Comment

Lichen planus is a very common skin disease and the overwhelming majority of the patients have oral lesions as well. However, lichen planus isolated to the conjunctiva is very rare. The cause of lichen planus is unknown. Available evidence points toward a T cell mediated immunological response to an induced antigenic change in the basal membrane zone of mucosa or skin.³ Conjunctival involvement, albeit rare, may be severe, causing cicatrix formation that is clinically indistinguishable from other forms of cicatrizing conjunctivitis.^{1,2} A conjunctival biopsy is the key to the correct diagnosis of the underlying aetiology. An irregular, coarse band of fibrin or fibrinogen deposition in the basement membrane, in the absence of immunoreactants demonstrated with direct immunofluorescein staining, strongly supports a diagnosis of lichen planus.²

Multiple therapeutic options including local and systemic corticosteroids, psoralens with ultraviolet A light, retinoids, ciclosporin, and cyclophosphamide have been recommended.⁴ Ciclosporin A is an immunomodulator that specifically inhibits T lymphocyte proliferation via inhibition of interleukin 2 receptor expression.⁵ Several clinical studies demonstrated favourable results with topical ciclosporin in the treatment of ulcerative or erosive oral lichen planus. Successful treatment of patients with isolated conjunctival lichen planus using topical ciclosporin A 2% in olive oil has also been reported.^{1,2}

We recommend that lichen planus be routinely included in the differential diagnosis of cicatrizing conjunctivitis, as this has important therapeutic and prognostic implications. Topical ciclosporin A may be of benefit in the treatment of selected patients with isolated conjunctival lichen planus.

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Bilateral peripapillary staphyloma, a clinicoradiological report

Peripapillary staphyloma is a rare non-hereditary congenital anomaly characterised by a relatively normal appearing optic nerve head located at the base of a staphylomatous excavation.^{1–3} It is generally not associated with other congenital defects or systemic diseases.¹

Peripapillary staphyloma is generally unilateral with bilateral cases being extremely rare.^{4–6} To the best of our knowledge, there is no previous report of neuroimaging findings in peripapillary staphyloma. We report a case of bilateral peripapillary staphyloma with the magnetic resonance imaging (MRI) findings.

Case report

An 11 month old boy was referred to the neuro-ophthalmology clinic by an ophthalmologist because of a suspected optic nerve mass noted on MRI. The patient was born to non-consanguineous parents at 39 weeks of

gestational age with a birth weight of 2800 g. The neonatal period was unremarkable. Development, other than ocular findings was normal. The boy was the first child of his parents and there was no history of significant ocular disease in his family. His parents' ocular examinations were normal.

Systemic physical examination including weight, length, and head circumference were normal for his age. Neurological examination was also normal except for vision.

He could only fix with his right eye. The eyes were orthotropic by Hirschberg examination. Cycloplegic refraction was –4.00 RE and –4.00 –2.5×180 LE. Slit lamp evaluation of both eyes was normal. Funduscopic examination (fig 1) revealed a deep posterior excavation with a normal appearing optic nerve head at its base in both eyes. Retinal vasculature was normal in both eyes. There was marked retinal pigment epithelium alteration surrounding the staphyloma especially in the left eye. No retinchoroidal coloboma was present.

MRI of the orbits (fig 2) disclosed a deep excavation of posterior wall of the globe with

Figure 1 Fundus photograph of right (A) and left (B) eyes, showing peripapillary staphyloma with surrounding mottled pigment epithelium and choroids. Normal appearing optic disc is located at the base of staphyloma. Retinal vessels are normal in number and distribution, emerging from the centre of the disc.

Figure 2 T1 weighted axial without contrast (top left), T2 weighted axial (top right), T1 weighted axial with contrast (bottom left), and T1 weighted sagittal view (bottom right) of MRI show a posterior excavation of the globe connected to the vitreous cavity with sharp borders and no enhancement on both eyes, which are compatible with a bilateral peripapillary staphyloma.

connection to the vitreous cavity without any structural deformity of other parts of the globe.

Comment

Peripapillary staphyloma denotes deep fundus excavation surrounding a relatively normal appearing optic disc with normal retinal vasculature and without any retinochoroidal coloboma.¹⁻⁶ The pathogenesis remains speculative. It is thought to be the result of a developmental failure of the posterior sclera from neural crest cells occurring near the fifth month of gestation.^{1,7} Peripapillary staphyloma is generally unilateral with only four bilateral cases reported previously.⁴⁻⁶ The staphylomatous depth varies in size, ranging up to about 10 mm with the fovea often involved by the excavation.⁶

Peripapillary staphyloma is often confused with other excavated congenital optic disc anomalies. In an optic disc coloboma, the optic disc itself is excavated while surrounding inferonasal retinochoroidal or iris tissue defects may also be present. In morning glory disc anomaly, the excavation may be shallower, cone-shaped and containing a centrally located peripapillary glial tuft. In addition, the optic disc may be anomalously. As opposed to the central origin of vessels in peripapillary staphyloma, in morning glory syndrome, the vessels radiate more distinctly from the disc periphery. Other vascular anomalies such as moyamoya disease affecting the carotid arteries, as well as pituitary hormone insufficiencies may be present in morning glory syndrome.⁸⁻¹⁰

Our patient had been referred for evaluation of a suspected optic nerve mass noted on MRI. Detailed examination of the fundi, however, revealed a bilateral peripapillary staphyloma. Furthermore, MRI findings, including the posterior scleral location and connection of the presumed "mass" with the vitreous cavity, associated with sharp internal and external margins, with absence of pathological enhancement all highlighted a peripapillary staphyloma.

Eyes with peripapillary staphyloma may sometimes achieve visual improvement by occlusion therapy, but compliance and visual outcome is often poor.⁶ Often macular involvement or "capture" by the staphyloma is present, limiting final visual outcome. A few reports of retinal detachment associated with peripapillary staphyloma have been published.²⁻⁶ A complete ophthalmic examination and regular follow ups are therefore necessary because of associated ocular disease and refractive errors. Unlike in patients with morning glory anomaly, however, MRI is unnecessary.

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Recurrent hypopyon in chronic anterior uveitis of pauciarticular juvenile idiopathic arthritis

The typical ocular manifestation of pauciarticular juvenile idiopathic arthritis (JIA) is an insidious, non-granulomatous chronic anterior uveitis (CAU). The main risk factors for the development of uveitis in JIA are being female, antinuclear antibody (ANA) positivity, and oligoarticular disease at onset.^{1,2} Even during acute exacerbations of JIA associated CAU with severe anterior chamber (AC) cellular reaction, it is rare for patients to have symptoms and the eye is invariably white.^{1,2} This is in distinct contrast to the acutely symptomatic red eye of acute anterior uveitis (AAU). Hypopyon can form within the AC in severe forms of anterior uveitis, particularly HLA-B27 associated AAU.³⁻⁵ We report a case of CAU in a patient with pauciarticular JIA, which was complicated by the development of recurrent hypopyon.

Case report

A 4 year old white girl first presented with blurred vision in her right eye and was found to have a unilateral, non-granulomatous CAU with extensive posterior synechiae and early band keratopathy. She was initially treated with intensive topical corticosteroids and mydriatics. She was ANA positive and had an ankle effusion, resulting in the diagnosis of pauciarticular JIA associated CAU. Repeated orbital floor steroid injections and systemic low dose methotrexate were required for the control of persistent anterior uveitis activity. Control of AC inflammatory activity was then maintained with regular topical prednisolone and oral methotrexate treatment.

At a routine follow up, 6 months after her initial presentation with CAU, the patient was found to have an asymptomatic inflam-

matory hypopyon and +4 cells in the AC of the right eye without conjunctival injection. There was no history of ocular surgery or trauma and the patient was well systemically. Topical steroids were increased to hourly leading to a significant improvement in the anterior uveitis activity with resolution of the hypopyon and the achievement of a quiet AC with no cells. Further investigations revealed that the patient was HLA-B27 positive and the angiotensin converting enzyme level was not elevated. There were no clinical features of a seronegative spondyloarthropathy. Two months later, the hypopyon recurred upon attempted tapering of topical steroid treatment, once again occurring in a painless "white" eye of CAU (fig 1). The hypopyon again demonstrated therapeutic response to hourly topical steroids and was completely resolved within 1 week. A quiet eye was eventually achieved and maintained with systemic methotrexate and regular topical prednisolone therapy, with last follow up being 9 months after the resolution of the recurrent hypopyon.

Comment

JIA is by far the most common identifiable clinical association of uveitis in children. There have been no previous reports describing the development of hypopyon in the typical clinical phenotype of CAU associated with pauciarticular JIA.^{1-2,5-6} Our case exhibited all of the characteristic clinical features of non-granulomatous CAU of ANA positive pauciarticular JIA, including its insidious onset and asymptomatic chronic disease course requiring long term anti-inflammatory treatment for uveitis control.

Although our patient was also found to be HLA-B27 positive, her clinical features were not consistent with HLA-B27 associated AAU, which includes an abrupt onset of red and painful anterior uveitis that is typically of short duration, predominantly occurring in boys who are ANA negative.³⁻⁶ HLA-B27 antigen is of clinical significance in AAU; however, in contrast, HLA-B27 positivity does not occur more frequently in CAU than in the general healthy population.³⁻⁷ JIA associated uveitis that is positive for both ANA and HLA-B27 appears to be a relatively rare clinical subgroup and whether these represent a distinct clinical entity remains unclear.^{2-6,8} Three patients with ANA positive CAU in early childhood who later developed HLA-B27 associated recurrent attacks of AAU during adolescence or adulthood have been

Figure 1 Slit lamp biomicroscopy examination demonstrating a hypopyon in a painless "white" eye with chronic anterior uveitis that is associated with ANA positive pauciarticular JIA. Note the lack of ciliary conjunctival injection.