Figure 2 Top row shows a representative normal trace with L.ocus dexter and L.ocus sinister responses of subject in rows 2 and 3.

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Combination treatment of intravenous immunoglobulin and cultivated oral mucosal epithelial transplantation for ocular cicatricial pemphigoid

Ocular cicatricial pemphigoid (OCP) is a progressive cicatrizing autoimmune disease affecting the skin and mucous membranes, including ocular surface epithelia.¹ Although systemic immunosuppression is advocated, both progression of ocular surface inflammation and systemic side effects remain a problem.² Intravenous administration of immunoglobulin (IVIg) is an immunomodulating treatment, and encouraging results have been reported for acute-phase OCP.³ However, corneal scarring and neovascularisation have also been reported. Cultivated oral mucosal epithelial transplantation (COMET) is a recently developed surgical method for ocular surface transplantation.⁴ Autologous oral epithelial cells were cultivated on preserved amniotic membranes, and the sheets were transplanted after fibrotic tissue excision. As autologous cells are used, immunological rejection is avoided, thereby eliminating complications associated with immunosuppression.⁵ In this report, we describe a patient with OCP successfully treated by a combination of IVIg and COMET.

Case report

A 55-year-old man with bilateral conjunctival inflammation, photophobia and bulla formation in oral mucosa was first seen in May 2003 with a 2-month history of systemic corticosteroids without improvements. The best-corrected visual acuity (BCVA) was hand motion in his right eye and 20/1000 in his left. Slit-lamp examination disclosed subconjunctival fibrosis, partial conjunctivalisation, foreshortening of fornices, and large corneal and conjunctival epithelial defects in both eyes (fig 1A). Immunohistology of the conjunctival and oral mucosal biopsy samples showed immunoglobulin (Ig)G deposition at the basement membrane zone, confirming the diagnosis of OCP.

Intensive treatments including ciclosporin, 1% methylprednisolone, preservative-free 0.3% hyaluronic acid, levofloxacin for topical treatments and prednisolone starting from 60 mg/day and cyclophosphamide 100 mg/day for systemic treatment were started. Amniotic membrane transplantation was also performed. Although some improvements were appreciated, recurrent epithelial defects and progress of fibrosis were noted. We started IVIg with the protocol of receiving immunoglobulin 20 g/day for 5 days, repeated as one cycle every alternate week, then tapered; after the 9th cycle, the applied dose was decreased gradually.³

Soon after starting of IVIg, the photophobia and pain improved. Repeated ulceration in the skin and oral mucosa, and nasal bleeding were also resolved. However, symblepharon and conjunctivalisation remained, the Schirmer

Figure 1 (A) Slit-lamp examination showing severe conjunctival inflammation and conjunctival shortening before intravenous administration of immunoglobulin (IVIg) treatment. (B) Ten months after the IVIg treatment, scarring and conjunctivalisation on the cornea can be observed with progressive cataract. (C) Six months after the cultivated oral mucosal epithelial transplantation, improvement in clarity of the cornea and conjunctival shortening can be observed.

test value was 0 mm and BCVA was decreased to hand motion in both eyes (fig 1B). To obtain visual improvement, surgery was performed in the right eye 10 months after the start of IVIg. Firstly, we removed all fibrotic tissue, then we applied 0.04% mitomycin C for 3 min, followed by irrigation with saline solution. Phacoemulsification and intraocular lens insertion were performed after the cornea had regained clarity. Cultivated autologous oral mucosal epithelial sheets prepared on preserved human amniotic membrane for 2 weeks⁴ were transplanted on the exposed ocular surface. Postoperatively, both the cornea and sclera were promptly epithelialised, and marked reduction in fibrosis was noted. One year after surgery, BCVA had improved to 20/300 and the corneal clarity was maintained (fig 1C). Improvements in vision and reduction in photophobia enabled the patient to return to social activities.

For the treatment of an active OCP, both control of autoimmune reaction and ocular surface reconstruction are necessary. We believe that the combination of IVIg and COMET is a powerful treatment modality while minimising the risk of postoperative consequences inevitably associated with immunosuppression.

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The potential role of testosterone in central serous chorioretinopathy

Risk factors for central serous chorioretinopathy (CSCR) are male gender, psychological stress, type-A personality, corticoid–steroid treatment and pregnancy. The reason for the presence of male gender as a risk factor is not yet clear. One possibility is a direct influence of androgens. We report a case of a female patient who developed CSCR under testosterone treatment.

Case report

A 45-year-old non-pregnant woman presented with a 1-day history of metamorphopsia and scotoma in the right eye. She had no eye problems in the past, but her current profession as a manager exposed her to several stress situations. On examination of the right eye, the best-corrected visual acuity (BCVA) was 20/25; paracentral scotoma and metamorphopsia

Figure 1 (A) Optical computerised tomography (OCT) confirming serous neurosensory and retinal pigment epithelium detachment in the right eye. (B) OCT of the right eye at 4-month follow-up showing reattachment of the retina.

were confirmed using the Amsler grid. Fundus examination disclosed a neurosensory serous detachment, and fluorescein angiography ruled out choroidal neovascular membrane. Optical coherence tomography (OCT) showed a typical CSCR (fig 1A). The left eye had a BCVA of 20/20 and did not show any pathology. At the second visit 3 weeks later and on being questioned again, the patient reported that she was on oral testosterone undecanoate 40 mg/day for 2 months because of a general loss of energy and symptoms of fatigue attributed to a low level of endogenous testosterone. At this stage, the plasma-testosterone level was markedly increased (table 1).

The testosterone treatment was stopped immediately. Oral oestradiol 1 mg/day was given for climacteric symptoms and for prevention of osteoporosis. This produced an increase in circulating oestradiol levels together with feedback suppression of both gonadotropins, luteinising hormone and follicle-stimulating hormone, as well as an increase in the sex-hormone-binding globulin. Sex-hormone-binding globulin enhances the binding of testosterone on this protein, thereby decreasing free circulating testosterone.

At 1 month follow-up, BCVA was 20/20, and angiography showed no signs of leakage or pigment epithelial detachment. Her blood level of testosterone dropped to values in the lower normal range (table 1). The neurosensory retina was reattached as confirmed by OCT at 4 months (fig 1B).

Table 1 Summary of the hormone levels

	Values on testosterone treatment*	Values after cessation of androgen treatment†	Normal range for females
Total testosterone	5.30	0.69	0.5–3.0 nmol/l
SHBG	37	141	20–118 nmol/l
Oestradiol	73	383	40–200 pmol/l
DHEA-S	3.6	1.2	3–12 µmol/l
LH	60	24	>10 E/l (menopausal)
FSH	56	29	>15 E/l (menopausal)

DHEA-S, dehydroepiandrosterone sulphate; FSH, follicle-stimulating hormones; LH, luteinising hormone; SHBG, sex-hormone-binding globulin.

*Oral testosterone undecanoate (40 mg/day).

†Oral oestradiol (1 mg/day).