Figure 2 Karyogram of cell line CM2005.1. A very complex karyogram with gains, deletions and rearrangements of almost all chromosomes was observed in the majority of cultured cells.

times of around 60 h for their conjunctival melanoma cell lines CRMM-1 and CRMM-2.³

This fourth conjunctival melanoma cell line is now available for research, and can hopefully contribute to expansion of our knowledge of conjunctival melanomas.

Sander Keijser, Willem Maat, Guy S Missotten, Rob J W de Keizer
Department of Ophthalmology, Leiden University Medical Center, Leiden, The Netherlands

Correspondence to: Professor R J W de Keizer, Department of Ophthalmology, Leiden University Medical Center, PO Box 9600, 2300 RC, Leiden, the Netherlands; r.j.w.de_keizer@lumc.nl

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Complications of management in primary acquired melanosis with atypia

The management of conjunctival primary acquired melanosis (PAM) with atypia is in flux. Lesions are treated with biopsy, excision, adjunctive cryotherapy, or topical mitomycin.¹ Acceptable, usually minimal, complications have been reported with these adjunctive therapies.² Adverse events associated with topical mitomycin are uncommon and punctal stenosis is observed, especially in cold windy climates in approximately 20% of cases. Limbal stem cell deficiency has been described after the use of this agent on a dose-related basis, but is uncommon.^{2,3}

We report a case of recurrent PAM with atypia treated with three courses of mitomycin and two courses of cryotherapy, with subsequent stem cell deficiency and vision loss that was reversed with an autologous stem cell transplant. We received human experimentation committee (CCPMC Human Experimentation Committee) approval for this study.

Case report

A 56-year-old woman presented with an enlarging area of right conjunctival and corneal pigmentation. It started to expand five years ago with marked growth over the past six months. She had 20/30 visual acuity in the involved right eye; the left eye was unremarkable. There was circumferential superficial right limbal pigmentation with a thickened area inferiorly to the inferior fornix, as well as pigment dusting the caruncle. We resected the inferior corneal-conjunctival area of thickened pigmentation, which showed PAM with atypia. The surgical margins were clear. Adjunctive double freeze-thaw cryotherapy was used. She received two one-week courses of postoperative mitomycin C, 0.04% three times a day. There was some regression of the remaining pigmentation over the next few months. Approximately six months later, there was an area of increased limbal pigmentation, which we treated with an additional one-week course of mitomycin. This produced marked diminution of pigmentation.

Five months later she developed increased pigmentation, which was treated with a one-week course of 0.4% mitomycin with a marked decrease of the pigmentation.

Pigmentation remained superficial and stabilized for approximately a year, and then she developed an additional spread of flat pigmentation without new vessels or increased

thickness. We treated this area of involvement in the superior conjunctiva with double freeze-thaw cryotherapy, with marked diminution of pigmentation.

Two and a half years from our initial intervention, she developed new areas of pigmentation that were flat, and these were treated with another two-week course of topical mitomycin. This produced a marked clearing of the pigment.

Five months later, she developed decreased vision in the right eye with corneal epithelial irregularity. This progressed, and on examination four months later the visual acuity was 20/200, with epithelial edema. We used an autologous limbal stem-cell transplant from the opposite eye, with return of vision to 20/20. There was no residual pigmentation noted.

Comment

There are few reports of severe toxicity with either mitomycin or cryotherapy in the management of superficial conjunctival neoplasms.¹ Limbal stem-cell deficiency has previously been reported by us and others.⁴ It is likely as some patients are managed with recurrent disease by repeated applications of these therapies, especially a combination of mitomycin and cryotherapy, that they are more likely to produce stem-cell deficiency. In this report we have shown again that stem-cell transplantation is useful in repairing visual loss in this setting.

Devron H Char

The Tumori Foundation, San Francisco, CA, USA and
Department of Ophthalmology, Stanford University,
Stanford, CA, USA

Correspondence to: Dr Devron H Char, The Tumori Foundation, 45 Castro Street, Suite 309, San Francisco, CA 94114, USA; devron@tumori.org

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Posterior scleritis: an ominous sign of occult Takayasu's arteritis

Takayasu's arteritis is a rare chronic obliterative vasculitis affecting the aorta and its major branches. Epidemiologically it is found mainly in females of reproductive age and is more prevalent in Asian and Latin American countries. Although the pathogenesis has not been entirely elucidated, Takayasu's arteritis (TA) is considered to be a T-cell mediated granulomatous vasculitis.¹

We herein report the case of a patient who presented with posterior scleritis that proved to be the only clinical manifestation of a fatal case of TA.

Case report

A 26-year-old healthy male Caucasian patient was referred to the Department of Ophthalmology of Aberdeen Royal Infirmary because of blurred vision, painful red eyes and photophobia. For 20 months prior to presentation he was treated with intermittent topical steroids and NSAIDs because of bilateral refractory steroid-dependent episcleritis. On examination visual acuity was 6/6 OD and 6/24 OS. Pupils were poorly reactive to light and no afferent pupillary defect was detected. He had significant episcleral injection bilaterally, but no signs of anterior uveitis were seen. Fundoscopy revealed a swollen right optic disc with choroidal folds in the posterior poles of both eyes (fig 1). B-scan echography revealed significant diffuse scleral thickening around the optic nerve extending to the extraocular muscle insertions with widening of Tenon's space. The patient was thus diagnosed to have posterior scleritis. His physical examination was unremarkable and he had normal blood pressure. Investigations that included haematological and biochemical tests as well as C-reactive protein (CRP) and chest x ray were normal. Serum angiotensin converting enzyme level was just above the normal range at 54 U/l (range 8–52 U/l), lower than the level reported 1 year earlier (60 U/l). Antineutrophilic cytoplasmic antibodies were not detected.

Treatment was initiated with pulse therapy of intravenous methylprednisolone (1 g/day for 3 consecutive days). The patient then received prednisone at 1 mg/kg/day. There was marked improvement in the ocular pain and redness. Two weeks later he had visual acuity of 6/6 in each eye, with no evidence of episcleritis, less optic disc swelling but still evident

Figure 3 Histology slide of sclera showing necrotising granulomatous inflammation.

choroidal folds in both posterior poles. B-scan echography revealed significant resolution of the scleritis with residual scleral thickening.

However, 6 days later he developed vomiting, diarrhoea and abdominal pain. This continued for approximately 24 h. Whilst awaiting transfer to hospital, he collapsed and cardiopulmonary resuscitation was unsuccessful. Autopsy revealed that the cause of death was ischaemic colitis. Histology disclosed mesenteric arteritis and a necrotising granulomatous aortitis and periaortitis, consistent with Takayasu's arteritis, affecting the thoracic and abdominal aorta, with the changes in the thoracic aorta more granulomatous and older than those in the abdominal aorta (fig 2). The eyes showed evidence of a bilateral necrotising posterior scleritis (fig 3).

Comment

We herein present a rare ophthalmic manifestation of Takayasu's arteritis. Posterior scleritis was not previously reported in association with TA. Our patient presented with posterior scleritis as the sole clinical feature of the disease. Three recent reports described the occurrence of anterior scleritis in TA patients.^{1–3} Those patients were symptomatic as a result of vascular insufficiency and two of them² subsequently underwent surgical repair under immunosuppressive cover with a favourable outcome. Our patient had silent disease and did not manifest three of the six criteria that define TA according to the American College of Rheumatology (onset by age 40, claudication of the extremities, decreased brachial pulses, upper limb systolic blood pressure difference greater than 10 mm Hg, aortic or subclavian artery bruit and angiographic narrowing of large arteries).⁴ Laboratory tests did not indicate an occult inflammatory condition as the acute-phase reactants, including CRP, were not elevated. CRP is exclusively made in the liver and during tissue injury its serum concentration rises and subsequently falls rapidly. It is thus a very sensitive index of ongoing inflammation. Once a diagnosis has been established, CRP may be used to monitor the patient's response to therapy. In many cases, CRP remains normal despite severe disease.⁵ The mechanism of this selective failure of the acute-phase CRP response is currently unknown as has been shown in the patient described here. However, serum angiotensin converting enzyme level was borderline and it could have raised the suspicion of an underlying granulomatous process had its level been more elevated.

Figure 1 Photograph of the right fundus showing a swollen hyperaemic optic disc with radial folds across the posterior pole.

Figure 2 Histology slide of aorta showing necrotising granulomatous inflammation consistent with Takayasu's arteritis.