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.

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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 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.

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.

Takayasu, an ophthalmologist, was the first to describe the disease in 1908.⁶

Takayasu's retinopathy is the most common ophthalmic manifestation. It reflects ocular hypoperfusion and is manifested by micro-aneurysms, arterio-venous anastomosis and neovascular complications. Hypertensive retinopathy is less frequently encountered. Retinal arterial occlusion was recently reported as well.⁷ Smith and Rosenbaum¹ suggested a real association between TA and scleritis because of the strong temporal relationship between the two conditions in their patient. We herein provide the first histopathological evidence of a necrotising inflammatory process in the sclera as part of a generalised necrotising granulomatous aortitis in TA.

Administration of corticosteroids is recognised as the first-line management of TA. Our patient received high-dose parenteral steroid therapy over 3 days. Thereafter, he manifested a remarkable improvement both clinically and echographically. He was then put on a tapering dose of prednisolone. We speculate that an occult systemic inflammation would also respond to this therapy, but unfortunately TA here proved fatal with ischaemic colitis being the direct cause of death. This again highlights an unusual feature of TA, as Mwipatayi *et al*⁸ reported that cardiac and renal failure accounted for 57% of deaths in 57 TA patients.

Disease patterns are influenced by many factors that are either inborn or acquired. Examples include genetic background, gender, ethnicity and environmental hazards. Moriwaki *et al*⁹ reviewed the clinical manifestations and angiographic findings of 102 Indian and 80 Japanese patients with TA. In Japanese patients, TA involved the ascending aorta, the aortic arch and/or its branches, whereas in Indian patients, vasculitis occurred in the abdominal aorta and involved renal arteries, subsequently extending into the thoracic aorta within one or two decades. Disease pattern is not well defined in Caucasians due to its rarity. Our patient showed evidence of granulomatous aortitis affecting both the thoracic and abdominal aorta, with the inflammatory changes in the thoracic aorta more granulomatous and older than those in the abdominal aorta. This pathology was clinically occult until the development of mesenteric arteritis, which proved refractory to treatment and resulted in fatal ischaemic colitis.

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Cyclical orbital eosinophilic myositis

Eosinophilic myositis is a rare inflammatory condition of skeletal muscle associated with peripheral eosinophilia.¹ Orbital eosinophilic angiocentric fibrosis² and idiopathic orbital myositis³ have been reported. We report the first case of cyclical orbital eosinophilic myositis (OEM).

Case report

A 38-year-old woman presented to the ocular motility service in January 2006 with a one-week history of left-sided ptosis and diplopia. In the past four years, she had reported alternating ptosis and retro-ocular pain that recurred at six-week intervals. She was systemically well. Cranial computed tomography, magnetic resonance imaging, tensilon testing, neurophysiology and anti-acetylcholine receptor antibody had been normal.

Ocular examination revealed left partial ptosis, left hypotropia (20 prism dioptres (PD)) ipsilateral limitation of elevation, and ipsilateral temporal conjunctival hyperaemia (fig 1A, B). Visual acuity was normal bilaterally. Full blood count, inflammatory markers, serum angiotensin-converting enzyme, autoimmune screen, thyroid antibody, Lyme and worm serology were normal. Chest X-ray, thorax and abdominal computed tomography did not demonstrate regional lymphadenopathy or metastases. Magnetic resonance imaging scan showed increased signal in all rectus muscles (fig 1C). Presumed bilateral orbital myositis was managed with intravenous methylprednisolone 1 g a day for two days.

The hypotropia and ptosis improved rapidly (2-PD esophoria). She developed two recurrences over a 10-week period, one affecting the right side (fig 2A). Incisional biopsy of the left lateral rectus muscle demonstrated extensive disruption of the skeletal muscle by lymphoid aggregates with abundant eosinophils (fig 2B). This was reviewed by the national centre for tropical medicine and the regional reference laboratory. There were no histological features to suggest parasitic infection, and a diagnosis of OEM was made. Maintenance steroid therapy was started; however, the condition has remained unresponsive (fig 2C). Topical

Figure 1 (A) A 38-year-old woman with a left partial ptosis (left upper lid is kept open during testing) and limitation of elevation. There is a 20-PD left hypotropia. (B) Severe engorgement of the conjunctival and episcleral vessels overlying the left lateral rectus muscle. (C) Magnetic resonance imaging scan performed on presentation in 2006. Coronal short τ inversion recovery sequence of the orbits. Increased signal is present within all recti muscles bilaterally, particularly the left lateral rectus and superior rectus muscles bilaterally. Informed consent was obtained for publication of this figure.