association with neurofibromatosis. Surgical correction involved a combination of superior rectus weakening with inferior oblique weakening and medial and lateral rectus tendon splitting procedures, as described by Hummelsheim.[10] All cases reported success. Cooper and Greenspan[3] were the first to use full horizontal rectus tendon transposition inferiorly for this indication, as decribed by Knapp,[11] with good outcome. Similar success was more recently reported by Taylor *et al.*[12] and Munoz.[13] Isolated superior rectus recession has been advocated by Vesly[14] in 1965 and by Ingham *et al.*[4] in 1986.

In a review of 21 patients, over 16 years, with isolated inferior rectus paralysis, without mechanical restriction of the globe, von Noorden and Hansell[15] found that 11 were of congenital origin. No patients were reported to have neurofibromatosis. Anomalous head posture or a disfiguring hypertropia were common presenting symptoms and surgery consisted of resection of the paralysed muscle, combined with or without recession of its antagonist and/or recession of the contralateral superior oblique.

We used isolated superior rectus recession to treat our patient, with good results. To our knowledge this is the first report of a patient associating neurofibromatosis with an absent rectus muscle, and the second linking the condition with absent extraocular muscle. We would be interested to hear of other similar cases and the surgical approach to management.

References

1. Helveston EM, Giangiacomo JG, Ellis FD. Congenital absence of the superior oblique tendon. Trans Am Ophthalmol Soc 1981;79:123–33.
2. Congenital absence of the superior oblique tendon in craniofacial dysostosis. Ophthalmic Surg 1985;16:375–7.
3. Cooper EL, Greenspan JA. Congenital absence of the inferior rectus muscle. Arch Ophthalmol 1971;86:451–4.
4. Ingham PN, McGovern ST, Crompton JL. Congenital absence of the inferior rectus muscle. Aust N Z J Ophthalmol 1986;14:355–8.
5. Lin PY, Yen MY. Congenital absence of bilateral inferior rectus muscles: a case report. J Pediatr Ophthalmol Strabismus 1997;34:382–4.
6. Mather TR, Saunders RA. Congenital absence of the superior rectus muscle: a case report. J Pediatr Ophthalmol Strabismus 1987;24:291–5.
7. Newman RM, Cogen MS. Congenital absence of the superior oblique tendon in a patient with neurofibromatosis. J Pediatr Ophthalmol Strabismus 1997;34:192–4.
8. Diamond GR, Katowitz JA, Whitaker LA. Variations in extraocular muscle number and structure in craniofacial dysostosis. Am J Ophthalmol 1980;90:416–8.
9. Kaiser-Kupfer MI. Ophthalmic manifestations. In: Mulvihill JJ, moderator. Neurofibromatosis 1 (Recklinghausen disease) and neurofibromatosis 2 (bilateral acoustic neurofibromatosis): an update. Ann Intern Med 1990;113:39–52.
10. Hummelsheim E. Weitere Erfahrungen mit partieller Sehnuberpflanzung an dem Augenmuskulen. Arch Augenheilkd 1908;62:71–4.
11. Knapp P. The surgical treatment of double elevator paralysis. Trans Am Ophthalmol Soc 1969;67:304–23.
12. Taylor RH, Kraft SP. Aplasia of the inferior rectus muscle. Ophthalmology 1997;104:415–8.
13. Munoz M. Congenital absence of the inferior rectus muscle. Am J Ophthalmol 1996;121:327–9.
14. Vesley L. Aplasia of the inferior rectus oculi. Ceskoslovenska Oftalmologie 1965;21:477–8.
15. von Noorden GK, Hansell R. Clinical characteristics and treatment of isolated inferior rectus paralysis. Ophthalmology 1991;98:253–7.

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Sir,

Optical coherence tomography and multifocal electroretinography of X-linked juvenile retinoschisis

X-linked retinoschisis (XLR) is a relatively rare bilateral vitreoretinal dystrophy which arises from mutations in the XLRS1 gene[1] and results in visual loss in young men. We report the case of a 35-year-old man who presented with XLR and underwent functional and anatomical imaging using conventional techniques as well as multifocal electroretinography (mfERG) and optical coherence tomography (OCT).

Case report

A 35-year-old man was diagnosed as having retinoschisis 20 years ago. One of his two brothers, a maternal uncle and his maternal grandfather have also been diagnosed with this condition. He had a visual acuity of 6/60 in the right eye and 6/36 in the left eye.

Functional information was obtained from conventional electroretinography and from mfERG using a custom-built system with a 61 hexagonal display digitally back-projected onto a polysilicon screen.[2] Anatomical images were obtained from fundus photography and optical coherence tomography.

Fundus photography revealed the typical stellate spoke-like pattern characteristic of this condition. Both eyes were affected; however, a larger area of affected macula was observed in the left eye.

OCT revealed splitting of the inner retinal layers in the macular areas of both eyes. This splitting was most marked in the region of the fovea (Fig. 1A, C). In other areas which appeared healthy on fundus photography, OCT revealed the presence of microcysts (Fig. 1B). A larger area of the left eye showed signs of damage. Condon *et al.*[3] observed occasional bridges of retinal

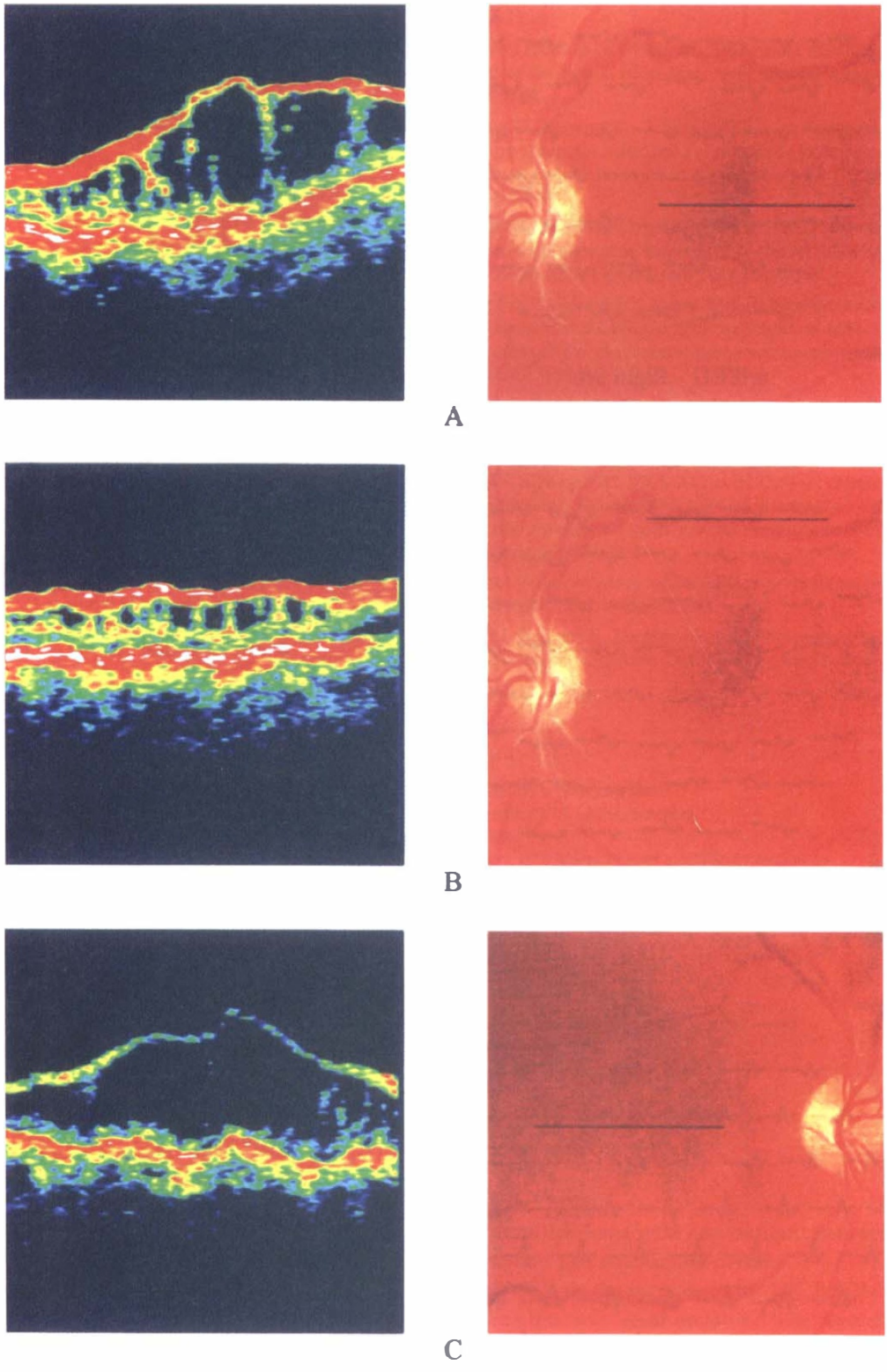
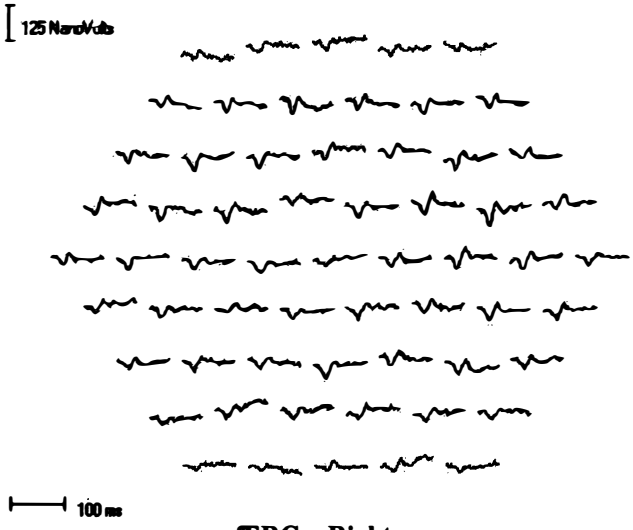
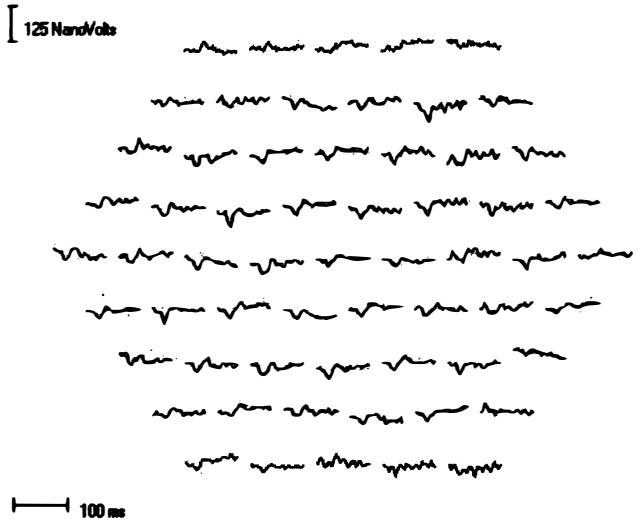


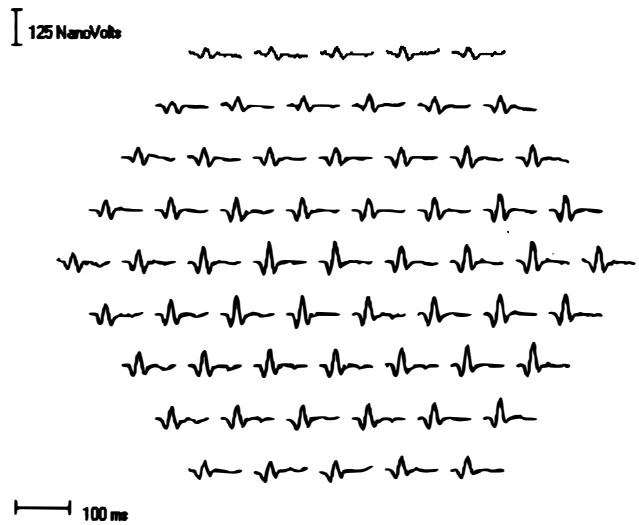
Fig. 1. *OCT images of XLR with corresponding fundus images to illustrate the position of the OCT scan. (A) Scan across the fovea, left eye; (B) microcysts in the left eye; (C) scan across the fovea, right eye.*



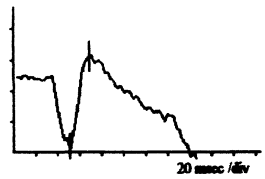
mfERG – Right eye



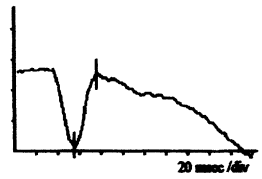
mfERG – Left eye



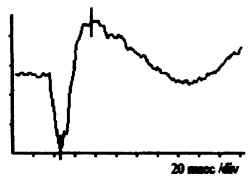
mfERG – Control



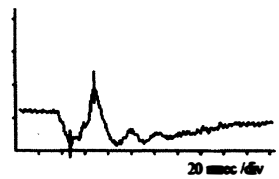
ERG maximal response – Right eye
***b-wave amplitude**: 257 μV; **implicit time**: 37 ms*



ERG maximal response – Left eye
***b-wave amplitude:** 192 μV; **implicit time:** 40 ms*

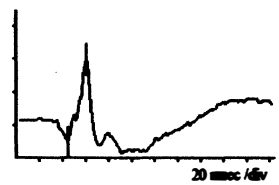


ERG maximal response – Control
***b-wave amplitude:** 241-709 μV; **implicit time:** 34-59ms*



ERG cone response – Right eye
***b-wave amplitude:** 71 μV; **implicit time:** 32 ms*

ERG cone response – Left eye
***b-wave amplitude:** 68 μV; **implicit time:** 34 ms*



ERG cone response – Control
***b-wave amplitude:** 68-222 μV; **implicit time:** 22-31ms*

Fig. 2. *Recordings obtained from mfERG and conventional ERG. Corresponding examples from control subjects are shown for comparison.*

tissue spanning the schisis gap. These can clearly be seen in the OCT images. The fact that there are fewer of these strands in the right eye and that the inner layer of the schisis cavity is much thinner may indicate a more advanced stage of the condition and could explain the slightly lower visual acuity in this eye despite the smaller affected area.

Results from the conventional ERG are shown in Fig. 2. The b/a ratios were 1.4 for the right eye and 1.0 for the left (normal range: 1.3–2.9). The maximal b-wave response was clearly reduced in the left eye but just within the normal limits for the right. Cone b-wave responses fell just within the normal limits for both eyes. Implicit times were within the normal range for the maximal responses but slightly delayed in the cone response.

The mfERG showed a marked reduction in the P1 component of the mfERG waveforms in a number of areas in both eyes (Fig. 2). This reduction was most marked for the central hexagons and a larger area of abnormal function was observed in the left eye. P1 amplitudes of the central response for right and left eyes were 41 and 44 nV respectively (normal range: 80–190 nV); P1 latencies were 50 ms and 49 ms respectively (normal range: 38–41 ms); and P1/N1 ratios were 1.5 and 1.6 respectively (normal range: 2.3–2.8).

Comment

The reduced maximal b-wave amplitude observed in the left eye of this patient is characteristic of patients with XLR;[4] however, there has also been a report of a patient known to have XLR but still retaining a normal scotopic b-wave,[5] as seen in the right eye of our patient. The conventional ERG is a measure of the response from the whole retina and in this case did not show an abnormal response in the right eye. The mfERG is capable of eliciting responses from localised areas of the retina and clearly demonstrated that there was abnormal retinal function in a number of macular areas in the right eye despite the normal result from conventional ERG. Thus the mfERG is a better tool for demonstrating the extent of this condition and may be very useful in diagnosing cases of XLR. There have been no publications on mfERG in XLR, and thus further work on a larger cohort of patients is required to establish the characteristics of mfERG recordings from this group of patients. Our findings from OCT are similar to those reported by other groups.[6,7]

To date there have been very few pathological studies on XLR,[8,9] since this condition rarely results in enucleation. Although the resolution currently achievable on OCT scans is not as high as that from microscope images, OCT nevertheless sheds light on the anatomical features of XLR and gives information which cannot be obtained from fundus photography.

References

1. Inoue Y, Yamamoto S, Okada M, *et al*. X-linked retinoschisis with point mutation in the XLRS1 gene. Arch Ophthalmol 2000;118:93–6.
2. Parks SW, Keating D, Evans AL. Wide field functional imaging of the retina. IEEE Med Appl Signal Processing 1999;107:91–6.
3. Condon GP, Brownstein S, Wang NG, *et al*. Congenital hereditary (juvenile X-linked) retinoschisis: histopathological and ultrastructural findings in three eyes. Arch Ophthalmol 1986;104:576–83.
4. Peachey NS, Fishman GA, Derlacki DJ, *et al*. Psychophysical and electroretinographic findings in X-linked juvenile retinoschisis. Arch Ophthalmol 1987;105:513–6.
5. Sieving PA, Bingham EL, Kemp J, *et al*. Juvenile X-linked retinoschisis from XLRS1 Arg213Trp mutation with preservation of the electroretinogram scotopic b-wave. Am J Ophthalmol 1999;128:179–84.
6. Azzolini C, Pierrot M, Codenotti M, Brancato R. OCT images and surgery of juvenile macular retinoschisis. Eur J Ophthalmol 1997;7:196–200.
7. Stanga PE, Chong NH, Reck AC, *et al*. Optical coherence tomography and electrophysiology in X-linked juvenile retinoschisis associated with a novel mutation in the XLRS1 gene. Retina 2001;21:78–80.
8. Manschot WA. Pathology of hereditary juvenile retinoschisis. Arch Ophthalmolol 1972;88:131–8.
9. Yanoff M, Kertesz E, Zimmerman LE. Histopathology of juvenile retinoschisis. Arch Ophthalmol 1968;79:49–53.

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Sir,

Bilateral papilloedema with concomitant neuroretinitis in a 7-year-old girl with Lyme disease

Lyme disease is on the increase in the UK. It is becoming regarded as the new 'great imitator'.[1] The diversity of ophthalmic manifestations combined with the need for prompt identification and treatment makes an increased awareness amongst ophthalmologists essential. We describe a patient who presented with disc swelling apparently secondary to raised intracranial pressure. However, an ophthalmic assessment subsequently recognised features suggesting an additional neuroretinitis. This prompted further serological tests resulting in a diagnosis of Lyme disease.