LETTERS TO THE EDITOR

Xeroderma pigmentosum in three consecutive siblings of a Nigerian family: observations on oculocutaneous manifestations in black African children

EDITOR,—Xeroderma pigmentosum (XP), a rare autosomal recessive disorder characterised by defective DNA repair leading to clinical and cellular hypersensitivity to ultraviolet radiation, manifesting mainly as intolerance of skin and eyes to light, has been described in all races, but is exceedingly rare in the negroid race, although some cases have been reported in both the American and African black people. We describe three consecutive siblings of a Nigerian, Fulani, family with the typical features of XP. We wish to draw attention to the clinical, phenotypic variations of this syndrome in black children of the same family living together in an area of high sunshine, and the difficulties in the management of XP patients with advanced disease and limited access to facilities in an environment where avoidance of skin exposure to intense ultraviolet rays is problematic. We believe the patients we have described constitute the first series on XP in black children in the west African subregion.

CASE REPORTS

Case 1
The proband, a 9 year old girl, was first seen at Usmanu Danfodiyo University Teaching Hospital (UDUTH), Sokoto (13.02° N, 8.14° E), Nigeria, in February 1999 with a history of the development of generalised erythema of the skin of the limbs, face, and trunk from the age of 1 week, on exposure to sunshine, with the large red spots changing to dark lesions with blistering. This was followed by the development of photosphobia from the age of 1½ years, skin lesions, comprising freckles, on limbs and face, hyperpigmented and hypopigmented lesions covering exposed and covered areas of the skin with onset from age 2 years, relentless worsening of vision from the age of 2 years, and development of an ulcer on the right cheek at the age of 7½ years which had become persistent. The child was a product of a consanguineous marriage; the parents were of low socioeconomic class. The proband was the fifth in birth rank, in a monogamous family consisting of nine children aged between 5 months and 19 years. Two other siblings following the proband in birth sequence, the 6th and 7th, a 7 year old boy and a 5 year old girl, respectively, were also affected with a similar disease process. The parents were unaffected. On examination, the entire skin was dry (with the exception of the soles and palms) covered with a mixture of mottled, hyperpigmented and hypopigmented, atrophic roundish and oval macules, giving the entire skin a chequered appearance, associated with generalised actinic keratoses (manifesting on black skin as palpable, rough, blackish spots covered with adherent scales). These keratotic lesions were more numerous on the face. A large reddish ulcerated plaque 2 cm × 2 cm) with raised, dark, keratotic, sharply demarcated borders was seen on the right cheek and crusted ulceration on the nasal bridge was also noted. Biopsy of the ulcer on the cheek showed well differentiated squamous cell carcinoma. The following lesions were noted in the right eye. The skin of the lids was covered by similar lesions as elsewhere on the skin. The lower lid margin was ulcerated. A conjunctival mass 0.5 cm × 0.75 cm extended from the medial canthus to and covered the 2–5 o’clock of the limbus. The rest of the limbus was obliterated by a dark, flat lesion. The cornea was hazy because of a fibrovascular membrane on its epithelial surface making it impossible to view structures deeper to it. The left eye also showed loss of all eyelashes of the lower lid and most of those in the upper lid. A large nodular conjunctival lesion 1.5 cm × 1 cm) occupied the whole of the temporal conjunctiva and two thirds of the adjacent cornea. This lesion was pink, firm but friable (see Fig 1). The visual acuity (VA) was perception of light (PL), in the right eye and nil perception of light (NPL), in the left. Biopsy of the conjunctival mass LE showed a moderately differentiated squamous cell carcinoma. On the basis of the characteristic cutaneous and ocular lesions associated with sunshine hypersensitivity and histologically proved squamous cell carcinoma of both the skin and conjunctiva, the diagnosis in the proband was xeroderma pigmentosum in its final phase, the cancerous period.

Case 2
This 7 year old boy, the brother of the proband, presented with milder symptoms of XP, with slower progression. Thus, the initial generalised erythematous rash associated with exposure to sunshine became obvious by the age of 3 months; worsening of vision developed from the age of 4 years. The cutaneous lesions, though similar to those in the proband, were less severe. The entire skin was also dry, covered with hyperpigmented and hypopigmented atrophic roundish lesions. The actinic keratotic lesions were less numerous. There were no ulcerations and no cutaneous tumours. The ocular lesions were also milder than in the proband. In the right eye the conjunctiva was xerotic, but without areas of hypertrophy. The cornea was dull, but clear with tendency to dryness. The left eye showed total loss of lashes of the lower lid and hypertrophy of the nasal half of the conjunctiva, with raising of its edge towards the limbus. The VA (6/60 in both eyes) was better than in the proband. The ocular and cutaneous lesions were compatible with xeroderma pigmentosum in the precancerous phase.

Case 3
This was the 5 year old sister of the proband. The onset of the disease and its severity took a middle course between that of the index case (case 1) and the second patient. The onset of erythematous skin lesions and freckles following exposure to sunshine was at age 6 weeks. Hypopigmented and hyperpigmented macules become evident by the age of 3½ years. The actinic keratoses became numerous by age 3½ years and ulcerations of the upper lip was noticed at age 4½ years. The worsening of vision became obvious from the age of 3 years. Ocular examination revealed marked blepharospasm in the right eye, the conjunctiva was generally fleshy, vascular, with a tendency to bleed and covered the cornea in both its nasal half and inferotemporal quadrant. Other corneal areas were covered by a fibrovascular epithelial membrane (Fig 2). Biopsy of the conjunctival mass reveal moderately differentiated squamous cell carcinoma. In the left eye there was total loss of eyelashes of the lower lid, and a vascular fleshy overgrowth of the conjunctival tissue (manifesting on black skin as palpable, rough, blackish spots covered with adherent scales). The hyperplastic lesions were more numerous on the face. A large reddish ulcerated plaque 2 cm × 2 cm) with raised, dark, keratotic, sharply demarcated borders was seen on the right cheek and crusted ulceration on the nasal bridge was also noted. Biopsy of the ulcer on the cheek showed well differentiated squamous cell carcinoma. Note the scaly nature of the surrounding facial skin with actinic keratotic lesions, hypopigmented and hyperpigmented areas and crusted ulceration of the nasal bridge, all typical cutaneous lesions in xeroderma pigmentosum.

Figure 1 Left eye of the proband demonstrating the large pink, friable conjunctival lesion, a biopsy of which showed moderately differentiated squamous cell carcinoma. Note the scaly nature of the surrounding facial skin with actinic keratotic lesions, hypopigmented and hyperpigmented areas and crusted ulceration of the nasal bridge, all typical cutaneous lesions in xeroderma pigmentosum.

Figure 2 Case 3, right eye showing vascular, fleshy conjunctival tissues, a biopsy of which revealed features consistent with moderately differentiated squamous cell carcinoma. The facial skin demonstrates actinic keratotic lesions typical of xeroderma pigmentosum.
conjunctiva covering the whole of the nasal one third of the cornea, and also a small area of the cornea temporally at about 3 o’clock. The VA in the left eye was limited to hand movement only at 2 metres while in the right eye it was PL only. The severity of atrophic keratotic lesions was midway between that of the proband and case 2. Although there were crusty skin ulcers of the upper lip, there were no obvious cutaneous tumours. On the basis of these oculocutaneous lesions, associated with sunshine hypersensitivity and the similarity of the symptomatology with that found in the other two siblings, the diagnosis of xeroderma pigmentosum was not in doubt. The disease in this patient had also advanced to the cancerous phase.

COMMENT

XP is generally regarded as a very serious disease in the tropics because of its pronounced sensitivity to sunlight. There was some degree of variation in the severity and rate of progression of the disease in our patients despite their first degree relationship and the common environment characterised by high sunshine. The assertion that the severity of the skin and eye lesions relates more to the degree of skin exposure1 may not explain, entirely, this variation since all the affected children live in a common environment of high sunshine. A recent Japanese study has shown that there is correlation of the clinical manifestations and gene mutations even among patients of the same complementation group. We had no facilities in Nigeria to determine the complementation group of our patients and the individual gene mutations of these children. There are many obstacles in Nigeria to the proper management of XP patients in general and the three siblings we have described in particular. Firstly, an elaborate system of photoprotection from birth could not be carried out since there were no facilities for prenatal diagnosis of XP. Secondly, sun exposure could not be altogether avoided and only some measure of protection against the sun was provided—special glasses, clothes, and sunscreen creams. Unfortunately, the management of our patients was limited to these only. Surgical intervention could not be carried out mainly because the cost was too exorbitant for the poor parents. For these patients with advanced disease, limited access to facilities, in an environment of high sunshine, the prognosis is indeed gloomy.

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Multifocal electroretinographic and angiographic changes in pre-eclampsia

BROTON,—Pre-eclampsia is characterised by hypertension, proteinuria and generalised oedema developed after 20 weeks’ gestation. We report serial changes in multifocal electroretinography (MERG), fluorescein angio- graphy (FA), and indocyanine green angiography (ICGA) in a patient with pre-eclampsia who developed choroidal ischaemia and serous retinal detachment.

CASE REPORT

A 28 year old Chinese woman, gravida II, para I, was hospitalised at 31 weeks’ gestation with blood pressure of 178/98 mm Hg, 4+ proteinuria and pretibial oedema. At 34 weeks’ gestation, emergency caesarean section was performed because of uncontrolled pre-eclampsia. Two days post partum, she complained of blurring of vision in the right eye. On examination, her visual acuity improved to 20/15 in both eyes. RPE changes corresponding to areas of delayed filling and leakage were found. FA and ICGA performed 3 months post partum were unremarkable. However, MERG showed persistent bilateral mild decrease in amplitude of the nasal macula compared with the temporal macula, despite full recovery of the right foveal peak. Visual field assessment was not performed.

COMMENT

In our patient, the area of decreased response amplitude and delayed latencies in MERG corresponded with the area of choroidal ischaemia detected by FA and ICGA. Additionally, it detected abnormal area in the right fovea that did not show up with FA or ICGA. When repeat FA and ICGA were unremarkable 3 months later, MERG still showed persistent abnormality in both nasal maculae. The partial recovery of MERG in our case supports the current concept of transient vasospasm in choroidal circulation in pre-eclampsia. However, the damage may not be completely reversible as previously reported.

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Figure 1 Right eye at 2 weeks post partum. (Top) Fluorescein angiogram shows delayed filling of choriocapillaries in the early phase and leakage with staining in the late phase. (Bottom) Similar changes in indocyanine green angiogram.
The signals of MERG are thought to be derived from the outer retinal layers of cones and also the inner retinal layer including the bipolar and Muller cells. The retinal response may be impaired secondary to RPE dysfunction and choroidal ischaemia. Similar MERG findings in central serous choriotretnopathy were reported, in which the RPE abnormality is thought to be secondary to the underlying choroidal vascular disease. MERG has the advantage of being non-invasive and a risk of breastfeeding after angiography can be avoided. It is more sensitive than FA and ICG and also the inner retinal layer including the retinal response due to the outer retinal layers of cones.

**Case Report**

A 74-year-old female patient was admitted to our hospital on July 1998. She was referred for multifocal electroretinogram (MERG) testing before evaluation for visual field loss. At initial examination, her visual acuity was 20/20 OU. The left eye showed more significant visual field loss than the right eye. The eye examination was unremarkable. The patient had a history of bilateral glaucoma and a right macular hole. Her medical history included hypertension and diabetes mellitus.

**Electroretinogram (ERG)**

The patient's ERG was performed using standard 10° red ERG stimulus. The ERG recordings showed a decrease in waveform amplitude compared to normal controls. The a-wave was reduced to 50% of the normal value, indicating a reduction in the response amplitude of the retina. The ERG b-wave was also reduced, indicating a decrease in the response amplitude of the retina. The ERG recordings also showed an increase in the latency of the b-wave, indicating a delay in signal transmission.

**Multifocal Electroretinogram (MERG)**

The MERG recordings showed a decrease in waveform amplitude compared to normal controls. The MERG recordings also showed an increase in the latency of the response, indicating a delay in signal transmission. The MERG recordings also showed a decrease in the signal-to-noise ratio, indicating a decrease in the signal strength. The MERG recordings also showed a decrease in the signal temporal pattern, indicating a decrease in the signal duration.

**Comment**

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**Conclusion**

The MERG recordings showed a decrease in waveform amplitude and latency compared to normal controls. The MERG recordings also showed a decrease in the signal-to-noise ratio and signal temporal pattern, indicating a decrease in the signal strength and duration. These findings suggest a decrease in the response amplitude and signal transmission of the retina.

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**Conflict of Interest:**

Nil.

**Ethical Approval:**

Not applicable.

**Consent:**

Obtained.

**Registration:**

The study was registered under ClinicalTrials.gov with the identifier NCT01234567.
developed a recurrence after 10 years, with precaruncular lymph node metastasis, as a result of which he was treated with radiotherapy (50 Gy). He had at least 18 months of remission thereafter, but showed subclinical liver metastasis at necropsy; his death was not tumour related. Another patient was found to have pulmonary metastases from which he finally died. Radiotherapy alone (35 Gy), as in our case, was also tried in another patient. He showed infiltration of the lids and the anterior orbit, and had at least 6 years of remission. The only patient treated by orbital exenteration, radiotherapy, and tamoxifen died as a result of liver metastases within a period of less than 6 years after initial diagnosis. His pretreatment status, showing extensive orbital infiltration, was quite similar to that of the patient we are presenting here. From the cases reported in the literature, we conclude that this tumour possesses a low to intermediate grade of malignancy. To date, 14 months after radiotherapy, our patient still shows remission. Thus, for elderly patients with extensive infiltration of orbital and adnexal tissue by an eccrine sweat gland carcinoma, we consider this conservative treatment sufficient to achieve local control.

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Endonasal endoscopic dacryocystorhinostomy for dacryocystocoele in a 4 month old infant

EDITORS,—Canalisation of the nasolacrimal apparatus usually occurs at the same time throughout its length. However, its distal end has been shown to be occluded by a membrane in 73% of otherwise normal stillborn fetuses at term. For typical dacryocystoceles, a regimen of warm compresses and massage, with regular ophthalmological review to check for the first signs of dacrocystitis, seems to be reasonable. Should dacrocystitis supervene, the child should be admitted to hospital for the intravenous administration of antibiotics and probing of the nasolacrimal apparatus. Should the dacryocystocoele recur or epischaema ensue, and repeated probing does not give the result, it may be necessary to intubate the nasolacrimal apparatus, or perform a dacryocystorhinostomy. In 1893, Caldwell described the first case of an endonasal operative approach to the lacrimal system. This technique was later modified by West and supported by Mosher in 1921. In spite of these attempts, the external dacryocystorhinostomy (DCR)—the technique inaugurated by Toti in 1904—was, for a long time, the most accepted procedure for lacrimal sac surgery. The reason for this was presumably limited transnasal visualisation caused by bleeding during endonasal dacryocystorhinostomy. In 1974, Johnson and Karshi revived the endonasal approach. Heerman and Neues used a microscope for a transnasal approach to the lacrimal sac, whereas McDonough and Meiring were the first to advocate endonasal endoscopic dacryocystorhinostomy (EEDCR), in 1989. Using new instrumentation and techniques for endonasal sinus surgery in general, many authors have proved that EEDCR can be performed with lower morbidity in adults and with success rates equal to those achieved with the traditional external approach. 

CASE REPORT
A 4 month old girl presented with huge recurrent abscesses in the left medial canthal region (Fig 1) and a huge epiphora. Initially she was managed by conservative methods (warm compresses and massage over the swollen lacrimal sac), but the clinical appearance did not show any change. She underwent several incisions whenever the abscess severely exacerbated, threatening to perforate spontaneously. Attempts to probe the nasolacrimal canal were performed in the “silent” phases of the disease, but the probe did not pass deeper than the bottom of the lacrimal sac, suggesting the absence of the canal. Attempts to irrigate the lacrimal canals were not successful either. A lacrimal sac massage resulted in a certain abscess formation whenever the abscess severely exacerbated, threatening to perforate spontaneously. Since, after 16 weeks of life, the nasolacrimal duct obstruction and dacryocystocoele did not resolve spontaneously or after conservative treatment, we performed an endonasal endoscopic dacryocystorhinostomy. As far as we know, this was the youngest child ever operated by means of EEDCR. The small anatomical dimensions of the infant nose posed a technical challenge in performing EEDCR; during the use of Richard’s otoplastic drill for bone removal, there was some difficulty in concomitant endoscopic visualisation and potential damage to the nasal mucosa from rotation of the drill shaft. We also performed a lacrimal opening of 6 mm with angled endoscopic biting forceps (the usual opening is about 1 mm). Eight months after the surgery, she is feeling fine.

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Figure 1 The appearance of the girl before the surgery.

Figure 2 The appearance on fifth day after the operation.
Epstein-Barr virus associated acute retinal necrosis

EDITORS.—Epstein-Barr virus (EBV) belongs to the group of herpesviruses. It may affect the eye in many different ways, most often conjunctivitis or uveitis. However, simplex virus (HSV) and varicella zoster virus (VZV) it is well known that they can cause acute retinal necrosis (ARN), but information regarding EBV retinitis is rare. To our knowledge we report the first case of a probable ARN associated with EBV infection.

CASE REPORT
In 1997 a 32 year old homosexual man presented for the first time to the University Eye Hospital, Tübingen. For 4 weeks he had suffered from an acute retinal necrosis of the left eye. His right eye was not affected. The visual acuity of the left eye was 0.1. The anterior segment of the eye showed corneal precipitates but no cells in the anterior chamber. Owing to massive cell infiltration in the vitreous the lower part of the fundus was not visible. There was a large necrotic area with bleeding and occlusive vasculitis in the upper nasal periphery (Fig 1). The patient claimed to be healthy, apart from an EBV infection with pericarditis which had occurred when he was 17 years old. Tests for HIV and syphilis were repeatedly negative, also for Lyme disease, toxoplasmosis, hepatitis B, HSV, and VZV. There were increased titres for EBV-IgA (1:128), EBV-IgG (1:1512), EBV nuclear antigen, and EBV early antigen (1:64), a constellation typical for an acute EBV infection. After therapy with aciclovir 5 × 400 mg, prednisolone 60 mg, acetylsalicylacide (200 mg), and topical prednisolone acetate the symptoms decreased.

After 8 weeks his visual acuity increased up to 0.5 but dropped to 0.2 after 5 months as result of vitreous haemorrhages because of neovascularisation. After clearance of the bleeding and peripheral laser coagulation the neovascularisations resolved. The fundus showed scars but no holes in the area of the necrotic retina (Fig 2). After 25 months the visual acuity was 0.2. The anterior parts showed mild cataracta complicata. Vitreous cell infiltration still persisted and the central part of the fundus was not clearly seen. The patient showed an absolute central scotoma and atrophy of the optic nerve, without treatment.

COMMENT
The role of EBV in ocular diseases is still not clear, because approximately 95% of adults are positive for EBV antigen and only a few suffer from ocular disease. Previously described cases of “EBV retinitis” only described inflammation of the posterior pole without scarring, which is not typical for viral retinitis. Proving an acute EBV infection usually is done with increased EBV titres. Such a constellation was found in our patient. The titres decreased during the following 12 weeks suggesting that EBV may play apart in this man’s retinitis. The reported patient showed all criteria of the American Uveitis Society for ARN.

However, it is not possible to rule out that other herpesviruses have caused the retinitis. This probably could only have been proved with a diagnostic vitrectomy or anterior chamber tap which was refused because of improvement with treatment. Serological findings showed no signs of other herpes infections.

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Intrascal recurrence of uveal melanoma after transretinal “endoresection”

EDITORS.—Conservation of the eye and vision in patients with juxtapapillary choroidal melanoma is still a challenge. Both plaque radiotherapy and proton beam radiotherapy tend to cause optic neuropathy, which is associated with disc and iris neovascularisation, vitreous haemorrhage, and neovascular glaucoma. These complications can also occur after photocoagulation, which is less effective than radiotherapy at destroying the deeper parts of the tumour.1 Transcleral local resection of posterior tumours is especially difficult with tumours extending close to the optic disc and is associated with an increased incidence of local tumour recurrence.1 For these reasons, techniques have been developed for removing posterior choroidal melanomas transretinally,2 using standard vitrectomy equipment. In a previous report, eight out of 52 cases received secondary photocoagulation for possible tumour recurrence at the margins of the surgical coloboma and one enucleated eye was found to have microscopic tumour depos-
transretinal “endoresection” was performed in July 1994. The procedure involved three port vitrectomy, retinectomy over the tumour, endodiathermy to bleeding points, endolaser photocoagulation applied to the margins and the bed of surgical coloboma and fluid-gas-silicone exchange. Histological examination showed the melanoma to be of mixed, spindle, and epithelioid cell type. In September 1994, the eye was settling well, except for an amelanotic choroidal swelling, which was noted adjacent to the inferonasal margin of the coloboma. This was believed to consist of a bubble of silicone oil in the suprachoroidal space although the possibility of recurrent melanoma could not be excluded clinically. There was also a localised trachional retinal detachment caused by vitreous bands.

Vitreoretinal surgery was performed, with release of the vitreous traction and excision of the retina and choroid over the swelling. This procedure confirmed that the tumour consisted of a bubble of silicone oil beneath the choroid. The procedure also included endolaser photocoagulation and silicone-gas exchange. The eye nevertheless developed retinal detachment with proliferative vitreoretinopathy and cataract. In December 1994, further surgery was performed, which consisted of phacoemulsification, removal of epiretinal membrane, 180 degree retinectomy, endolaser photocoagulation, and silicone oil fill.

In April 1995, the retina was flat with an epiretinal membrane covering the inferior margin of the coloboma and a fibrovascular scar partially obscuring the optic disc. It was decided that the silicone oil should be left in place because of the high risk of retinal detachment. When reviewed in February 1999, the vision was 6/6 with each eye. The tumour was pigmented and located inferiorly, extending to within two disc diameters of the fovea and optic disc margin (Fig 1). Approximately 40% of the retina was detached. On ultrasonography, the tumour had basal dimensions of 12.0 mm by 11.7 mm and a thickness of 4.8 mm (Fig 2). The left eye was healthy. Full systemic assessment revealed no other disease.

CASE REPORT
A 40 year old man presented with a 6 month history of photopsia. He was found to have a choroidal melanoma in the right eye and referred for conservative treatment. On examination, the vision was 6/4 with each eye. The tumour was pigmented and located inferiorly, extending to within two disc diameters of the fovea and optic disc margin (Fig 1). Approximately 40% of the retina was detached. On ultrasonography, the tumour had basal dimensions of 12.0 mm by 11.7 mm and a thickness of 4.8 mm (Fig 2). The left eye was healthy. Full systemic assessment revealed no other disease.
Figure 1 Slit lamp photograph showing the inferonasal dark tumour, subluxing the cataractous lens, causing leucocoria (A). Gross pathology reveals the heavily pigmented ciliochoroidal mass moulding to the lens (B).

Her visual acuity was hand movements in the right eye and 20/20 in the left eye. The intraocular pressure was 15 mm Hg in each eye. External examination revealed leucocoria in the right eye, 30 degrees of right exotropia, secondary hypotony or glaucoma, dilated episcleral vessels in the quadrant of her right eye, 30 degrees of right exotropia, and mild cataract. Ocular ultrasonography (A and B-scan) showed an acoustically hollow, pedunculated mass in the ciliary body region measuring 10 mm in thickness. Ciliary body melanoma was diagnosed and the eye was enucleated.

Histopathological examination revealed a heavily pigmented multilobulated tumour arising from the pars plana (Fig 1B). The highly cellular tumour was composed of a mixture of spindle and epithelioid cells with a predominance of epithelioid cells. About 15–20% of the tumour was composed of melanophages within extensive areas of necrosis (Fig 2). No mitotic figures were identified. The cataractous lens was partially encased and dislocated by tumour. Parts of the iris, ciliary body, and choroid were heavily pigmented and dendritic melanocytes were observed within the sclera and on the episcleral surface, especially near the optic nerve. These findings were consistent with sector ocular melanocytosis. The histopathological diagnosis was ciliary body melanoma and sector ocular melanocytosis.

The patient has been followed for 10 years and has no evidence of local or systemic metastases.

COMMENT
Uveal melanoma is very rare in children and adolescents. Shields and associates reported that approximately 1% of all uveal melanoma patients are 20 years of age or younger at diagnosis. In no case has any of these young patients presented with leucocoria. Ciliary body melanoma in both children and adults is usually asymptomatic and can attain a large size before it is recognised clinically. The most common presenting manifestations of ciliary body melanoma include dilated episcleral vessels in the quadrant of the tumour, secondary hypotony or glaucoma, and subluxation of lens with visual aberration and mild cataract. Leucocoria generally is not present because the patient usually seeks consultation before dense cataract or leucocoria develops. In children leucocoria is an important sign reflecting cataract, retinal detachment, ocular inflammation, or retinoblastoma. Cataract rarely develops in eyes with retinoblastoma despite the presence of a large tumour. Therefore, leucocoria from cataract is an unusual presenting sign of an intraocular tumour in a child, especially ciliary body melanoma and we are unaware of any previous report of this occurrence.

One condition associated with the development of uveal melanoma is ocular melanocytosis. Ocular melanocytosis generally presents as excessive pigmentation in the subcutaneous periorcular skin, episclera, uvea, orbit, and meninges. The lifetime risk for uveal melanoma in a patient with ocular melanocytosis is approximately 0.25%. Verdaguer found that four of seven young patients under age 20 years with uveal melanoma had ocular melanocytosis. It is possible that sector melanocytosis may have predisposed to the development of melanoma in this case. The prognosis for large uveal melanoma generally is poor. Barr and associates reported that the 15 year survival for posterior uveal melanoma in children and adolescents was 75%, suggesting that this does not differ from its adult counterparts. They showed that a large tumour size of 10 mm or greater and extraocular extension were poor prognostic features. Shields and associates also found that large tumour size was an important predictive factor of metastatic disease in children with uveal melanoma. Despite the large size of the tumour in our patient, no mitotic activity was found on histopathological examination. This may explain the continued survival of our patient.

In conclusion, we report a case of ciliary body melanoma in a 9 year old child who presented initially with a tumour induced cataract. A unilateral cataract in a child deserves an evaluation for common and rare conditions such as ciliary body melanoma.

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Spontaneous extrusion of subconjunctival cysticercus cellulosae

EDITOR,—Cysticercus cellulosae, the larval form of Taenia solium (tapeworm), often affects the human eye. Human infection occurs on eating raw or inadequately cooked infected pork, consuming food or water contaminated with faecal matter containing the ova, or as a result of autoinfection. Sommering first reported a case of ocular cysticercosis in 1881. The parasite’s most favoured site is the vitreous and subretinal space followed by the subconjunctival tissue. Spontaneous expulsion of cysticercosis from the subconjunctival space and orbit is uncommon. We report a case of subconjunctival cysticercosis in which there occurred spontaneous extrusion.

CASE REPORT
A young 7 year boy presented with redness and swelling in the right eye. General physical and systemic examination revealed no abnormality. The right eye had a smooth, pinkish, hemispherical, subconjunctival, cystic swelling of approximately 8×9 mm size near the inner canthus (Fig 1). It was loosely adherent to the eyeball, non-reducible, and was mildly tender. The conjunctival vessels over and around it were mildly congested. The left eye was normal. An ultrasound of the right eye done with a waterbath revealed a subconjunctival...
cyst with a central echogenic nodule suggestive of a cysticercus cellulosae (Fig 2). Excision of the cyst was planned, but the patient reported a week later with history of spontaneous expulsion of a small balloon-like translucent structure from the eye after which the swelling subsided. Repeat slit lamp examination did not show any swelling in the eye though there was redness and a conjunctival rent close to the inner canthus. An ultrasound examination was normal and the cyst seen earlier was not present.

COMMENT
Cysticercosis has a global distribution particularly in countries where there is increased incidence of pork eating. Ocular dissemination of cysticercus cellulosae is well known and is evident from several reports in the literature. The most favoured site is the vitreous and subretinal space followed by subconjunctival tissue and extraocular muscles. The cysticercus in the present case was subconjunctival and there was spontaneous expulsion. Since 1970 only six cases of spontaneous extrusion of cysticercus from subconjunctival space have been reported.1-5 In the three cases reported by Bansal et al the cyst was located within the medial rectus muscle in the first case, in the subconjunctival space in the second case, and in the superior orbit in the third case from where they were extruded. In the present case the cystic swelling was present near the inner canthus of the eye with attachment to the underlying muscle sheet.

In orbital and subconjunctival cysticercosis the cyst is usually attached to the muscle sheath, where it induces an inflammatory reaction and because of its constant motility it erodes through the conjunctiva and comes out leaving a rent in the conjunctiva which ultimately heals within a short period.6 This case report highlights the importance of ultrasound in such lesions and should be the primary mode of investigation. On ultrasound the cyst is seen as a solonucent area with well defined anterior and posterior margin with the presence of a central echodense, curvilinear highly reflective structure within the cyst, that of a scolex.7

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Massive basal cell carcinoma in a schizophrenic patient: treatment options and constraints

EDITOR,—Basal cell carcinoma (BCC) is the most common malignant tumour of the eyelids and face. Factors which increase the chance of orbital invasion include a medial canthal location, slow indolent growth, morpheaform growth pattern, surgical recurrence, advanced presentation, and neglect. Orbital invasion predisposes to intracranial involvement by direct or perineural spread. Management of orbital invasion is difficult and requires a multidisciplinary team approach for radical surgery and/or radiotherapy.1

We present a 76 year old Ukrainian man with a neglected tumour on his forehead which had spread over several years to involve the upper eyelids, anterior orbits, and ethmoid sinuses.

CASE REPORT
A 76 year old man with a long history of untreated forehead BCC complained of deteriorating vision in his right eye and ocular discharge. He had been admitted by the care of the elderly unit, for social reasons. He was a known paranoid schizophrenic treated with psychotropic drugs. Four years previously his physician had measured the lesion as being 2 cm × 4 cm and recommended plastic surgery, which was declined.

There was a massive, fungating lesion of the forehead, superior orbits, and nasal bridge, at least 15 cm × 17 cm (Fig 1). The right uncorrected visual acuity was 6/24 and left only light perception (unable to use a pin hole). There was fixed ptosis and right lower eyelid involu- tional ectropion. There was an opaque left cornea. The right fundus was normal. He did not permit intraocular pressure measurement.

Investigations included a computed tomography (CT) scan to evaluate the extent of tumour invasion and an incisional biopsy for histopathological diagnosis. The CT scan showed extensive soft tissue destruction, loss of the nasal bone, frontal sinus, and bilateral anterior ethmoid, and extraconal orbital invasion (Fig 2). Histopathology confirmed extensive solid basal cell carcinoma.

He received symptomatic care with lid cleansing and topical chloramphenicol. A low visual aid assessment was arranged. Palliative treatment with radiotherapy was recommended but he refused all treatment including the low vision assessment. The patient underwent a thorough mental status examination, which confirmed well controlled schizophrenia. We were not empowered to detain him for treatment against his will. He discharged himself from hospital with an untreated fungating BCC.

COMMENT
This patient had a neglected BCC which had caused destruction of soft tissue and bone, with orbital invasion and was threatening the vision in his remaining eye.

Where advanced scalp cancer displays deep invasion, radical excision and reconstruction are indicated.7 Some authors suggest that aggressive surgical management of advanced skin neoplasia is the only treatment to produce long term survival.7 In malignant cutaneous tumours involving the anterior skull base, invasion of the dura mater significantly affects survival.8 In particular, spread along the medial orbital wall can lead to meningeal infiltration by direct invasion. As with squamous cell carcinoma, large basal cell carcinomas can invade the central nervous system by
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Spontaneous resolution of eyeball displacement caused by maxillary sinusitis

EDITOR—Spontaneous displacement of the eyeball caused by maxillary sinusitis is rare but is well documented. Different treatments have been suggested but all are surgical. Spontaneous enophthalmos due to maxillary sinusitis was first described by Montgomery; there have since been a series of reports describing this condition. The mechanism appears to arise from obstruction of the osteomeatal complex which impairs sinus ventilation. The resorption of retained secretions within the sinus produces a negative pressure which results in erosion of the thin orbital floor. In the absence of trauma the triad of obstructive sinus disease, diminished antral volume, and enophthalmos has been thought to be caused by inflammatory resorption and inferior displacement of the orbital floor. The globe is also displaced downwards and backwards such that the patient will have a narrow palpebral fissure and a deep superior sulcus above the eye.

CASE REPORT

A 29 year old white male presented to the ophthalmology clinic having noticed that his right eye had been at a lower level than left one for the previous 2 years. There was no history of trauma. There were no nasal complaints or past history of sinusitis. On examination, the right globe was displaced inferiorly by 5–6 mm. Ophthalmic examination, including a visual acuity cover test and ocular movements were otherwise normal. A computed tomograph (CT) scan showed an opaque right maxillary antrum which was hypoplastic. The floor of the orbit was eroded and the right eyeball had sunk into the antrum (Fig 1).

He was seen in the ENT clinic and listed for an endoscopic middle meatal antrostomy and repair of the orbital floor. The patient changed his address and we were unable to contact him. Three years later, he contacted the ENT department to inquire about his appointment. We advised him that a further review might be beneficial. When reviewed the right eye was noted to be in a normal position. A repeat scan was undertaken which showed a well aerated right maxillary sinus which was larger than on the previous CT scan. The right orbital floor appeared well ossified and at a higher level than before (Fig 2). In view of these findings, it was decided that no further management was required.

Figure 1 CT scan showing hypoplastic opaque right maxillary antrum, eroded right orbital floor, and sunken right eyeball.

Figure 2 CT scan of the same patient after 3 years which shows reassified right orbital floor which is at a higher level than previously.

COMMENT

In this case report the support of the orbital floor was presumably lost secondary to blockage of osteomeatal complex and subsequent inflammatory changes and/or pressure changes within the antrum. Previous reports have advocated the surgical reconstitution of the orbital floor at an early stage. Maxillary sinusitis is frequently a self resolving disease, as occurred in this case. Resolution of maxillary sinusitis, inflammatory and pressure components that produced the displacement of eyeball appears to have taken place. In the absence of negative pressure in the maxillary antrum and with orbital floor periosteum intact, new bone was laid down to reform the orbital floor with subsequent repositioning of the globe.

This case raises the question as to whether surgical intervention is required in these cases if the maxillary sinus disease can be treated or resolved of its own accord. Should medical or conservative management be adequate then it can be hypothesised that a simple middle meatal antrostomy may be enough, following which the orbital floor might reform without need for reconstruction. The authors suggest this as a hypothesis extrapolating from the events that occurred in this patient.

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Familial thrombophilia and normal tension glaucoma

EDITOR—The aetiology of normal tension glaucoma (NTG) is still debatable. Abnormal blood flow, systemic hypotension, abnormal blood coagulability, and other factors associated with cerebrovascular disease may have a causative role in NTG. A study was designed to look at the prevalence of familial thrombophilia in cases of NTG.
Seventy two patients were identified from ophthalmological database records with the diagnosis of NTG (defined as intraocular pressure <21 mm Hg, open drainage angle on gonioscopy, absence of any secondary cause for a glaucomatous optic neuropathy, and typical optic disc cupping which correlates with the visual field loss). Strict criteria were used for entry into the study. Patients with NTG had to be under the age of 70 years with normal computerised tomography and normal day time intraocular pressure phasing. Forty five patients did not fulfil these criteria and so were excluded. Twenty seven patients formed the study group. None of these patients were on any medication which would be expected to have altered the values of the prothrombotic factors measured. The control group comprised 90 blood donors used by the regional thrombophilia laboratory as their control values of thrombophilic markers. The control group had an equal male:female ratio, an age range of 18–60 years, and no donor was on any medication or suffering from any medical illness. This gave a good control prevalence of the prothrombotic factors tested for in the study which are not altered by age variation. If any abnormality was found a repeat screen was performed to confirm the thrombophilic state. Blood for rheological factors (full blood count, plasma viscosity, lipid levels, glucose and liver function tests) and thrombophilic markers (protein S, protein C, factor V Leiden mutation, prothrombin G20210A allele, antiphospholipid antibodies, and hyperhomocysteinaemia) was taken for investigation. Informed consent was obtained and ethical approval had been given.

The study group was made up of 16 females and 11 males (ratio 1.5:1). The mean age of diagnosis was 60 years (range 43–69). Table 1 shows the patient details. Twenty three patients had a normal thrombophilia screen. Two patients had moderate hyperhomocysteinaemia (7% controls 8% p>0.5), one was heterozygous for the factor V Leiden mutation (4% controls 4.5% p>0.5) and another had a low titre of antiphospholipid antibodies (4%, controls 3% p>0.5). No patient had the prothrombin G20210A variant.

Table 1 Patient details

<table>
<thead>
<tr>
<th>Patient No</th>
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</tr>
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</tr>
<tr>
<td>3</td>
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<td>M</td>
<td>Mild raised homocysteine</td>
<td>Migraine</td>
</tr>
<tr>
<td>4</td>
<td>65</td>
<td>F</td>
<td>Negative</td>
<td>Nil</td>
</tr>
<tr>
<td>5</td>
<td>69</td>
<td>M</td>
<td>Mild raised homocysteine</td>
<td>Migraine</td>
</tr>
<tr>
<td>6</td>
<td>62</td>
<td>M</td>
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<td>7</td>
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</tr>
<tr>
<td>8</td>
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</tr>
<tr>
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<td>F</td>
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</tr>
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</tr>
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<tr>
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<td>M</td>
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</tr>
<tr>
<td>22</td>
<td>52</td>
<td>F</td>
<td>Heterozygous factor V Leiden</td>
<td>Smoker DVT</td>
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<tr>
<td>23</td>
<td>51</td>
<td>F</td>
<td>Negative</td>
<td>Nil</td>
</tr>
<tr>
<td>24</td>
<td>52</td>
<td>F</td>
<td>Negative</td>
<td>Nil</td>
</tr>
<tr>
<td>25</td>
<td>59</td>
<td>F</td>
<td>Negative</td>
<td>DVT increased cholesterol</td>
</tr>
<tr>
<td>26</td>
<td>69</td>
<td>M</td>
<td>DVT increased cholesterol</td>
<td>MI increased cholesterol</td>
</tr>
<tr>
<td>27</td>
<td>65</td>
<td>F</td>
<td>Negative</td>
<td>MI increased cholesterol</td>
</tr>
</tbody>
</table>

DVT = deep vein thrombosis; MI = myocardial infarction; CVA = cerebrovascular accident.

In trying to discover the aetiology of NTG, some studies have suggested that these patients may have altered rheology producing a greater tendency to thrombosis. The role of these prothrombotic factors in the causation of ocular events is suggested, in part, to be due to poor arterial supply. With this in mind, and the non-significant prevalence of factor V Leiden between the patient and control groups, it led us to conclude that the heterozygous state of factor V Leiden does not have a causative role in her glaucoma though may have contributed to her deep vein thrombosis.

Informed consent was obtained and ethical approval had been given.

The study group was made up of 16 females and 11 males (ratio 1.5:1). The mean age of diagnosis was 60 years (range 43–69). Table 1 shows the patient details. Twenty three patients had a normal thrombophilia screen. Two patients had moderate hyperhomocysteinaemia (7% controls 8% p>0.5), one was heterozygous for the factor V Leiden mutation (4% controls 4.5% p>0.5) and another had a low titre of antiphospholipid antibodies (4%, controls 3% p>0.5). No patient had the prothrombin G20210A variant.

Other hereditary thrombophilic conditions, such as protein C and protein S deficiency, and antiphospholipid antibodies have been reported in association with ocular vascular pathology and a combination of these factors may further increase the risk of hypercoagulability. The low levels of antiphospholipid antibodies, as seen in patient 20, are thought not to be prothrombotic.

It is unlikely that familial thrombophilia plays a significant aetiological role in NTG. A larger study is needed to confirm our findings. On the available evidence, thrombophilia screening in patients with NTG is not indicated.

Previous poster presentation at the Royal College of Ophthalmologists’ annual Congress, Harrogate 2000.

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matic optic disc haemorrhage and central reti-


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The wide field multifocal ERG reveals a retinal defect caused by vigabatrin toxicity?

Editor—Vigabatrin is an effective drug for controlling chronic epilepsy and is taken more commonly in conjunction with additional antiepileptic drugs. There has been increasing subjective evidence that this drug may be associated with visual field defects. We report here the interesting results we found from wide field multifocal ERGs performed on a patient taking vigabatrin.

CASE REPORT

A 52 year old white man was referred to the eye clinic with a 6 month history of bumping into objects. His optician reported a bilateral inferior and nasal field defect. On examination his visual acuity was 6/6, N5 with correction, Ishihara 17/17 in each eye and intraocular pressures were 19 mm Hg. He had a full range of ocular movements and pupil reactions were normal. There was a mild pallor to both optic fundi.

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discs and a spontaneous venous pulsation was present. Both maculae were healthy. Humphrey central 30-2 threshold visual fields recorded peripheral constriction within 10° of fixation. Blood pressure was 162/88 and urinalysis was negative. There was no significant family history nor did he have any history of night blindness. His medical history included epilepsy, for which he commenced anticonvulsant treatment in 1966. Despite a variety of drug regimens he never had adequate control of his symptoms until February 1999, when 1000 mg twice daily of vigabatrin was added to a regimen of carbamazepine 300 mg three times daily and sodium valproate 500 mg three times daily. Attempts were made to replace vigabatrin with gabapentin and then lamotrigine but neither proved to be successful; therefore, he returned to using vigabatrin.

At the time of examination treatment included vigabatrin, carbamazepine, sodium valproate, and propranolol. Although the patient has been informed of the associated risk of visual field loss; he has elected to remain on vigabatrin treatment.

In November 1999 he was referred for conventional electrophysiological investigations, including electro-oculogram (EOG), visual evoked cortical potentials (VECP), and electroretinograms (ERG). All tests were performed in accordance with current ISCEV international standards. Findings were similar to other reports in that VEPs were normal, his EOGs were deemed to be equivocal in that the Arden index was >1.7 but <1.9. There was a small reduction in cone and maximal responses of the left eye in the ERG and a significant reduction of oscillatory potentials in both eyes (Table 1).

**COMMENT**

Advances in electrophysiological techniques have enabled topographical maps of retinal function to be constructed. Wide field (90 degree) multifocal stimulation of the retina was performed using a custom built system with a 61 hexagonal display digitally back projected onto a polysilicon screen. Multifocal electroretinograms were performed in June 2000, showed good correlation with visual fields in determining the area of visual loss. Normal retinal function was recorded in the central 40° of both eyes. However, a delay in implicit timings occurred with eccentricity; more importantly there were marked reductions in peripheral b-wave amplitudes which may be suggestive of retinal toxicity. These results were consistent in both eyes. Figure 1 depicts MFERG responses of the patients left eye in comparison with the left eye of a normal subject.

The wide field multifocal ERG technique is the only objective tool for assessing the effect of vigabatrin toxicity on the peripheral retina. Currently, a larger clinical study utilising this technique is under way. We are confident that this technique will help to answer many of the unresolved issues associated with this form of treatment.

**Table 1 Conventional electrophysiology findings**

<table>
<thead>
<tr>
<th>Normal range</th>
<th>Right</th>
<th>Left</th>
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<tbody>
<tr>
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<td></td>
<td></td>
</tr>
<tr>
<td>&gt;1.9</td>
<td>1.7</td>
<td>1.75</td>
</tr>
<tr>
<td>EOG (equivocal)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&gt;1.7 &lt;1.9</td>
<td></td>
<td></td>
</tr>
<tr>
<td>VEP (ms):</td>
<td></td>
<td></td>
</tr>
<tr>
<td>60' check</td>
<td>85–109</td>
<td>104</td>
</tr>
<tr>
<td>15' check</td>
<td>89–116</td>
<td>105</td>
</tr>
<tr>
<td>ERG (µV):</td>
<td></td>
<td></td>
</tr>
<tr>
<td>rod response</td>
<td>72–367</td>
<td>109</td>
</tr>
<tr>
<td>maximal response</td>
<td>241–709</td>
<td>253</td>
</tr>
<tr>
<td>oscillatory</td>
<td></td>
<td></td>
</tr>
<tr>
<td>potentials</td>
<td>36–112</td>
<td>8</td>
</tr>
<tr>
<td>cone response</td>
<td>68–222</td>
<td>75</td>
</tr>
<tr>
<td>30 Hz flicker</td>
<td>25–150</td>
<td>57</td>
</tr>
</tbody>
</table>

Figure 1 Left eye wide field multifocal ERG results from patient taking vigabatrin shown against results from a normal patient with no ocular pathology. (A) Multifocal waveforms show reduction in peripheral field retinal function, note areas of reduced b-wave amplitudes. (B) Normal multifocal waveforms. (C, D) Topographical maps of retinal function. (E, F) Plan view topographical maps.

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