PostScript

CORRESPONDENCE

The naming of parts

Many deplore the journalistic trend to label well recognised conditions by acronyms, or by recently invented names—commonly to no useful purpose. Thus neurologists may not welcome yet another two names, recorded in past literature but not in general currency. Unquestionably they give a most valuable and timely review of the dropped head and bent spine syndromes.1 Why perpetuate the un anglais phrase head ptosis? Ptosis, Greek πτωσις = falling, has traditionally been applied only to the upper eyelid and to prolapse of any of the viscer or of the breasts. Head drop is short and its meaning is unequivocal.

Un questionably, Greek κορµος = trunk of a tree: but, like a few others,2 Umapathi et al apply it to signify a bent spine. Camptocormia was first an illness occurring among soldiers in World Wars I and II, and was regarded as a sign of hysteria. Ankylosing spondylitis is a more frequent cause. It is an ostentatious word that portends something more mysterious than a bent back or neck. To be similarly pretentious, can we not survive without such euphuisms?

J M S Pearce

Anlaby, East Yorks, UK; jmps@freenet.co.uk

References


Head drop and camptocormia

The article by Umapathi et al in this journal referred to the original use of the term camptocormia by Souques in 1915,3 though functional bent back was first described by Brodie in 1837. Mlle Rosanoff-Saloff supported Soutard’s fine collection of shell shock cases; this soldier was wounded five months previously by a bullet that entered along the spine. Hence the title “Head drop and camptocormia, the spectrum of bent spine disorders”.4

It would be difficult to doubt the probability that psychological factors influenced these men’s recuperation. To describe these soldiers as being “sick” though this was the terminology used during this period, or indeed that they suffered functional bent back, is probably unfair. They may well have suffered acute traumatic spinal injury and reactive muscle spasm (and contractures). Persistent stooping in shallow trenches, in appalling conditions of deprivation and danger, may have been contributing factors weakening the tone of paraspinal muscles. However, these case reports suggest that the traumatic injury alone may be sufficient explanation for the bent spines. The management of camptocormia in the first world war was to provide biomechanical supports, such as corsets, apparently with good results. The psychological therapies of “presuasive re-education” were additive rather than pivotal, and faradisation (and other tortures) used only “if necessary.”

The Sandler trial of low self esteem with confusion of identity, sadomasochistic behaviour towards military authorities, and impotence were, in 1947, proposed as being an essential part of camptocormia. Umapathi’s recognised causes of camptocormia and the contributing factors however implicate organicity, as indeed do the original case reports.

A D Macleod

Psychiatric Consultation Service, Christchurch Hospital, Private Bag 4710, Christchurch, New Zealand. PCS@cdhb.govt.nz

References


Author’s reply

We would like to thank Dr J M S Pearce for his comments. We agree with him on the proliferation of medical terms referring to similar if not identical conditions. One of the chief aims of writing this paper is to thread a line of commonality through the various names in literature, which in essence refer to an anterior curvature of the spine. Hence the title “Head drop and camptocormia, the spectrum of bent spine disorders”.

However, we would like to disagree with Dr Pearce labelling the spinal deformity seen in ankylosing spondylitis as camptocormia. In arthritic conditions and diseases that affect bone, the spinal deformity is fixed. In the bent-spine disorders referred to in the paper, the deformity may reduce considerably and even disappear with change in position, for example when supine. We would therefore prefer to reserve the phrases head drop (used interchangeably with head ptosis) and camptocormia to neurological conditions that affect the spine or back of the muscles controlling spinal posture.

As aficionados of medical history, we very much enjoy Dr A D Macleod’s letter. We agree that organic factors were probably present when the camptocormia in soldiers believed to have been suffering from hysteria. It would have not been unexpected for patients, like the man described by Southard with a bullet wound near the spine,5 to have developed spasm or even denervation of thoracic paraspinal muscles.

T Umapathi

Department of Neurology, National Neuroscience Institute, 11 Jalan Tan Tock Seng, 308433 Singapore. tumapathi@yahoo.com

Reference


Infection and multiple sclerosis

The article by Hawkes6 and the editorial commentary about the role of infectious agents in multiple sclerosis (MS) examined this question from a new viewpoint based on epidemiological observations.4 Several infectious agents, most not sexually transmitted, were reported to be associated with MS according to epidemiological data, serology in CSF and blood, or demonstration of pathogens in tissue. A relation with measles virus (MV) has been an early and most consistent finding. More recently, higher prevalence and higher titres of antibodies against human herpesvirus 6 (HHV6), but not other herpesviruses, were shown in MS patients compared to control groups, suggesting different exposure to HHV6 in MS.7 HHV6, like vaccine strain MV and certain wild type MV, uses the membrane cofactor protein (MCP; CD46) as a receptor for entry into cells. This suggests a possible involvement of CD46 in MS.

The possibility of a particular isoform of CD46 predisposing MS patients to infection is unlikely because all isoforms have similar affinity to MV. Increased levels of soluble CD46 have been reported in the serum and cerebrospinal fluid of MS patients more in those who have HHV6 DNA.8 One interpretation of these findings involved increased activity of the complement system in MS. However, experimental studies show no influence of inflammatory cytokines on CD46 expression and do not support inflammation.

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as a cause of increased CD4. Incorporation of CD46 in the viral envelope, or a possible genetic propensity in MS patients, has also been considered as causes of increased CD4. While its origin in MS is unclear, soluble CD46 might be involved in viral pathogenesis by binding the virus in the viremic phase and allowing another to attach to CD46 and spread from cell to cell. Both HHV6 and MV are infectious agents encountered in early childhood, and HHV6 can indeed become reactivated a few weeks after primary MV infection. On the other hand, because HHV6 and MV downregulate CD46 expression on the infected cell, they may diminish the efficiency of each other, delaying the time of infection. Therefore, they might produce increased antibody levels in young adults through delayed infection with, or reactivation of, each other. These suggest increased antibodies against these two viruses in MS may be interrelated.

The question remains whether a cause-effect relation exists between infectious organisms and MS, or whether viruses are just a consequence of the activation of the inflammatory-immune sequence or increased susceptibility of MS patients to infection. Studies of CD46 and other viral receptors seen warranted in MS.

Infection and multiple sclerosis

The paper by C H Hawkes (Is multiple sclerosis a sexually transmitted infection?) has caused predictable distress to people with multiple sclerosis (MS) and their families. Living with MS is a difficult enough experience without such sudden and avoidable alarm. The UK Multiple Sclerosis Society's national helpline and local branches have been inundated with calls expressing anger and anxiety. It is hard to understand the motive for publication when your own expert editorial commentary specifically referred to the paper's “pure speculation” and “potential to cause harm”. Did the sensational nature of Dr Hawkes' hypothesis and the virtual guarantee of extensive publicity it could receive outweigh proper consideration of its scientific merit?

There is also the worrying question of what damage may have been caused to the reputation of MS research in the UK by the lay media coverage which was attracted. The MS Society has a current forward commitment of around £12 million to nearly 70 research projects. That money is raised by voluntary donation. Anything which could discredit the quality of research here is of material concern to us.

M O'Donovan
Chief Executive, The Multiple Sclerosis Society, M S National Centre, 372 Edgware Road, London NW2 6ND, UK; modonovan@msociety.org.uk

Reference

Delirium in old age

Ed: The journal regrets any distress caused to patients with MS as a result of the widespread publicity this article received in the media. However, we wish to emphasise that the article was subject to the usual peer review process.

BOOK REVIEWS

Delirium in old age


Delirium is an extremely important condition for a number of reasons. It is very distressing and frightening for those who experience the symptoms, and descriptions of the effects on the brain as a result of high fever have been well described. There is a high mortality associated with the development of delirium, and it is often associated with behavioural disturbance that can be troublesome for carers and attendants. Finally, it presents a unique opportunity to look at the interface between psychiatric symptoms caused by organic disease and functional disorders.


Neurophysiology in neurosurgery. A modern intraoperative approach

Edited by Vedran Deletis and Jay L Shils (Pp 469, $125.00), Published by Academic Press, California, 2002. ISBN 0-12-209036-5

This book comprises 17 chapters contributed by 24 authors. It has clearly benefited from most of the chapters being written in a more or less homogenous style and formed into seven parts mainly based on surgical procedures: motor evoked potentials/ neurophysiological base; intraoperative neurophysiology (ION) of the spinal (spinal cord monitoring); ION of peripheral nerves, nerve roots and plexuses; ION of cranial nerve and brainstem; ION of supratentorial procedures; ION during stereotactic neurosurgery for movement disorders; and ION and anaesthesiatherapy management. Most of the chapters cover the background of methodological description of the surgical procedure, and the related neurophysiological procedure, personal experience, and case reports, which gives a balanced theoretical and practical view on the topic of each chapter. One interesting summary approach taken in this book will ensure it has a wide range of readers across “neurosurgery, neurology, orthopaedic surgery, neurophysiology, anesthesiology, interventional radiology, and biomedical engineering”.

Chronic deep brain stimulation or neuro-modulation has extended the role of clinical neurophysiology beyond its traditional diagnostic role. This new field is touched upon briefly in the part on ION during stereotactic neurosurgery. An interesting feature of this book is that it is accompanied by a CD that certainly enhances its value. Cross references are given at the end of the corresponding chapter rather than in the list of contents in the book, and at the front page of the display.

In conclusion, it is an authoritative review of intraoperative neurophysiology much weighted on the motor system for a wide range of surgical procedures. Perhaps, in its present form, those hoping for a more systematical (informed) review of intraoperative neurophysiology of the sensory system may feel slightly disappointed.

X Liu, T Z Aziz

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Role of proteases in the pathophysiology of neurodegenerative diseases


This volume would be an extremely useful addition to the bookshelf of anybody with an active interest in the biochemical and pathophysiological processes that underlie some of the more common neurological diseases. In the past the role of proteinases in these disorders has been largely neglected because it was assumed that it represented a general non-specific metabolic process. In terms of attracting research interest the field also suffered from the confusion in the literature concerning the naming of these enzymes and the fact that the same enzyme might have many different names. However, as the editors point out in their preface, this is no longer the case and they have therefore brought together an impressive array of current research on the involvement of proteinases in a wide variety of disorders. From what individually might have been regarded as disparate studies, one can now start to see common themes not least of which is the potential therapeutic value of targeting specific proteinases and the development of specific inhibitors.

If, like me, you don’t have specialist knowledge of this area I would recommend going straight to the last chapter on the mammalian proteinase genes. Here you will find a clearly laid out summary of the classification and characteristics of the four main groups of proteinases (serine, cysteine, aspartic, and metallo-proteinases). I also found the chapter on the ubiquitin/proteasome system and the normal physiological breakdown of proteins particularly informative. Having read these two chapters you then have a wide choice of disorders and proteinases to choose from. Perhaps the most widely discussed is Alzheimer's disease, undoubtedly because of the huge advances that have been made in understanding the biochemical processes underlying this disease over the past 15 years. Papain-like cysteine proteinases (cathepsins), caspases, calpains, and a novel metalloendopeptidase (EC 3.4.24.15) all appear to have some role in the pathology of Alzheimer's disease and may, therefore, be potential targets for drug development. There is also a group of Alzheimer's disease specific proteases that affect the processing of the Amyloid Precursor Protein (β, β', and γ Secretase) and presenilin (presenilinase).

Both of these proteins are central to the development of pathology and so these enzymes in particular are key targets for current drug research company.

Apart from the interest in Alzheimer's disease, there are other chapters covering the role of matrix metalloproteinases and calpain in the demyelination of multiple sclerosis and the key role of calpain in the pathology of traumatic brain and spinal cord injury.

Further chapters describe the loss of calcium homeostasis and the subsequent pathological activation of calpain, resulting in the breakdown of key structural proteins in some neuromuscular disorders.

In summary, this book has something for everyone in an area of research that holds huge promise for the future in terms of developing useful therapies for treating neurodegenerative disorders.

S Gentleman

CORRECTIONS

The following abstract was not printed with the article by E L J Hoogervorst, M J Elkermenboom, B M J Uitdehaag, and C H Polman (One year changes in disability in multiple sclerosis: neurological examination compared with patient self report) in the April issue of JNNP (J Neurol Neurosurg Psychiatry 2003;74:439–42).

Objective: To characterise the relationship between one year changes in neurologist rating of neurological exam abnormalities as measured by the EDSS and changes in patient perceived disability as measured by the GNDs in patients with MS.

Methods: 250 patients with MS were recruited at an outpatient clinic. Disability at baseline and one year follow up was assessed using the EDSS and GNDs. Correlations between change in EDSS, GNDs–sum score, functional systems, and GNDs subcategories were studied as well as the significance of changes in EDSS associated with changes in perceived disability.

Results: The correlation between one year changes in EDSS vs GNDs was substantially lower (0.19) than cross–sectional correlations between EDSS and GNDs, either at baseline (0.62) or at follow up (0.77). Notably, changes in functional system scores that are based on neurological examination are poorly or not at all correlated with changes in disability as perceived by the patient. Analysing the impact of a significant worsening in EDSS score we found that this was associated with significant worsening, insignificant change, and significant improvement in the patients’ perceived disability in 45%, 39%, and 15% of patients, respectively.

Conclusion: Patients’ perception of change in disability differs not only quantitatively but also qualitatively from that of an examining physician. There are true differences in change as perceived by the patient and measured by the physician and changes in many dimensions of disability are relevant to the patient and have no measurable impact on the EDSS.

The authors of the short report entitled Paraneoplastic ophthalmoplegia and subacute motor axonal neuropathy associated with anti-GQ1b antibodies in a patient with malignant melanoma, published in the April issue of JNNP 2003;74:507–9), were listed in the incorrect order. The author order should read as follows: L Kloos, C W Ang, W Kruit, G Stoter, and P Sillevis. 