coeruleus, but not in the dorsal vagal nucleus, nucleus basalis, cerebral cortex, spinal cord, superior cervical sympathetic ganglia, and ciliary ganglia.

The second case was a nine year old boy presenting with intellectual deterioration and an epileptic seizure. A month later, when admitted to Hôpital Ste-Justine, Montreal, he was cooperative and able to communicate, his affect was inappropriate and there was mild oro-facial chorea. The left planter was extensor and his gait ataxic. He developed generalised myoclonus with occasional opisthotonus, and subnormal decorticate posturing. Serum measles virus titre was 1024. CSF protein was 0.54 g/l, with no cells, and the measles complement fixation titre was 128. An EEG was dominated by low voltage slow and periodic high voltage slow-wave complexes. His condition stabilised, but he remained bed-bound and died at the age of 19 years. The brain showed generalised atrophy, with profound neuronal loss and gliosis in the cerebral cortex and hippocampus with a few microglial nodules and tangles. Some remaining neurons showed intranuclear inclusions. All central grey nuclei showed severe neuronal loss with glial and microglial reaction and several tangles. There was severe neuronal loss in the substantia nigra and locus coeruleus with tangles and Lewy bodies (fig). Lewy bodies were also found in the dorsal vagal nucleus, cerebral cortex, spinal cord or autonomic nervous system.

These patients with SSPE showed long survival of 13 and 10 years respectively. Tangles occur even in patients dying at a young age,1 but Lewy bodies have not been described in other cases. They were present in the substantia nigra and locus coeruleus, but not in other areas usually susceptible to Lewy bodies. We know of a third case of SSPE with onset at 21 years, and a nine year survival. In this case there were very few nerve cells in the substantia nigra, and some pale bodies, which are normally seen in Lewy body diseases.4

Apart from the Lewy body–Parkinson’s disease spectrum Lewy bodies are confined to a small group of rare degenerative disorders.1 They may be by-products of attempted regeneration, rather than degeneration, thus explaining their occurrence in long-standing cases of SSPE showing periods of relative stability. SSPE is a destructive inflammatory process due to persistent viral infection, but the aberrant immunological mechanisms are not fully understood. Although there is no direct evidence for an infectious etiology for Parkinson’s disease, there are parallels with SSPE which results from an infectious agent acquired in early life, leading to a progressive disorder associated with disconordance in identical twins,1 and Lewy bodies pathologically.


Combined neuroleptic malignant syndrome and the central anticholinergic syndrome

The neuroleptic malignant syndrome (NMS) is an idiosyncratic reaction to neuroleptic drugs characterised by encephalopathy, rigidity, dysautonomia and hyperthermia.1 Dopamine receptor blockade appears central to the pathogenesis of rigidity,2 and possibly the encephalopathy. Because cholinergic receptor blockade can also cause encephalopathy, the differential diagnosis of NMS includes the central anticholinergic syndrome (CAS).3 The central anticholinergic syndromes, which are used to treat Parkinsonism associated with neuroleptic use, are often used to test the rigidity in NMS.3 However, anticholinergic syndromes could theoretically exacerbate the encephalopathy of NMS, possibly spawing the development of combined NMS and CAS. We report the first case of combined NMS and CAS.

After four months of lithium therapy, a 28 year old man with a suggestive disorder was given 15 mg of daily haloperidol. One week later he was brought to the emergency room confused, agitated and hallucinated. Neurological examination disclosed a fever of 101°F, cogwheel rigidity, resting tremor, and dystymria. Serum glutamic oxaloacetic transaminase 128 IU/l (normal 5-35), serum glutamic pyruvate transaminase 65 IU/l (normal 5-30), lactic dehydrogenase 373 IU/l (normal 90-220) were all increased in concentration. His creatinine phosphokinase was 7700 IU/l (normal 25-145). All other laboratory studies, including computed tomography of the brain and lumbar puncture were normal. The haloperidol and lithium were stopped.

Treatment included levodopa/carbidopa 25/250, and 4 mg of intramuscular benzhexol. Twenty hours after receiving the benzhexol, his mental status deteriorated from being agitated and confused to coma, and his temperature increased from 101°F to 104°F. The rigidity was unchanged, but he developed urinary retention, decreased bowel sounds and large pupils, suggesting anticholinergic toxicity.

Six mg of intravenous physostigmine was administered. Within minutes he was able to follow simple commands. His temperature decreased from 102°F to 99.5°F over the next hour.

The differential diagnosis of NMS includes CAS. In a recent review, Guze and Baxter state that a response to physostigmine is sufficient to differentiate between NMS and CAS.1 However, this case suggests that the two disorders can coexist in the same patient. Furthermore, since drugs that cause NMS have anticholinergic properties, cholinergic blockade by neuroleptic drugs may make patients more susceptible to CAS from even low doses of anticholinergic agents.

Two points are illustrated by this unusual case. First, anticholinergic agents, which do not have demonstrable efficacy in NMS are probably contraindicated in NMS. Second, patients with NMS should be carefully monitored for signs of anticholinergic toxicity.

DAVID A BENNETT
Rush Alzheimer’s Disease Center, and Department of Neurology, Rush Presbyterian-St Luke’s Medical Center, 710 S Paulina Street, 8 North JRB, Chicago, Illinois, 60612


Intraoperative aneurysms rupture during the prediscision stage

Much has been written about intracranial aneurysms which rupture as they are being dissected, probably because of the frequency of this event (24%, in one well known series).1 Less has been written about rupture of aneurysms during the pre- and post-craniotomy stages.2 We present a case to illustrate this event.

A 59 year old right handed white woman experienced the acute onset of severe headache 48 hours before admission. She had no previous neurological symptoms and the only abnormal finding was neck stiffness. Computerised tomography (CT) of the head was normal but lumbar puncture disclosed grossly bloody fluid under an opening pressure of 210 cm H2O. Left carotid angiogram showed an anterior communicating artery aneurysm 3 mm in diameter pointing forward and downward.

During opening the patient was given 50 g intravenous mannitol, but spinal fluid was not drained. A slow drip of nitroprusside was started. As the chiasmatic cistern was opened, and before the aneurysm was displayed, the aneurysm ruptured and the brain herniated massively into the craniotomy opening. A hand held brain retractor was left in place to allow escape of blood. At the time of rupture the patient’s blood pressure was 100/80 mm Hg. Within a few minutes the BP had risen to 150/ 95 mm Hg. Immediately after rupture the patient was given 40 mg of nitroprusside and 20 mg of dexamethasone intravenously. Five minutes after rupture nitroprusside was stopped and a nitroglycerin drip (40 mg in 50 cc of 5% dextrose and water) was started. Within two minutes, that is seven minutes after rupture, the brain suddenly returned to its pre-rupture position and the BP fell to 60/40 mm Hg. Frontal lobe amputation, which had removed only 5 cc of brain, was terminated and the BP was allowed to rise slowly over the next 45 minutes. No further attempt to

W R G GIBB
F SCARAVILLI
Department of Neurology,
National Hospitals for Nervous Disease,
Queen Square, London WC1N 3BG,
United Kingdom

J MICHAUD
Department of Neurosurgery,
Hôpital Ste-Justine,
Montreal, Canada

expose the aneurysm was made and the craniotomy was closed.

Postoperatively the patient had a prolonged period of unconsciousness but gradually improved. When seen six months later she had only a left homonymous hemianopia and mild lack of concentration. The family refused a second craniotomy.

Several laboratory and clinical studies have begun to elucidate the physiological events during the first minutes after subarachnoid haemorrhage. Nornes has presented a teleological argument to explain the beneficial effects caused by the rapid increase and rapid fall of intracranial pressure when an aneurysm ruptures and distinguished two patterns. In Type I, "ischaemic-oedematous", haemorrhage results in an increase in ICP for a few minutes but then ICP falls to baseline level within 15 minutes. During the period of high ICP there is "brain tamponade" so that forward blood flow is mainly during systole; at the end of diastole there is flow arrest. This reduces the pressure gradient across the aneurysm wall and allows two to four minutes for the formation of a platelet plug. This is the scenario described in the present case.

The second example is a Type II "haemorrhage-compressive lesion" with haematoma producing a sustained increase in ICP and leading to death. Support for this concept came with puncturing the internal carotid artery through the sphenoid, thus simulating a bleed with the dura closed. Epidural pressure over both hemispheres increased immediately. Within one minute it approached the diastolic BP, plateaued for 90 seconds, and declined within minutes to double normal values. Transcranial Doppler observations made within minutes of the occurrence of a subarachnoid haemorrhage during surgery, before the dura was opened, were reported by Grote and Hassler. The patient had zero end-diastolic flow followed in seconds by negative diastolic flow, a situation of cerebral circulatory arrest which lasted for 100 seconds. End-diastolic flow returned to normal after 260 seconds. They postulated that the acute rise in ICP and cerebral circulatory slowing or arrest were due to acute vasospasm of distal cerebral arterioles.

At variance with this hypothesis is a study by Picard, Lovick, and Read which showed that cerebral blood flow increased after subarachnoid haemorrhage. They postulated that an increase in cerebral blood volume was responsible for rapid brain swelling.

I propose the following management algorithm if an aneurysm ruptures before the dura is opened. The BP should be lowered pharmacologically and even though, theoretically, this should decrease cerebral perfusion pressure it may cause massive cerebral swelling to subside, as described here. Mannitol, hyperventilation, and other measures to decrease ICP should be applied intensively. If dural tension does not decrease within five to 10 minutes, the surgeon should suspect an intracranial haematoma. The surgeon can then either terminate the operation or make a small dural opening to evacuate a haematoma. Internal decompression might be life saving in these circumstances and Tsementzis and Hitchcock were able to save five of eight patients, three in good condition, who bled during the induction of anaesthesia. The decision should take into account the pre-operative condition of the patient, the location of the aneurysm, and the experience of the surgeon and surgical team.

If the dural tension decreases within five to 10 minutes, the operation can probably proceed, and there are many reports of this being successful. Direct observation of the aneurysm area has most often disclosed remarkably little fresh blood.

When an aneurysm ruptures after the dura is opened, measures to decrease BP and ICP should be intensified. At this point if a retractor is in place, it should be left in place to allow for egress of blood which might otherwise form a haematoma. If, after five to 10 minutes, brain herniation is still present, the surgeon should proceed with an internal decompression if only to facilitate craniotomy closure. Again, when sufficient room in which to operate is achieved, the surgeon must decide whether to terminate the operation or proceed.

ROBERT A BEATTY
Department of Neurosurgery, University of Illinois, College of Medicine, Chicago, Illinois, United States
