illustrates the importance of obtaining a pertinent history and review of systems.

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Rituximab for the treatment of extranodal marginal zone B-cell lymphoma of the lacrimal gland

Extranodal marginal zone B-cell lymphoma of the mucosa-associated lymphoid tissue (MALT lymphoma) of the lacrimal gland is a rare condition. Treatment options chiefly include radiation of the tumour, chemotherapy, surgical removal or a combination of these strategies.[1] Radiation therapy is associated with a high risk of ocular morbidity, especially dry eye.[1] Here, we report a case of a biopsy-proven extranodal MALT lymphoma of the lacrimal gland treated with two courses of four weekly cycles of rituximab.

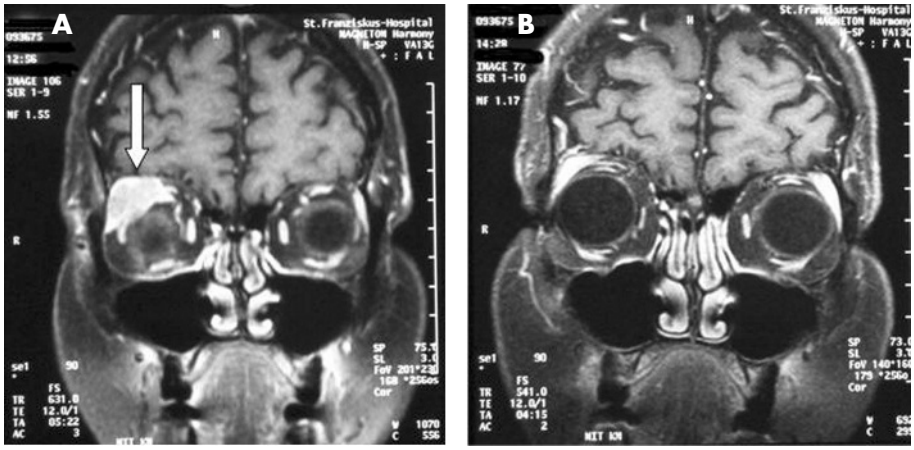

Figure 1 Coronal contrast-enhanced T1-weighted MR image shows MALT lymphoma (arrow) before therapy (A) and 12 months later (B).

Case report

A 64-year-old woman presented with unilateral ptosis and swelling of her right lacrimal gland. Best-corrected visual acuity was 20/25. Slit-lamp examination showed severe punctate superficial keratopathy (basal secretion 0 mm OU). MRI scan detected a tumour of the lacrimal gland (fig 1A). Histopathological and immunohistochemical studies of a lacrimal gland biopsy specimen disclosed an extranodal MALT lymphoma (fig 2); typical extraorbital manifestations were excluded. Treatment was started with four weekly cycles of rituximab (each dose 375 mg/m^2).

The MRI scan at 2 months showed a subtotal involution of the tumour. Schirmer values were 2 mm OU. Exophthalmus, which according to Hertel readings was 3 mm before treatment, had resolved 3 months after initiating therapy. Another four cycles of rituximab

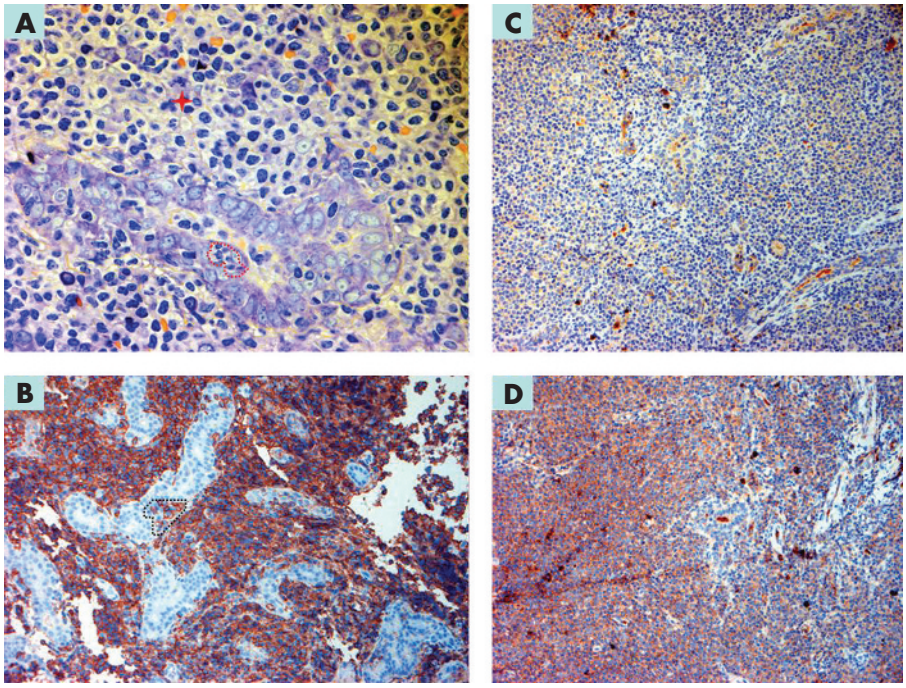

Figure 2 (A) Orbital soft tissue with destruction of the lacrimal gland by an atypical lymphoid infiltrate. Remnants of the glandular structures surrounded by atypical centrocytoid or monocytoid lymphatic cells with an increase in blasts (one atypical mitotic figure is highlighted with a cross) and scattered plasma cells can be seen. A typical intra-acinar lymphoepithelial lesion is shown surrounded by a dotted line. Giemsa 630×. (B) The neoplastic lymphoid infiltrate is mainly composed of strongly CD20-positive cells. Several destructive intraglandular lymphoepithelial lesions are clearly visible, one of which is highlighted by a surrounding dotted line. The complete phenotype of the lymphoma was CD20+, CD5−, CD23−, cyclinD1−, kappa+, lambda−; 200×. (C, D) Lambda and kappa light chain stainings show monoclonal light chain restriction of the neoplastic lymphoid cells, with a polyclonal background of plasma cells; 200×.

therapy were given 4 months after the first course to treat the residual lacrimal gland tumour. After 12 months the lacrimal gland tumour was no longer visible on MRI (fig 1B) and the Schirmer test score was 7 mm. Vision was unchanged and the corneal slit-lamp appearance was normal. In addition to topical lubricants, the patient underwent temporary punctual occlusion with resorbable collagen plugs as therapy. No extraorbital manifestation of the MALT lymphoma occurred during the subsequent follow-up period.

Comment

Patients treated with radiation for orbital MALT lymphomas often suffer from eye-related side effects. In a recent paper, four of nine patients who received additional radiotherapy for orbital and/or conjunctival lymphoma suffered from dry eye, conjunctivitis and cataract. One of the patients eventually lost his sight due to radiation-induced retinopathy.[1] Particularly when lymphoma develops in the lacrimal gland, the therapeutic approach should aim to prevent further damage to the that gland.

Treatment with the anti-CD20 monoclonal antibody rituximab for ocular adnexal MALT lymphomas has been reported primarily in recurring conjunctival lymphomas.[2] Only a few studies also report on the treatment of lacrimal gland involvement.[1 3 4] In a recent report, two patients with lacrimal gland lymphomas had a relapse after a median time of 5 months following primary rituximab therapy with one cycle of four weekly infusions.[3] Rituximab therapy also had a beneficial effect on the salivary and lacrimal gland function in patients with primary Sjögren syndrome associated with MALT lymphoma of the parotid gland.[5] Our patient presented with severe dry eye and a very low Schirmer test score. Notably, rituximab treatment led to significant relief of the patient's complaints and increased tear secretion. Rituximab may be considered as a first-line therapy for lacrimal MALT lymphoma when radiation therapy is expected to aggravate the dry eye symptoms or when a reduction in the tumour size is anticipated before radiation therapy. In our patient two courses of four weekly cycles of rituximab therapy led to a prolonged remission period.

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Efficacy of intravitreal triamcinolone for macular oedema due to CRVO after anti-androgen therapy for hirsutism in a young monocular female

After diabetic retinopathy, retinal venous occlusion (RVO) is the most common cause of retinopathy leading to severe visual loss in all age groups. Generally, RVO is associated with systemic diseases with significant morbidity and mortality. The risk of RVO is increased in patients older than 65 years of age with systemic hypertension, cardiovascular disease, diabetes and obesity.[1] Patients younger than 50 years of age may have other underlying conditions requiring appropriate investigation and treatment.[2 3] We describe the development of central retinal venous occlusion (CRVO) and the efficacy of intravitreal triamcinolone acetonide injection (IVTA) for associated cystoid macular oedema (CME) in a young healthy woman after she was started on systemic multiple anti-androgen therapy for treatment of hirsutism.

Case report

A 24-year-old woman with a 4-month history of painless loss of vision and a diagnosis of CRVO in her left eye (OS) was referred for further evaluation and management. Her ocular history revealed squint surgery and amblyopia in her right eye (OD) since childhood. Since puberty she had had acne and hirsutism that were unsatisfactorily treated conservatively. Seven months earlier she had sought treatment for the hirsutism and was started on systemic multiple anti-androgen therapy that included Noractone (spironlactone) 25 mg three times daily, Androcur (cyproterone acetate) 25 mg twice daily and Diane-35 (cyproterone acetate 2 mg and ethinyl oestradiol 35 μg) once daily. These medications were discontinued by a local ophthalmologist after the discovery of CRVO. The patient reported no other systemic disorder and family history was non-contributory. Her visual acuity (VA) was 20/300 in OD and 20/200 in OS with normal intraocular pressures. Slit-lamp examination was unremarkable with no evidence of rubiosis iridis in OS. Dilated fundus examination of OD was within normal limits; however, OS showed flame-shaped haemorrhages with tortuous dilated retinal veins (fig 1A). There was evidence of significant CME but no evidence of retinal neovascularisation on the disc or elsewhere. Intravenous fluorescein angiography of OS fundus showed delayed fillings and diffuse CME without any evidence of capillary non-perfusion or abnormal neovascularisation (fig 1B–D). Central macular thickness by optical coherence tomography was 736 μm (fig 1E). A diagnosis of non-ischaemic CRVO with extensive CME in OS was established. Her medical evaluation revealed normal blood pressure and physical examination. Mild microcytic hypochromic anaemia was noted in her laboratory work-up. Her erythrocyte sedimentation rate, protein electrophoresis, plasma viscosity, lipid profile, plasma homocysteine, sickling test, anti-phospholipid antibodies (cardiolipin antibodies IgG, IgM), plasma fibrinogen, lipoprotein A, abnormal factor V Leiden, factors VII, VIII, XII, antithrombin III, protein S, and protein C were normal. She had negative anti-nuclear antibody with non-reactive TPHA. Application of grid laser therapy to the macular area did not resolve her CME. However, a single injection of IVTA (4 mg/0.1 ml) resulted in a dramatic decrease in CME (fig 2A) and improvement of her VA to 20/50. The improvement in CME was verified by intravenous fluorescein angiography (IVFA; fig 2B,C) and optical coherence tomography (OCT; fig 2D), and was sustained over a 3-year follow-up period.

Discussion

Hirsutism (excessive hair growth), acne and seborrhea are distressing and common endocrine problems in women which may be difficult to manage conservatively. One cause of hirsutism is excessive androgen production by the ovaries. Anti-androgen regimes effective for hirsutism include spironolactone, cyproterone acetate and cyproterone acetate with ethinyl oestradiol (Diane-35).[4] These medications suppress the release of gonadotrophin hormone, which reduces androgen production. Cyproterone used alone has a minor effect on blood clotting factors, however in combination with ethinyl oestradiol it enhances coagulation. Numerous reports have described vascular complications due to drugs after use of the oral contraceptive pill (OCP). Use of the OCP is known to be a risk factor for cardiovascular and cerebrovascular diseases and, as this report suggests, CRVO.[5] Other serious ocular complications include branch RVO, artery occlusion and acute ischaemic optic neuropathy.[5 6] Investigation in a 28-year-old CRVO patient treated with an anti-androgen drug revealed an abnormality of the haemostatic system; however, 1 month after the primary episode CRVO resolved and the fundus became normal without macular oedema.[7] Our case suggests that anti-androgen therapy in young patients may result in CRVO. A single injection of IVTA (4 mg/0.1 ml) may be effective to resolve CRVO-associated CME and improve VA. To the best of our knowledge this is the first report