

MATTERS ARISING

Alexander Ypsilante (1792-1832)

We read in the neurological stamp column of the *Journal* the comments of Dr Haas¹ regarding the disease of Alexander Ypsilante (1792-1832). As the writer noted the picture on the stamp in honour of Prince Alexander Ypsilante, issued by the Greek state in 1930, fails to show that he probably had myotonic dystrophy, a disease which was diagnosed in 1932 in London by Professor Caughey in a female descendant of Alexander Ypsilante^{2,3}; another four siblings from her family of eight also had the illness.

Recent in depth research on the medical history of the family of Alexander Ypsilante, ongoing in the Department of Neurology, University of Patras^{4,5} support Caughey's hypothesis, according to which Alexander Ypsilante, the leader of the Greek revolution—and probably some of his brothers—had myotonic dystrophy. Our studies, based on correspondence, state archives, and medical documents found in Greece, Rumania, and Austria have shown that Alexander Ypsilante, his younger brother Demetrious, also a hero of the Greek struggle for independence against the Turks in the last century, and two more brothers of the remaining five children of the family, often manifested symptoms highly suggestive of dystrophia myotonica; these included early frontal baldness, severe weakness and atrophy of face and limb muscles, hypofertility, pulmonary infections, heart problems, and vision failure. Despite these data and the fact that necropsy was carried out on Alexander Ypsilante, Ypsilante brothers' disease still remains an enigma because the disease, unknown during their time, was only described later (1909), by Steinert. Hopefully with the help of modern molecular genetic techniques (DNA technology)⁷ and bones from the tomb of Demetrious Ypsilante, located in Nafplion, the first post-revolutionary capital of Greece, it would be possible to precisely state the nature of the Ypsilante family illness.

TH PAPAPETROPOULOS
Department of Neurology,
Medical School of Patras University,
PO Box 1045, 26 500 Rion,
Greece

- 1 Haas LF. Alexander Ypsilante (1792-1832): neurological stamp. *J Neurol Neurosurg Psychiatry* 1994;57:885.
- 2 Gaughey JE. Diseases of the lens. Cataract in dystrophia myotonica. *Transactions of the Ophthalmological Societies of the UK* 1933;5: 60-70.
- 3 Caughey JE, Myriantopoulos NC. *Dystrophia myotonica and related disorders*. Springfield Ill: Charles C Thomas, 1963:VII-VIII.
- 4 Soteriadou C, Papapetropoulos Th. D Ypsilante disease. *Historica (Athens)* 1986;5: 97-106.
- 5 Soteriadou-Vayenas C. "Myotonic dystrophy: hypotheses and facts about the Ypsilante brothers disease" [thesis]. Medical School of Patras University, Patras, 1988.

- 6 Soteriadou-Vayenas C, Papapetropoulos Th. Myotonic dystrophy: hypotheses and facts about the illness of the Ypsilante brothers. *J Neurol Sci* 1989;90:231-7.
- 7 Galloway J. Can these bones speak? *Helix, Angen's Magazine of Biotechnology* 1994;5: 52-7.

Angiotropic lymphoma in the differential diagnosis of systemic vasculitis

We were interested to read the report by Roux *et al* of angiotropic lymphoma with mononeuritis multiplex mimicking systemic vasculitis.¹ We have recently published a case with striking similarities to this one.² Two other cases not mentioned by the authors have also mimicked vasculitis.^{3,4} In our patient even the antineutrophil cytoplasmic antibody titre test was repeatedly positive emphasising the point made by Roux *et al* that this condition may mimic a systemic necrotising vasculitis extremely closely. In both cases the patient had a rash suggestive of livedo reticularis, a progressive flaccid paraplegia, loss of sensation in the legs, urinary retention, and anal incontinence. Both patients also developed panhypopituitarism, massive oedema, and hypotension with profound hyponatraemia. Antineutrophil cytoplasmic antibody titres are found in several inflammatory conditions and are often present in systemic vasculitis.⁵ In our case, the combination of such titres and clinical features pointed us incorrectly towards this diagnosis. We agree strongly with the authors that the differential diagnosis of systemic necrotising vasculitides must include angiotropic lymphoma, particularly as this condition is difficult to diagnose before death, had a dismal prognosis if undiagnosed, and is potentially curable.

AMMAR AL-CHALABI
University Department of Neurology
and Neuroscience,
Institute of Psychiatry and
Kings College School of Medicine and Dentistry,
Denmark Hill,
London SE5 8AF, UK

RICHARD J ABBOTT
Department of Neurology,
Leicester Royal Infirmary,
Leicester LE1 5WW, UK

Correspondence to: Dr Al-Chalabi.

- 1 Roux S, Grossin M, De Bandt M, Palazzo E, Vachon F, Kahn MF. Angiotropic lymphoma with mononeuritis multiplex mimicking systemic vasculitis. *J Neurol Neurosurg Psychiatry* 1995;58:363-6.
- 2 Al-Chalabi A, Sivakumaran M, Holton J, West K, Wood JK, Abbott RJ. A case of intravascular malignant lymphomatosis (angiotropic lymphoma) with raised perinuclear antineutrophil cytoplasmic antibody titres—a hitherto unreported association. *Clin Lab Haematol* 1994;16:363-9.
- 3 Kamesaki H, Matsui Y, Ohno Y, Amano H, Imanaka T, Takahashi Y, *et al*. Angiocentric lymphoma with histological features of neoplastic angioendotheliomatosis presenting with predominant respiratory and haematological manifestations. *Am J Clin Pathol* 1990;94:768-72.
- 4 Krieger C, Robitaille Y, Jothy S, Elleker G. Intravascular malignant histiocytes mimicking central nervous system vasculitis: an immunopathological diagnostic approach. *Ann Neurol* 1982;12:489-92.
- 5 Ramirez G, Khamashta MA, Hughes GRV. The ANCA test: its clinical relevance. *Ann Rheum Dis* 1990;49:741-2.

SHORT NOTICES

Readers may be interested in:

Drug Treatment in Psychiatry. Fifth Edition. By Trevor Silverstone and Paul Turner. (Pp 309 Pb £16.99). Published by Routledge, London 1995. ISBN 0-415-10610-9.

Companion to Clinical Neurology. By William Pryse-Phillips. (Pp 1009; \$99.95). Published by Little, Brown and Company, Boston 1994. ISBN 0-316-72041-0.

Prescription Drug Abuse and Dependence. Edited by Daniel P Greenfield. (Pp 170; \$42.95). Published by Charles C Thomas, Illinois 1994. ISBN 0-398-05931-4.

The Human Brain Circulation. Edited by Rosemary D Bevan and John A Bevan. (Pp 456; H/b \$89.50). Published by The Humana Press Inc, Totowa 1994. ISBN 0-896-03271-X.

BOOK REVIEWS

All titles reviewed here are available from the BMJ Bookshop, PO Box 295, London WC1H 9TE. Prices include postage in the United Kingdom and for members of the British Forces Overseas, but overseas customers should add £2 per item for postage and packing. Payments can be made by cheque in sterling drawn on a United Kingdom bank, or by credit card (Mastercard, Visa or American Express) stating card number, expiry date, and your full name.

Cytopathology of the Central Nervous System. Edited by SANDRA H BIGNER and WILLIAM W JOHNSTON. (Pp 190; £95.00). Published by Edward Arnold, London 1994. ISBN 0-340-59490-X.

Cytopathology of the Central Nervous System is a textbook and atlas which will appeal to anyone working in the clinical neurosciences. It can be highly recommended to the practising neuropathologist or cytopathologist as the only high quality and comprehensive reference for everyday diagnostic use. The authors have taken the original step of presenting normal neuroanatomy and gross neuropathology as a prelude to the histo- and cytopathology preparations. For this reason, the book will be of interest to all clinicians who wish to know how cytological examination of cerebrospinal fluid and brain aspirates might help them in practice. The approach was ambitious and it proves highly successful as these components are skilfully integrated in

a way which facilitates both detailed reading and quick reference; each of these activities is rendered interesting and enjoyable.

Throughout this book, the text is pertinent and the illustrations superb. One of the introductory chapters on normal cells and contaminants in CSF will be particularly useful to trainees. A chapter on general cytological principles would have helped clinicians. Inflammatory disease, vascular disorders, degenerative and demyelinating diseases, leukaemia and lymphoma, metastatic tumours, and primary central nervous system neoplasms are discussed in subsequent chapters so that the entire field of diagnostic neurocytology is covered. The cytology of AIDS-related illnesses, particularly opportunistic infections is a feature. In all these chapters, colour photomicrographs correlate CSF cytological appearances with wet tissue smear preparations and paraffin-embedded tissue sections. The application of immunohistochemistry to CSF and brain aspirates are discussed and illustrated. Another chapter outlines cytogenetic and molecular genetic aspects of nervous system tumours.

This really is a superb and unrivalled volume which can be recommended to clinicians as well as pathologists. It is pricey but this is justified by the quality of the illustrations.

J H XUEREB

Occupational Neurology and Clinical Neurotoxicology. Edited by MARGIT L BLEECKER. (Pp 387 £67.00.) Published by Waverly Europe Ltd, London 1994. ISBN 0-683-00848-X.

Many are concerned that toxins play an under-recognised role in causing neurological disease. Whenever that question is raised, for instance by medicolegal cases, it is often hard to obtain a balanced and succinct review of the relevant literature, much of which is secreted away in unfamiliar journals often not stocked by our usual libraries. Dr Bleecker's book fills this void with success and neurologists will find it useful. The wide scope ranges from industrial neurotoxicology to the mechanical consequences for nerves of various occupations and activities. The book does not cover pharmaceutical neurotoxicology, a subject about which information is already more easily available. Practising clinicians will enjoy the case vignettes which illustrate the diagnostic dilemmas posed by patients in whom a neurotoxic disorder is suspected.

Dr Bleecker is to be congratulated for her success in disciplining twenty five authors to produce contributions of similar weightings. The excellent tables, comprehensive indexing, and careful use of headings enable chapters to be scanned quickly for information about suspected toxins. The toxicology coverage ranges from assessment of the degree of exposure, to the accepted biological exposure limits, and to the clinical and neuropsychological consequences of toxicity. Chapter four reviews methods of quantifying neurological dysfunction. Much of this chapter will be of little interest to practising neurologists, since it largely reiterates aspects of the neurological assessment with which we are already well-versed; I would question whether the MRC grading scale for muscle power has much application in

this setting. There is some unnecessary overlap of subject matter between the chapters on neurotoxicology of the visual system and on quantitative neurological examination. The chapter by Thomas on primary brain tumours associated with chemical exposure made me reflect disturbingly on the impression that I encounter gliomas increasingly commonly.

The sections on occupational neurology (mainly spinal problems and peripheral nerve compression syndromes) are somewhat overshadowed by the preceding neurotoxicological chapters. This section of the book would have been enhanced by a discussion of neurological disorders in sportsmen or musicians. There is some overlap in the coverage of causation of entrapment neuropathy in chapters 11 and 12; amalgamation of that information into a single chapter might have been better.

Chapter 10 considers some of the medicolegal issues which are involved. Naturally, it is biased towards North American legal requirements. However, it does address certain principles necessary for proof that exposure has resulted in toxic damage. Expert witnesses practising in other countries will find this section useful when advising on cases of suspected toxic or occupational damage to the nervous system.

MICHAEL DONAGHY

Functional Neuroimaging. Edited by ROGER E KELLEY. (Pp 470; \$98). Published by Futura Publishing Inc, Armonk 1994. ISBN 0-87993-590-1.

This is a challenging and exciting title and I approached it with eager anticipation. A quick look through seemed to suggest that the title was reasonable but a number of points caught my eye which dampened my enthusiasm.

A very large image demonstrating "subtle haemorrhage" is shown but omitting mention of the large bilateral subdurals. There are many anatomical images but relatively few functional images (though there is an appendix with colour plates), long lists of the causes of clinical disorders and even the International Classification of Headache. Closer inspection of areas of personal interest raised more doubts. It is stated that "blood brain permeability can be measured non-invasively with MR by the injection of Gadolinium". This is a non-sequiter and only partially true as the relationship to signal intensity and Gadolinium concentration is not as easy to define as with CT, where there is substantial body of literature which has been ignored. The chapter on the spine includes a whole page on the history of prolapsed intervertebral disc herniations, fascinating but irrelevant. "Infections" almost completely ignored functional aspects.

If I had seen this in a bookshop it would probably have been returned to the shelves. However there is much to commend in this book. The various techniques of functional imaging are described and to a degree, put into context clinically. This provides a good introduction to bridging the gap between form and function. Unfortunately the title owes more to marketing than its content. In spite of the criticisms I strongly recommend it for those who wish to go beyond the basic

texts on conventional neuroradiology. I have already put it in the "priority purchase" category for our departmental library.

CHARLES FREER

Minimally Invasive Neurosurgery II. Edited by B L BAUER AND D HELLWIG. (Pp 166 DM150). Published by Springer-Verlag, New York 1994. ISBN 3-211-82593-2

The prospect of having to undergo neurosurgery, and in particular intracranial surgery, is so horrifying that most patients idea of minimally invasive neurosurgery will be something they watch on television with the option to switch off if it gets too much. There is a clear perception that intracranial surgery is inherently dangerous, however carefully performed. Who, if offered the choice between minor surgery through a small opening, would not prefer that to major operation with an extensive opening of the head, provided the same result could be achieved. There is an expectation that smaller operations are less likely to cause damage and complications than larger operations. But that may only apply if the abnormality for which the operation is done is one that requires a small operation. The real question is whether major abnormalities, conventionally treated by large operations, can be managed at least equally effectively and with smaller risk by the smaller operations now being developed.

It is clear that if patients choose smaller operations, they expect them to be safer, but the experience of minimally invasive surgery within the peritoneal cavity, indicates quite clearly that these procedures are by no means free of risk and that complications can occur which are inherent in the smaller procedures and which would either not have occurred in the larger procedures, or if they did, could have been recognised and dealt with at the time. In the introduction to this book, there is a plea for scientific evaluation of new surgical treatment methods by randomised controlled trials and in the same way as for new drugs. The book deals largely with endoscopic neurosurgery and describes methods which are producing impressive results in the treatment of abnormalities that can be reached through the lateral ventricles, and also the treatment of some intracerebral lesions. There are sections on laser technology, magnetic field-guided endoscopic dissection and stereotactic surgery. This is only part of the armamentarium which is now available and which includes stereotactic radiosurgery and endovascular surgery for aneurysms and arteriovenous malformations. Hormonal control of meningioma growth may resolve the problems of management of many recurrent or difficult basal meningiomas. The book is an impressive statement of the current state of development of this field which should be of interest to all neurosurgeons who wish to keep in touch with current progress, even though they may not wish to practice it themselves. Training is clearly an important part of the development of these services and neurosurgeons will need to satisfy their peers, patients, and occasionally the lawyers, that they have the necessary skills and experience. This book makes an important contribution to a developing field.

RD ILLINGWORTH