a novel phenotype in our patient in the Alström locus. As this is a single case, we cannot be certain that the retinal exudation is definitely caused by this mutation.

Our patient however had unilateral exudative retinopathy. This may be due to an asymmetry of the retinal dystrophy between the eyes. The natural history of this retinopathy is unknown and the beneficial results of treatment varying from photocoagulation,[8] cryotherapy[9] and pars plana vitrectomy[10] are difficult to quantify.

It is possible that in time, the other eye may become affected, probably as the retinal dystrophic changes progress.

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doi: 10.1136/bjo.2005.088781

Accepted 30 August 2006

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Inverse relationship between age and severity and sequelae of acute corneal hydrops associated with keratoconus

Acute hydrops occurs in approximately 3% of eyes with keratoconus.[1] The incidence increases dramatically in eyes with associated vernal keratoconjunctivitis.[2 3] Neovascularisation rarely develops after resolution of acute hydrops although it has been reported with large area of involvement with hydrops associated with either close proximity to the limbal vascular arcades[4] or intrastromal clefts.[5] The association between patient age and area of involvement of hydrops, with the subsequent risk of development of neovascularisation, has not been adequately addressed in a prospective clinical trial.

In Saudi Arabia, patients present with severe keratoconus at a much younger age than in Western populations and have a higher incidence of associated atopic eye disease.[2 3] The average age at the time of penetrating keratoplasty in our patient population is 19 years, with nearly one quarter of all cases performed in children 15 years of age or younger.[3] Approximately one fifth of cases have severe vernal keratoconjunctivitis or seasonal allergic conjunctivitis, 30% have a history of hydrops prior to corneal transplantation,[3] and one quarter of the cases have penetrating keratoplasty at 5 years of age or younger.[3] In this study we evaluate the influence of early age on the severity and sequelae of acute hydrops.

Methods

After obtaining approval of the Institutional Review Board of the King Khaled Eye Specialist Hospital, all patients with keratoconus who presented to the emergency room with acute hydrops between February 1, 2004 and July 31, 2004 were enrolled in a prospective, observational clinical trial. A comprehensive ocular examination was performed at the time of presentation and after resolution of the hydrops. The location of acute hydrops was identified and measured using slit lamp examination of the hydrops and its relation to the distance from the limbus. Also, the presence or absence of vascularisation was noted at the time of hydrops. All patients were treated with a tapering regimen of topical fluoromethalone 0.1% and lubrication.

Results

Thirteen patients (eight males, five females) were enrolled in the study (table 1). The median age was 20 years (range, 11–36). Complete resolution of hydrops occurred after a mean of 10.2 weeks (range, 6–13). Seasonal or vernal keratoconjunctivitis was present in nine (69.2%) eyes. There was an inverse correlation between age and area of involvement of corneal hydrops and the likelihood of developing neovascularisation after resolution (table 1, fig 1). Based on the Fischer exact test, patients 16 years of age or younger (5 out of 13) were significantly more likely to have an area of involvement of more than 90 mm^2 ($p = 0.03$), to have involvement less than 1.0 mm of the limbal vascular arcades ($p = 0.02$), and to develop neovascularisation after resolution of the hydrops ($p = 0.03$) than older patients. Proximity of the hydrops of less than 1.0 mm from the vascular arcades was significantly correlated with the risk of development of neovascularisation ($p = 0.03$). Although atopic disease was present in all 3 eyes that developed neovascularisation, there was no significant correlation between the presence of atopic disease and the likelihood of development of neovascularisation.

Table 1 Case Summaries

Case	Age, gender	Hydrops size Max., Min. (mm)	Area (mm^2)	Limbal Vascular arcade proximity (mm)	Duration of hydrops resolution (weeks)	Vascularisation after resolution	VKC/SAC
1	11, F	10, 9	90	0.2	13	Yes	Yes
2	12, M	8, 8	64	1.0	11	No	Yes
3	13, M	10, 9	90	0.2	12	No	Yes
4	14, F	10, 10	100	0.1	12	Yes	Yes
5	16, M	8, 8	64	0.1	12	Yes	Yes
6	19, F	6, 5	30	1.5	8	No	Yes
7	20, M	7, 7	49	1.5	9	No	No
8	22, M	9, 7	63	0.2	9	No	Yes
9	23, M	7, 7	49	1.7	6	No	No
10	24, M	8, 8	64	1.0	11	No	Yes
11	31, F	5, 5	25	2.0	10	No	No
12	33, F	7, 6	42	1.5	9	No	Yes
13	36, M	6, 4	24	1.3	11	No	No

VKC, vernal keratoconjunctivitis; SAC, seasonal allergic conjunctivitis; Max, maximum; Min, minimum

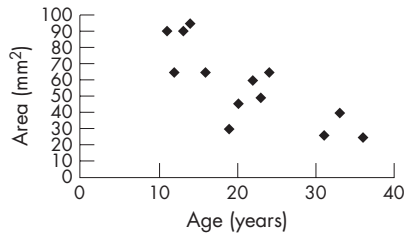

Figure 1 Relationship between patient age and area of involvement of acute hydrops

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 neovascularisation[7] 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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doi: 10.1136/bjo.2005.085878

Accepted 7 November 2005

Competing interests: None.

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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 H_2O. 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 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.

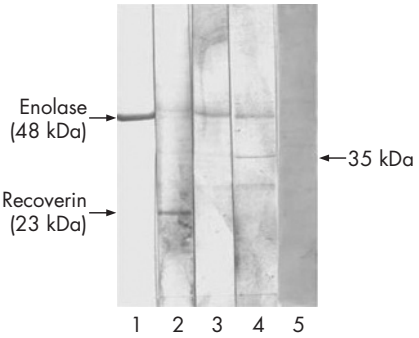

Figure 2 Western blot against optic nerve protein extract. A blot of optic nerve protein extract was probed with: lane 1, anti-enolase antibody; lane 2, anti-recoverin antibody; lane 3, serum from a healthy control; lane 4, serum from our patient at the initial presentation; and lane 5, serum from our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

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

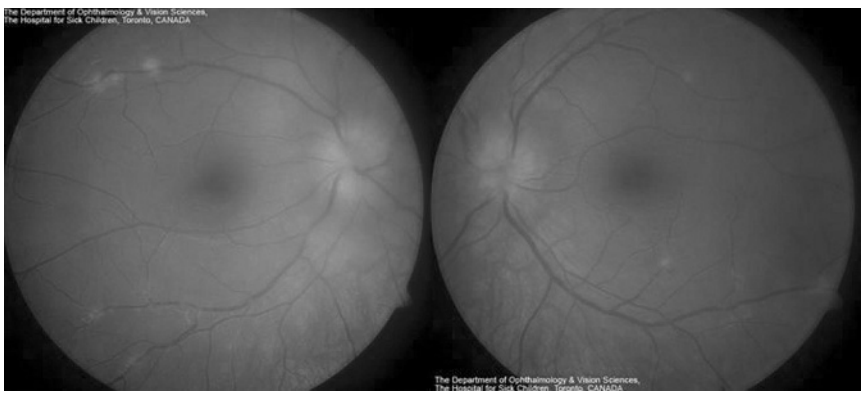

Figure 1 Initial presentation of patient. Colour fundus photos demonstrate bilateral disc oedema and periphlebitis.