

presence of neoplasms or hyperplasia involving endocrine glands. The organs most often affected are the parathyroids, pancreatic islets and pituitary gland.<sup>1</sup> Occasionally neoplasms of the thymus gland have been reported in this syndrome.<sup>2-4</sup> Myasthenia gravis is frequently associated with thymic hyperplasia and approximately 10-15% of the patients have a thymic tumour.<sup>5</sup> Our patient presenting with myasthenia gravis had a thymoma and characteristic features of MEN-1 syndrome.

A previously healthy 28 year old male was admitted to hospital with a six weeks history of progressive speech disturbance, problems with mastication and nasal regurgitation, followed by diplopia. There was a progression of symptoms during the day and after exertion and reduction after rest. There was no weakness in the extremities. He suffered from nocturnal dyspnoea. Micturition and sexual functions were normal. There were no symptoms of hypoglycaemia. The patient's mother had been operated on in the past because of hyperparathyroidism.

Physical examination showed a slightly overweight man (height 1.72 m, weight 85 kg) with little facial hair. Internal investigations including the genital organs were normal. Neurological investigation revealed slight dysarthria, bilateral ptosis, facial weakness and paresis of abduction and elevation of both eyes. These symptoms fluctuated, depending on the degree of exertion. Leg raising was possible for 45 seconds, no other limb muscle weakness was present. Deep tendon reflexes were normal. The functional capacity of the lungs by spirometry was within normal limits.

Laboratory investigations demonstrated serum calcium level of 3.19 mmol/l; phosphate 0.84 mmol/l; chloride 101.4 mmol/l; chloride/phosphate ratio 102 mol/mol (N: less than 103 mol/mol); parathyroid hormone level of 11.7 pmol/l (N: less than 5 pmol/l); prolactin 4.68 U/l (N: less than 0.3 U/l); glucose 4.4 mmol/l; insulin 29 IU/l (N: 5-25 IU/l); insulin/glucose ratio 10 (N: less than 9); serum gastrin and glucagon were normal, as 5HIAA in daily urinary excretion. Circulating antibodies to acetylcholine receptors were elevated 97 nM (N: less than 2 nM). Anti-striated muscle antibodies were positive. There was a normal EMG pattern of the abductor digiti quinti muscle after repetitive (3 Hz) and tetanic (20 Hz) stimulation. Edrophonium chloride (Tensilon) 10 mg given intravenously resulted in a marked temporary improvement of signs and symptoms. Computerised tomography (CT) of the thorax revealed a large dense, partly cystic, anterior mediastinal mass. CT of the sella turcica showed a pituitary tumour. On echocardiography and CT of the abdomen no tumour of the pancreatic islets was seen.

The patient had a median sternotomy. A mediastinal tumour (12.5 × 8 × 3 cm) was completely excised. Microscopically the classic histological features of a benign thymoma were present with both epithelial and lymphoid cells without argentaffinity or mitoses. The tumour cells stained positively with cytokeratin and antibodies to insulin, no other endocrinological activity could be demonstrated. Furthermore, a parathyroid adenoma was removed (1.5 × 0.5 × 0.5 cm) in the same session, the other parathyroid glands were normal. The patient made a slow recovery from thymectomy; some days he required intubation and mechanical ventilation because of respiratory insufficiency. On

repeated postoperative examination his calcium, phosphate, insulin level and insulin/glucose ratio had normal values, the antibodies to acetylcholine receptors were reduced to 24 nM; anti-striated muscle antibodies were still positive. Nine months later he was considerably improved taking pyridostigmine 60 mg four times daily, and bromocriptine 2.5 mg twice daily.

This case represents the combination of myasthenia gravis, a benign thymoma, hyperparathyroidism and a pituitary tumour, probably a prolactinoma. The coexistence of hyperparathyroidism and a pituitary tumour is characteristic of MEN-1 syndrome. The evolution of this syndrome may take years and it is inherited as an autosomal dominant trait with a very high degree of penetrance.<sup>1</sup> The mother of our patient also had hyperparathyroidism. Less common findings reported in patients with MEN-1 syndrome included lipoma, carcinoid tumours, thymic tumours and pinealoma.<sup>1-4</sup> The neoplasms of thymic origin mainly concern carcinoid tumours with a relatively poor prognosis.<sup>2-4</sup> None of these cases had myasthenia gravis.

Only one other patient has been described with the combination of myasthenia gravis, thymoma and hyperparathyroidism; a sporadic *forme fruste* of the MEN-1 syndrome was suggested by the author.<sup>6</sup> Moreover, our patient had a pituitary tumour which underlines the diagnosis of MEN-1 syndrome. The insulin level and insulin/glucose ratio were slightly elevated, these values, however, did not prove to be a pancreatic islet tumour. Perhaps the thymoma was responsible for this disturbance as the tumour cells reacted positively with antibodies to insulin and the value returned to normal post-operatively.

The importance of this remarkable finding is not clear, but cell surface receptors for some polypeptide hormones have been mentioned in thymoma cells.<sup>7</sup> This reaction was negative in five other thymomas that we have investigated in recent years. The association of tumours of the thymic and parathyroid gland is not surprising as these structures have a common embryonic origin from the third pharyngeal pouch.

LJHM VROEGINDEWEIJ-CLAESSENS  
CC TIJSSSEN  
Department of Neurology,  
GJM CREEEMERS  
JHM LOCKEFEEER  
Department of Internal Medicine  
JLJM TEEPEN  
Department of Pathology,  
St Elisabeth Hospital, Tilburg,  
The Netherlands

Correspondence to: LJHM Vroegindewei, Department of Neurology, St Elisabeth Hospital, PO Box 90151, 5000 LC Tilburg, The Netherlands.

- 1 Lips CJM, Vasen HFA, Lamers CBHW. Multiple endocrine neoplasia syndromes. *CRC Crit Rev Oncol Hematol* 1984;2:117-84.
- 2 Floros D, Dosios TH, Tsourdis A, Yiatromanolakis N. Carcinoid tumor of thymus with multiple endocrine adenomatosis. *Path Res Pract* 1982;175:404-9.
- 3 Rosai J, Higa E, Davie J. Mediastinal endocrine neoplasms in patients with multiple endocrine adenomatosis. A previously unrecognized association. *Cancer* 1972;29:1075-83.
- 4 Manes JH, Taylor HB. Thymic carcinoid in familial multiple endocrine adenomatosis. *Arch Pathol* 1973;95:252-5.
- 5 Oosterhuis HJGH. Myasthenia gravis. A survey. *Clin Neurol Neurosurg* 1981;83:105-36.

- 6 Palmer FJ, Sawyers TM. Hyperparathyroidism, chemodectoma, thymoma and myasthenia gravis. *Arch Intern Med* 1978;138:1402-3.
- 7 Arya S, Gilbert EF, Hong R, Bloodworth Jr GMB. The thymus. In: Bloodworth Jr GMB, ed. *Endocrine Pathology*. 2nd ed. Baltimore: Williams and Wilkins, 1982:801.

## MATTERS ARISING

### Korsakoff's psychosis in the presence of multiple sclerosis: an unusual cognitive state

After the publication of our letter in your journal,<sup>1</sup> some inaccuracies were brought to our attention by Professor E K Warrington in whose department the patient was tested.

The difference between the WAIS IQ and the estimated premorbid IQ (NART) indicates a decline in the performance on non-verbal tests rather than general intellectual ability as stated in the article. A score of 19/30 in the McKenna and Warrington Graded Naming Test is within the average range and not indicative of nominal dysphasia. The statement: "in both Korsakoff's syndrome and multiple sclerosis verbal IQs decline more than performance IQs ..." is erroneous and it would be correct to say that the performance IQ is more likely to deteriorate.

Finally, although not an inaccuracy, the Camden Memory Tests quoted in the letter are still in the process of development and low scores should be interpreted as corroborating poor performance on the standard memory tests. These corrections in no way detract from the conclusions of our letter and we feel that it is worth highlighting the need to search for other pathologies in patients with alcohol related brain damage if their clinical presentation is in anyway unusual.

MARIA A RON  
ANTHONY FEINSTEIN  
The National Hospital for Nervous Diseases,  
Queen Square,  
London WC1N 3BG, United Kingdom

- 1 Ron MA, Feinstein A. Korsakoff's psychosis in the presence of multiple sclerosis: an unusual cognitive state. *J Neurol Neurosurg Psychiatry* 1989;52:540.

### Magnetic resonance imaging in patients with progressive myelopathy following spinal surgery

The explanation by Mr Adams<sup>1</sup> of the post-operative myelopathy with a biomechanical mechanism producing postoperative traction of the dura and the spinal cord during cervical movements is an adjunct to our paper,<sup>2</sup> in

which merely an observation was made, rather than an explanation.

We agree that the biomechanical mechanism can explain the progressive myelopathy, beginning late in the post-operative course. However, it is hard to believe that myelopathy beginning five years or more following surgery can be caused by arachnoiditis. On the other hand, the possibility of spinal cord injury during operation is unlikely. The biomechanical mechanism of postoperative myelopathy, so well described by Mr Adams, can explain that the myelopathy develops more slowly after surgery of spinal stenosis and disc disease, rather than after intramedullary surgery.

We also agree with the great importance given by Mr Adams to the careful preoperative evaluation of the candidates for cervical spine surgery by conventional radiography. In addition, we advise pre- and postoperative magnetic resonance imaging (MRI) examinations of these patients. MRI has the advantage of demonstrating the spinal cord and its relation to the bony spinal canal, and is easier to recognise the problems of the postoperative spine and spinal cord. Further experience in this field is necessary to solve these problems.

E AVRAHAMI  
R TADMOR  
DF COHN  
Department of Radiology,  
Ichilov Hospital,  
Tel-Aviv Medical Center,  
Tel-Aviv, Israel

- 1 Adams CBT. Magnetic resonance imaging in patients with progressive myelopathy following spinal surgery. *Matters Arising. J Neurol Neurosurg Psychiatry* 1989;52:1016.
- 2 Avrahami E, Tadmor R, Cohn DF. Magnetic resonance imaging in patients with progressive myelopathy following spinal surgery. *J Neurol Neurosurg Psychiatry* 52:176-81.

## BOOK REVIEWS

**Essentials of Clinical Neurology.** By CARL H GUNDERSON. (Pp 550; Price \$69.00.) New York: Raven Press. 1989.

Dr Gunderson is chief of the Neurology Service at Walter Reed Army Medical Centre in Washington DC. In the preface to his new publication the author explains how it was that "brief handouts" to help his students became expanded over a period of 20 years to become, in 1982, his first book entitled *Quick Reference to Clinical Neurology* (J B Lippincott Co). The *Essentials of Clinical Neurology* represents a further expansion and revision of that work.

Dr Gunderson's stated aim is to avoid a book that espoused "to become encyclopaedic" but he hoped, rather, to "restrict the

material" to that which he "would like every medical student to have been exposed to during a four week clinical neurology rotation". (I presume he means by this "heard about" rather than "seen" because the subject index includes reference amongst others to such things as acid maltase deficiency, barognosia and Hollenhorst plaques.) The book is designed for the use of undergraduate, junior house officer or "practising physician" alike. A tall order? I rather think so—the many lists and tables do indeed read like handouts for undergraduates and I would doubt whether this approach is either comprehensive enough or sufficiently attractive in its literary style for the post-graduate. Indeed, it is rather military—the section on "Physical Examination" begins A. Inspection—1. The patient should disrobe (!).

Other chapters cover specific complaints (eg, The Dizzy and Deaf Patient), management of specific syndromes (Neuropathies and Stroke), for example, and selected diseases (eg Infections and Tumour). Whilst there is an immense amount of valuable information, indeed encyclopaedic in undergraduate terms, unfortunately there are also important omissions and, I think, errors. For example, routine examination of the upper limb omits the testing of muscle tone (page 10) and there is no mention of the characteristic pyramidal distribution of upper motor neurone weakness, only that "hemiparesis, quadriparesis or paraparesis" form the usual pattern. Patients who trip over often have spasticity, not "foot weakness" (page 67). Does muscle biopsy (page 48) only "occasionally differentiate" muscle disease from neuropathy? It is stated (on page 255) that "about 25 per cent of patients will have status epilepticus as their first symptom of epilepsy". Surely this is not true? The chapter on epilepsy, in fact, raises another problem with the book and that is that old and new nomenclature is mixed side by side.

Chapter 26 (Disorders of Myelin) closes with a few paragraphs on "Brachial Neuritis" (page 474). Nowadays called Neuralgic Amyotrophy or Shoulder-girdle Neuritis, this disorder was never, as stated, "attributed to the design of the British rucksack". It was indeed initially described during the Second World War but the majority of patients were serving in the United Kingdom at the time. Many sufferers were soldiers, but the authors of the original papers all noted the association of the condition with incidental operations, convalescence from infective illness or prophylactic inoculations. The close similarity to "serum neuritis" was also noted. The possibility that this disorder was an immune based demyelinating disease was recognised very early on—not recently, as implied. I don't think the condition ever spreads, as stated, to a leg, although there is a complimentary form of radiculopathy of "idiopathic" variety affecting the lumbo-sacral outflow.

I doubt whether the style of this book will attract many students or junior doctors in the UK. There is too much for the undergraduate, and the book is not sufficiently comprehensive or accurate for the discerning postgraduate. I am sure the latter would be irritated by the numbering and lettering of every paragraph. I think they are more likely to be encouraged by the good selection of more attractive, compact text books of this kind already available with further reference to be made to the larger comprehensive text.

J A SPILLANE

**Comprehensive Neurologic Rehabilitation Vol. 1 The Management of High Quadriplegia.** Edited by: G WHITENECK, C ADLER, R E CARTER, D P LAMMERTSE, S MANLEY, R MENTER, K A WAGNER, C WILMOT. New York: Demos Publications (pp 367; Price \$69.95) 1989.

Patients with spinal injuries have only been kept alive since the development of spinal centres in 1944 at the end of the Second World War. Initially only paraplegic patients survived but with improved treatment morbidity and mortality decreased and paraplegic patients began to experience a nearly normal life expectancy. Initially few tetraplegic patients were admitted to spinal centres and those that were, died soon after injury. However, following the same principles tetraplegic patients were kept alive and since 1980 even those on ventilators have been kept alive and discharged home.

In the rehabilitation of high quadriplegics the United States is ahead of the United Kingdom and this book reports the combined experience of centres that are leading the world in this field. The Craig Hospital, Santa Clara Valley Medical Center and the Institute of Rehabilitation and Research (Houston, Texas) have information on 216 individuals who have been discharged home, many of whom were on ventilators, an extraordinary record of achievement.

The priority in patient care in the United States differs from that in the United Kingdom in that patients are admitted to neurological and neurotrauma centres initially and only some weeks later come to rehabilitation centres; thus the authors' experience of the initial treatment of spinal injury is limited—in the United Kingdom patients are admitted immediately to spinal centres.

The book is split into four sections, Acute Care, Rehabilitation, Discharge Planning and The Real World. Unfortunately the chapters on acute care do not deal with the associated complications found in some 75% of cases, the management of head injuries, the problem of hypercalcaemia and temperature regulation. In mentioning the various forms of anticoagulation they suggest that the patient should be anticoagulated at 1.5 times the control level, a view that would not gain universal acceptance.

Also, the description of a teardrop fracture being due to an extension injury is inaccurate "The Radiograph may show a posterior displacement of the superior vertebral body on the inferior one, but more often one sees a separation of the disk space at one motion segment or an avulsion fracture of the anterior superior part of the inferior vertebrae. This is called a teardrop fracture." A teardrop fracture is a crush fracture with extrusion of fragment. On the other hand the chapters by Carter on Available Respiratory Options, Anderson on Psychological Issues, the chapters on the medico-legal aspects and the whole approach to discharge home, are excellent. The chapter on the long term outlook, though short, is an extraordinary tribute to care whereby after nine years 63% of their initial group of respiratory dependent patients are still alive. This is an exceptional piece of work.

The whole book is fragmented with respiration management dotted about in different places. It does not follow conventional presentation and there is no discussion of the fundamental problems of chest wall movement, postural problems associated