body types and traces of liquids in the form of numerous drops and vacuoles (Figure 1b). The brown spherules are erythrocytes that have been altered and clumped by the action of the petroleum.[4] There are several reports of foreign body giant cell reaction to insoluble tetracycline preparations and tetracycline powder in petroleum-based carrier vehicles, mostly in the otolaryngology literature.[5,6] There are few reports of ophthalmic manifestations. The lesions must be removed as extensively as possible as they can recur, requiring repeat procedures as seen in two of our patients.

Swelling of the eyelid during lacrimal irrigation indicates trauma to the canalicular mucosa with subcutaneous infiltration. Probing should be halted immediately. If the irrigating solution is saline or fluorescein, the problem resolves within 24 h.[7] At the end of probing, the presence of the probe in the nose should be confirmed by a second probe; metal-on-metal contact provides confirmation that the first probe is in the nose. Then irrigation with saline or fluorescein can be safely performed. Sometimes a thin membrane over the tip of the probe can fool the doctor into thinking it was truly metal-on-metal, and retrieval of irrigating fluid from the nose confirms a patent passage.

Similarly in dacryocystography, if a cannulation has been traumatic and a false passage is suspected, the use of oil-based agents should be avoided. Lipiodol is the most commonly used contrast agent for this procedure. It is an iodised oil-based contrast agent. It too causes lipogranuloma formation if it extravasates into tissues via a false passage.[8]

Other case reports highlight equally disastrous consequences of irrigating other solutions through false passages. One report described permanent visual loss and optic atrophy following irrigation of the lower canaliculus with 20% chloramphenicol. A false passage had been created allowing the solution to infiltrate orbital tissues. The oedema that ensued caused a central retinal artery occlusion.[9] A further report described occlusion amblyopia in an infant caused by complete occlusion of an eye for 1 month from persistent periorbital swelling after methylene blue infiltrated the periorbital tissues inadvertently through a false passage during probing.[10]

Excellent results can be obtained in treating infants with lacrimal system obstruction without routinely irrigating antibiotic or steroid ointments through the lacrimal system. Most antibiotic/steroid solutions that are manufactured for topical ophthalmic use are specifically contraindicated for use intramuscularly or intradermally. Petrolatum-based substances should not be used after lacrimal irrigation because disfiguring scars, permanent lacrimal drainage problems, and repeat surgical interventions can be the unwanted result.

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S Fenton[1], MR Canninga-van Dijk[2] and MPh Mourits[1]

[1]Oribital Unit, Department of Ophthalmology
University Medical Centre, PO Box 85500
3508 GA, Utrecht, The Netherlands

[2]Department of Pathology
University Medical Centre, PO Box 85500
3508 GA Utrecht, The Netherlands

Correspondence: Dr MPh Mourits
Tel: +31-30-2567-888
Fax: +31-30-2505-417
E-mail: m.p.mourits@oogh.azu.nl

Sir,

The diagnosis of autosomal dominant late-onset retinal degeneration in two sisters

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Late-onset retinal degeneration (L-ORD) is an autosomal dominant disorder characterised by onset of night vision problems in midlife and progressive loss of peripheral visual field and central vision over ensuing decades.[1–4] The late onset of symptoms mimics other more common

eye disorders in the elderly in this condition and can lead to misdiagnosis. We report cases of sisters with L-ORD, who were initially diagnosed as age-related macular degeneration and normal tension glaucoma.

Case reports

Case 1 A 63-year-old woman presented in 1986 with floaters in her left eye. Her best corrected visual acuity was 6/9 N5 in both eyes. Her father was blind in later life. A posterior vitreous detachment was diagnosed, but she was noted to have cupping of the right optic disc despite normal intraocular pressures. After 2 years, she had developed bilateral visual field defects. Despite ocular hypotensive treatment, her visual fields continued to deteriorate and she had bilateral trabeculectomies.

Her visual acuity then started to deteriorate and pigmentary changes at the macula were noted. This progressed to atrophy with widespread peripheral retinal degenerative changes in both eyes (Figure 1a). An electroretinogram showed no measurable scotopic or photopic responses and led to the diagnosis of L-ORD. Her vision is now hand movements in both eyes with very restricted visual fields.

Case 2 A 78-year-old woman was referred in 1997 with a gradual deterioration in her vision. On examination, her visual acuity was hand movements in both eyes. Fundal examination showed atrophic maculae. A diagnosis of age-related macular degeneration was made and the patient was registered blind.

She returned to the clinic 1 year later with worsening sight. Her visual acuities remained at hand movements but she had reduced visual fields. On examination, the peripheral retina showed widespread pigmentary changes with chorioretinal atrophy (Figure 1b). She reported that her sister (case 1) had recently been registered blind and her father and grandfather had poor sight in later life. A diagnosis of L-ORD was made on this clinical picture. An electroretinogram showed unmeasureable responses.

Comment

The late onset of this disease is the defining feature of L-ORD. Initially, there can be no ophthalmoscopic abnormalities, which is unusual for most inherited dystrophies. The earliest symptoms occur in the sixth decade and after, with problems with dark adaptation[1] although formal dark adaptation testing can show abnormalities a decade before symptoms.[2] The first signs can be clusters of fine yellow–white dots in the mid-periphery.[1,2] The disease then progresses to loss of central and peripheral vision with peripheral pigmentary retinopathy and chorioretinal atrophy. The macula usually becomes atrophic but occasionally there is a disciform scar. The optic disc becomes pale and this coupled with the visual field abnormalities may lead to the diagnosis of normal tension glaucoma. The standard ERG may be normal in the early stages. At the end stage of the disease, only a reduced amplitude, delayed cone ERG is detectable.[1] There is some evidence that the early dark adaptation abnormalities are partially responsive to vitamin A therapy[2] but how this affects the poor

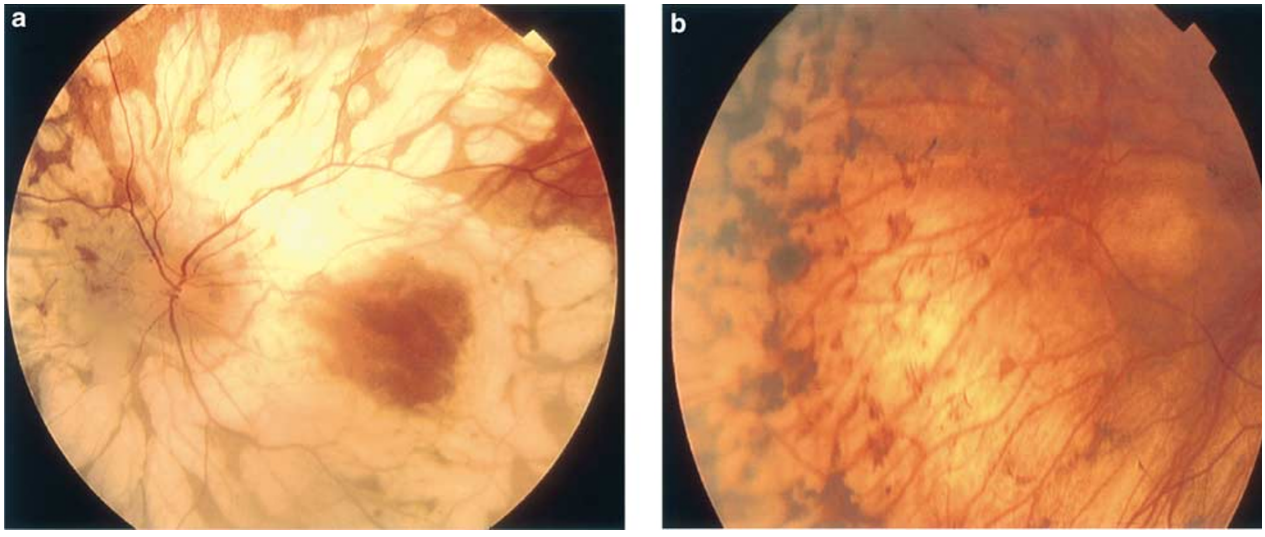

Figure 1 (a) Fundus appearance of case 1. (b) Peripheral retinal appearance of case 2.

long-term prognosis is still unknown. The diagnosis of L-ORD is either based on the above clinical picture or on the pathological appearance of a thick sub-RPE deposit throughout the retina[1,3,4] like that found in Sorsby's fundus dystrophy[6] and sometimes in age-related macular degeneration.[7] It is postulated that this deposit may act as a barrier to nutrient transport to the retina. In the later stages, there is widespread loss of RPE and photoreceptors.[4]

In the early stages, the disease can mimic other ophthalmic conditions as in our two cases. Our first patient was seen relatively early in the disease course but the diagnosis was not made in the patient or her sister until the peripheral retinal changes were present. Asking about night vision problems, family history and careful examination of the fundus including peripheral retina can be helpful in detecting this unusual disease. Our patients' diagnosis was further confirmed when they were found to be distantly related members of a fully investigated L-ORD family.[2]

These cases are important to raise awareness of L-ORD in the ophthalmic community. We often try to reassure our patients with ARM that they will not lose their peripheral vision. This rare subgroup of patients does lose this vital visual function with devastating visual results.

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CJ Styles[1], B Dhillon[1] and AF Wright[2]

[1]Princess Alexandra Eye Pavilion
Chalmers Street, Edinburgh, EH3 9YW, UK

[2]Cell and Molecular Genetics Section
MRC Human Genetics Unit
Western General Hospital, Crewe Road
Edinburgh, EH4 2XU, UK

Correspondence: CJ Styles
Tel: +44 131 536 1674
Fax: +44 131 536 1574
E-mail: meadow.house@virgin.net

Sir,

Ocular coloboma and radial aplasia: syndrome or association

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Ocular colobomata are relatively common malformations and are often associated with microphthalmos. A typical coloboma is located in the inferonasal quadrant, and may affect any part of the globe traversed by the fissure from the iris to the optic nerve. The majority of cases are isolated defects; however, sometimes they may be seen with other defects. Patients with multiple malformations and coloboma often present diagnostic and therapeutic challenges especially in infants.

Case report

The affected proband is a female born at 41 weeks gestations. The delivery was by emergency caesarean section because of meconium stained liquor and prolonged rupture of membranes. Apgar scores were 8 at 1 min and 10 at 5 min. The birth weight was 2420 g (below third centile) and head circumference was 33 cm (third centile). The baby was admitted to the neonatal unit because of respiratory grunting, and received oxygen for 24 h and 3 days of intravenous antibiotics. Clinical examination revealed a congenital right upper limb abnormality with an absent thumb.

Radiography of the right upper limb showed absence of the radius and thumb with only four metacarpals (Figure 1). Chest X-ray showed normal mediastinum and