Paediatric neuro-Behçet’s disease presenting with optic nerve head swelling

EDITOR,—Involvement of the central nervous system in Behçet’s disease (BD) occurs in approximately 10–49% of patients and the frequency of ocular manifestations is believed to be in the range of 28–80%.

Neuropapillitis has been reported very rarely as a manifestation of Behçet’s disease.1 As far as we know, there has been no report of paediatric onset neuropapillitis in BD. Here we report a case of neuropapillitis in a 10 year old girl.

CASE REPORT
A 10 year old Omani girl presented in August 1998 with complaint of right sided headache and blurring of vision in the right eye for 10 days and complete loss of vision in the same eye for 5 days. There was no history of vomiting, loss of consciousness, convulsion, or any other neurological deficit. On admission, examination revealed total vision loss in the right eye. The vision was 20/20 in the left eye.

Examination of right eye showed relative afferent pupillary defect and fundus examination revealed blurred disc margins with no apparent cup. There was a 2 dioptre elevation of the disc (Fig 1).

Visual evoked potentials (VEP) and brainstem auditory evoked potentials (BAEP) on the right side were prolonged in latency and poorly formed. Magnetic resonance imaging of the brain revealed left sided temporal and parietal lesion suggestive of inflammation/demyelination. The child was started on methylprednisolone 500 mg intravenously in two doses followed by 15 mg/day for 2 weeks. Vision returned on day 2 with perception of hand movements, finger counting day 4, and complete recovery on day 8. VEP was repeated and showed normal response.

She was admitted in September 1999 with complaint of headache, diplopia, and visual blurring. She had meningeval signs, left lateral rectus palsy, and bilateral papilloedema. Cerebrospinal fluid (CSF) examination showed opening pressure of 210 mm Hg, closing pressure of 140 mm Hg, with CSF protein 0.36 (0.15–0.50 g/l), glucose 3.8 (2.2–3.9 mmol/l). White blood cells (WBC) 23, 60% lymphocytes, 40% polymorphs, RBC 10, and no organisms. Chest x ray was normal, Mantoux test was negative, blood culture and CSF had no growth after 48 hours. The clinical picture was suggestive of aseptic meningitis with raised intracranial pressure. The child was treated with systemic steroids over 2 weeks and improved with complete recovery of the sixth nerve palsy and resolution of papilloedema. In November 1995, she presented with left supranuclear facial nerve palsy and left hemiparesis, with hemihypospasticity. There was no papilloedema. CT showed right side thalamic and posterior internal capsule infarct. There was complete recovery after 1 month. During follow up the child had aphthous ulcer and skin rash. Skin biopsy showed prominent lymphocytic infiltration of the vessels in the dermis consistent with vasculitis, possibly Behçet’s disease.

COMMENT
Neuro-Behçet’s disease is an uncommon presentation in childhood. This patient had four episodes of neurological involvement and a skin rash once. Initial manifestation was raised intracranial pressure with aseptic meningitis-like picture and no underlying cause was found. The child recovered with steroids (prednisolone) and diuretics (acetazolamide), which are recommended in the severe form of Behçet’s disease. Later the child had a similar incident that also recovered with steroids. There was no underlying cause found. It was on follow up, when the child had aphthous ulcer and skin rash, that diagnosis of Behçet’s disease (neuro-Behçet’s) was made. With time (last admission) she presented with loss of vision in right eye and investigations—for example, VEP, suggested neuropapillitis.2 The diagnosis of Behçet’s disease was made on two major criteria oral aphthous ulcer, eye involvement and minor criteria of skin biopsy suggestive of lymphocytic vasculitis,3 CNS involvement and abnormal changes in the magnetic resonance imaging (MRI).4 We recently reviewed the ocular manifestations of BD in 32 patients. Inflammatory involvement of the optic nerve in the form of papillitis was reported in their three patients. Our case here is the first reported case of inflammatory optic nerve involvement in the paediatric age group, and clinically resembles multiple sclerosis (MS) as reported in adults by Kansu et al.2 Multiple sclerosis is rare at this age and the presentation of aseptic meningitis, brain edema, skin lesion, and aphthous ulcer, with MRI showing a single patch of demyelination, excludes this possibility. The MRI of the optic nerve was normal despite abnormal VEP in our case.

Unless the patient presents with clear symptoms, the diagnosis of Behçet’s disease is difficult initially. The diagnosis in children is more difficult as the disease is uncommon and usually the diagnosis is made after several systemic presentations and the exclusion of other common diseases.

SANDIP MITRA R L KOUL
Department of Ophthalmology, Sultan Qaboos University Hospital, PO Box 18, Alkhod, Muscat 123, Sultanate of Oman


Figure 1 The right eye fundus picture shows blurred disc margins with full cup and elevation of 2 disc diameters.

REFERENCES

Reply
EDITOR,—We are grateful to the chairman of the Royal College Working Party responsible for the 1997 guidelines on ocular hypertension and glaucoma,1 Mr S Vernon, for highlighting our inadvertent omission of the Blue Mountains eye study paper2 which was available to the working party before the publication of the guidelines. This paper from our review was unintentional and unfortunate.

The strengths of this rigorous study are its population based sampling frame, its careful definition of diabetes, and its definition of glaucoma to include field loss matching disc changes regardless of intrasaccular pressure. The overall prevalence of diabetes was 7.0% of whom 98% had type 2 diabetes and 15% were newly diagnosed during the course of the study (by fasting glucose test). The prevalence of POAG was 3.0% (108 patients).3

Exactly half the glaucoma cases were newly diagnosed by the researchers. The prevalence of diabetes in the 54 pre-study diagnosed
POAG patients was almost twice that in the 54 "newly diagnosed" cases (nine cases in the former compared with five in the latter; age/sex adjusted odds ratio (OR) 2.12). The prevalence of diabetes in those with POAG differed significantly from that of the general population without glaucoma (6.9%) only in the latter group (age/sex adjusted OR 2.82, 95% CI 1.35–5.87). However tempting to suggest that unmasking bias may be operative in this group, it is noteworthy that six of the nine patients did not know they had diabetes at the time of diagnosis of POAG. Unmasking bias will only have been operative if the eye clinic is as likely to diagnose diabetes in referrals with possible POAG as it is to diagnose unmasking diabetes in referrals for possible retinopathy or diabetes related cataract. This is unlikely since random blood glucose estimation is probably not a routine test in a busy eye clinic.

Our review asked whether the published information available to clinicians and health planners could support targeted screening of diabetic patients. The suggested method was measurement of intraocular pressure. It is informative that in the Blue Mountain eye study only 25.5% of POAG patients had raised pressure. This then would seem an inappropriately insensitive means for screening.

The paper by Mitchell et al adds convincing support to the existence of an association between diabetes and POAG. The problem of how best to cater for this group remains. In light of the available evidence the advice of the Royal College of Ophthalmologists seems apposite; to be aware of the increased risk of POAG in this group of patients who attend for retinopathy screening.

Hertel exophthalmometry: the most appropriate measuring technique

EDITOR.—The common method used in Hertel exophthalmometry is to align the corneal vertex with the reference cone on the exophthalmometer and noting the position of this alignment on the millimetre scale (method 1). Keeler, the manufacturers of the exophthalmometer, recommend that the corneal vertex should be aligned with the reference line on the millimetre scale and the position of the anterior surface of the cornea on the millimetre scale noted (method 2).

Our study, in which two masked observers of differing experience examined 50 patients using these two methods, was set up to compare the results obtained using the two methods, determine if they could be used interchangeably, and whether we could recommend one method over the other.

Readings obtained by each observer from each eye with one method were compared with the readings of that same observer from the same eye using the different measurement method. A Wilcoxon statistical analysis was performed. Observer 2 was the more experienced.

There was a statistically significant difference between the readings made by observer 2. This is most probably clinically relevant and does show that both methods of measurement cannot be used interchangeably.

There was a 60–70% (method 1) and 30–40% (method 2) interobserver percentage agreement for readings from each eye and each measurement method. Agreement was present if the interobserver readings were within 1 millimetre of each other. These results may reflect the fact that both observers were more familiar with method 1. A follow up study, once the observers were more familiar with the Keeler method, should show an improvement in the agreement rate if this was the case.

We believe this study shows that a failure to make clear the measurement method used in exophthalmometry is both statistically and clinically significant false positive results which could have serious implications in the management of these patients.

BOOK REVIEWS


This is the long awaited second edition of this classic volume in the WHO international histological classification of tumours series. With the exception of Dr Sobin, only two of the participants in the second edition were involved in the preparation of the first and, inevitably, this reviewer turned to the first edition to compare it with the second. The second edition is now in a much more user friendly paperback format and the material has been reorganised in a more logical and simpler way than in the first edition. Further improvements can be seen in the end of the book rather than halfway through as it was in the 1980 version. Another major improvement in the second edition is the addition of the TNM classification of tumours of the eye and its adnexa. It makes sense to have the TNM classification included along with histological typing of tumours rather than having to refer to the separately published TNM classifications of malignant tumours whenever one is reporting. The photomicrographs are also generally of a higher quality than in the 1980 edition.

However, the second edition is not entirely an improvement on its predecessor. I would have to take issue with the claim on the back cover that it “is more extensive and detailed than the previous edition”. It is certainly more extensive with regard to the number of tumours mentioned, but not more detailed, as there are far fewer explanatory notes than in the first edition, and the explanatory notes suffer from a degree of selectivity for which is not always obvious. Furthermore, the classification suffers from the “overlap syndrome” and descriptions of skin tumours and CNS tumours would be better left to the relevant histological typing volumes. For example, the explanation of glioblastoma multiforme as “anaplastic pleomorphic cells with or without necrosis replace the normal optic tissue” is hardly a helpful description of what is, in reality, an extremely rare tumour of the optic nerve.

Although the photomicrographs are generally of higher quality, there are in fact fewer (112 £ 150) in the second edition and once again they are highly selected for reasons which are not immediately obvious—for example, there are four figures of a phacomatoses choristoma of the eyelid including immunocytochemistry but only one of the much commoner and more difficult to diagnose conjunctival neoplastic neoplasms. Final glimpses about the photographs relate to the absence of arrows to point out subtle features and inconsistencies in some of the figure legends—for example, “undifferentiated sebaceous adenocarcinoma” when, by definition, there must be sebaceous differentiation in order for the diagnosis of sebaceous carcinoma to be made.

All in all, I think the second edition is something of an improvement on the first edition. It is now more user friendly and the addition of the TNM classification is very helpful. However, the book still suffers from the problems which bedevil the whole series of WHO histological classifications—selectivity, “lowest common denominator” classification, and the construction of the classification by a small group of monospecialists. Nevertheless, the book will still be useful as a common language for use in ophthalmic oncology and a way of resolving disputes between pathologists at meetings.
Each chapter begins with a section on clinical evaluation including presenting symptoms, signs, and guidelines on investigations. Considerable emphasis is placed on pretreatment planning and the preparation of patients, not only in deciding which course of treatment to take, but also what the patient should be told for the purposes of consent. This information is often presented in handy checklists, which stand out in the text. The authors take the reader through all the steps in planning a treatment session from the choice of laser, delivery system and lens, to the location and spot characteristics of the laser burn. For each condition, an impressive array of fundus photographs and fluorescein angiograms, both pre- and post-treatment are presented. Suggestions are made concerning the frequency and nature of post-treatment follow up and this is often illustrated with the inclusion of real clinical examples.

The chapters on diabetic retinopathy are well referenced and treatment guidelines closely follow the ETDRS and DRS protocols. An extensive reading list is provided at the end of the chapters. One possible criticism is the lack of indexing of references within the text, which could make accessing a particular reference difficult.

The authors add their own well reasoned suggestions for managing more complicated clinical situations and a section on “special cases”, where there are other coexistent ocular conditions, is included.

An superb set of stereo fundus photographs and angiograms in slide form complement the chapter on choroidal neovascularisation. A stereo viewer is provided, which is very easy to use. At the start of the chapter, the reader is guided through the interpretation of both fluorescein and indocyanine green angiography.

Advice is given on the management of recurrent choroidal neovascularisation post-treatment, and this is illustrated with clinical examples. The authors give their own guidelines for treating lesions near the fovea. There is an extensive presentation of the Macular Photocoagulation Studies’ findings in summary form, with references. This is a very useful feature and will no doubt be helpful to those preparing for clinical exams.

Guidelines on the management of retinal tears and a variety of retinal vascular and chorioretinal conditions are presented in a separate chapter. The book concludes with a number of self assessment MCQs. These are chapter based and broadly cover the information presented. Detailed explanations are provided with the answers.

In summary, the book is compact, succinct, and easy to read with the essential information readily accessible through a well designed “contents” section. The treatment guidelines that are suggested are evidence based and follow the major multicentre trial findings. Important practical points are often included in highlighted checklists for easy reference. For those wishing to read further about a particular subject, the references are very comprehensive. With its practical emphasis, it would be a welcome addition to any eye department library.

I think individuals preparing for clinical examinations would find this book helpful, as would anyone wanting to learn how to perform laser treatment safely and effectively.

MARK COSTEN
The appendices contain practice guidelines issued by the College of Optometrists and more self assessment.

The book is written in a readable, chatty style with anecdotes and cautionary tales used to illustrate important clinical messages. The book is not a primer for the novice refractionist as it assumes basic knowledge and clinical method, nor is it a comprehensive manual of optics and refraction techniques, but rather an "experienced practitioner" offering a pragmatic companion to revision and refinement of technique "in the real world". While some techniques are expounded at length, others are mentioned only in passing or merely referenced, with some references quite historical. The ophthalmologist will find some of the references unfamiliar, but will be clearly directed to the relevant material to pursue. The text emphasises the need for flexibility of approach and the importance of familiarity with more than one method of examination or approach to achieve a desirable outcome and to satisfy the foibles of colleagues (examiners!). Thus it brings together material which might be used in vivo or examinations.

Overall, I found this an interesting read, with useful practical advice, and the case histories illuminating. I would direct the reader to peruse the introduction first so as to gauge the flavour of the text.

J A SCOTT

NOTICES

**Ageing and the eye**

The latest issue of Community Eye Health (no 29) discusses ageing and the eye. Included are papers on ageing and the eye from a global perspective; epidemiology; delivery of eye care to the elderly; and age related macular disease. For further information please contact Community Eye Health, International Centre for Eye Health, Institute of Ophthalmology, 11–43 Bath Street, London EC1V 9EL. (Tel: (+44) 171 608 6098/6910/6923; fax: (+44) 171 250 3207; email: eyeresource@ucl.ac.uk) Annual subscription £25. Free to workers in developing countries.

**Residents’ Foreign Exchange Programme**

Any resident interested in spending a period of up to one month in departments of ophthalmology in the Netherlands, Finland, Ireland, Germany, Denmark, France, Austria, or Portugal should apply to: Mr Robert Acheson, Secretary of the Foreign Exchange Committee, European Board of Ophthalmology, University College Dublin, 60 Eccles Street, Dublin 7, Ireland.

**International Agency for the Prevention of Blindness**

The sixth general assembly of the International Agency for the Prevention of Blindness will be held on 5–6 September 1999 at the Conference Centre, Beijing Friendship Hotel, Beijing, People’s Republic of China. The theme is "The right to sight". Further details: IAPB Secretariat, LV Prasad Eye Institute, LV Prasad Marg, Banjara Hills, Hyderabad 500 034, India (tel: 091-40-215389; fax: 091-40-248271; email: IAPB@lveye.stph.net).

**Ophthalmological Clinic, University of Creteil**

An international symposium on the macula will be held on 1–2 October 1999 at the Ophthalmological Clinic, University of Creteil. Further details: Professor G Soubrane, Chef de Service, Clinique Ophthalmologique Universitaire de Creteil, Centre Hospitalier Intercommunal, 40 Avenue de Verdon, 94010 Creteil, France (fax: 01 45 17 52 27).

**Jules François Prize**

The 2000 Jules François Prize of $100 000 for scientific research in ophthalmology will be awarded to a young scientist who has made an important contribution to ophthalmology. All topics in the field of fundamental and/or clinical research in ophthalmology will be considered. The application should be sent jointly with a curriculum vitae, the list of all publications, and three copies of the candidate’s 10 most relevant publications to Jules François Foundation Secretary, Professor Dr M Hansens, Dienst Oogheelkunde, de Pintelaan 185, B-9000 Gent, Belgium. Deadline for applications 31 December 1999.

**Joachim Kuhlmann Fellowship for Ophthalmologists 2000**

The Joachim Kuhlmann AIDS Foundation, Essen, Germany, is sponsoring two fellowships per year for ophthalmologists at a well known institute, who want to train in CMV retinitis and other HIV related ophthalmological diseases. The fellowships are valued at $US5000 each. Deadlines for applications are 31 January and 31 July. Detailed applications, including CV and publication list, should be sent to the Joachim Kuhlmann AIDS Foundation, Bismarckstrasse 55, 45128 Essen, Germany (tel: 0201 87910-87; fax: 0201 87910-99; email: jk-stiftung@t-online.de).

**XXXIV Nordic Congress of Ophthalmology**

The XXXIV Nordic Congress of Ophthalmology will be held in Reykjavik, Iceland, 18–21 June 2000. This meeting celebrates the 100 year anniversary of the Nordic Ophthalmology Conference. Further details: Iceland Incentives Inc, Hambrarborg 1–3, Is-Kopavogur, Iceland (tel: +354 554 1400; fax: +354 554 1472; email: incentiv@itn.is).

**5th International Viteoretinal Meeting–IV 2000**

The 5th International Viteoretinal Meeting–IV 2000 will be held in Parma, Italy, on 26–27 May 2000. The main topics will include “Hypotony and glucoma in vitreoretinal surgery”, “Internal limiting membrane surgery”, “Macula oedema”, “Open globe injuries”, and “News in retinal pigment epithelium”. Further details: C Cantu, MA De Giovanni, or S Tedesco, Scientific Secretariat, Institute of Ophthalmology, University of Parma, Via Gramsci 14, 43100 Parma, Italy (tel: ++39 0521 259106; fax: ++39 0521 292358; email: nuzzi@ipruniv.cce.unipr.it).

**DR-2000, International Forum on Diabetic Retinopathy**

The International Forum on Diabetic Retinopathy will take place on 7–9 September 2000 at the Palazzo Reale, Naples, Italy. Further details: Francesco Bandello, Congress Secretariat, MGR Congressi, Via Servio Tullio, 4, 20123 Milano, Italy (tel: 39 02 430071; fax: 39 02 48008471; email: dr2000@mgr.it).

**12th Afro-Asian Congress of Ophthalmology**

The 12th Afro-Asian Congress of Ophthalmology (Official Congress for the Afro-Asian Council of Ophthalmology) will be held on 11–15 November 2000 in Guangzhou (Canton), China. The theme is “Advances of ophthalmology and the 21st century”. Further details: Professor Lezheng Wu, Zhongshan Eye Center, SUMS, New Building, Room 919, 54 Xianlie Nan Road, Guangzhou 510060, PR China (tel: +86-20-8760 2402; fax: +86-20-8777 3370; email: lwuicv@gzsums.edu.cn).
Should diabetic patients be screened for glaucoma?

S A VERNON

Br J Ophthalmol 1999 83: 1096
doi: 10.1136/bjo.83.9.1096a

Updated information and services can be found at:
http://bjo.bmj.com/content/83/9/1096.2

These include:

References
This article cites 4 articles, 1 of which you can access for free at:
http://bjo.bmj.com/content/83/9/1096.2#BIBL

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/