LETTERS TO THE EDITOR

Retinopathy associated with pancreatitis in a child with maple syrup urine disease

EDITOR,—Retinopathy associated with pancreatitis is an uncommon condition first described in 1975.1 To date, fewer than 50 cases have been reported, all involving adults. We report a case of pancreatitis with retinopathy in a young child with maple syrup urine disease, a rare metabolic disorder.

CASE REPORT
A 7 year old Indian female presented with maple syrup urine disease (MSUD) diagnosed in infancy. She has been maintained on a special diet since then, enjoying normal development. However, during December 1995 she was admitted to the hospital for management of acute gastroenteritis and dehydration. Laboratory studies included serum amylase and lipase which were within normal limits. She received hyperalimentation with glucose and intralipids through a femoral vein. On the third hospital day she was noted to have mental status changes which improved with hydration. However, on her sixth hospital day her mental status deteriorated again. Her abdomen became diffusely tender. Laboratory studies revealed serum amylase 453 U/l (normal limits 40–128), calcium 8.2 mg/dl, phosphorus 2.2 mg/dl, albumin 2.6 g/dl, total protein 5.1 g/dl, and a white cell count of 12.4 × 10^6/l. The serum levels of leucine were 12.9 mg/dl (normal limits 1.0–5.2). During a neurological examination white lesions were noted in both fundi. In the next 48 hours, the patient’s general status improved. Her serum amylase levels declined while lipase levels increased to reach a peak of 2485 U/l (normal limits 23–208).

An ophthalmic evaluation 2 days after the initial elevation in serum amylase revealed visual acuity of 20/50 in both eyes (Snellen E) both at distance and at near. No afferent pupillary defect was recorded. Ocular versions were intact. Slit lamp biomicroscopy was unremarkable. Dilated fundus examination revealed the presence of bilateral white fluffy lesions and areas of haemorrhage (Fig 1).

Findings were concentrated in the posterior poles only. Fluorescein angiography showed minimal capillary non-perfusion (Fig 2).

The patient slowly improved spontaneously. On follow up examination 1 month later, visual acuity was 20/25 in both eyes with no correction. Fundus examination was completely normal, with full resolution both of cotton wool spots and of haemorrhages.

COMMENT
Maple syrup urine disease is caused by a defect in branched chain ketoacid (BCKA) dehydrogenase.2 Ocular complications of untreated disease or late diagnosis include optic atrophy, nystagmus, ophthalmoplegia, strabismus, and cortical blindness.3

Pancreatitis is a known complication of branched chain organic acidemias (BCOA).

In a series of 108 paediatric patients with BCOA, nine cases of pancreatitis were found during a 10 year period. One of these cases was in a patient with MSUD.4 Retinopathy as a rare complication of acute pancreatitis was first described in 1975.5 It has been reported to occur either before6 or after the manifestation of acute pancreatitis.7,8 All reported cases to date have been in young adults (ages 25–40 years), often associated with a history of ethanol misuse. None of the previously reported cases had associated systemic metabolic diseases.

The aetiology of retinopathy of pancreatitis is debatable. An older theory, supported by some experimental data,9 holds that fat emboli found in many organs in cases of acute pancreatitis10,11 cause ischaemic retinal infarcts. Retinal fat emboli have also been found in a patient’s retinopathy,12 a condition remarkably similar to pancreatitis associated retinopathy.

Another theory proposes embolisation of retinal vessels by complement induced fibrin clots and leucocaggregates as the causative mechanism.13 However, experimental studies have failed to reproduce various features of the clinical picture of retinopathy of pancreatitis.14,15

The presence of lipaemia in our patient argues for the lipid embolism theory. Hypovolaemia and hypoxia did not occur and are thus unlikely to have been involved in the pathogenesis of the retinal picture. Although our patient maintained almost normal levels of leucine, isoleucine, and valine throughout the above episode, it is unclear whether the presence of MSUD predisposed her to the development of retinopathy of pancreatitis.

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8 Jacob HS, Goldstein IM, Shaprio I, et al. Sudden blindness in acute pancreatitis. Possible role of...
Immediate postoperative objective refraction as an indication to final refraction in phacoemulsification surgery

EDITOR,—The goal of cataract surgery is the rapid attainment of good visual acuity, ideally unaided for the intended focal distance and therefore an accurate intraocular lens (IOL) power calculation. Refractive prediction errors, if substantial, can be problematic for the patient. If the problem is one of symmetrical ametropia, spectacle correction is usually successful. Contact lens fitting may be necessary for those patients with anisometropia in whom spectacle wear would induce significant and intolerable astigmatism. IOL exchange as a secondary procedure may have to be considered in cases refractory to these measures.

In our study, we aimed to ascertain whether objective refraction at the end of surgery was feasible and if so, to determine how this refraction related to the final refraction at 4–7 weeks postoperatively with a view to suggesting guidelines for immediate exchange of implant in cases of gross refractive prediction errors.

CASE REPORT

Consecutive patients undergoing phacoemulsification cataract surgery after continuous curvilinear capsulorhexis with capsular bag implantation of Chiron C10UB injectable IOLs were refracted at the end of surgery on the table objectively with a streak retinoscope, and at 4–7 weeks postoperatively with a view to determine the best visual acuity. Viscoelastic used was Provvisc (sodium hyaluronate 1%). The immediate postoperative refraction results were not available to the ophthalmologist performing the final refractions. Patients with glaucoma, diabetic retinopathy, macular degeneration, peroperative, and/or postoperative complications and patients who failed to attain 6/9 or better Snellen visual acuity at final refraction were excluded.

Twenty six eyes of 26 patients were included (six males and 20 females, 16 right eyes and 10 left eyes). Ages ranged from 62 to 86 years with a mean of 78.9. Twenty four patients had topically administered anaesthesia and two had peribulbar injections followed by application of Honan balloon. IOL powers ranged from 16 D to 27 D with a median of 22.5 D.

Data were analysed using statistical software on a Macintosh computer.

We were unable to refract two patients owing to the presence of corneal epithelial haze. Figure 1 shows a few values clustering around the neutral—that is, “no change” line, and a single significant myopic change but the predominant feature is the shift towards hypermetropia. The single significant myopic change occurred in a patient who had topical anaesthesia.

The mean change in refraction was 1.11 D hypermetropia with a standard deviation of plus or minus 0.94 D. The range was from 1.63 D myopic change to 2.75 D hypermetropic change. The 95% confidence interval was 0.73 D, 1.48 D. Paired two t test performed on the immediate postoperative and final refraction results gave a p value of <0.001.

Figure 1 Scattergram of refractive change from immediate postoperative refraction to final refraction.

We would advise a fundal examination be performed to exclude any posterior pole lesions hitherto undetected.

In conclusion, objective refraction immediately following phacoemulsification surgery is feasible; if a gross refractive prediction error is found, immediate implant exchange may be considered, taking into account the mean hypermetropic shift of about 1 dioptre from immediate postoperative objective refraction to the final refraction in the postoperative period in cases where the Chiron C10UB lens has been implanted.

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Retinal haemorrhage caused by "ecstasy"

EDITOR,—“Ecstasy” (3,4-methylenedioxymethamphetamine (MDMA)) is an amphetamine derivative classified as a class A drug under the Misuse of Drugs Act 1971. It is a recreational drug and can be bought under a variety of names (XTC, Adam, E, yellow burger, etc). The drug is a white powder and comes in coloured tablets or capsules. Usual doses are from 30 to 150 mg.

A patient is described who developed retinal haemorrhage after taking one ecstasy capsule.

CASE REPORT

A 22 year old white woman took an ecstasy capsule while sitting down in a night club. After 30 minutes she felt the effects of the ecstasy. During this time she had sat at a table and had not exerted herself, vomited, danced,

Figure 1 Photograph of the left fundus showing a submitial limiting membrane haemorrhage at the centre of the macula. The haemorrhage has broken through the internal limiting membrane into the vitreous.
or performed any valsalva manoeuvre. She denied sudden loss of the central vision in her left eye and felt unwell. She then left the club. She presented at the casualty department 2 days later. On examination her right visual acuity was 6/9 and left visual acuity was 1/60. She had no relative afferent pupillary defect. The anterior segments were normal and intraocular pressure was 15 mm Hg in the right eye and 17 mm Hg in the left eye. The right fundus was normal. The left fundus revealed a 3 disc diameter subinternal limiting membrane haemorrhage at the centre of the macula (Fig 1). There were retrohyaloid haemorrhages inferior and superonasal to the macula. General examination including cardiovascular and neurological assessment revealed a 3 disc diameter subinternal limiting membrane haemorrhage without evidence of other vascular abnormalities (Fig 1). Fundus fluorescein angiography (FFA) showed a petaloid appearance typical of cystoid macular oedema (Fig 2). The latanoprost was stopped, and within 2 weeks the symptoms of visual blurring and the cystoid macular oedema had improved. The patient continued on betaxalol to the left eye but inadequate intraocular pressure control made further filtering surgery necessary.

COMMENT
This eye had undergone retinal detachment repairs including scleral buckling, cryotherapy, vitrectomy, and then phacoemulsification and lens implantation. There was a 3 year gap before latanoprost was prescribed. These procedures may each be associated with cystoid macular oedema and selective use of pars plana vitrectomy used in its treatment, but all procedures occurred over 2 years previously with 6/6 vision in the intervening period. While we cannot exclude pre-existing subclinical, angiographically positive cystoid macular oedema, the timing of onset and relief of clinical symptoms and signs with latanoprost administration was striking.

Hoyng et al administered latanoprost (0.006%) twice daily for a month to un-complicated pseudophakic patients and fluorescein angiography showed no retinal leak. Six aphakic cynomolgous monkeys given seven times the usual daily dose for 6 months failed to develop cystoid macular oedema. However, intravitreal injections of PGF-2α to pigmented rabbits showed a small but statistically significant leak by vitreous fluorophotometry. Animal work needs to be interpreted circumspectly, particularly regarding the rabbit which shows an atypical response to inflammation. These experimental results suggest that aphakic or pseudophakic priamtes do not normally develop cystoid macular oedema in response to prostaglandins.

Miyake and colleagues’ work on rabbits and baboons confirmed that aphakia and pseudophakia may be associated with impaired removal of prostaglandins by the ciliary processes (Bito’s pump), with resulting accumulation in the eye, since prostaglandins are not broken down intraocularly. Possibly Bito’s pump was impaired in our patient, further raising prostaglandin levels and increasing the concentration gradient.

This patient had already had a peripheral anterior vitrectomy during the second left retinal detachment repair, and in such a case it probably would facilitate diffusion of prostaglandins posteriorly.

Furthermore, there were extensive and repeated vitrectinal procedures performed before administration of the medication and this was a complex case of posterior chamber pseudophakia. This case confirms the

1 Dowling GP, McDonough ET, Bost RO. ‘Eve’ and ‘ecstasy’ a report of five deaths associated with the use of MDEA and MDMA. JAMA 1997;278:1615-7.

Figure 1 Left fundus photograph shows supernasal peripapillary haemorrhage and cystoid macular oedema.

Figure 2 Left fundus fluorescein angiogram shows cystoid macular oedema.
Factor V Leiden mutation in association with idiopathic intracranial hypertension

EDITOR—Idiopathic intracranial hypertension has an association with prothrombotic conditions. The recently described thrombophilic defect of activated protein C (APC) resistance, caused by the factor V Leiden mutation, has been postulated to be a factor in some cases of our knowledge, we describe the first two cases reported of the factor V Leiden mutation identified in association with idiopathic intracranial hypertension. Both patients had been previously well and thus should have a past history to suggest a clotting disorder.

CASE REPORTS

Case 1

A 31-year-old obese white female was referred by her general practitioner complaining of transient visual loss in her left eye for the previous 5 months and non-specific headaches. She had started on warfarin 2 years previously because of the factor V Leiden mutation identified. She was referred to the haematology department for consideration of anticoagulation, bearing in mind the increased bleeding risk and thrombogenicity of warfarin. First degree blood relatives of the patient should be warned to avoid smoking and to seek medical advice before taking oral contraceptives or accepting elective surgery: the factor V Leiden mutation is of dominant inheritance.

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COMMENT

Prothrombotic abnormalities have recently been implicated in the pathogenesis of “benign intracranial hypertension” (BIH). It has been suggested that CSF reabsorption due to damaged arachnoid villi, secondary to microthrombus formation, could be the cause of raised intracranial pressure. Arachnoid villi dysfunction causing BIH has also been postulated in SLE and may be the result of venulitis, aseptic meningitis, or immune complex deposition and, in the antiphospholipid syndrome, possibly due to microocclusion. Such mechanisms affecting the optic nerves in addition could cause the poor outcome in case 1.

APC resistance is gaining greater recognition in the pathogenesis of ophthalmic disorders and has been implicated as a cause of central retinal vein occlusion in patients younger than 50 years. The factor V Leiden mutation may be heterozygous or homozygous and carries a lifelong increased thrombotic risk. In the homozygous, and 50–100 fold respectively. As 5% of the population has the factor V Leiden mutation, it is felt that most individuals carrying the defect will never experience a thrombotic event unless they carry another genetic defect, such as a deficiency of protein C or C4 or are exposed to a precipitating factor such as oral contraceptives, pregnancy, or surgery. Currently the criteria for anticoagulation in idiopathic intracranial hypertension are unknown owing to the absence of controlled clinical trials. It is reasonable for haematologists to offer warfarin as a short course or continuous therapy to symptomatic patients, especially if the event is severe, and in those exposed to other risk factors who have a familial thrombotic tendency. Without further studies addressing the association of idiopathic intracranial hypertension and thrombophilia, routine testing for the factor V Leiden mutation and other thrombophilic factors cannot be recommended in patients with idiopathic intracranial hypertension outside a clinical trial basis. However, if the mutation is found, referral should be made to a haematologist for advice concerning anticoagulation, bearing in mind the increased bleeding risk and thrombogenicity of warfarin. First degree blood relatives of the patient should be warned to avoid smoking and to seek medical advice before taking oral contraceptives or accepting elective surgery: the factor V Leiden mutation is of dominant inheritance.
reactivations of their retinitis despite having no specific anti-CMV therapy. However, the factors underlying this improved immunity to CMV are not entirely clear. We report the case of a patient on highly active antiretroviral therapy (HAART) who has had no reactivation of CMV retinitis after 6 months without anti-CMV treatment. The unusual feature of this case is that the CD4 count has remained persistently low.

We discuss factors that may be relevant in the improved immune response to CMV infection and may be useful in isolating a group of patients on HAART who do not require lifelong maintenance with anti-CMV therapy.

CASE REPORT

A 33 year old man with a CD4 count of 27 cells \( \times 10^3/\text{L} \) who had been diagnosed with AIDS in 1992 developed CMV retinitis in zones 1, 2, and 3 of the left eye in March 1996. Treatment was commenced with intravenous ganciclovir 10 mg/kg and maintenance therapy consisted of oral ganciclovir. After 3 months reactivation and progression of the retinitis necessitated a further induction course of intravenous ganciclovir with subsequent maintenance therapy with daily intravenous foscarin.

Further reactivation led to the use of intravitreal ganciclovir and further re-induction course of intravenous foscarin. As a result of difficulties with intravenous line sepsis intravenous cidofovir was commenced. In March 1997 the patient was started on HAART including protease inhibitor but despite this the CD4 count remained low at 22 cells \( \times 10^3/\text{L} \).

The patient decided to discontinue anti-CMV therapy in April 1997 and has undergone frequent ophthalmological review since then. He has received no anti-CMV therapy for 6 months and despite a continuing low CD4 count of 28 cells \( \times 10^3/\text{L} \) and a HIV-1 viral load of 201 637 RNA copies/ml there has been no further reactivation of CMV retinitis.

COMMENT

Before the use of HAART, maintenance therapy with anti-CMV medication was required in all patients with CMV retinitis. Without therapy the average time to progression of CMV retinitis was about 2–3 weeks.

Several cases have been reported which describe lack of progression of CMV retinitis in patients treated with HAART and in each of these cases there was an associated rise in CD4 counts. There have also been reports of newly diagnosed CMV retinitis after commencement of HAART with improving CD4 counts, although considerable doubt has been shed on whether these were in fact previously undetected CMV retinitis and not just reactivation of previously undetected disease. In this case the CD4 count remains consistently low and the HIV viral load high despite HAART.

This suggests that the immune response to CMV is not solely related to the CD4 count and that other factors are also involved in the recovery of immunity to CMV following treatment with HAART.

Bowen et al have demonstrated by quantitative and qualitative measurements of CMV DNA by polymerase chain reaction based assays that CMV viral load may be an indicator of patients at increased risk of reactivation of CMV retinitis. They suggest that this measurement can help with clinical management. It has also been shown that the detection of the early CMV antigen (p65 antigen) suggests elevated susceptibility to CMV infection.

Transforming growth factor-ß (TGF-ß) inhibits the IL-2 induced proliferation of T lymphocytes, and expression of TGF-ß may be increased in CMV infection. Measurement of TGF-ß and other cytokines may also be useful in the monitoring of patients at risk of CMV disease.

By performing a controlled prospective trial involving measurement of CMV and HIV viral load, together with these immunological markers it may be possible to identify a subgroup of patients on HAART who do not require long term anti-CMV therapy.

However, early reports from Martin et al on the results of their study of the use of combined oral ganciclovir and intravitreal implant for treatment of CMV retinitis suggest that there is still a survival benefit from continued use of oral ganciclovir in the HAART environment.

This once again raises the debate over whether CMV is just an opportunist taking advantage of immunosuppression or is it a cofactor acting in partnership with HIV.

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REFERENCES


Spontaneous and sustained resolution of CMV retinitis in patients receiving highly active antiretroviral therapy

Environ.—Cytomegalovirus (CMV) retinitis predominantly occurs in severely immunocompromised patients with CD4+ cell counts below 50 cells \( \times 10^3/\text{L} \). Before the use of combinations of protease inhibitors and other antiretroviral agents, CMV retinitis invariably progressed in the absence of specific anti-CMV therapy. This highly active antiretroviral therapy (HAART) can decrease human immunodeficiency virus (HIV) load and increase CD4 T cell counts in patients with AIDS. In this study we describe two patients receiving HAART, who present with probable inactive CMV retinitis although they never received anticytomegaloviral therapy. In one of these patients, CMV retinitis recurred shortly after his CD4+ cell counts fell below 50 cells \( \times 10^3/\text{L} \) despite continued combination antiretroviral therapy.

CASE REPORTS

Patient no 1

A 28 year old man with AIDS and a history of one positive blood culture for CMV and a CD4+ cell count of 13 cells \( \times 10^3/\text{L} \) was placed on ritonavir, zidovudine, and lamivudine in April 1996. Within 2 months the CD4+ cell count increased to 65 cells \( \times 10^3/\text{L} \) and the patient developed blurred vision and floaters in his left eye. Examination of the left eye revealed mild vitritis and a large area of retinal atrophy with several small haemorrhages at the border, consistent with inactive CMV retinitis (Fig 1). The right eye was normal. He was placed on no anti-CMV therapy. The haemorrhages resolved and no new progression of the retinitis was observed.
COMMENT

We describe two patients who presented with mild uveitis and an area of retinal atrophy consistent with inactive CMV retinitis despite never having received specific anti-CMV therapy. Both patients had prior CD4+ cell counts in the natural history of AIDS. Both patients had prior CD4+ cell counts below 50 cells per μl. A 44-year-old white man with AIDS had a CD4+ cell count of 25 cells per μl in March 1996 and 1 month later the patient developed a temporal scotoma in his left eye. Examination of the left eye showed mild anterior uveitis, vitritis, and a large area of retinal atrophy in the nasal midperiphery consistent with inactive CMV retinitis (Fig 2). The area of inactive retinitis has remained stable over the past 12 months despite the fact that he has received no anti-CMV therapy. A CD4+ T-cell count in October 1996 was 111 cells per μl.

Successful penetrating keratoplasty in an infant after extended storage of infantile donor cornea

Euston.—Despite the fact that infantile corneas are such a rare and valuable material in our eye banks they are not always properly utilised.

Infantile corneas, because of their characteristics (steepness, flexibility, elasticity), are not preferred for transplantation in emmetropic adults if other tissue is available. On the other hand it has been suggested that for infants undergoing penetrating keratoplasty, donor and recipient age should be matched as closely as possible. Such age matching is highly not always possible in the existing corneal storage system.

According to the United Kingdom Transplant Support Services Authority (UKTSSA) statistics, in 1996 there were 15 recipients in the 0–5 age group. Forty-four infantile corneas were retrieved in the same period. Only five infantile recipients, however, received corneas from the same age group, while the other 10 received corneas from older donors, in some cases the age difference exceeded 50 years (Fig 1A). Of the available infantile corneas, apart from the five transplanted into infants, 17 were transplanted to recipients of various ages (11–40), and 22 were not used at all because they could not be allocated to suitable recipients within the required time (which for most eye banks is 4 weeks) or for other reasons such as inadequate endothelial cell density (Fig 1B).

CASE REPORT

Keratec Eye Bank recently obtained donor tissue from a child aged 13 months. After 22 days in culture one cornea was transplanted into a recipient aged 5 years. For the second cornea, however, no suitable recipient could be found, either through UKTSSA or Bio-Implant Services (BIS, Eurotransplant). Rather than discard the tissue after the standard 4 weeks in culture we placed it into 75 ml of fresh medium in the hope of extending the preservation period until a suitable recipient could be found.

Within a few weeks a male infant aged 3 months with bilateral Peters' anomaly (Fig 2a,b) was referred to St George's Hospital for penetrating keratoplasty. The left penetrating keratoplasty was carried out with the infantile cornea which, at that time, had been

maintained in culture for 68 days (over 9 weeks), the final endothelial assessment, by vital staining and light microscopy, having confirmed its suitability for use. The right eye was operated on 1 week later using a cornea from a 25 year old donor as no infantile cornea was available.

At 2 months postoperatively at examination under anaesthesia both corneal grafts were clear (Fig 2c,d). Examination under anaesthesia was repeated 1 year later. Again both grafts were clear, and specular microscopy of the graft endothelium revealed a cell density of 2235 cells/mm² in the graft from the infantile donor, and 1410 cells/mm² in the graft from the older donor (Fig 2e,f). Retinoscopy showed high myopia in both eyes, but both fundi appeared normal.

COMMENT

In most eye banks cultured corneas that are not transplanted within 4–5 weeks of retrieval are discarded. This is based on the finding that the rate of decline of endothelial cell density increases after 35 days of culture in a single aliquot of culture medium.1 Pels et al reported that storage of corneas in culture for 3–7 weeks induced a mean cell loss of about 11% while preservation for 9–17 weeks (medium changed after 6 weeks), resulted in a mean cell loss of about 43%. The actual cell loss, however, varied significantly among examined corneas and therefore it has been suggested that the suitability of an individual cornea for transplantation should be based on the quality of the endothelium during final assessment rather than the length of storage.1 It has also been suggested that the increased cell loss after 40 days of culture might be caused by the depletion of nutrients and accumulation of waste products in the culture medium. A larger volume of medium, or the renewal of the medium, may postpone or prevent this process. However, with prolonged organ culture endothelial survival is not the only factor in determining the suitability of tissue for transplantation. If there is epithelial overgrowth onto the posterior corneal surface this can potentially lead to problems of implantation of epithelial cells into the anterior chamber.

There has been a report of corneas safely preserved in culture for 7 weeks before transplantation,2 and we have found that an infantile cornea preserved for 9 weeks in organ culture can be transplanted successfully.3 There is usually no need to extend the storage time of adult corneas, but it may be appropriate to extend the preservation time of some infantile corneas in order to maximise the chance of their most appropriate utilisation, and further research is needed to optimise storage methods and evaluate the cost/benefit of longer storage times.

Choroidal melanoma in a patient with retinitis pigmentosa and Usher’s syndrome

EDITOR,—Usher’s syndrome is an autosomal recessive condition characterised by retinitis pigmentosa and hearing loss. It is the most common cause of combined blindness and deafness in the USA. Usher’s syndrome has two well defined subtypes including type I and type II.4 Type I has more severe and early onset findings.

CASE REPORT

A 39 year old woman with retinitis pigmentosa and type I Usher’s syndrome developed blurred vision in the left eye over a 4 month period. The best corrected visual acuity was 20/50 in the right eye and 20/80 in the left eye. Anterior segment examination and intraocular pressures (16 mm Hg) were normal bilaterally. Ophthalmoscopic examination showed mild optic disc pallor, marked vascular attenuation, and retinal pigmentary changes in both eyes. Inferotemporally in the left eye, there was a 15 × 10 mm dome-shaped choroidal melanoma with a secondary retinal detachment affecting the inferior half of the fundus (Fig 1). There was no evidence of retinal invasion or seeding. A-scan and B-scan ultrasonography demonstrated a 5.0 mm thick choroidal mass with low to medium internal reflectivity, acoustic hollowness, and a secondary retinal detachment (Fig 2). Goldmann perimetry showed marked constriction of visual fields with only 5 degrees of central field remaining in both eyes. Electroretinographic testing under photopic and scotopic conditions revealed that the b-wave was isoelectric.

The patient was treated with iodine-125 plaque radiotherapy with tumour apex dose of 9000 cGy, base dose of 43 000 cGy, and optic disc dose of 3700 cGy. The tumour showed a...
satisfactory response to radiation. At 10 months’ follow up, the subretinal fluid dried up completely but radiation papillopathy developed. The papillopathy resolved over 5 months, leaving more optic disc pallor. At 22 months’ follow up, the patient underwent cataract surgery with posterior chamber lens implantation in both eyes because of advanced posterior subcapsular cataract. At 40 months’ follow up, the vision was hand movements in the right eye and tumour thickness was 2.8 mm. There was no radiation retinopathy or metastatic disease.

COMMENT
Abnormalities involving chromosomes 1 (type II), 11, and 14 (type I) have been observed in patients with Usher’s syndrome. Reported chromosome alterations in uveal melanoma involve chromosomes 3, 6, 8, and 9; therefore, the simultaneous occurrence of these two conditions, as in our patient, is probably coincidental.

The development of retinal detachment in a patient with retinitis pigmentosa is rare because of adhesions between the retinal pigment and sensory epithelium. Only a few cases of retinitis pigmentosa with retinal detachment have been reported. Our patient illustrates a non-rhegmatogenous retinal detachment with retinitis pigmentosa.

Our patient developed radiation papillopathy with an optic disc dose less than 5000 cGy, which is unusual. The atrophy/gliosis of the optic disc in retinitis pigmentosa might increase the susceptibility of the disc to irradiation. On the other hand, she did not develop radiation retinopathy despite a high dose of radiation to the retina. Perhaps the atrophic retina in retinitis pigmentosa was unable to elicit a radiation response.

Choroidal melanoma can rarely lead to a pseudoretinitis pigmentosa picture secondary to retinal invasion and dispersion of melanoma cells on the retina. Retinal invasion was not present in our patient and retinal pigmentary changes were noted in both eyes, ruling out pseudoretinitis pigmentosa. Retinitis pigmentosa can be associated with several fundus tumours including giant drusen resembling astrocytoma and acquired vasoproliferative tumours. To our knowledge, this is the first report of choroidal melanoma in a patient with retinitis pigmentosa.

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