NJS London¹, C Siverio Jr² and ET Cunningham Jr^{1,3}

¹California Pacific Medical Center, San Francisco, CA, USA

²The Instituto Nacional de Oftalmologia, Lima, Peru

³Department of Ophthalmology, Stanford University School of Medicine, Stanford, CA, USA
E-mail: emmett_cunningham@yahoo.com

Eye (2010) **24**, 1294–1295; doi:10.1038/eye.2009.333;
published online 15 January 2010

Sir,
Coats' disease associated with muscular dystrophy treated with ranibizumab

Coats' disease can be associated with muscular dystrophy. We present a case in which ranibizumab was used for Coats' disease associated with a muscular dystrophy.

Case report

An 18-year-old man came to our department because he had noted decreased vision in his left eye. The patient had no systemic complaints. The best-corrected visual acuity (VA) was 20/20 in his right eye (RE) and count

fingers in his left eye (LE). The anterior segment was unremarkable. Fundus examination showed hard circinate exudates, telangiectatic vessels, microaneurysms, and venous dilatation only in the LE (Figures 1a and b). The patient underwent fluorescein angiography (Figures 1c and d) and optic coherence tomography (OCT) (Figure 2). Coats' disease was confirmed.

A complete blood count, liver and muscle enzymes and coagulation testing was performed. The patient showed a consistently elevated creatine phosphokinase on repeat testing of 3000 UI/ml. We referred the patient to a neurologist, who performed the diagnosis of facioscapulohumeral dystrophy (FSHD).

Peripheral argon laser photocoagulation and three ranibizumab injections were administered to the LE. The VA improved to 20/60 and has stayed stable with 1 year of follow-up (Figure 3).

Comment

Coats' disease is characterized by vascular abnormalities, commonly telangiectatic vessels and retinal exudation. Normally only one eye is involved and predominantly men are affected before 20 years of age.^{1,2} Usually, it is not necessary to do a systemic study with typical signs of Coats' disease but there are some cases of muscular and facioscapulohumeral dystrophies associated with Coats'

Figure 1 (a) Funduscopy of left eye with the presence of hard circinate exudates. (b) Funduscopy of periphery of left eye in which telangiectatic vessels, venous dilatations, microaneurysms can be observed. (c) Angiogram of left eye showing exudation in posterior pole. (d) Angiogram of temporal periphery of left eye showing telangiectasia and tortuosity of retinal vessels.

Figure 2 First OCT with macular oedema.

Figure 3 Last OCT, after treatment (peripheral laser and ranibizumab injections).

disease in the literature.³ FSHD is a muscular dystrophy characterized by weakness of the muscles of the facial and scapulohumeral regions. The age of presentation is in early adulthood. FSHD has an autosomal dominant inheritance and it maps to chromosome 4.⁴ Coats' disease, hearing loss, and mental retardation may be extramuscular manifestations of FSHD.⁵ Most cases of FSHD are associated with bilateral Coats' disease.³ There are a few cases of isolated Coats' disease in which other anti-angiogenics were used but, to the best of our knowledge, this is the first case, especially with FSHD, treated with ranibizumab.²

Conflict of interest

The authors declare no conflict of interest.

References

- 1 Retina and Vitreous. *Basic and Clinical Science Course Section 12*. American Academy of Ophthalmology: Singapore, 2007.
- 2 Venkatesh P, Mandal S, Garg S. Management of Coats disease with bevacizumab in 2 patients. *Can J Ophthalmol* 2008; **43**: 245–246.
- 3 Shields CL, Zahler J, Falk N, Furuta M, Eagle Jr RC, Espinosa LE *et al*. Neovascular glaucoma from advanced Coats disease as the initial manifestation of

facioscapulohumeral dystrophy in a 2-year-old child. *Arch Ophthalmol* 2007; **125**(6): 840–842.

- 4 Brouwer OF, Padberg GW, Ruys CJ, Brand R, de Laat JA, Grote JJ. Hearing loss in facioscapulohumeral muscular dystrophy. *Neurology* 1991; **41**: 1878–1881.
- 5 Fitzsimons RB, Gurwin EB, Bird AC. Retinal vascular abnormalities in facioscapulohumeral muscular dystrophy: a general association with genetic and therapeutic implications. *Brain* 1987; **110**: 631–648.

T Diago¹, B Valls¹ and JS Pulido²

¹Department of Ophthalmology, Hospital of Sagunto, Valencia, Spain

²Department of Ophthalmology, Mayo Clinic, Rochester, MN, USA

E-mail: tediase@gmail.com

Eye (2010) **24**, 1295–1296; doi:10.1038/eye.2009.336; published online 15 January 2010

Sir, Eyelid necrosis after local anaesthesia for lacrimal sac surgery

A 74-year-old woman (C1) and an 81-year-old woman (C2) with no history of atopy each received a standard injection of 3 ml lidocaine hydrochloride 2% and 1/100 000-epinephrine into the medial canthal region in preparation for dacryocystectomy. For each injection, the contents were obtained from a recently opened sterile bottle and delivered with a single sterile needle. Each woman had received lidocaine local anaesthesia (LLA) in the past, peribulbar administration in the case of C1 and non-ophthalmic administration for the implantation of a pacemaker in C2. During the injection, each of them experienced sudden faintness and local pain, and within a few hours, an extensive periorbital erythema and swelling was observed. Within 8 days, skin necrosis involving the medial canthus occurred and was accompanied by a serous fluid-filled bulla (Figures 1a and 2a). There was no sign of periorbital/orbital involvement on computerised tomography, no abnormal laboratory findings, and microorganisms were absent in direct microscopy and in the culture of bullous fluid. Treatment with intravenous amoxicillin and local and systemic corticosteroids was given. After 15 days, swelling and necrotic skin changes decreased (Figures 1b and 2b), and 6 months later, a retractile pigmented scar with a lower lid cicatricial ectropion (Figures 1c, d and 2c) was noted.

In ophthalmic surgery, side effects of LLA are uncommon, but they have been reported after subconjunctival injection or retrobulbar anaesthesia.^{1–3} Inadvertent intravascular injection, leading to platelet–fibrin emboli or vasoconstriction by epinephrine, resulting in occlusion of the lacrimal artery, is also reported.^{3,4} Among the amide anaesthetics, lidocaine shows a higher toxicity to the central nervous and cardiovascular systems; prilocaine produces less vasodilatation and toxicity but causes methaemoglobin formation.¹ In the case of these two women, there was no evidence of peripheral vascular disease or diabetes, but