Sir,

Coats' disease and Duchenne muscular dystrophy

Coats' disease of the retina and the association with muscular dystrophy was first described by Small in 1968.[1] Coats' disease was first described by George Coats in 1908.[2] It is a non-inherited retinal vasculopathy affecting males 3 times more commonly than females and it presents in the paediatric age group. Most cases are unilateral but bilateral cases have been reported.[1] Duchenne muscular dystrophy is an X-linked recessive disorder characterised by a deficiency of dystrophin. Dystrophin is a protein that maintains calcium haemostasis and deficiency leads to progressive necrosis of all striated muscles except the extraocular muscles.[3]

Although there is an association between the muscular dystrophies and Coats' disease, no genetic link has been determined.

Case report

An 8-year-old Caucasian male was referred by his babysitter who is an optician. The optician had noticed a left leukocoria 2 months prior to referral. Seven months prior to presentation the patient had been noted to have a Snellen visual acuity of 6/6 in both eyes at a school eye examination.

At presentation visual acuity was 6/5 in the right eye and there was inaccurate light projection in the left eye. Examination of the left eye revealed an exotropia, a normal anterior segment, leukocoria and a subtotal exudative retinal detachment (Fig. 1). Oral fluorescein angiography demonstrated aneurysmal dilation, capillary dropout, vessel leakage, lipid deposition and a subneural retinal exudate with detachment. Fluorescein angiography of the right eye was normal. Ultrasonography and computed tomography of the left globe confirmed these findings and ruled out a mass lesion.

The patient had been diagnosed with Duchenne muscular dystrophy at the age of 3 years and had mild lower limb weakness. No other family members were affected with either Coats' disease or Duchenne muscular dystrophy. Audiometry was within normal limits.

Comment

Subclinical retinal vasculopathy, retinal telangiectasia, pigmentary retinopathy and Coats' disease have all been described in association with different types of muscular dystrophy.[1,4,5] The clinical features seen in our patient are classical features of Coats' disease. It was unusual, however, not to have observed iris neovascularisation at the time of presentation. The patient did become blind in that eye 2 months after presentation, and subsequently developed a painful, rubeotic glaucoma 6 months later.

Coats' disease has been described in association with a myriad of diseases, including retinitis pigmentosa, muscular dystrophy, deafness, mental retardation, Turner's syndrome and the Senior–Loken syndrome.[1,6] There is no definite genetic link described between these. A deficiency of norrin, a protein product of the NDP gene (X-linked), has been described in the retinal tissue of eyes affected with Coats' disease.[7] It has been postulated that this protein, whose tertiary structure is similar to transforming growth factor β,[8] may be involved in the previously described non-inherited Coats' disease. Norrin is involved in the terminal differentiation of the inner retinal cell layers, in antiproliferative cellular interactions in the vitreous and in retinal vasculogenesis.[9] The exact role in Coats' disease has not been ascertained.

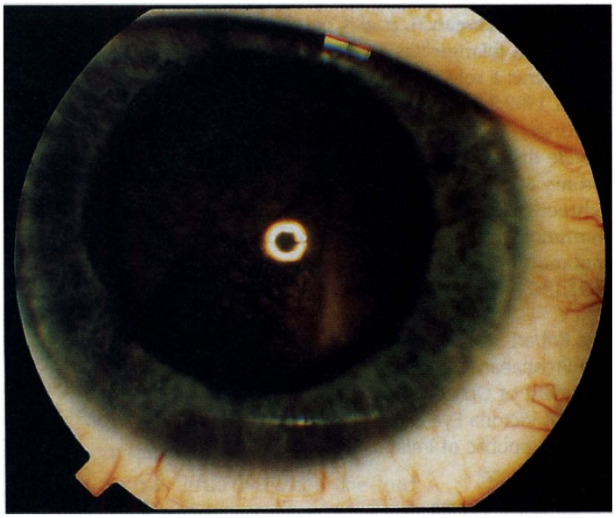

Fig. 1. *Exudative retinal detachment in Coats' disease.*

Facioscapulohumeral muscular dystrophy is the type most frequently described in association with Coats' disease, whether inherited or sporadic. This is the first reported case of a sporadic Duchenne muscular dystrophy patient presenting with a non-rubeotic, subtotal exudative retinal detachment secondary to Coats' disease.

The association between Coats' disease and any of the muscular dystrophies has not been determined to have a genetic link. A larger series of patients needs to be reviewed to determine the prevalence of both sporadic and familial cases.

References

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Errata

Ng SGJ et al. Necrotising orbital cellulitis. Eye 2001;15:173–7.

The name of the third author of this paper is C.P. Subudhi.

Curi ALL et al. Retinal toxicity due to Efavirenz. Eye 2001;15:246–8.

In the final paragraph of this letter it should state that the other drugs being used by the patient were AZT (Zidovudine) and 3TC, *not* azathioprine and 3TC.