Figure 1 Relationship between patient age and area of involvement of acute hydrops

Discussion

The present study demonstrates a clear inverse correlation between age and the severity of acute hydrops, as well as the likelihood of developing neovascularisation after its resolution. This correlation could be explained on the basis of severity of allergy and rubbing or other reasons related to structural differences in developing corneas such as variations in the distribution and orientation of proteoglycan glycosaminoglycans complexes with time.⁶ Like previous reports of neovascularisation following involvement of hydrops near the limbal arcades,⁴ the use of topical steroids was ineffective in preventing neovascularisation in our three cases. Topical cyclosporine A may play a role in suppressing the development of corneal neovascularisation⁷ and should be evaluated prospectively, along with a more aggressive regimen of intensive topical corticosteroids, in a clinical trial of treatment of severe hydrops that extends within 1 mm of the limbal vascular arcades.

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Paraneoplastic optic disc oedema and retinal periphlebitis associated with pineal germinoma

Cancer-associated retinopathy (CAR) is a well-described paraneoplastic syndrome that is mediated by antiretinal antibodies.¹ Inflammatory changes such as optic disc oedema and retinal vasculitis have not been reported in CAR. Although CAR has been reported with various tumours, there have been no reports of paraneoplastic retinopathy or optic neuropathy with pineal gland tumours.² We report a case of a novel paraneoplastic syndrome consisting of bilateral disc oedema and retinal periphlebitis in a patient with pineal gland germinoma.

Case report

A 14-year-old boy with a pineal gland tumour was referred because of blurred vision. Best-corrected visual acuity was 20/20 in the right eye and 20/20 in the left eye. Examination of the pupils and eye movements verified the presence of a dorsal midbrain syndrome. Anterior segment examination and intraocular pressures were normal. Dilated fundus examination showed bilateral disc oedema and retinal periphlebitis (fig 1). A complete systemic investigation for vasculitis performed by the rheumatology service was negative. Full-field electroretinography was normal. The neurosurgical team performed a ventriculostomy with endoscopic biopsy of the lesion. The biopsy results were consistent with pineal gland germinoma. At the time of ventriculostomy, the opening pressure was <10 cm H₂O. Cytological examination of cerebrospinal fluid obtained during ventriculostomy was negative for malignant cells. Taken together, these findings indicate that the optic nerve swelling was inflammatory in nature and not secondary to raised intracranial pressure or leptomeningeal tumour spread. After biopsy, the patient was started on a course of focal irradiation and chemotherapy (carboplatin, etoposide and ifosfamide).

We decided to observe the patient, and no treatment for the ocular inflammation was initiated. Before the initiation of radiotherapy

Figure 2 Western blot against optic nerve protein extract. A blot of optic nerve protein extract was probed with: lane 1, anti-enolase antibody; lane 2, anti-recoverin antibody; lane 3, serum from a healthy control; lane 4, serum from our patient at the initial presentation; and lane 5, serum from our patient 6 months after successful treatment of pineal gland germinoma.

and chemotherapy, serum was analysed for autoantibodies. Western blot analysis of the patients' serum against retinal protein extract was negative (data not shown), whereas the blot against optic nerve protein extract revealed a single band of approximately 35 kDa (fig 2). During follow-up 6 months after initial treatment of the tumour, retinal periphlebitis and optic disc swelling were resolved (fig 3). Western blot analysis repeated with the patients' serum obtained during remission failed to reveal the 35 kDa band (fig 2). Three years after the initial presentation, the patient remains in remission of his tumour without any evidence of optic disc oedema or retinal periphlebitis. There has been no evidence of metastatic disease, as confirmed by serial MRI scans.

Comment

We have reported the case of a patient with pineal germinoma who was found to have bilateral optic disc oedema and retinal periphlebitis. The inflammatory fundus findings, along with the presence of antibodies against a 35 kDa optic nerve protein, resolved with successful treatment of the tumour. CAR is known to be secondary to autoantibodies specific for aberrantly expressed retinal

Figure 1 Initial presentation of patient. Colour fundus photos demonstrate bilateral disc oedema and retinal periphlebitis.

Figure 3 Follow-up of patient. Six months after successful treatment of the pineal gland germinoma, the bilateral disc oedema and periphlebitis had resolved.

proteins in systemic tumours that cross react with proteins in retinal photoreceptors.³ Interestingly, the pineal gland is known to express ocular antigens.^{4,5} Cross reactivity between autoantibodies specific for tumour antigens with proteins in the optic nerve and retinal vessels could explain the inflammatory fundus findings observed in our patient.

Chang *et al*⁶ have previously reported a case of bilateral optic disc oedema with retinal periphlebitis in a 14-year-old boy with pineal germinoma. As in our patient, the inflammatory fundus findings resolved after successful treatment of the tumour. Paraneoplastic optic neuropathy has been associated with autoantibodies against 62 and 66 kDa antigens.² We propose that the paraneoplastic syndrome of optic disc oedema and retinal periphlebitis, which we and others have described in patients with pineal germinoma, represents a novel paraneoplastic syndrome. The 35 kDa autoantibody target identified in this report may be implicated and in fact unique to this syndrome.

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MAILBOX

The social impact of visual impairment

We read with interest the extended report by Hassell *et al*¹ about the adverse effect of quality of life in mild visual impairment (<6/12) and worse. Their demonstration of the impact of age-related macular degeneration on quality-of-life indices is an important illustration of the difficulties experienced by visually impaired people.

We have reported a similar experience in West Glamorgan (Wales, UK).² Our study was comprised of 66 patients registered as blind/partially sighted (59.1% because of age-related macular degeneration). The demographic characteristics mirrored this study (mean age 81.33 (SD 9.87) years; 69.7% female). We used an alternative index to the impact of vision impairment (IVI) questionnaire, the National Eye Institute visual function questionnaire (NEI-VFQ 25).³ This has similar categories, including general health, general vision, ocular pain, near activities, distance activities, social functioning, mental health, role difficulties, dependency, colour vision and peripheral vision.

Hassell *et al* showed a statistically significant restriction in leisure and work, social and consumer interaction, and household and personal care difficulties between those with a mild/moderate impairment and those with severe impairment. This was our experience in the analogous categories of social functioning (NEI-VFQ score 36.49% for the blind vs 50% for the partially sighted; $p < 0.001$) and depen-

dependency (NEI-VFQ score 30.07% for the blind vs 46.98% for the partially sighted; $p < 0.001$). Furthermore, percentage scores in our study were <50% for all categories except general health, ocular pain and colour vision for blind and partially sighted people. None of the studies that have used the NEI-VFQ to determine the extent of visual function, have previously demonstrated mean scores as low as in our study.^{4,5} Hassell *et al*¹ have, therefore, shown a worse quality of life in severely visually impaired people (<6/60), and also reinforced the important fact that all people with visual impairment experience difficulties.

A second interesting finding in their paper was the absence of correlation between duration of visual impairment and adaptation. We found that the NEI-VFQ scores for those living alone were better than those for individuals living with someone for numerous categories including near vision activities (20.9% vs 15.3%, $p = 0.03$), distant vision activities (27.9% vs 20.1%, $p = 0.056$) and dependency (46.3 vs 31.4%, $p = 0.004$; Williams GP, Pathak-Ray V and Austin MW, unpublished data, 2001).

This may imply that living alone forces people to adapt. We interpreted this finding with caution, however, as a number of people “living with someone” resided in a nursing/residential home. This may have occurred as a result of living alone, causing difficulties with coping in the first instance.

Finally, the paper by Hassell *et al* was undertaken before people had received low vision services. Depressingly, our study found that there was incomplete delivery of formal low visual aid assessment ($n = 44$, 66%); 70% of these people found the aids to be of use. We, therefore, agree with Hassell *et al* referral to low visual services should be considered, and adequate delivery and support is required for it to be effective.

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