Merkel cell carcinoma of the eyelid in association with chronic lymphocytic leukaemia

Merkel cell carcinoma (MCC) is a rare skin neoplasm. Tang and Toker first described MCC in 1978 and since then 19 cases in association with chronic lymphocytic leukaemia (CLL) have been reported. To the best of our knowledge, involvement of the eyelid by MCC has never been reported in the literature in association with CLL.

Case report

An 84 year old white man was referred with an 8 week history of a painless lump on his right upper eyelid (Fig 1A). He was complaining of visual obscuration secondary to a mechanical ptosis. Ophthalmic history was unremarkable and specifically there were no previous chalazions or trauma. On examination a firm lesion of the right eyelid measuring 2 × 1 cm with overlying telangiectatic vessels and sparing of the eyelashes was noted (Fig 1A). Further ophthalmic examination was unremarkable. General examination did not reveal any abnormalities.

General medical history revealed that the patient had been diagnosed with CLL 11 months previously and was being treated with pulsed chlorambucil. His condition was considered to be stable by his oncologist. At the time he had a white cell count of 15.7 × 10^9/l.Histopathological examination of the biopsy sample showed an intact epidermis with the underlying dermis being infiltrated by clumps of a small cell tumour (Fig 2A).

Immunostaining showed the tumour cells were negative for LCA (leucocyte common antigen), CD3 (T cell marker), CD20 (B cell marker), chromogranin, and S100 antigens. The tumour cells were positive for NSE (neuron specific enolase), EMA (epithelial membrane antigen) and CAM 5.2, which showed characteristic paranuclear accentuation (Fig 2B). Other staining techniques showed 50% of the tumour cells to be in cycle. All these features are consistent with the diagnosis of MCC.

Further investigation revealed no systemic metastasis. We opted for radiotherapy as the patient was reluctant to have surgical intervention. The patient was given a total of 40 Gy in 15 fractions. This caused the tumour to reduce in size relieving the mechanical ptosis (Fig 1B).

Comment

The recent surveillance, epidemiology, and end results (SEER) programme in the United States has estimated the incidence of MCC at 0.23/100 000. MCC is very rare below the age of 50 and is more common on sun exposed sites. It is an aggressive tumour with 12–45% being lymph node positive at presentation. This increases to 55–79% during the course of the disease. The 5 year survival has been reported at 30–64%. Involvement of the eyelid occurs in only 0.8% of MCC, and has not been reported in the literature in association with CLL.

Secondary tumours are common in B cell neoplasia with the relative risk of non-melanotic skin cancer being 4.7 in men and 2.4 in women. The frequency and aggressiveness of MCC and other skin neoplasms increases with immunosuppression, organ transplantation, as well as B cell neoplasia. The precise reason for such an association is not fully understood. Quaglino et al suggested that a depressed immunological system as well as exogenous oncogenic factors may, in various degrees, contribute to the development of neoplastic processes at different sites.

The treatment is wide local excision with or without adjuvant therapy consisting of block dissection of lymph nodes or radiotherapy. Adjuvant therapy reduces local recurrence and regional failure from 39% and 46% to 26% and 22% respectively. Most patients die from causes directly related to the disease. Potentially there is an increased risk of all skin tumours including MCC in patients suffering from CLL and this diagnosis should be considered when evaluating an eyelid lesion in such patients. In a patient with reduced immunity it would be best practice to send all surgical specimens for histology even if a simple chalazion is thought to be responsible for the lid lesion.

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Steatocystoma simplex of the caruncle

The caruncle has a non-keratinised epithelium lining similar to the conjunctival epithelium. However, unlike the conjunctiva, the caruncle harbours skin elements such as hair follicles, sebaceous glands, sweat glands, and accessory.
adnexal tumour, thought to originate from a naevus malformation of the pilosebaceous duct junction.^

Lesions are described on the forehead, nose, scalp, neck, axillae, chest, upper limbs, back, legs, and even intraorally. To our knowledge though, steatocystoma has not previously been reported in the caruncle.^

Thirty-two cases of SS are reported in the literature, divided evenly between men and women and ranging in age from 15 to 70 years.^

Clinical and histological features in SS are usually identical to those seen in the individual lesions of SM. Lesions are described as asymptomatic, flesh coloured or yellowish, intracutaneous, well circumscribed, soft, mobile, and non-tender. On incision they are found to contain an oily substance composed of sebum.^

However, it is important to confirm the solitary nature of a steatocystoma. SM can be familial and autosomal dominantly inherited (steatocystoma multiplex congenita). Several familial cases have been linked to pachyonychia congenita and ectodermal dysplasia through a mutation in keratin 17.^

Steatocystoma is histologically characterised by a cystic structure with sebaceous glands within the cyst wall and epithelium that displays an eosinophilic cuticle. It is possible to make a diagnosis of steatocystoma if the characteristic hyaline luminal cuticle is present, even in the absence of sebaceous elements.^

The differential diagnosis included sebaceous gland hyperplasia, sebaceous gland adenoma, and lipogranuloma. Clinically they can usually be excluded histologically. Oncocytomas are asymptomatic, slowly progressive, solid or cystic masses but usually described as reddish blue/tan.^

Most treatment regimens for steatocystoma reflect the multiplicity and widespread extent of lesions of SM. Oral isotretinoin has been used to reduce associated inflammation. Cryosurgery, carbon dioxide laser, and incision with removal of the cyst wall have been employed.^

We felt the best way to manage this solitary lesion arising in the caruncle was by simple excision with removal of the cyst wall intact, thereby reducing the risk of recurrence. We were able to confirm the unique nature of this lesion and rule out malignancy.^

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References
myopia in both eyes (right eye −4.00, left eye −9.00). There was a history of subretinal macular neovascularisation and cataract extraction in his left, surgical, eye, with an ECC of 950 cell/mm. Preoperative BSCVA was 0.5 in the right eye and 0.2 in the left. The follow up was done during 14 months (Fig 1D).

Examinations in all cases were at day 1, 1 week, 3, 6, 9, and 12 months. All Nylon sutures were removed before the 6 month control. All surgeries were technically uneventful. The immediate and late postoperative controls showed transparency of the cornea and no signs of rejection. In case 1 at the time of suture removal, a separation was noted between the anterior cap and the recipient eye in both corneal layers, and our cases showed a significant astigmatism and very low visual results.

We think that the time of graft deswelling was not as expected because at the time of suture removal a separation was noted between the anterior cap and the recipient eye in cases 1 and 2. We placed sutures in this site but the time of suture removal was extended to 12 months. Another contributing factor would be host-graft disparity, trephination, and suture technique.

In our experience this technique shows that it is possible to change only the posterior layers of the cornea with successful anatomical result. Nevertheless, from a functional perspective penetrating keratoplasty has been a much better and faster approach and, in fact, in both techniques we are replacing the endothelium using an open sky technique.

### Table 1

<table>
<thead>
<tr>
<th>Case</th>
<th>Best corrected visual acuity before and after the endokeratoplasty</th>
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<tr>
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### Table 2

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<td>Case 3</td>
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Persistent accommodative spasm after severe head trauma

Spasm of accommodation is the sudden, sometimes paroxysmal, and often bilateral, increase in accommodation that occurs in response to an increased demand for accommodation. It is usually accompanied by blurred vision, photophobia, and sometimes by systolic headache. The causes of spasm of accommodation are varied and include cataracts, glaucoma, myopia, astigmatism, and a variety of other ocular and neurological conditions. In this article, we report two cases of persistent accommodative spasm after severe head trauma.

Case 1

A 34-year-old female patient presented to our refractive surgery service for correction of myopia and astigmatism. She had a history of a severe head injury at the age of 24, and 3 months later, she noticed difficulties in her distant vision and injury at the age of 24, and 3 months later, she had myopia and astigmatism. She had been comatose for 2 months and had been referred to the refractive surgery service. The patient had been treated with intracranial pressure monitoring and had recovered normal accommodation 10 years after head trauma.

On examination visual acuity was 20/20 in both eyes without correction and 20/25 with −1.00 sph in each eye. The patient was prescribed 1% atropine every other day for 2.5 years associated with myopia and astigmatism. She had been comatose for 2 months and had been referred to the refractive surgery service. The patient had been treated with intracranial pressure monitoring and had recovered normal accommodation 10 years after head trauma.

Case 2

A 24-year-old male patient had been referred to our refractive surgery service for correction of myopia and astigmatism. He had been comatose for 2 months and had been referred to the refractive surgery service. The patient had been treated with intracranial pressure monitoring and had recovered normal accommodation 10 years after head trauma.

On examination visual acuity was 20/20 in both eyes without correction and 20/25 with −1.00 sph in each eye. The patient was prescribed 1% atropine every other day for 2.5 years associated with myopia and astigmatism. He had been comatose for 2 months and had been referred to the refractive surgery service. The patient had been treated with intracranial pressure monitoring and had recovered normal accommodation 10 years after head trauma.

References

Extramedullary plasmacytoma of the eyelid

A 74 year old man presented with a foreign body sensation in the right eye superimposed on a slowly growing enlarging lump in the right eye. He had no history of recurrent infections, bleeding, weight loss, or night sweats. His only other symptom was chronic backache secondary to osteoarthrosis. Past medical history included cataract extraction from the right eye 4 years previously and medical history included cataract extraction infections, bleeding, weight loss, or night sensation in the right eye superimposed on a slowly growing enlarging lump in the body. Chan and Trobe reported a retrospective review of six patients with post-traumatic pseudomyopia but did not include MRI studies. In our patients, MRI scan failed to show abnormalities in the mid-brain. Both of them had lesions in the left temporal lobe and the first patient also had abnormalities in the frontal and parieto-occipital lobes bilaterally and the cerebellum. Although it is possible that they have small mesencephalic lesions, not detected by MRI scan, the findings is our cases suggest a higher origin for accommodative dysfunction in some patients with closed head trauma.

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Figure 1 Photomicrograph showing infiltration of the conjunctiva by the neoplastic plasma cells with strong immunocytochemical staining for immunoglobulin IgG (>355).

Figure 2 Whole of upper eyelid treated by radiotherapy using a customised lead cutout with internal shielding of the eye.


Figure 1 Extramedullary plasmacytoma of the eyelid.

Figure 2 Whole of upper eyelid treated by radiotherapy using a customised lead cutout with internal shielding of the eye.

Reference
and C reactive protein. She also had a chest x-ray, erythrocyte sedimentation rate (ESR), myalgia, and hiatus hernia. Her only medications were calcium supplements. Her vision was well controlled with oral prednisolone. She had no headaches. Her medical history included oesophagitis and hiatus hernia. Her only medical investigations revealing a normal full blood picture, haematological investigations revealing that they are areas of calcification associated with abnormalities of electrolytes. She had bilateral nucleosclerotic cataracts and both fundi revealed minimally elevations along the superotemporal arcades and nasally in both fundi. Her visual acuity was 6/12 right eye improving to 6/9 with pinhole, and 6/18 left eye improving to 6/12 with pinhole. She had bilateral visual acuity was 6/12 right eye improving to 6/9 with pinhole, and 6/18 left eye improving to 6/12 with pinhole. She had bilateral nucleosclerotic cataracts and both fundi revealed numerous pale elevated lesions clustered around the superotemporal and inferotemporal arcades (Fig 2A). Haematological investigations and chest x ray were unremarkable. Ultrasound scanning revealed that these were areas of calcification with high reflectivity (Fig 2B).

Case 3
A 71 year old man attended routinely for review 2 weeks after cataract extraction. He had no past medical history and vision was 6/9 right eye and 6/6 left eye. He was noted to have a small optic disc haemorrhage coincidentally and therefore dilated fundal examination was performed. He had bilateral pale visual acuity was 6/12 right eye improving to 6/9 with pinhole, and 6/18 left eye improving to 6/12 with pinhole. She had bilateral nucleosclerotic cataracts and both fundi revealed numerous pale elevated lesions clustered around the superotemporal and inferotemporal arcades (Fig 2A). Haematological investigations and chest x ray were unremarkable. Ultrasound scanning revealed that these were areas of calcification with high reflectivity (Fig 2B).

Figure 1 Case 1 (A) Elevated yellow areas along the superotemporal arcades bilaterally and demonstration of autofluorescence of these lesions. (B) Ultrasound scan showing the lesions to be highly reflective with orbital shadowing.

Figure 2 Case 2 (A) Pale yellow elevated lesions in the mid-periphery clustered around the superotemporal arcade. (B) Ultrasound of lesions revealing that they are areas of calcification with high reflectivity.

Idiopathic sclerochoroidal calcification
Sclerochoroidal calcification is a relatively rare condition characterised by yellow-white irregular subretinal lesions usually in the superotemporal mid-periphery of the fundus. It is usually asymptomatic and has a classic clinical appearance. Most cases are idiopathic but a few reports have associated this condition with abnormalities of electrolytes. We present three cases of idiopathic sclerochoroidal calcification.

Case reports
Case 1
A 71 year old woman was referred by her optician after attending for routine glasses update. On questioning she did complain of a “slight blurring of vision” gradually for several months. She had a history of left amblyopia. Her medical history included asthma, osteoarthritis, lymphoedema, fibromyalgia, and hiatus hernia. Her only medications were inhalers and paracetamol. She had previously taken calcium supplements. Her visual acuities were 6/6 in the right eye and 6/12 in the left eye with a hypermetropic correction. She had early cataract lens opacities and no vitritis. Both fundi revealed minimally elevated yellow areas in the choroid along the superotemporal arcades (Fig 1A).

The patient had haematological investigations revealing a normal full blood picture, urea and electrolytes, ACE level, urate, liver function tests, bone profile, immunoglobulins, erythrocyte sedimentation rate (ESR), and C reactivity protein. She also had a chest x-ray which was unremarkable. Her fundal photography showed autofluorescence of these lesions and ultrasound scanning aided the diagnosis in that the lesions were highly reflective with orbital shadowing (Fig 1B).
Stereotactic irradiation of biopsy proved optic nerve sheath meningioma

The role of conventional external beam radiotherapy in the management of optic nerve sheath meningiomas (ONSM) has been controversial because of limited radiation sensitivity of these tumours and radiation damage to surrounding tissues. Recently, in a study of 64 patients with ONSM managed with observation, surgery, radiotherapy, or surgery and radiotherapy, Turbin and colleagues found that patients treated by (conventional) radiotherapy alone demonstrated the best long term visual outcome, and suggested fractionated external beam radiation (5000–5500 cGy) as the initial treatment in selected cases, when preservation of visual function is a reasonable goal.

The collateral damage secondary to conventional radiotherapy may be minimised by better focusing and shaping of the radiation beams, as in stereotactic radiotherapy (SRT). We report on a woman whom we treated with fractionated SRT for a biopsy proved, large ONSM.

In April 2000 a 41 year old woman was referred with a 1 month history of proptosis of her left eye (Fig 1, top left). She had been treated for a presumed orbital “pseudotumour” with oral prednisone (initial dose 90 mg/day) without effect. At referral, she had no history of diplopia or retrobulbar pain. On examination, the visual acuity (VA) was 1.25 (unaided) of the right eye and 0.8+ (cc S+2) of the left eye. The intraocular pressure was 18 and 21 mm Hg in the right eye and left eye, respectively. There was left relative afferent pupillary defect (RAPD). The motility of the left eye was mildly restricted in upgaze. The left eye showed mild periorcular swelling and conjunctival chemosis. There was 5 mm of left proptosis without upper eyelid retraction or lid lag. Funduscopy of both eyes showed no abnormalities. Visual field testing (Humphrey field analysis (HFA II 730) showed relative scotomas of the left eye, mainly in both lower quadrants. Visual evoked potential (VEP) examination of the left eye showed prolonged latency and decreased amplitudes, suggestive of optic nerve dysfunction.

Orbital MRI (T1 weighted) scans showed a proptosis of the left eye and a large retrobulbar, intracranal mass that stained intensely with gadolinium contrast (Fig 1, bottom left). Computed tomography (CT) imaging also showed an intensely staining retrobulbar tumour without calcifications, that encased and slightly displaced the optic nerve. There was no “tram-tracking” sign or bone involvement. No tumour extension into the optic canal or intracranially was noted. Orbital colour Doppler ultrasound imaging showed a highly vascularised retrobulbar mass with a vertical diameter of at least 25 mm (Fig 2, top left).

Since, on imaging, no evident diagnosis could be made, we decided to perform a biopsy on the lesion through a lateral orbitotomy. At surgery, the tumour was pale and solid. Histopathological examination of the incisional biopsy specimen showed whorls of meningothelial cells, with small nuclei and inconspicuous nucleoli, consistent with a meningioma (Fig 2, bottom).

After surgery we observed the patient for 9 months. During this period her left (corrected) VA deteriorated to 0.2 and her left visual field showed progression of her scotomas. This prompted us to treat her with fractionated SRT in March 2001. The radiation, delivered with a 6 MV linear accelerator (Varian), was given 5 days a week at 1.8 Gy per fraction, with a cumulative dose of 34 Gy. Treatment planning was based on orbital MRI matched with CT scans. A non-invasive stereotactic frame was fixed with an external coordinate system (one isocentre). Target and surrounding tissues at risk were defined as volume of interest on contrast media enhanced T1 weighted MRIs and transferred to CT by the stereotactic localisation technique using a three dimensional planning system (X Plan, Radionics). Portals were optimised using a beam’s eye view technique. Five irregularly shaped non-coplanar beams (arcs) per treatment were used. Beam shapping was done with a mini-multileaf collimator (Radionics). No early complications of the radiation treatment were noted. At 6 months after SRT, the (corrected) VA of her left eye had recovered to 0.8, while no RAPD was observed.

Figure 1 Top left. Appearance of a 41 year old woman with a biopsy proved optic nerve sheath meningioma before SRT. Note the left exophthalmos and periorcular swelling. Top right. Post-treatment appearance. Note the decrease of the fullness of the left eyelids. Also note the right upper lid retraction secondary to left upper lid ptosis. Bottom left. Orbital MRI scan (T1 weighted with fat suppression and gadolinium contrast enhancement) at presentation. Bottom right. Six months after radiotherapy. A decrease of both tumour size and proptosis is clearly visible.

Figure 2 Top left. Ultrasound examination at presentation. A large, heavily vascularised retrobulbar mass is visible. Top right. Six months after radiotherapy, the tumour has diminished in size and vascularisation. Note that a different depth setting of the ultrasound system has been used. Bottom. Histopathology of the optic nerve tumour, showing whorls of meningothelial cells, with small nuclei and inconspicuous nucleoli (haematoxylin and eosin, ×200 original magnification).

Her periorcular swelling had markedly diminished (Fig 1, top right). Compared to previous measurements, the protrusion of the left eye had diminished by 4 mm. Funduscopy, however, showed mild pallor of the left optic nerve head. Visual field testing showed unchanged loss of the left visual field compared to pretreatment values, with a higher foveal threshold. VEP measurements showed improved amplitudes, but prolonged latency compared to previous examination. Orthoptic examination showed ductions similar to those before treatment. Post-treatment MRI revealed a markedly decreased tumour size and a decrease of exophthalmos (Fig 1, bottom right). Colour Doppler ultrasonography showed a decrease in tumour size with markedly diminished vascularisation (Fig 2, top right). At the last follow up visit, 16 months after treatment, her left VA and visual fields were stable.

Comment
As in the recent report on a presumed ONSM by Moyer et al.,5 fractionated SRT in our biopsy-proved case gave a remarkable visual recovery without detectable side effects. Both the size and the blood flow of the tumour regressed within the first 6 months, leading to reduced exophthalmos and periorcular swelling. The effect of restored cosmesis was important to this young woman whose main complaint was her unilateral exophthalmos.

Since our follow up is limited to 16 months, no conclusions with regard to long term outcome can be made. More cases of SRT for ONSM need to be studied over a longer period of time to assess the efficacy of this treatment.

Acknowledgements
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Severe interferon associated retinopathy
Interferon alfa is used in various human malignancies for its antitumour activity. One of its oculocutaneous effects is retinopathy. Interferon associated retinopathy is generally mild and resolves completely. We describe a severe retinopathy in a hypertensive patient treated with interferon for multiple myeloma.

Case report
A 56 year old man presented with a 3 week history of deterioration and distortion of right vision. VA were 6/60 right and 1/60 left. Funduscopy revealed bilateral extensive peripapillary cotton wool spots, retinal thickening, optic disc hyperaemia, and blot haemorrhages. Arteriolar changes were minimal.

He was anaemic (Hb 10.6 g/dl) and slightly thrombocytopenic (platelets 93 × 10^3). Plasma viscosity was 1.59 (normal 1.5–1.7). Renal function was normal at presentation.

He underwent peripheral blood stem cell transplant for multiple myeloma 8 months previously after having melphalan 110 mg/m^2 and total body irradiation (including the head) in a total dose of 1200 cGy given in six fractions over 3 days. He then had interferon alfa therapy for 4 months, initially 3 mega units three times a week, later reduced to twice a week. It was stopped immediately after visual deterioration.

Five years previously, he had macular laser treatment following left inferotemporal branch retinal vein occlusion. On discharge 1 year later, VAs were right 6/5, left 6/36. He was a known hypertensive, taking lisinopril, but control was poor around the time his VA began to deteriorate, with readings up to 150/100 mm Hg. He was not diabetic. His myeloma status was stable. Cytomegalovirus (CMV) antigen checked by polymerase chain reaction (PCR) and pretransplant HIV status were negative.

One week after presentation at the eye clinic, VAs dropped to right 2/60, left finger counting and did not improve after a course of intravenous methylprednisolone (1 g/day for 3 days). He was registered blind.

A further 3 weeks later, bilateral cotton wool spots and haemorrhages were more numerous and both foveas showed gross thickening with exudates (Fig 1). Fundus fluorescein angiography revealed retinal ischaemia with capillary non-perfusion, pruning, and tortuosity of vessels, vessel wall staining, and leakage (Fig 2).

Five months later preproliferative retinopathy was noted and subsequent proliferative changes were treated with bilateral panretinal laser photocoagulation. At 9 months, VAs were 1/60 right and left.

Comment
In a review article on interferon retinopathy, initial interferon alfa doses ranged from 3–9 mega units three to six times per week for several weeks. In a prospective randomised placebo controlled trial of interferon alfa therapy for macular degeneration, retinopathy was noted with increasing frequency in the highest dose group (5% of the patients taking 6 mega units three times a week). The interferon doses in our patient were at the lower level of these regimens.

Severity of retinopathy was found to be related to the presence of the following risk factors: large initial dosages, long duration of treatment, and systemic diseases like diabetes mellitus or hypertension. Early onset of retinopathy was also a good indicator of severity.
and fundal examination up to 8 weeks from start of treatment was advocated for those at risk. Interferon therapy was associated with macular oedema in a hypophalumininaemic but non-hypertensive, non-diabetic patient; and in two other cases in a case series report, one of whom had poor control of blood pressure and the other an occasional mildly elevated blood pressure level. All three patients had resolution of the lesions by 2 months and subsequently made good visual recoveries, unlike in our case. In another case series report, two out of six patients who received interferon alfa-2b suffered permanent visual loss after developing macular oedema. Both patients were hypertensive and one had radiation treatment to the brain. The latter later developed proliferative retinopathy as well. Deposition of immune complexes in the retinal vasculature has been postulated as a pathogenetic mechanism for the retinopathy. Interferon alfa was also found to induce leukocyte capillary trapping in rat retinal microcirculation. Radiation may have contributed to the development of the clinical picture although its use in the treatment of myeloma is frequent and ocular side effects have not been widely recognised in the past. Low doses to the eye similar to that used in our patient were associated with retinopathy after treatment of age related macular degeneration. Retinal oedema is an indicator of severity in interferon associated retinopathy. Early detection of it, especially in hypertensives and diabetics, may help avoid progression to permanent visual loss.

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Figure 1 Fundus photograph of the left optic disc shows diffuse swelling and increased surface vascularity. Additionally, a more focal elevation at the inferotemporal quadrant gives a nodular-like appearance to the disc swellings (arrows). The irregular white spot at the lower left corner of the figure is a photographic artefact.

Mass lesions of the posterior segment associated with Bartonella henselae

Bartonella (previously Rochalimaea) henselae is the infectious agent causing cat scratch disease, a self limited regional lymphadenopathy associated with flu-like symptoms. In approximately 10% of cases, extranodal dissemination of the organism results in a variety of intraocular inflammatory lesions. We report a patient with acute Bartonella henselae infection in whom the only physical manifestation was multiple, mass-like lesions in the eye which resembled ocular metastases.

Case report

A 12 year old boy developed daily headaches and blurred vision. Four weeks later, he noted a central grey spot in his left eye. Visual acuity was 20/20 in his right eye and 20/400 left eye. Goldmann perimetry of the left eye revealed a dense central scotoma with steep margins nasally and moderate constriction of the temporal perimeter. An afferent pupillary defect was present on the left side. Slit lamp examination was normal in both eyes. Funduscopy of the left eye showed nodular optic disc swelling with peripapillary subretinal elevation (Fig 1). A large creamy-coloured mass (approximately one and one half disc diameter) without subretinal exudates or haemorrhages was present in the macula. Several smaller but similar, elevated lesions of the choroid were detected in the periphery as well as an inferior serous retinal detachment. There was no evidence of ocular inflammation. An orbital ultrasound showed the macular lesion to be a solid mass without associated subretinal fluid (Fig 2).

Complete blood count, electrolytes, and glucose were normal. A sedimentation rate was marginal at 18 mm/hour (normal less than 15 mm/hour). Antinuclear antibodies were elevated at 1:320, speckled pattern. Rheumatoid factor was negative. Evaluation for various infectious aetiologies was negative, including Bartonella serologies (IgM and IgG). A systemic evaluation for a primary tumour causing possible ocular metastases was negative. The patient was empirically treated with doxycycline 100 mg twice daily for 10 days. The macular mass and choroidal lesions resolved in 4 weeks. Shortly thereafter, the disc swelling resolved. Because a diagnosis remained lacking, serologies were repeated 1 month later and revealed a marked rise of Bartonella titres, confirming Bartonella henselae infection (B henselae IgM = 24, normal less than 16, and B henselae IgG = 1024, normal less than 256). Final visual acuity was 20/70 left eye due to residual peripapillary and macular gliosis.

Comment

The range of ocular findings associated with B henselae continues to expand. The classic follicular conjunctivitis described with lymphophenopathy and fever (Parinaud’s ocular glandular syndrome) is due to direct inoculation of the conjunctiva. Neuroretinitis, a syndrome of acute visual loss and optic disc swelling and macular star, was the first established intraocular complication from disseminated Bartonella. Since then, other reported intraocular findings include inflammatory chorioretinal white spots, papillitis, serous detachment, vitritis, uveitis, vasculitis, retinal vasculo-oedematous disease, and vitreous haemorrhage. A mass lesion at the optic nerve head has been reported in several instances. A solitary maculae lesion without other ocular inflammatory findings has also been reported. Such lesions in the posterior pole have been presumed to represent a mass effect of Bartonella infection.

We report a patient with a circumscribed, elevated lesion in the macula as well as other mass-like lesions at the optic nerve head and in the choroid. These lesions occurred in the absence of systemic or ocular inflammation and clinically resembled ocular metastases. This case highlights the importance of recognising the wide spectrum of ocular bartonellosis. Furthermore, clinicians are reminded that cat exposure is not essential for contracting this bacteria and, therefore, Bartonella titres should be obtained whenever there is a clinical index of suspicion, regardless of cat exposure.

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References


Optic neuritis with marked distension of the optic nerve sheath due to local fluid congestion

Distension of the subarachnoid space of the optic nerve is not a common feature of optic neuritis. We describe a patient with optic neuritis with swelling of the optic nerve head of the right eye. On magnetic resonance imaging (MRI) there was marked distension of the optic nerve sheath due to an increase of fluid in the subarachnoid space. The location of the lesion in the optic nerve and concurrent inflammatory changes of the arachnoid trabecula and septae may have had a role in the pathophysiology of this condition.

Case report

A 38 year old man was admitted with pain in eye movements and loss of vision in the right eye. Best corrected visual acuities measured 20/40 on the right and 20/20 on the left. The patient identified 16 out of 18 Ishihara plates with the right eye and 18/18 with the left eye. There was a relative afferent pupillary deficit (RAPD) on the right. Funduscopy demonstrated a swollen optic disc on the right. The left optic disc was normal. Spontaneous venous pulsations (SVP) were detectable bilaterally. Laboratory examinations, including red and white blood counts, C reactive protein, sedimentation rate, serologies for syphilis, HIV, herpes, toxoplasmosis, Lyme disease and cytomegalovirus, as well as collagen vascular disorders and coagulopathies were all in normal range. The right visual field (Octopus programme G2) demonstrated a wedge-shaped defect in the inferior nasal and temporal visual field pointing towards the macula. The left visual field was normal. Neurological examination was normal. MRI of the brain was normal but showed enhancement of the right optic nerve in the T1 weighted axial and coronal (not shown) image and hyperintense fluid in the expanded optic nerve sheath on the T2 weighted image (Fig 1A and B). Two days after admission the visual acuity in the right eye decreased to 20/100 and only two out of 18 Ishihara plates were identified. SVP were no longer present on the right. The swelling of the right optic disc progressed and temporal peripapillary Patton folds appeared, suggesting the diagnosis of papillodema (Fig 2). Within 2 weeks visual acuity improved to 20/25 right eye and colour vision returned to normal. A repeat MRI of the orbits 7 weeks later demonstrated normal diameters of both periopical subarachnoid spaces (Fig 1C).

Comment

Distension of the periopical subarachnoid space is a hallmark MRI feature of papillodema due to an intracranial mass lesion, inflammatory disease, and pseudotumour cerebri. Unilateral distension of the optic nerve sheath due to increased fluid volume of the subarachnoid space of the optic nerve has previously been reported in some patients with optic hydrops, anterior ischemic optic neuropathy, and anatomical anomalies such as arachnoid cysts. This patient with optic neuritis demonstrated marked distension of the subarachnoid space of the right optic nerve, presumed to be caused by an increase of total fluid following optic neuritis. As all cerebrospinal fluid compartments are thought to communicate, equalisation of fluid via the chiasmal cistern would have been expected to occur. The MRI scan of the brain and orbits, however, demonstrated localised and isolated stasis of fluid in the right optic nerve subarachnoid space only. The reason for this fluid congestion causing a optic nerve sheath compartment syndrome could not be identified by neuroimaging. The site of inflammation of the optic nerve and local anatomical variations and alterations of the subarachnoid space—for example, the amount and number of trabecula and septae in the subarachnoid space—may have a crucial role in the pathophysiology of unilateral papillodema.

References


Corneal opacification following keratoplasty in the rat model

I read with great interest the excellent perspective by Plsková et al. in which they raise the issue of transient corneal opacification following corneal transplantation in the mouse model and argue that it might be due to a sufficient number of endothelial cells regaining function. What the authors describe for the mouse model also occurs in the rat model. In fact,
Surgery for glaucoma in the 21st century

The authors of the article “Surgery for glaucoma in the 21st century” should be commended for attempting to tackle this issue. Nevertheless, we do feel that their fundamental points and principal arguments merit reconsideration.

The authors state categorically that “This finding of a higher ‘failure’ rate based on intraocular pressure after ‘non-penetrating’ surgery compared with trabeculectomy has been found by the majority of randomised trials comparing the two procedures” and then go on to quote three references allegedly supporting this remark.

One of the three studies’ reports lower mean IOP with deep sclerectomy compared to trabeculectomy (although not statistically significant) and almost identical success rates. What was significant was the dramatically lower complication rates with deep sclerectomy.

When discussing the other two papers it is of paramount importance to understand that given the long learning curve associated with deep sclerectomy, it is neither fair nor scientifically sound to compare a surgeon’s first 20 cases of trabeculectomy with his first 20 cases of deep sclerectomy. As an example, one group reported 0% success rate in their first series of viscoanastomolysis patients and then presented their second series with a success rate of 95%. The same group also analysed the depth of their dissection of the deep sclera to find that they dissected too superficially in 48% of their cases and too deeply in 17%; meaning that the proper depth of dissection, which should reflect transversally the Schlemm’s canal deroofing it, was not achieved in the majority of their cases.

The authors also failed to cite published long term (3.2, SD 14.3 months) results for deep sclerectomy with collagen implant. The study provided a qualified success rate of 94.8% and the complete success rate, 61.9% after 60 months (survival analysis), with a mean IOP at the last follow-up of 11.8 (SD 5) mm Hg. Although the study reports a non-randomised consecutive series of patients, it should be taken as a proper indication of results achieved by experienced surgeons.

It should be emphasised that consideration that non-penetrating surgery is a broad genre of surgery, under which different surgeons perform fundamentally different procedures that include sinusotomy, ab externo trabeculectomy, deep sclerectomy with or without the use of an implant, viscoanastomolysis, performance of postoperative gonipuncture, and the use of antimetabolites. The different techniques have one thing in common, the element of non-perforation.

What is true is that this type of surgery is continuously evolving, so it is unlikely that a proper judgment can be made yet. At the risk of sounding dramatic, it is valid to say that editors like the one by Khaw et al seem to indirectly sign a death certificate of non-penetrating surgery. It is much more useful to encourage research in non-penetrating surgery, including mixed up follow-on randomised studies, to see if trabeculectomy will remain king.

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References

NOTICES
Role of optometry in Vision 2000

The latest issue of Community Eye Health (No 43) discusses the mobilisation of optometry to deal with uncorrected refractive error, which is now a major cause of functional blindness. For further information please contact: Journal of Community Eye Health, International Centre for Eye Health, Institute of Ophthalmology, 11–43 Bath Street, London EC1V 9EY UK (tel: +44 (0)20 7608 6910; fax: +44 (0)20 7250 3207; email: eyeresource@ucl.ac.uk; web site: www.jceh.co.uk). Annual subscription (4 issues) UK£25/US$40. Free to workers in developing countries.

International Centre for Eye Health

The International Centre for Eye Health has published a new edition of the Standard List of Medicines, Equipment, Instruments and Optical Supplies (2001) for eye care services in developing countries. It is compiled by the Task Force of the International Agency for the Prevention of Blindness. Further details: Sue Stevens, International Centre for Eye Health, 11–43 Bath Street, London EC1V 9EY UK (tel: +44 (0)20 7608 6910; email: eyeresource@ucl.ac.uk).

Second Sight

Second Sight, a UK based charity whose aim is to eliminate the backlog of cataract blind in India by the year 2020 and to establish strong links between Indian and British ophthalmologists, is regularly sending volunteer surgeons to India. Details can be found at the charity web site (www.secondsight.org.uk) or by contacting Dr Lucy Matljen (lucymatjen@yahoo.com).

Specific Eye Conditions (SPECs)

Specific Eye Conditions (SPECs) is a not for profit organisation which acts as an umbrella organisation for support groups of any conditions or syndrome with an integral eye disorder. SPECs represents over fifty different organisations related to eye disorders ranging from conditions that are relatively common to very rare syndromes. We also include groups who offer support of a more general nature to visually impaired and blind people. Support groups meet regularly in the Boardroom at Moorfields Eye Hospital to offer support to each other, share experiences, and explore new ways of working together. The web site www.eyeconditions.org.uk acts as a portal giving direct access to support groups own sites. The SPECs web page is a valuable resource for professionals and may also be of interest to people with a visual impairment or who are blind. For further details about SPECs contact: Kay Parkinson, SPECs Development Officer (tel: +44 (0)1803 524238; email: k@eyeconditions.org.uk; web site: www.eyeconditions.org.uk).

The British Retinitis Pigmentosa Society

The British Retinitis Pigmentosa Society (BRPS) was formed in 1975 to bring together people with retinitis pigmentosa and their families. The principle aims of BRPS are to raise funds to support the programme of medical research into an eventual cure for this hereditary disease, and through the BRPS...
welfare service, help members and their families cope with the everyday concerns caused by retinitis pigmentosa. Part of the welfare service is the telephone helpline (+44 (0)1280 860 363), which is a useful resource for any queries or worries relating to the problems retinitis pigmentosa can bring. This service is especially valuable for those recently diagnosed with retinitis pigmentosa, and all calls are taken in the strictest confidence. Many people with retinitis pigmentosa have found the Society helpful, providing encouragement, and support through the Helpline, the welfare network and the BRPS branches throughout the UK. (tel: +44 (0)1280 821 334; email: lynda@brps.demon.co.uk; website: www.brps.demon.co.uk)

**Detachment Course with international faculty on: Retinal and Vitreous Surgery with Case Presentations preceding Retina Meeting**

The detachment course with international faculty on: Retinal and Vitreous Surgery with Case Presentations and the Retina Meeting will be held 14–15 March 2003 and 16 March 2003 respectively, in Mexico City, Mexico. Further details: Scientific programme: Prof Ingrid Kreissig, University of Tuebingen, Schleichstr. 12, Breuningerbau, 72076 Tuebingen, Germany (tel: +49 7071 295209; email: ingrid.kreissig@med.uni-tuebingen.de). Local organisation: Prof. Quiroz-Mercado, Prof. Munoz, and Prof. Gonzalez “Hospital la Ceguera en Mexico Vicente Garcia Torres #46, Coyoacan, Mexico DF 04330 (fax: 5253 5639 5928; email: retinamex@yahoo.com).

**16th Annual Meeting of German Ophthalmic Surgeons**

The 16th Annual Meeting of German Ophthalmic Surgeons will be held 8–11 May 2003 in Nürnberg, Germany, Messezentrum. Organised by the Professional Association of German Ophthalmologists Ophthalmic Surgery Group the conference will cover cataract surgery, refractive surgery, glaucoma surgery, vitreoretinal surgery, corneal surgery, eye surgery in developing countries, and orbita, lacrimal and lid surgery. Further details: MCN Medizinische Congress organisation Nürnberg AG, Zerzabelshofstr 29, 90478 Nürnberg, Germany (tel: +49 911 3931621; fax: +49 911 3931620; email: doc@mcnag.info; website: www.doc-nuernberg.de).

**3rd British Oculoplastic Surgery Society Meeting**

The 3rd British Oculoplastic Surgery Society Meeting will be held 18–19 May 2003 in Birmingham, UK. For further details please contact the Secretary of the British Oculoplastic Surgery Society Jane Olver (tel: +44 (0)121 424 5464; fax: +44 (0)121 424 4464; email: MartiDi@heartsol.wmids.nhs.uk; website: www.bopss.org).

**13th Meeting of the EASD Eye Complication Study Group**

The 13th Meeting of the EASD Eye Complication Study Group will be held on the 23–25 May 2003, in Prague, Czech Republic. The scientific programme includes keynote lectures from Professor John H Fuller (UK) on The epidemiology of diabetic retinopathy; Dr P Martin van Hagen (The Netherlands) on Growth factors and diabetic retinopathy; Professor Jeretic Pelikanova (Czech Republic) on Pathophysiology of diabetic microvascular complications; Dr Tomas Sosna (Czech Republic) on Risk and protective factors of diabetic retinopathy. Three travel grants of 1000 each, sponsored by GlaxoSmithKline for young scientists (under 35 years at the time of the meeting). Applications should be made with the submission of abstracts. The deadline for abstracts is 14 February 2003. Further details: Ortopedické Centrum, s.r.o., Strekovské nábřeží 51, 400 03 Usti nad Labem, Czech Republic (tel: +420 47 521 6588; fax: +420 47 533 40 77; email: ortcentrum-ul@volnv.cz; website: www. ortopedické-centrum.cz).