the effects of anticholinergic drugs and other cause in Parkinson's disease. 4

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 found to have multiple communications, his affect was inappropriate and there was mild orofacial chorea. The left plantar was extensor and his gait ataxic. He developed generalised myoclonus with occasional opisthotonus, and there was an accentuated plantar response. 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 waves 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 a neuronal loss with glial and microglial reactions 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, 5 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. 6

Apart from the Lewy body–Parkinson's disease spectrum Lewy bodies are confined to a small group of rare degenerative disorders. 7 They may be by-products of other degenerative processes, whether of degeneration, or the de novo formation of Lewy bodies, and this explains their occurrence in the post-mortem brains of patients with SSPE showing periods of relative stability. SSPE is a destruc- tive inflammatory process due to persistent measles infection, but the abnormal immunological mechanisms are not fully understood. Although there is no direct evidence for an infectious aetiology 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 discon- connection in identical twins, 2 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 NMS, 2 and possibly the encephalopathy. 3 Because cholinergic receptor blockade can also cause encephalopathy, the differential diagnosis of NMS includes the central anticholinergic syndrome (CAS) due to anticholin- ergics, which are used to treat Parkinsonian patients with neuroleptic use, are often used to treat the rigidity in NMS. 4 However, anticholinergics could theoretically exacerbate the encephalopathy of NMS, possibly sparing 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 malesonic affective 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 dysmetria. Serum glutamic oxaloacetic transaminase 128 IU/l (normal 5–35), serum glutamic pyruvate transaminase 65 IU/l (normal 5–30), lactate 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 and lumbar puncture were normal. The haloperidol and lithium were stopped.

Treatment included levodopa/carbidopa 25/200, and 4 mg of imipramine benzo- tropine over two hours. Several hours after receiving the benzotropine, his mental status deteriorated from being agitated and confused to comatose, 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. 8 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.

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Intraoperative aneurysm rupture during the predisssection 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). 9 Less has been written about rupture of the aneurysm during the period after the cran- iotomy is started and before the aneurysm is exposed, the predisssection stage, discussed by Batjer and Samson. 10 Fortunately, this is uncommon. (Yasargil 90, Cadrin G, Drake J, King L, Fortin, personal communications). Batjer and Samson encountered it four times in a series of 307 consecutive aneurysm operations and questioned whether the operations should have been aborted. I report an experience of rupture of an aneurysm before dissection.

A 59 year old right handed white woman experienced the acute onset of severe headache 48 hours before admission. She had no neurological symptoms and the only abnor- mal 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 H 2 O. Left carotid angiogram showed an anterior communicating artery aneurysm 3 mm in diameter pointing forward and down. During opening the patient was given 50 g of 20% 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, Hgb 10.5 g/dl, Prednisolone was started after 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


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