This case illustrates how Chiari malformations may present at any age and should be considered even in septuagenarians. The onset of symptoms in the eighth decade is extremely unusual, but presentation at the age of 74 has been reported on other occasions. Symptoms may precede diagnosis by several years, as in this patient. However, it is unclear why a congenital malformation should suddenly become symptomatic. Several reports have highlighted the role of trauma, degenerative changes in the cervical spine and cerebrovascular disease that may also contribute to neurological deterioration. Pathal paresis, dysphagia, dyspnoea and ataxia are well recognised symptoms of type 1 Arnold-Chiari malformations,12 and recent reports have highlighted the respiratory problems that these patients may encounter.13 The presence of downbeat nystagmus strongly implied an abnormality at the cranio-cervical junction and this was confirmed, non-invasively, by imaging. Despite their ages both our patient and the one reported by Hosford and Spector made the one recovery following surgical decompression showing that age alone should not be regarded as a barrier to active management.

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Ramsay Hunt syndrome: to bury or to praise

"If there is one disease in the neurological literature that is difficult to define and diagnose, it is the cerebellar dysmyelination of Ramsay Hunt." Radermecker, 1974.

Our recent suggestion that the Ramsay Hunt syndrome (dysmyelination cerebellaris myelococica) is a distinct and useful diagnostic category1 has provoked considerable controversy.12 We reached this conclusion following a study of 84 cases of progressive myoclonus epilepsy (PME), of which 13 were previously regarded as Ramsay Hunt syndrome. Review and restudy of this material established the diagnosis of mitochondrial encephalomyopathy (MERRF) in 11 of the 13 cases; the remaining two cases were not available for reexamination.11 Those patients who wish to preserve the Ramsay Hunt syndrome differ widely in their concept of the disorder,11 which only reinforces our view that the term should be buried.

Tassinari et al recently reported a series of 13 patients with "Ramsay Hunt syndrome" who had onset of myoclonic or tonic-clonic seizures at ages 6 to 15 years with a mild cerebellar syndrome. Family studies suggested autosomal dominant inheritance and muscle biopsies failed to show evidence of mitochondrial disease. Tassinari’s patients are different from the cases that we reclassified as MERRF and we agree that they do not have mitochondrial disease. The clinical, electroencephalographic and genetic features of their patients are, however, identical to those of Unverricht-Lundborg disease (Baltic myoclonus) as described by the original authors and based on the earlier studies of Koskineni.11 There is no doubt that this disorder occurs outside the Baltic region. Although there is as yet no diagnostic laboratory in the Unverricht-Lundborg laboratory, the clinical picture is distinctive and a clinical diagnosis can be made with considerable certainty.11,12 We find the use of the term "Ramsay Hunt syndrome" for such patients is historically inaccurate and diagnostically misleading.

Tassinari et al have emphasised the electroencephalographic features of their patients with normal or mildly slow waking background activity, fast spike-wave discharges, photosensitivity and lack of activation during slow wave sleep.11 These findings are not different from those of Unverricht-Lundborg disease.11 Indeed, critical study of the EEG patterns in all the PMEs, including MERRF and Unverricht-Lundborg disease, reveals more similarities than differences.11 Tassinari et al have also described vertex spikes during REM sleep.13 Unfortunately, REM studies of Unverricht-Lundborg disease have not been reported. These spikes are, however, also seen in MERRF14 and we suspect they may be common to all the PMEs, much like giant evoked potentials, which they may in fact represent.

Unlike the situation previously, the vast majority of patients with PME can now be accurately diagnosed during life.13 Whilst occasional undiagnosed patients remain, there is no residual homogeneous group of cases for which the term Ramsay Hunt syndrome is appropriate. Radermecker’s frustration at attempting to define the Ramsay Hunt syndrome can at last be put to rest. In retrospect it was a true syndrome, with many causes, although we suspect that most reported cases were probably examples of MERRF. Its validity as a clinical category has been overtaken by clinical, genetic, biochemical and pathological advances in specific diagnosis.11

"We come to bury, not to praise..."

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6Marsden CD,Osojo JA. The Ramsay Hunt Syndrome is a useful clinical entity. Movement Disorders 1989;4:6-12.

Tassinari et al reply: We thank Drs Berkovic and Andermann for the kind comments they made on our recent paper. We are glad to know that the patients previously referred to as Ramsay Hunt syndrome (RHS) by these authors and subsequently found to have a mitochondrial encephalopathy (MERRF) were significantly different from those we have described under the eponym of RHS. This fact supports our statement that RHS and MERRF are not different clinical, EEG and evolutive features. Drs Berkovic and Andermann however criticise the term RHS applied to our cases. In these patients the main clinical complaint was action myoclonus combined with rare generalised epileptic seizures: indeed this association was described by Ramsay Hunt under the heading "Dysnynergia Cerebellaris Myoclonica." Ramsay Hunt also emphasised the coexistence of "cerebellar ataxia" but, in such cases, the cerebellar component was difficult to define because of the presence of severe intention myoclonus, as recently pointed out by Harding. Thus, in our opinion, the use of the term RHS for our patients is justified,
with the viral, bacterial, fungal and protozoal infections seen in AIDS. The final section is on the use of diagnostic laboratories.

This structure has unfortunately led to frequent repetition. The epidemiology and clinical features of infections appear in both the second and third sections of the book. The classification of HIV infection appears in both section 1 and 2. The laboratory features of opportunistic infections appear in sections 2, 3 and 4. These problems of balance. In section 2 the nervous system, skin, mouth and paediatric patients are all dealt with in separate chapters. The other systems, gastro-intestinal, respiratory, cardiovascular, endocrine and renal are all discussed in a single chapter. This chapter is further crowded by the author’s uncertainty as to whether he is addressing the diagnosis of HIV infection in general medical patients or the diagnosis of complications in HIV infected patients. In section 3 rarer infections, such as the endemic mycoses, are also given space disproportionate to their importance.

The delay between writing and publication (there are occasional references from 1988) is less of a problem than might be expected in this fast moving field. Examples of new developments would indicate that the confusion in proctocutaneous meningitis or of desaturation or immunofluorescence in the diagnosis of Pneumocystis carinii pneumonia. More frustrating, however, is the reliance on abstracts from AIDS conferences, rather than published sources, as references.

This book is designed to be a practical guide to the management of opportunistic infections in AIDS. There are however some inconsistencies. For example, there is a long chapter on the epidemiology of HIV infections and a chapter on acute HIV infection but not on other complications attributed to HIV itself. The book has also excluded some areas which are of indirect interest in the diagnosis and management of opportunistic infections. For example the use of zidovudine as an adjunct in opportunistic infections, the place of visceral Kaposi’s sarcoma, lymphoma and HIV itself in the differential diagnoses of opportunistic infections. This raises the question of the validity of the separation of the management of opportunistic infections from that of other complications of HIV infection, particularly when some “non-infective” complications, such as Kaposi’s sarcoma or lymphoma may be related to various viral infecions.

Each chapter in this well produced book provides a good introduction to its subject by an authority in the field. The neurological chapter is good, though neurologists would find it brief and prefer the book “AIDS and the Nervous System” edited by the same authors. However the whole is less than the sum of its parts. It is expensive and individuals or cash restricted libraries wanting an introduction to AIDS would be better advised to buy one of the more comprehensive texts on the subject which are now available.

Geraint Fuller


In the late seventies the problems created by a failure of the autonomic nervous system were more clearly recognised and described in the current literature. In 1982 in the first edition of “Autonomic Failure”, Sir Roger Bannister suggested that the protean symptoms of autonomic failure would lead patients towards specialists in many disciplines, for example the cardiologist, the neurologist, the general physician, and since many of the disorders occur late in life, the geriatrician. It was the editor’s aim at this time to provide a book which would guide these specialists to an understanding of the many problems when the autonomic nervous system failed.

The first edition was very well received and now the second edition appears aimed at providing a comprehensive scientific basis for the diagnosis and treatment of the wide range of autonomic disorders which are being recognised with increasing frequency.

Sir Roger edits what is now a multi-author volume with contributions from some 30 new authors of whom 14 are from countries outside the UK. The many advances in the basic science of autonomic function have been considered and special attention is given to recent progress in peptide chemistry and immunocytochemical staining.

In an introductory chapter the editor outlines the problems of autonomic failure, classifies the disease processes responsible for the disintegration of the system and summarises the new techniques used in the investigation of such patients. The embryo, the basic anatomy and physiology of the autonomic system are described in great detail with chapters devoted to the influence of the autonomic system on cardiovascular function, blood pressure regulation, heart rhythm and cardiac function.

The second part of the book, which is of main interest to the clinician, is devoted to the clinical and pathophysiological features of autonomic failure. The lead chapter by the editor is followed by a succession of essays on the investigation of the various disorders, their clinical phenomena and the management of symptoms. A large section is devoted to the clinical presentation of diabetic autonomic neuropathy. It would seem that nothing has been excluded from this very comprehensive essay.

This book must remain the outstanding contribution to the study of the autonomic nervous system. Not only is it up to date, it is accurate, well edited and will provide a useful reference source for any clinician who suspects that his patients have autonomic neurological problems. I feel the essays are uniformly consistent and therefore would select no particular author for mention. This reflects upon the overall competence of the editing. Each essay is followed by a comprehensive list of references, some as recent as 1987, suggesting that the book, published in 1988, certainly does discuss truly recent advances.

J B Foster


This new textbook seeks to emphasize the importance of neurology in general medicine and to provide a practical approach to diagnosis and treatment for general physicians, MRCP candidates and neurologists in training. Set against the mass of recent texts on

BOOK REVIEWS

Opportunistic Infections in Patients with the Acquired Immunodeficiency Syndrome. (Infectious Disease and Therapy Series 3). Edited by: curroro s and Edward F. Mills. (Pp 476; Illustrated; Price: £89.75 (US and Canada) £68.00). New York: Marcel Dekker Inc. 1989.

“Opportunistic infection in patients with the acquired immunodeficiency syndrome” is the latest in the infectious disease and therapy series from Marcel Dekker. The stated aim of this book is to provide a practical approach to the management of opportunistic infections in HIV infected patients. The good response to appropriate treatment of some infections is stressed. It is a multi-author book principally from the San Francisco group.

The book is divided into four sections. The first deals with the epidemiology and immunology of HIV infection. The second section is on patient assessment. The third section deals