LETTERS TO THE EDITOR

Spasms

It all happened a long, long time ago. I had seen the first case shortly after my arrival in Baghdad. A colonel in his early fifties, he positively overflowed an already ample uniform. He was a leading member of the ruling Party and spoke reasonable English, displaying in conversation that sinister mixture of urbanity and circumspection that I had come to expect of "political" officers in various parts of the Middle East.

His twitch had come on some two years earlier, fairly abruptly, under circumstances he could not or would not recall. At times, it was most distressing, seizing him and compelling him against his wish, to point his third chin in the general direction of his right shoulder. He was most aware of his impediment during political discussions with those in authority. It afflicted him little in the Staff College to which he was attached. The twitch had disappeared completely during a recent visit to London, where he had felt faintly ridiculous consulting neurologists for an involuntary movement he had been unable to show them.

In the hotel, later that evening, I hardly gave the matter a second thought. I was tired, had seen many patients that day, and the colonel had left no deep impression upon me, either as a person or as a problem. Spasmodic torticollis was a rare condition, singularly refractory to treatment. It had never excited my clinical curiosity. Neuropathologists and neuropharmacologists had had little of substance to say concerning its aetiology and my psychiatric colleagues, over the years, had proved quite unable to help those few cases I had guided their way.

The Tigris flowed quietly through the city, its banks a double necklace of delightful design. The beads—houses and gardens of buff and ochre, grey and pink—were enlivened at intervals by nuggets of brilliant green or turquoise: the fronds of palms and the domes of various mosques. But the peace was deceptive. The city had known only a few months of civilian rule since the revolution of 1958. One group of officers after another had held power in the name of varying creeds, all dutifully—if somewhat implausibly—proclaimed to be radical variants of the same belief. Deposed rulers often met bloody ends. The corpse of the hated Nuri Said had been dragged by the feet through the streets of the city, to reassure the multitude that he was totally and irreversibly dead.

The second case had been shown to me about a week after the first. He was a much younger man, twenty-five (if that), lean and tense, and with that periodic glint of fanaticism in the eye that proclaimed the true believer. I found it hard to elicit his precise occupation. He had something to do with the Ministry of the Interior. Given the machine gun turrets I had seen rather obviously displayed outside that particular building I probed the point no further.

His spasms were of a slower kind. A long writhing movement seemed to start at the root of his neck and gradually worked its way up, throwing the musculature on either side into extraordinary relief and making his eyes protrude, before petering out, somewhere near his occiput. We had only conversed through an interpreter. I dislike such interviews, for they virtually preclude the proper assessment of any relevant psychogenic factors. The patient seemed resigned to his disability. "How strange the ways of the Merciful" I almost heard myself say, when told that this patient too had been jerking for about two years.

The next few days passed quickly enough, and I thought no more about nuchal spasms. I lectured on basic neurological topics. I saw Assyrian ruins and the ravages of leishmaniasis. I admired the arch of Ctesipon, coped with comas in the ward, and learned a lot about the Hammurabi code. I mused cynically about contrasts: the sustained equalitarian rhetoric of the regime and its extremes of wealth; its talk of internationalism and its ubiquitous xenophobia. But then who were we, to criticise others on such grounds?

On the last evening we were invited to a farewell dinner. We ate stuffed peppers, masgouf (grilled Tigris fish) and roasted sheep from an enormous buffet. The whisky and arak flowed freely as did the conversation.

Shortly after midnight I spotted the third case: a small, bald-headed man in his early forties. He was sitting in a corner, talking medical shop with some colleagues. Every now and then his head would give two or three little jerks to the left, a sort of minispasm quite obvious to the trained observer but which the patient himself sought rather half-heartedly to disguise as a response to a tight collar.

At first I felt reluctant to intrude and to broach such a personal matter. We were in the drawing room of a private house, not in the out patient department of some neurological hospital. But my curiosity was now aroused and the arak made it articulate. I walked up to him and, taking full advantage of the privilege accorded visitors in such circumstances, introduced myself. He was a psychiatrist, had travelled widely and spoke fluent English and passing French. I eventually came to the point. He smiled sardonically and looked straight through me, running a podgy finger between shirt and neck. Yes, he knew of the other two cases. But he was surprised that they had been brought to me. Baghdadis didn't really like washing their dirty linen in public. To tell the truth, he had known them very well indeed. He too was a Ba'athist militant and in that capacity all three had enjoyed a stretch in prison together, some two years earlier, during the last few days of the previous junta.

Sensing an imminent denouement I got him another drink. "Well", he said philosophically, "those were violent times. And somatic responses to stress are unpredictable, even to us doctors. Five of us had been condemned to death. Two were executed. By hanging. We were due for the drop on the following day. The new regime came to power in the very nick of time. It was a close shave. Believe me, friend, my neck—since then—has never felt quite the same".

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This article was written for the special supplement in honour of Professor C D Marsden published in June 1989 and we apologise to Dr Pallis and Professor Marsden for not including it in that supplement.

EDITOR

Pontine myelinolysis presenting with acute parkinsonism as a sequel of corrected hyponatraemia

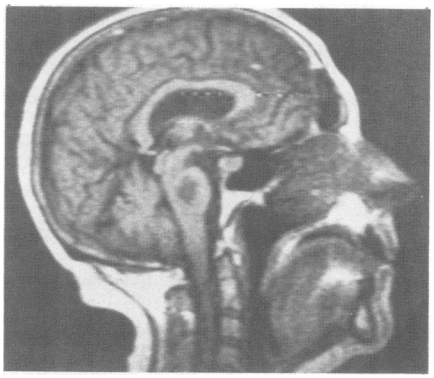
We report an unusual case of pontine myelinolysis, associated with rapid correction of hyponatraemia. The presentation was one of acute parkinsonism, despite the presence of a large pontine lesion on magnetic resonance imaging (MRI).

A 66 year old woman presented with a 'flu-like illness and was treated with oral erythromycin. Six days later she became confused and drowsy. She was referred to hospital and found to have a chest infection associated with the syndrome of inappropriate antidiuretic hormone secretion (in serum: albumin 33 g/l, sodium 94 mmol/l, potassium 1·6 mmol/l, urea 2·3 mmol/l, osmolality 204 mmol/kg; in urine: sodium 45 mmol/l, potassium 35 mmol/l, osmolality 407 mmol/kg). There was no history of alcohol abuse and she had not received diuretics. She was treated with twice normal saline with added potassium, intravenously, until her serum sodium was 122 mmol/l and her serum potassium was 3·3 mmol/l. She became orientated and cooperative, intravenous fluids were stopped and she was treated with oral fluid restriction. The rate of serum sodium correction was 19 mmol/l in the first 24 hours (6 mmol/l in the first 10 hours and 13 mmol/l in the subsequent 14 hours), 10 mmol/l in the next 24 hours and then an average of 3 mmol/l per day over the next five days.

Twelve days after admission she again became confused and agitated, with paranoid ideas. She had developed impassive facies, bradykinesia, rest tremor, cogwheel rigidity in the limbs and a parkinsonian gait. No biochemical abnormality was detected and she had not received any neuroleptic drugs. CT scan was normal but MRI showed a large area of pontine myelinolysis (fig). Imaging of the basal ganglia was inadequate due to movement artefact. A small dose of levodopa (Madopar 62·5 mg four times daily) improved her gait and abolished the tremor. The levodopa was withdrawn after two months and she is now neurologically normal and fully independent. During her hospital admission she was found to have mild primary hypothyroidism and was treated with thyroxine.

This case illustrates the dangers of rapid correction of hyponatraemia.[1,2] Hypernatraemia did not occur at any time and the suggestion that myelinolysis will not occur providing hypernatraemia is avoided[3] appears invalid.

Only one previous case of pontine myelinolysis presenting with dopa-responsive park-



MRI scan (sagittal section, spin echo, repetition time 160 ms, echo time 30 ms) showing large area of abnormal signal in the pons.

insonism has been reported; that patient also had pyramidal signs.[4] A further patient exhibited dopa-responsive tremor and facial impassivity during recovery from a more typical presentation of pontine myelinolysis with stupor, abnormal eye movements and tetraparesis.[5] Pathological changes in the basal ganglia have been well documented in typical cases of pontine myelinolysis,[6] and it has been suggested that the pontine lesion masks the extra-pontine clinical features. In our case the large lesion in the pons was clinically silent. MRI is clearly the investigation of choice in patients presenting with neurological syndromes associated with hyponatraemia; subclinical or clinically atypical pontine myelinolysis may be more common than is currently realised.

We are grateful to Dr S P Kane for permission to report a patient under his care.

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Visual hallucinations and the cholinergic system in dementia

Cholinergic deficits are associated with various forms of dementia, including Alzheimer's and Parkinson's disease, in which they relate to the degree of cognitive impairment.[1] New findings on the neurochemical pathology of senile dementia of Lewy body type (SDLT) suggest that particularly extensive cholinergic abnormalities in certain neocortical regions are associated with visual hallucinations. SDLT may be the second most common form of dementia in the elderly (after Alzheimer's disease).[2] It is characterised, clinically, by acute presentation with confusion frequently accompanied by hallucinations and, neuropathologically, by the presence of Lewy bodies, particularly in archicortical areas. The relative absence of neocortical neurofibrillary tangles in SDLT[2] also suggests such patients might be more amenable to transmitter replacement therapy.

In 12 SDLT cases with frozen tissue available for neurochemical analyses, visual hallucinations had been noted in six at presentation and continued throughout the course of the disease; in the remainder this symptom was not recorded. Choline acetyltransferase (ChAT) activities were reduced in both SDLT sub-groups compared with normal levels but were significantly lower in those with hallucinations compared with those without for two of three cortical areas examined (see table). Thus in hallucinating cases ChAT activities in parietal and temporal cortex were reduced by 80–85% compared with 50–55% in the non hallucinating cases. No other neurochemical or neuropathological parameter so far examined (including dopamine, serotonin, cholinergic receptors, Lewy bodies or plaques) divided the two groups.

Table Cortical choline acetyltransferase activities[a] in senile dementia of Lewy body type[b] mean, SD

	Parietal	Temporal	Occipital
Normal SDLT	10·8, 5·2 (7)	8·7, 5·6 (11)	11·6, 3·4 (8)
Without hallucinations	5·1, 2·7 (5)	4·0, 1·7 (5)	9·2, 4·0 (6)
With hallucinations	1·6, 1·4 (6)	1·5, 1·6 (6)	6·0, 4·2 (4)

[a]ChAT expressed as nmol/mg protein[3] in Brodmann areas 39 or 40 (parietal), 21 or 22 (temporal) and 17, 18 or 19 (occipital). Case numbers in parentheses.
[b]In parietal and temporal but not occipital cortex, ChAT was significantly different between the two subgroups ($p = 0{\cdot}009$ and 0·015, respectively, Mann Whitney U test). The normal and SDLT groups were matched for age and postmortem delays mean (SD) 78 (6), 81 (6) and 76 (7) years; 27 (12), 41 (24) and 23 (15) hours, respectively).

The suggestion that extensive ChAT loss in SDLT cases with hallucinations may be related to this clinical feature is supported by psychopharmacological data. Thus, although no patient was receiving any anticholinergic medication at the time they presented, drugs such as scopolamine are known in certain cultural[4] or medical[5] situations to induce hallucinations. Severe degeneration of cholinergic neurons innervating certain cortical areas in dementing disorders such as SDLT may similarly give rise to this symptom. If confirmed, this observation is important in relation to the function of cholinergic innervation of human neocortex (innervation of other areas such as hippocampus being involved in memory) and in relation to selecting suitable cases of dementia for cholinergic therapy (those with hallucinations having a more profound cholinergic defect).

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Type 1 Arnold-Chiari malformation in a 77 year old woman

Chiari malformations may present in a variety of ways, most commonly between the ages of 20 and 60 years.[1-3] Onset of symptoms after the seventh decade is distinctly unusual.

A 77 year old woman had fallen and was found unconscious by her general practitioner. She was thought to have aspirated. She stopped breathing before reaching hospital but was successfully resuscitated and needed artificial ventilation for a week. As she recovered, frequent choking and aspiration of fluids were noted. On closer questioning she admitted having dysphagia with nasal regurgitation over a two year period. During the preceding six months she had become unsteady and dizzy, with a tendency to fall, and there had been fleeting episodes of diplopia. Some years before she had had bilateral total hip replacement and had experienced several serious falls.

The right hip prosthesis had loosened in a recent fall and she was unable to walk. Her eye movements were full and there was no nystagmus in the primary position. There was down beating nystagmus on downgaze and obliquely down beating nystagmus on lateral gaze to either side. Palatal movement and gag reflexes were absent although pharyngeal sensation and tongue movements were preserved and speech was normal. Fluid aspiration occurred consistently and she was fed through a nasogastric tube. Her limbs were ataxic, but there were no pyramidal features.

A sleep study demonstrated considerable nocturnal hypoxia ($SaO_2 < 80\%$). MR imaging showed cerebellar hypoplasia and tonsillar herniation down to the level of the second cervical vertebral body (fig). The medulla was elongated and kinked over the odontoid peg. There was mild associated hydrocephalus. Her foramen magnum was decompressed, the dura incised and a fascial graft was inserted. Although there was no change in the physical signs following operation, she was able to swallow fluids normally without choking, aspirating or regurgitating. The nasogastric tube was successfully removed and she was discharged from hospital.

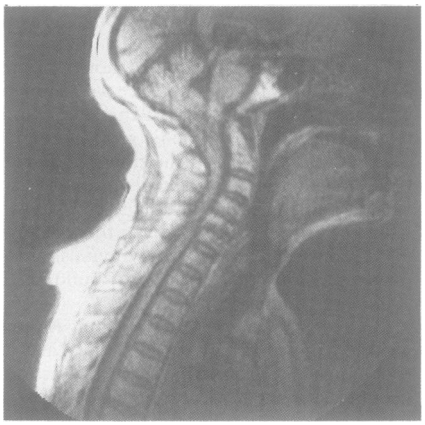

Figure Sagittal MR image.