Figure 3 (a) Fundus fluorescein angiogram of case 2 showing the subretinal neovascular membrane in the right macula. (b) Angiogram of left macula.

Figure 4 Pedigree of the family with autosomal dominant inheritance with retinal and macular phenotypes.

References

- 1 Wells J, Wroblewski J, Keen J, Inglehearn C, Jubb C, Eckstein A *et al.* Mutations in the human retinal degeneration slow (RDS) gene can cause either retinitis pigmentosa or macular dystrophy. *Nat Genet* 1993; **3**(3): 213–218.
- 2 Wang DY, Chan WM, Tam PO, Baum L, Lam DS, Chong KK *et al.* Gene mutations in retinitis pigmentosa and their clinical implications. *Clin Chim Acta* 2005; **351**(1–2): 5–16.
- 3 Sohocki MM, Daiger SP, Bowne SJ, Rodriguez JA, Northrup H, Heckenlively JR *et al.* Prevalence of mutations causing retinitis pigmentosa and other inherited retinopathies. *Hum Mutat* 2001; **17**(1): 42–51.
- 4 Trujillo MJ, Bueno J, Osorio A, Sanz R, Garcia-Sandoval B, Ramos C *et al.* Three novel RDS-peripherin mutations (689delT, 857del17, G208D) in Spanish families affected with autosomal dominant retinal degenerations. Mutations in brief no 147. *Online Hum Mutat* 1998; **12**(1): 70.

TS Kalyanasundaram, GC Black, J O'Sullivan and PN Bishop
Department of Ophthalmology, Manchester Royal
Eye Hospital, Manchester, UK
E-mail: tskalyan@yahoo.com

Proprietary interests or research funding: None.

This work has not been presented anywhere.

Eye (2009) **23**, 237–239; doi:10.1038/eye.2008.33;
published online 21 March 2008

Sir, Necrobiotic xanthogranuloma masquerading as posterior scleritis

Necrobiotic xanthogranuloma is a rare skin disease that may herald eye complications. We report the presentation of posterior scleritis in a patient 2 years after a skin biopsy-proven diagnosis of necrobiotic xanthogranuloma.

Case report

A 47-year-old Caucasian lady presented with a 3-day history of reduced vision in her right eye, redness, watering, photophobia, pain, and headache. She was already under regular follow-up for blepharitis. She had necrobiotic xanthogranuloma skin lesions proven by biopsy on her upper and lower limbs (Figure 1). She also had raised ESR, IgG- κ paraproteinaemia, neutropenia, lymphopenia, and low complement C4.

Visual acuities were 6/18 right eye and 6/5 left eye. She had bilateral anterior blepharitis and anterior segments were otherwise unremarkable. Fundoscopy revealed bilateral choroidal folds and right cystoid macular oedema.

Ultrasonography showed irregular thickening of the sclera in both eyes (Figure 2). Magnetic resonance imaging of the orbits showed bilateral diffuse-enhanced thickening of posterior sclera but no orbital abnormality. A diagnosis of bilateral posterior scleritis associated with

Figure 1 Necrobiotic xanthogranuloma on the skin of the right thigh.

Figure 2 B-scan ultrasonography of the eye before treatment. Note the irregular lumpy thickening of the globe.

necrobiotic xanthogranuloma was made. She was commenced on prednisolone 60 mg daily. Her vision, symptoms, and signs improved dramatically and after 1 month, she was asymptomatic. Vision in both eyes was 6/5 2 months after starting treatment. Repeat ultrasonography showed complete resolution of the scleral thickening. Her skin lesions regressed and the paraproteinaemia also resolved. Prednisolone was tapered and finally discontinued after 4 months. She has remained asymptomatic for the past 3 years.

Comment

Necrobiotic xanthogranuloma is a rare progressive disease usually affecting patients in their fifth to seventh decades. It is characterised by red or yellow skin papules or indurated plaques with surface telangiectasia, which subsequently ulcerate and scar. Ophthalmological complications affect 50% of patients and include orbital and subconjunctival masses, keratitis, episcleritis, scleritis, and uveitis. Other organs may rarely be involved. Approximately 80% of patients have a

monoclonal gammopathy and 10% of these patients develop multiple myeloma. Hepatosplenomegaly, high ESR, leucopenia, hypocomplementemia, and cryoglobulinaemia are other common findings.¹ Necrobiotic xanthogranulomas are composed of histiocytes and Touton giant cells, forming palisades around zones of necrotic tissue, degenerated collagen, and cholesterol clefts. They are triggered by deposition of immunoglobulins and lipid complexes in affected tissues.²

Improvement of skin lesions has been seen with systemic corticosteroids, chlorambucil, melphalan, radiotherapy, interferon α -2a, and plasmapheresis. Intralesional injection of triamcinolone may be beneficial for accessible lesions.³⁻⁵ The scleritis in our patient was thought to be related to necrobiotic xanthogranuloma. Scleral biopsy was not carried out because we felt that it would have been technically difficult, potentially hazardous, and unlikely to add useful information. Her systemic diagnosis confirmed by skin biopsy and the beneficial response to steroid treatment supports our diagnosis.

Patients with necrobiotic xanthogranuloma may present to many specialties. Posterior scleritis is an uncommon but treatable sight-threatening complication of this disease. Prompt ophthalmological referral is warranted if eye symptoms develop.

References

- Robertson DM, Winkelman RK. Ophthalmic features of necrobiotic xanthogranuloma with paraproteinaemia. *Am J Ophthalmol* 1984; **97**(2): 173-183.
- Matsura F *et al.* Activation of monocytes *in vivo* causes intracellular accumulation of lipoprotein-derived lipids and marked hypocholesterolaemia: a possible pathogenesis of necrobiotic xanthogranuloma. *Atherosclerosis* 1999; **142**: 355.
- Chave TA, Chowdhury MM, Holt PJ. Recalcitrant necrobiotic xanthogranuloma responding to pulsed high-dose oral dexamethasone plus maintenance therapy with oral prednisolone. *Br J Dermatol* 2001; **144**: 158.
- Char DH, LeBoit PE, Ljung BM, Wara W. Radiation therapy for ocular necrobiotic xanthogranuloma. *Arch Ophthalmol* 1987; **105**: 174-175.
- Elner VM, Mintz R, Demirci H, Hassan AS. Local corticosteroid treatment of eyelid and orbital xanthogranuloma. *Trans Am Ophthalmol Soc* 2005; **103**: 69-73.

Ayad Al-Bermani¹, F Figueiredo¹, EL Speight², GH Jackson³ and R Pandit¹

¹Department of Ophthalmology, Royal Victoria Infirmary, Newcastle upon Tyne, UK

²Department of Dermatology, Royal Victoria Infirmary, Newcastle upon Tyne, UK

³Department of Haematology, Royal Victoria Infirmary, Newcastle upon Tyne, UK
E-mail: ayadberman@hotmail.com

Eye (2009) **23**, 239-240; doi:10.1038/eye.2008.37;
published online 22 February 2008