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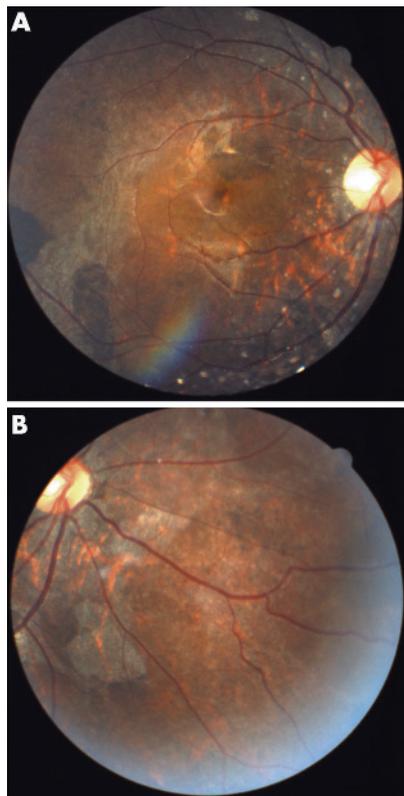


Figure 1 Fundus photograph of the right eye showing (A) deferoxamine induced toxic retinopathy with diffuse retinal pigmentary changes and areas of hyperpigmentation and hypopigmentation in the macula, with (B) extension of the pigmentary changes to the equatorial region and the peripheral retina areas.

when the therapeutic index was briefly elevated to 0.030.¹ Davies *et al* also reported the development of pigmentary retinopathy in a patient who had intravenous deferoxamine infusion with a therapeutic index of 0.039 but the visual symptoms resolved after cessation of therapy and the patient's final vision was not affected.² Our case highlighted the importance of frequent serum ferritin level monitoring and maintaining the deferoxamine dosage with therapeutic index <0.025 as severe irreversible retinopathy can develop rapidly. Frequent regular ophthalmic assessments should also be carried out in patients who are on continuous intravenous deferoxamine infusion, as prompt drug discontinuation might potentially arrest the retinal damage.

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Epstein-Barr virus infected cells in the aqueous humour originated from nasal NK/T cell lymphoma

Nasal natural killer (NK)/T cell lymphoma is a definitive diagnostic entity in the World Health Organization lymphoma classification.¹ In many cases, NK/T cell lymphoma is invariably associated with Epstein-Barr virus (EBV). Although ocular involvement is found in less than 10% of patients with systemic lymphoma,² because of its anatomical proximity, nasal NK/T lymphoma can sometimes complicate uveitis and orbital infiltration.³ We experienced a case of nasal NK/T lymphoma, and the cells collected from the aqueous humour originated from the lymphoma which was infected by EBV.

Case report

A 53 year old man noticed nasal obstruction and imaging demonstrated a mass lesion in the nasal cavity. The biopsy specimen from the cavity demonstrated a variety of diffuse

atypical lymphoid cell infiltration. Immunohistochemically, the atypical cells showed cytoplasmic reactivity for CD56, CD3, granzyme B and perforin, but not for CD20. Many EBV encoded small RNA (EBER) positive cells were detected in the lymphoma tissues. The patient was diagnosed with nasal NK/T cell lymphoma in May 2004. Magnetic resonance imaging (MRI) showed multiple metastatic lesions in the brain in June 2004. He underwent combined chemotherapy and radiotherapy. Since he had blurred vision in both eyes during the treatment, he visited our clinic on 1 September 2004. Visual acuities were 20/13 in both eyes. Intraocular pressures were 10 mm Hg in the right eye, and 11 mm Hg in the left. Biomicroscopy showed anterior segment inflammation with 1+ flare and 2+ cells in the anterior chamber, and small iris nodules were also sporadically seen in both eyes. A fundus examination showed no particular findings. Since then, although he was treated with eye drops of a mydriasis agent and betamethasone for 3 weeks, his symptoms had not improved. Moreover, the number of cells in the anterior chamber increased and pigmentation on the lens was noted. After informed consent was obtained, we performed anterior chamber paracentesis to obtain aqueous humour for cytological diagnostic analysis. May-Giemza staining disclosed a variety of severe atypical lymphoid cells with a high nuclear/cytoplasmic ratio (fig 1A, arrowheads) mimicking lymphoma cells in the nasal cavity. Only a few macrophages in intracytoplasmic phagocytosed nuclei were intermingled (fig 1A, arrow). The EBV genome was detected by polymerase chain reaction (PCR) using primers targeting the infrared region in the aqueous humour. Furthermore, EBV DNA was clearly detected by PCR Southern blot analysis (fig 1B). Despite continuous chemotherapy, he died as a result of multiple organ dysfunction associated with lymphoma invasion on 13 December 2004.

Comment

Nasal NK/T cell lymphoma can sometimes lead to ocular complications such as orbital infiltration owing to its anatomical proximity.³ Although it is reported that NK/T cell lymphomas complicate anterior uveitis,³ invasion of lymphoma cells into the ocular

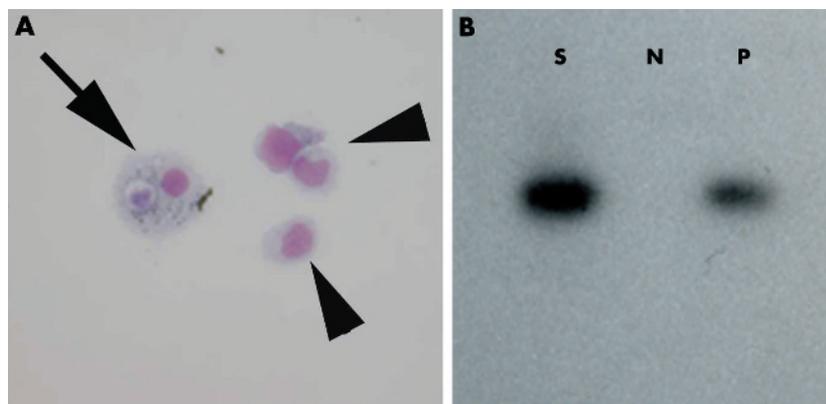


Figure 1 Cytological examination (A) and detection of EBV DNA by Southern blot analysis (B) in the aqueous humour. (A) A variety of atypical lymphoid cells proliferated with a high nuclear/cytoplasmic ratio (May-Giemza staining (A) arrowheads). Macrophages with intracytoplasmic phagocytosed nuclei are intermingled (A, arrow). (B) EBV DNA was markedly detected in the aqueous humour. Lane S: aqueous humour sample aspirated from the patient; lane N: distilled water as an EBV DNA negative control; lane P: EBV DNA integrated plasmid as a positive control.

tissue has not been demonstrated. In this case, cytological examination obtained from the aqueous humour demonstrated a variety of severe atypical lymphoid cells mimicking lymphoma cells in the nasal cavity. These results indicated that anterior chamber cells did not originate from endogenous uveitis, but had invaded from the nasal NK/T cell lymphoma using this method.

Many NK/T lymphoma cases are invariably associated with EBV, as reported previously.^{4,5} In fact, it is likely that EBV infects NK/T lymphoma cells in humans.⁵ In this case, EBER was detected in the nasal NK/T lymphoma cells, and EBV DNA was markedly detected in the aqueous humour by PCR Southern blot analysis. These results suggest that lymphoma cells detected in the aqueous humour originated from the nasal lymphoma as a primary lesion. The primary routes by which nasal NK/T lymphoma cells invaded to the anterior chamber remain unclear. In this case, MRI indicated that the lymphoma cells had already formed metastatic lesions in the brain 3 months before the onset of ocular symptoms. This suggests that the route of the lymphoma invasion from the nasal cavity to the anterior chamber may be indirectly systemic.

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Phakomatous choristoma of the eyelid: a case with associated eye abnormalities

Phakomatous choristoma is a rare lesion, first reported in 1971¹ with a further 18 cases reported to date. Previously reported cases have not been identified preoperatively, nor associated with ocular abnormalities except

as a secondary effect.^{2,3} We report here a case which was suspected on clinical examination, and which is associated with other eye abnormalities.

Case report

This healthy female patient first presented at the age of 2 days with a red left eye, and she was noted to have a lump in the nasal aspect of the left lower eyelid which was initially diagnosed as a dacryocystocele. The red eye resolved, but the eyelid swelling persisted despite regular massage. There was also concern about left amblyopia, astigmatism, and myopia. On specialist oculoplastic review, the possibility was raised of phakomatous choristoma, and excision with ocular examination under general anaesthetic was carried out at age 10 months.

At surgery, the eyelid mass was removed with no complications. Examination showed a normal right eye, but the left eye showed a colobomatous hypoplastic disc with a staphyloma. Refraction was approximately emmetropic.

Following surgery, the eyelid mass did not recur. However, the patient has developed progressive myopia in both eyes, with left amblyopia. Her most recent refraction at the age of 5 years is –11.25D right eye and –22.0D left eye, and her visual acuity is 6/18 right eye, 3/60 left eye.

Histology of the eyelid mass showed small nests and cords of epithelial cells within a dense fibrous stroma. The cells were bland with pale cytoplasm, and in areas formed vesicle-like structures containing degenerate swollen cells (fig 1A). Around the nests was a prominent basement membrane (fig 1B), and occasional psammoma-like bodies were present. The morphological appearance and immunohistochemical staining pattern (cells positive for vimentin and S100, negative for the pancytokeratin AE1/AE3) were typical of phakomatous choristoma (fig 2A and B).

Comment

Phakomatous choristoma is a lesion first described over 30 years ago. It is viewed as a proliferation of ectopic lens tissue, which presents at a young age and may enlarge because the abnormal cells are attempting to proliferate and differentiate. The clinical presentation is with a mass located at the nasal aspect of the medial eyelid and/or orbit. The histological appearance is of lens-type epithelium, with degenerate areas reminiscent of the Wedl or “bladder-like” cells of cataract. Previously reported cases have shown immunohistochemical and ultrastructural appearances consistent with a lenticular origin.^{3–7}

Theories of pathogenesis include (1) surface ectoderm “dipping down” into the mesoderm of the developing eyelid, (2) migration of putative lens tissue through the closing optic fissure, and (3) the site of origin of the invaginating lens being located in the area destined to form the nasal lower lid.^{1,5,8–10}

None of the previously reported cases has had any other ocular or orbital abnormalities, apart from astigmatism or epiphora attributable to the eyelid lesion that resolved post-operatively. The patient we report here also has a colobomatous hypoplastic optic disc with staphyloma, and severe myopia. Although the eye abnormalities here may be coincidental, the presence of a left optic disc coloboma and staphyloma suggests that there

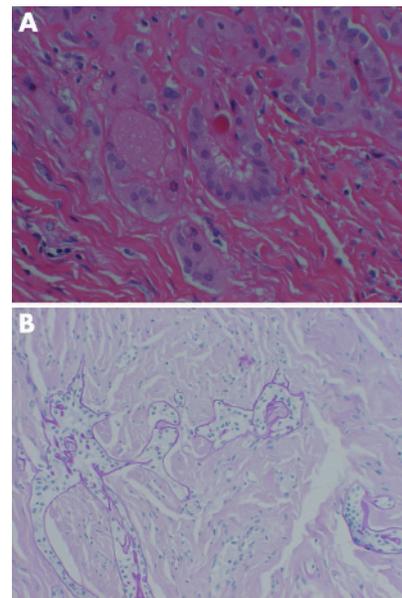


Figure 1 (A) Vesicle-like structures are present with thick basement membrane-like material and a swollen cell (haematoxylin and eosin $\times 20$). (B) The basement membrane surrounding the epithelial islands is accentuated (DPAS $\times 10$).

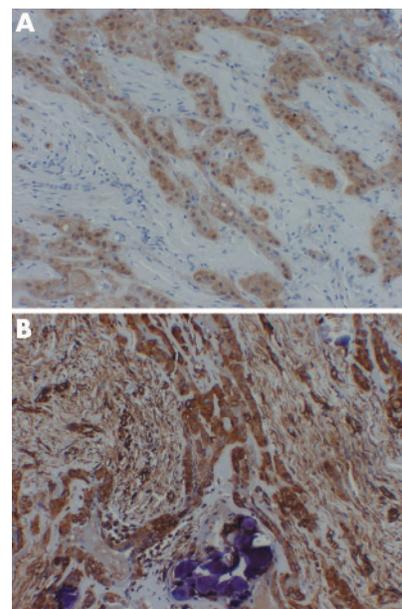


Figure 2 (A) S100 immunoreactivity is restricted to epithelial cells (S100 $\times 10$). (B) The epithelial cells are strongly positive for vimentin (Vimentin $\times 10$). There is also vimentin positivity in stromal cells.

may have been an abnormality of closure of the optic fissure.

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