Clinicians and patients should be aware that immunosuppression significantly increases risk of skin cancer, and that any suspicious skin lesion should undergo urgent excision biopsy, in an attempt to prevent development and growth of these aggressive lesions. Patients should be advised to take sensible precautions against UV light exposure, such as wearing sun blocking creams and hats, in an attempt to reduce the risks of developing skin carcinoma.

References

1 Westburg SP, Stone OJ. Multiple cutaneous squamous cell carcinomas during immunosuppressive therapy. *Arch Dermatol* 1973; **107**: 893–895.
2 Stockfleth E, Ulrich C, Meyer T, Arndt R, Christophers E. Skin diseases following organ transplantation—risk factors and new therapeutic approaches. *Transpl Proc* 2001; **33**: 1848–1853.
3 Digiovanna JJ. Post-transplantation skin cancer: scope of the problem, management, and role for systemic retinoid chemoprevention. *Transpl Proc* 1998; **30**: 2771–2775.
4 Euvrard S, Kanitakis C, Pouteill-Noble F, Disant G, Finaz de Villaine J, Claudy A *et al*. Aggressive squamous cell carcinomas in organ transplant recipients. *Transpl Proc* 1995; **27**: 1767–1768.
5 Liddington M, Richardson AJ, Higgins RM, Endre ZH, Venning VA, Murie JA, Morris PJ. Skin cancer in renal transplant recipients. *Br J Surg* 1989; **76**: 1002–1005.
6 Hartevelt MM, Bavinck JN, Kootte AM, Vermeer BJ, Vandenbroucke JP. Incidence of skin cancer after renal transplant in the Netherlands. *Transplantation* 1990; **49**: 506–509.
7 Reifler DM, Hornblass A. Squamous cell carcinoma of the eyelid. *Surv Ophthalmol* 1986; **30**: 349–363.
8 Rao NA, Dunn SA, Romero JL, Stout W. Bilateral carcinomas of the eyelid. *Am J Ophthalmol* 1986; **101**: 480–482.
9 Stewart WB, Nicholson DH, Tenzel RR, Spencer WH. Eyelid tumours and renal transplantation. *Arch Ophthalmol* 1980; **98**: 1771–1772.
10 Bernstein SC, Lim KK, Brodland DG, Heidelberg KA. The many faces of squamous cell carcinoma. *Dermatol Surg* 1996; **22**: 243–254.
11 Henkind P, Freidman A. Cancer of the lids and ocular adnexia. *Cancer of the Skin*. WB Saunders: Philadelphia, 1976, pp 1345–1371.

CM Knapp, JHY Tan and R Sampath

Department of Ophthalmology
Leicester Royal Infirmary
Leicester, UK

Correspondence:
CM Knapp
Department of Ophthalmology
Leicester Royal Infirmary
Infirmary Square
Leicester LE1 5WW, UK
Tel: 0116 2541414
Fax: 0116 2585927
E-mail: christopher_knapp@lineone.net

Sir,

Surgical management of anterior lenticonus in Alport's syndrome

Eye (2002) **16,** 798–800. doi:10.1038/sj.eye.6700218

Alport's syndrome is a rare basement membrane disorder characterized by progressive hereditary nephritis, sensorineural hearing loss and ocular abnormalities. The inheritance is predominantly X-linked (85%), although it can be autosomal recessive (10%) or autosomal dominant (5%).[1] Homozygote males are usually severely affected but females tend to have a mild form, often with only microscopic haematuria and normal renal function.[1] The ocular abnormalities in the X-linked form are characteristically a dot-and-fleck retinopathy, less often anterior lenticonus and rarely a posterior polymorphous corneal dystrophy.[1,2] Additional ocular abnormalities are not uncommon, including cataracts, posterior lenticonus and retinal detachment.[2]

This case report of a female patient has several unusual features. There was no positive family history, renal function was severely affected and prior to developing visual symptoms, the cause of renal failure was unclear. In addition the ocular features included a unilateral dot-and-fleck maculopathy and bilateral anterior lenticonus, known to occur in approximately 85% and 25% of affected males respectively, but both rarely reported in females with the syndrome.[1]

Case report

A 40-year-old lady was referred by her optometrist with bilateral visual failure and a large myopic shift over a 2-week period. Within this period her refractive status had changed from −7.75DS / −1.25DC × 105 right eye, −6.75DS left eye, to −10.00DS / −1.75DC × 115 right eye, −8.25DS / −0.50DC × 30 left eye.

The patient had no significant past ocular history other than wearing spectacles to correct myopia for many years. Her past medical history however, included bilateral hearing loss for which she wore hearing aids and a previous renal transplant (1994) for renal failure secondary to a 'nephritis' of unknown aetiology. She was on long-term immunosuppressive therapy, including oral steroids. There was no family history of ocular or systemic conditions.

On examination the Snellen visual acuity with

spectacles was 6/60 right eye and 6/24 left eye. The ocular surface and corneas were normal, with intraocular pressures of 12 mmHg in each eye. There was marked bilateral anterior lenticonus (Figure 1), with a positive 'oil droplet' sign on retinoscopy. Both lenses were clear, with no evidence of cataract. Fundoscopy revealed subtle pale perifoveal flecks at the right posterior pole, but no abnormalities in the left eye.

The ocular signs in conjunction with her hearing loss and renal disease were compatible with a clinical diagnosis of Alport's syndrome. The visual failure appeared to be due to the development or exacerbation of bilateral anterior lenticonus. A right clear lens phacoemulsification and intraocular lens (IOL) implantation was performed under general anaesthetic. The implant used was a foldable, hydrophobic, acrylic IOL. The right eye was 6/9 unaided (6/5 with pinhole) the following day. The postoperative period was uneventful, although topical steroids were required at a slightly increased frequency for anterior uveitis. The left eye underwent a similar procedure 8 weeks later, the unaided vision settling at 6/12, improving to 6/6 with a small myopic correction.

A review, 12 months postoperatively, disclosed the following findings on examination. The best-corrected Snellen visual acuity had remained 6/6, in each eye, with a small myopic correction. The anterior chambers were quiet, the IOL implants were well-centred and the intraocular pressures were normal. No significant posterior capsular opacification was present. The perifoveal flecks at the right posterior pole were unchanged.

Comment

Alport's syndrome was first described as an acute haemorrhagic nephropathy with deafness in 1927, the association with anterior lenticonus being recognized later.[3,4] The underlying abnormality is a defective type IV collagen, the synthesis of which is coded for on the X chromosome. Histopathological features of anterior lenticonus include marked thinning of the anterior capsule centrally, decreased number of lens epithelial cells which may also be atypically arranged and large numbers of partial capsular dehiscences containing fibrillar material and vacuoles.[5–8]

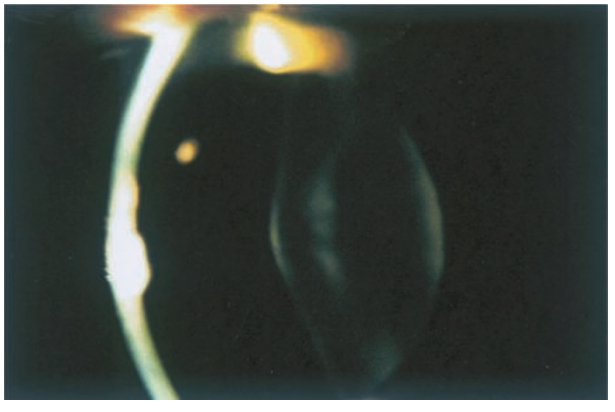

Figure 1 Anterior lenticonus in a 40-year-old patient with Alport's syndrome.

The association of 'capsular fragility' with anterior lenticonus secondary to Alport's syndrome is well documented[6–8] and is supported by rare reports of spontaneous rupture of the capsule.[9,10] In our case, however, we found that the anterior capsule was highly elastic, making the capsulorhexis technically more difficult in both eyes. The capsule elasticity was far greater than one would expect in relation to the patient's age. John *et al* have also reported a case where the anterior capsules were 'tough'.[11] It is possible that fragile capsules represent a more severe form of ocular disease.

There are various precautions that can be taken to help complete a successful capsulorhexis in anterior lenticonus. It is worth considering a general anaesthetic in challenging cases, as the procedure may take longer and akinesia is more important. Making a small paracentesis initially allows for a complete aqueous-viscoelastic exchange in a 'sealed' anterior chamber. Use of a high molecular weight viscoelastic agent may further help deepen the anterior chamber and flatten the anterior capsule. We found it necessary to perforate the capsule with a 27G needle and then complete the capsulorhexis using microforceps. The capsule was highly elastic and far more prone to tear out towards the lens periphery.

Having completed the capsulorhexis a gentle, but thorough, hydrodissection from several sites was made to ensure that the lens rotated well within the capsular bag. This is particularly important in younger patients when the lens can be extremely soft. Extreme caution is required when there are associated posterior subcapsular opacities or posterior lenticonus, since hydrodissection could result in a posterior capsular rupture. Under these circumstances good hydrodelineation to ensure easy nucleus rotation within the epinuclear shell is recommended. A standard 'bowl technique' was used during the phacoemulsification, the centre of the soft lens being debaulked and then imploded with high vacuum during the second stage of nucleus removal. The lens cortex was aspirated in a circumferential fashion in order to avoid unnecessary tension on the zonules. The capsule was polished thoroughly to avoid capsule phimosis and possible posterior capsule opacification

and a foldable, hydrophobic, acrylic IOL was inserted into the capsular bag.

Clear lens phacoemulsification is an excellent surgical procedure for the treatment of anterior lenticonus in patients with Alport's syndrome, allowing for rapid visual rehabilitation.

References

1 Colville DJ, Savige J. Alport syndrome. A review of the ocular manifestations. *Ophthal Genet* 1997; **18**: 161–173.
2 Colville D, Savige J, Morfis M *et al*. Ocular manifestations of autosomal recessive Alport syndrome. *Ophthal Genet* 1997; **18**: 119–128.
3 Alport AC. Hereditary familial congenital haemorrhagic nephritis. *Br Med J* 1927; **1**: 504–506.
4 Arnott EJ, Crawford MD'A, Toghill PG. Anterior lenticonus and Alport's syndrome. *Br J Ophthalmol* 1966; **50**: 390–403.
5 Brownell RD, Wolter JR. Anterior lenticonus in familial hemorrhagic nephritis. *Arch Ophthalmol* 1964; **71**: 481–483.
6 Streeten BW, Robinson MR, Wallace R *et al*. Lens capsule abnormalities in Alport's syndrome. *Arch Ophthalmol* 1987; **105**: 1693–1697.
7 Junk AK, Stefani FH, Ludwig K. Bilateral anterior lenticonus: Scheimpflug imaging system documentation and ultrastructural confirmation of Alport syndrome in the lens capsule. *Arch Ophthalmol* 2000; **118**: 895–897.
8 Kato T, Watanabe Y, Nakayasu K *et al*. The ultrastructure of the lens capsule abnormalities in Alport's syndrome. *Jpn J Ophthalmol* 1998; **42**: 401–405.
9 Olitsky SE, Waz WR, Wilson ME. Rupture of the anterior lens capsule in Alport syndrome. *J AAPOS* 1999; **3**: 381–382.
10 Ignat'ev AN. Case of bilateral anterior lenticonus with spontaneous rupture of the capsule of the lens and a swelling cataract in one eye. *Oftalmol Zh* 1980; **35**: 375–376.
11 John ME, Noblitt RL, Coots SD *et al*. Clear lens extraction and intraocular lens implantation in a patient with bilateral anterior lenticonus secondary to Alport's syndrome. *J Cataract Refract Surg* 1994; **20**: 652–655.

I Mavrikakis, C Zeilmaker and MJ Wearne

Department of Ophthalmology
Eastbourne District General Hospital
Eastbourne, UK

Correspondence: IMJ Wearne, FRCOphth
Department of Ophthalmology
Eastbourne District General Hospital
Kings Drive, Eastbourne
East Sussex, BN21 2UD, UK
Tel: 01323 417400
Fax: 01323 414908
E-mail: mwearne@eyetext.net

Sir,

Bilateral idopathic optic nerve sheath meningocele associated with unilateral transient cystoid macular oedema

Eye (2002) **16,** 800–802. doi:10.1038/sj.eye.6700212

Optic nerve sheath dilatation or meningocele is a rare condition describing an 'enlargement and dilation of primarily the optic nerve sheath'.[1] There is an expansion of the cerebrospinal fluid space around the optic nerve with no associated inflammation, orbital or cerebral neoplasm at the apex of the orbit. We describe a case of bilateral meningoceles with an associated unilateral cystoid macular oedema (CMO).

Case report

A 59-year-old Caucasian male presented with a 10-month history of blurring of vision in the right eye. He had experienced a 6-month period of retro-orbital and facial pain. His past medical history included presumed tuberculoma of a cervical gland at the age of 15 years, hypertension, chronic obstructive airways disease and angina.

His best corrected visual acuity was 6/9 in the right eye and 6/6 in the left on presentation. Colour vision was tested with Ishihara colour plates and was normal on both sides. On examination he had bilateral symmetrical proptosis but the patient indicated that his appearance had been stable for many years. His pupil reactions were normal. His ocular motility was unrestricted and his anterior segment was normal. Contact lens fundoscopy revealed CMO in the right eye. There was no disc swelling. The left eye showed no fundal abnormalities.

Investigations including thyroid function tests, full blood count, serum B12, folate, urea and electrolytes, erythrocyte sedimentation rate and a treponemal screen were normal. Anti-nuclear antibodies were negative. Humphrey visual field testing showed non-specific depression in the periphery of both eyes which was not consistent with any neurological defect. A lumbar puncture was performed with an opening pressure of 24 cm of water.

Magnetic resonance imaging (MRI) scans revealed bilateral optic nerve sheath dilatation and enlargement but with normal sized optic nerves (Figure 1).

The patient was followed up for the next 5 years and demonstrated a gradual deterioration in visual acuity in the right eye. Colour vision was also reduced in the right eye with a score of 10 correct responses out of the first 21 Ishihara plates. Visual evoked responses at