not explain the opsonoclonus. The opsonoclonus was considered paraneoplastic because it was not associated with an infectious or tumor-related hyperergic syndrome. Other possible causes (toxic, metabolic, degenerative, and vascular)5 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,6 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 adults7 and are sometimes discovered during necropsy.8 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 excluded 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 hyperal hyperal not in the Klüver-Bucy syndrome

Clinicians have not sufficiently appreciated the danger of hyperal not in the Klüver-Bucy syndrome. A clear-cut clinical syndrome of hyperal not behaviour is the Klüver-Bucy syndrome.1 Originally described in monkeys after anterior bitemporal lobectomies, this syndrome includes indistinguishable dietary behaviour and a tendency to examine objects by mouth.2 The complete syndrome also results in placidity, hypersexuality, hypermetamorphosis or a tendency to attend to any visual stimuli, and visual agnosia. We report two patients with the Klüver-Bucy syndrome who died as a consequence of their hyperal not behaviour.

Patient No 1 was a 40 year old man with epilepsy who developed persistent hyperal not behaviour after prolonged status epilepticus lasting several hours. On resolution of the seizures and recovery of consciousness, he

Figure 2. Lymphoid cells with azurophil granules. Myelogram (originaly × 100).