LETTERS TO THE EDITOR

Sudden bilateral visual loss in acute myeloid leukaemia

EDITOR,—Intraocular manifestations of leukaemia are not uncommon and may be related to manifestations of the disease itself or to its complications including those of treatment. There may be direct infiltration by neoplastic cells of ocular tissue, including optic nerve, choroid, retina, iris, ciliary body, signs in the retina of associated haematological abnormalities such as anaemia, thrombocytopenia or hyperviscosity states, or retinal destruction by opportunistic infections such as that caused by herpetic viruses. In addition, occlusive retinal microvasculopathy has been reported in patients with acute leukaemia, with radiation considered to be a contributing factor in some cases.1 2

CASE REPORT

A 20 year old woman with a diagnosis of acute myeloid leukaemia FAB (French, American, British) classification M1, was started on BF-12 chemotherapy consisting of a combination of cytarabine, etoposide, and idarubicin (British) and cytosine arabinoside, etoposide and teniposide (American). The patient had complete remission of his acute lymphocytic leukaemia (ALL).

Serous retinal detachment caused by leukaemic choroidal infiltration during complete remission

EDITOR,—Various ocular complications in leukaemia are due to direct invasion by leukaemic cells or haematological abnormalities associated with leukaemia—for example, anaemia, thrombocytopenia, and hyperviscosity states.3 4 These complications usually occur when the disease is clinically and haematologically active but rarely during complete remission. Moreover, serous retinal detachment is a less common complication, while dilated and tortuous vessels, vascular sheathing, white centred retinal haemorrhages, intraretinal haemorrhages, and cotton wool spots are often seen in the fundus. We describe an uncommon case of a young boy who showed a serous retinal detachment during the first complete remission of his acute lymphocytic leukaemia (ALL).


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CASE REPORT
A 17 year old boy presented with gradually blurring vision and metamorphopsia in his right eye. ALL had been diagnosed and treated with chemotherapy 9 months earlier. His disease was in complete remission at the time of presentation. Corrected visual acuity was 20/250 in the right eye and 20/15 in the left eye. Fundus examination of the right eye showed a serous retinal detachment involving the fovea in the posterior pole (Fig 1A). There were no retinal holes. The left eye was normal. Fluorescein angiography showed numerous hyperfluorescent spots beneath the retina and diffuse subretinal accumulation of fluorescein with time (Fig 1B). B-scan ultrasonography demonstrated diffuse leukemic infiltration of the choroid beneath the retinal detachment. Simultaneous A-scan indicated thickening of the choroid to be 2.5 mm (Fig 2). The white blood cell count was 4.8 × 10^9/L with a normal differential but bone marrow aspiration showed 91.6% lymphoblasts. The patient received systemic chemotherapy immediately. Three weeks later, visual acuity in the right eye improved to 20/20 and the serous retinal detachment decreased markedly.

COMMENT
Main fundus manifestations of leukemias are round or flame-shaped haemorrhages with a white component, intraretinal haemorrhages, and cotton wool spots, comprising what is called “leukemic retinopathy”. These retinal findings are observed commonly but serous retinal detachments are unusual in leukemias and even much less common during complete remission. Stewart et al postulated that leukemic infiltration of the choroid caused decreased blood flow in the choriocapillaris, resulting in ischaemia to the overlying retinal pigment epithelium and disruption of the intercellular tight junctions.

If leukemic choroidal involvement is evident clinically, it usually presents as a serous retinal detachment. However, none of those reporting serous retinal detachments in leukemias could demonstrate leukemic infiltration of the choroid even with ultrasonography. In contrast, Abramson et al reported that leukemic involvement of the choroid could easily and reliably be detected with contact ultrasonography. Although we could also detect leukemic infiltration of the posterior choroid with ultrasonography in our case, we could not have found it if it had been much thinner. It may be difficult to detect thin diffuse choroidal infiltration even with ultrasonography, just as it is difficult to diagnose a flat melanoma or diffuse intraretinal pseudotumour.

Intraocular manifestations of leukemias usually are not treated directly. First of all, systemic chemotherapy is attempted. When definite leukemic infiltrates fail to respond promptly to systemic chemotherapy, ocular radiation is usually recommended. In our case, only systemic chemotherapy was administered but the serous retinal detachment was resolved promptly and the visual acuity in the affected eye improved to 20/20. This case suggested that systemic chemotherapy alone could preserve visual acuity if performed early. As the rate of remission induction in leukemic patients increases, ophthalmic examination is becoming more important during remission. Serous retinal detachment caused by leukemia clinically may mimic a simple central serous chorioretinopathy. Ophthalmologists should bear this in mind in leukemic patients even in apparent remission.

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Observations on time course changes of the cherry red spot in a patient with Tay-Sachs disease

COMMENT

Involvement of cranial nerves, such as abducens and oculomotor nerve, leading to diplopia. Both pupils are pharmacologically dilated. With intravenous methylprednisolone (250 mg twice a day) was started immediately. Microscopic examination of the biopsy specimen revealed florid giant cell arteritis (Fig 2B).

Within 3 weeks the oculomotor dysfunction improved rapidly along with normalisation of the ESR, CRP, and all liver factors.

CASE REPORT

The subject was a boy who was born weighing 1600 g at a gestational age of 36 weeks. Two weeks after birth, ophthalmoscopy disclosed a favourable stretch of retinal blood vessels to the peripheral area without any abnormality of the optic disc and macula in both eyes. Following pursuit movement was observed 5 months after birth. Mental and emotional retardations were manifested beginning at 6 months of age which were precipitated with the onset of afebrile tonic convulsions at the age of 1 year 1 month. Following the convulsions, funduscopy revealed chalk-white macular areas with a cherry red spot in the centre of both eyes. Optic atrophy was present in the left eye and mild paleness in the right eye. Nyctagmus with no light fixation was present.

Quantiative analysis of plasma cells and cultured skin fibroblasts revealed a deficiency of β hexosaminidase A enzyme. Immunoelectron microscopy of a biopsy specimen from the rectum disclosed lamellar inclusion body positive for anti-GM, antibody, where a diagnosis of Tay-Sachs disease was made.

At the age of 1 year 6 months, no alterations were observed in the cherry red spot in both eyes and the optic nerve atrophy in the left eye. However, optic nerve atrophy was quite evident in the right eye. Thereafter, there were no funduscopic changes as evidenced in the fundus photograph at the age of 2 years 10 months (Fig 1).

The diagnosis of Tay-Sachs disease is attributable to the accumulation of GM, trihexosylceramide secondary to defects of β hexosaminidase A enzyme. GM, trihexosylceramide accumulates predominantly in the retinal ganglion cells whereby retina becomes turbid with a milky-white coloration. The pattern of the coloration is in conformity with the density of the ganglion cells. Strong opacity is observed in the macula area which is characterised by the multilayered ganglion cells, and the opacity is not found in the foveal pit that is devoid of ganglion cells. Eventually, a cherry
red opacity develops. In our patient, the macular opacity, in all likelihood, was induced by the accumulation of GM1 trihexosylceramide in ganglion cells that then decreased over time. It has been reported from cerebral biopsy findings at various phases of this disease that there is a ballooning of the neurons with vacuolisation in the cytoplasm with progression of the disease process. Eventually, there is a disappearance of the neurons resulting in gliosis.

The retina is an extension of the central nervous system, and ganglioside fractions parallel those of the brain. Lipids stored in the ganglion cells of the retina have similar histochemical reactivity in the retina as in the brain. Accordingly, it is postulated that, as with the cerebral neurons, hyper trophy is also observed in the retinal ganglion cells, followed by their death and disappearance over time. Finally, there is proliferation of glia cells. These progressive changes probably account for the alleviation of retinal opacity and the loss of the cherry red spot in our patient.

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CASE REPORT
A 37 year old Sri Lankan woman presented complaining of intermittent red eyes. The patient’s vision was good and there were no abnormalities in her past general health. She reported having an unusual bluish appearance in her eyes since the age of 15 and reported that her two sisters had a similar corneal appearance, though neither of her brothers or parents were affected. Her parents’ marriage was non-consanguineous. On examination her uncorrected visual acuity was 6/5 in both eyes. Both corneas had a dense arcus extending for 360° of the peripheral cornea. The bilateral corneal stromal opacity was slightly denser anteriorly than posteriorly (Fig 2). Corneal thickness was normal. The left eye was mildly injected. Intracorneal pressure was normal (15–16 mm Hg) in both eyes throughout the follow up. Dilated fundus examination revealed a normal posterior pole. Specular microscopy, pachymetry, and photography were conducted. Specular microscopy revealed cell counts of 2776/2562 cells/mm² right eye and 2836/2456 cells/mm² left eye in two successive measurements. Epithelium with no coincidental systemic disease was provisionally diagnosed. No abnormality was detected on examination of the chest, heart, lungs, neck, and tonsils.

Blood pressure was 120/70. Fasting blood samples were taken for lipids, plasma protein, thyroid function, immunoglobulins, VDRL, FBC, ESR, and IEPG. The patient was treated in both eyes with topical prednisolone 1% and 0.1% (0.0–0.5), ESR was high (36 mm in the first hour, normal 0–30). A diagnosis of severe homozygous lipoprotein A-1 deficiency syndrome was confirmed. There was no corneal change throughout the 18 months of follow up.

COMMENT
The differential diagnosis of our patient’s bilateral corneal stromal opacity with advanced arcus senilis included Tangier disease, fish eye disease, Schnyder central crystalline dystrophy, and apolipoprotein A-1 deficiency. Schnyder central crystalline dystrophy is characterised by elevated serum cholesterol. A disc-shaped pattern of cholesterol crystals deposits are located in the centre of the cornea at the level of Bowman’s layer and the anterior part of the stroma. Our patient had bilateral diffuse stromal involvement. Tangier disease is characterised by low levels of HDL, apolipoprotein A-1 and LDL, mild hypertriglyceridaemia and cholesterol ester deposition in the tonsils (orange tonsils), liver (hepatomegaly), spleen (splenomegaly), lymph nodes (lymphadenopathy), Schwann cells (peripheral neuropathy), and bone marrow. 1

References

Figure 1 Right eye. Corneal appearance: showing increased corneal deposits.

Figure 2 Left eye. Slit lamp appearance: showing increased corneal deposits.

Figure 1 The fundus photograph of the left eye at the age of 5 years 8 months. This shows the cherry red spot and the optic nerve atrophy.

Figure 2 The fundus photograph of the left eye at the age of 5 years 8 months. This shows the reduction in the retinal opacity surrounding the cherry red spot.

Effects of homozygous apolipoprotein A-1 deficiency on the cornea

Eptrop-Apolipoprotein A-1 (Apo A-1) plays a central part in the metabolism of high density lipoproteins (HDL). Apo A-1 and apolipoprotein A-2 (Apo A-2) make up 80–90% of the protein content of HDL. The characteristics of this deficiency are low levels of HDL serum and Apo A-1, normal levels of triglycerides serum and high levels of LDL serum and total cholesterol. 2 HDL concentration is inversely correlated with a risk of coronary heart disease (CHD). 3 However, there is disagreement about the importance of normal Apo A-1 and HDL serum levels in preventing atherosclerosis. Signs of advanced atherosclerosis and early coronary heart disease were only found in some patients with Apo A-1 deficiency, 4 and did not appear in other patients. 5 Thus, in addition to low Apo A-1 levels, other cardiovascular risk factors must be present to cause premature atherosclerosis. 6 Retinopathy, neuropathy, and corneal opacity are associated with this deficiency. 7,8 Our report describes the corneal condition of a 37 year-old Sri Lankan woman with homozygous Apo A-1 deficiency.


diabetes and cholesterol ester deposition in the tonsils.

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Acquired capillary haemangioma of the eyelid in an adult treated with cutting diathermy

Editor,—Capillary haemangiomas usually present within the first weeks or months of life. Less commonly they can present at birth. To our knowledge only one other report exists of the occurrence of acquired capillary haemangioma of the eyelid in an adult.1 Our case is the first reported acquired capillary haemangioma to be treated with cutting diathermy.

CASE REPORT

A 40 year old man was referred to the oculoplastics clinic for evaluation of a left upper eyelid mass. The lesion first appeared 9 months earlier. It had grown in size. The patient was otherwise fit and well—in particular, there was no history of other cutaneous lesions or antecedent trauma. On examination the visual acuities were 6/6 unaided bilaterally. Examination of the adnexa revealed a dusky red pedunculated mass of the left upper eyelid and a mechanical ptosis (Fig 1). Telangiectatic vessels were noted above the lesion. Clinically the appearance was consistent with a capillary haemangioma. The remainder of the ophthalmic and orbital examination was normal. Excision of the lesion was performed with the Ellman cutting diathermy. Haemostasis was maintained throughout the procedure. The wound was sutured with interrupted 6–0 Prolene.

Histopathological examination of the 1×0.7×0.6 cm pedunculated nodular mass revealed numerous capillary lumina lined by endothelial cells. No cellular atypia or mitotic figures were noted (Fig 2).

The patient was assessed 2 weeks postoperatively. The wound had healed and the sutures were removed. The previously noted telangiectatic vessels were now not clinically evident. There has been no recurrence of the lesion after 6 months of follow up.

COMMENT

Capillary haemangiomas are the most common congenital vascular tumours of the periorbital region.1 The majority of capillary haemangiomas appear over the first weeks or months of life. The natural history of the lesion is that of increasing size to 1 year of age and then gradual regression during the next 4–5 years. Intralesional corticosteroid injection is the current treatment of choice. Other treatment methods employed include excisional biopsy and carbon dioxide laser.

Acquired capillary haemangioma of the eyelid in an adult is a very rare occurrence and has been reported only once in the literature.1 Our diagnosis was made after exclusion of other similar presenting lesions. The differential diagnosis would include Kaposi’s sarcoma, cavernous haemangioma, angiosarcoma, varix, acquired tufted angiomia, and intravascular papillary endothelial hyperplasia.

Our choice of treatment proved to be very successful. The lesion was removed in its entirety and the postoperative cosmetic result was excellent. The technique was easy to perform for two reasons; firstly, the Ellman cutting diathermy allowed cutting precision and haemostasis was maintained throughout the procedure. The size of the capillary haemangioma and its location in this case made it essential to incise around the edge of the lesion accurately and therefore avoid the necessity for skin grafting. The Ellman cutting diathermy was particularly well suited to this purpose. We would recommend this method for the treatment of similar lesions.

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Figure 2 Low power histological section of mass stained with haematoxylin and eosin.

Figure 1 Pedunculated red mass of left upper eyelid.

Idiopathic sclerosing inflammation of the orbit: a new finding of calcification

Editor,—Idiopathic sclerosing inflammation of the orbit (ISIO) is a rare but well described condition.2 4 There is controversy as to whether it is a condition at one end of the spectrum of non-specific orbital inflammatory syndrome (pseudotumour)3 5 or whether it is...
A 75 year old woman with bilateral proptosis, marked photophobia, and severe ocular discomfort was referred to the ophthalmology service for an orbital biopsy. She was being investigated for an IgM paraproteinaemia, and the proptosis combined with a haematological disorder suggested orbital lymphoma. There was a history of hypertension and, at presentation, she also had a lower respiratory tract infection. She had a history of dry eyes, treated with hypromellose eye drops. On examination proptosis measured 23 mm on the right and 21 mm on the left. She had marked restriction of extraocular movement in all directions of gaze in both eyes (Fig 1). Visual acuity with pinhole was 6/9–1 right and 6/12+3 left. Often there are no features of active inflammation, and there may be aggressive local destruction of bone with invasion of adjacent structures including brain, meninges, and sinuses. Reliable differentiation from lymphoma is only possible with immunohistochemical studies. Management is difficult because of poor response to both steroids and radiotherapy. Kennerdell recommends early, aggressive treatment with surgery, steroids, radiotherapy, or a combination. Results of the use of systemic chemotherapeutic agents such as azathioprine and cyclophosphamide, in addition to the above modalities, have been reported by Rootman et al.

CASE REPORT

A 75 year old woman with bilateral proptosis, marked photophobia, and severe ocular discomfort was referred to the ophthalmology service for an orbital biopsy. She was being investigated for an IgM paraproteinaemia, and the proptosis combined with a haematological disorder suggested orbital lymphoma. There was a history of hypertension and, at presentation, she also had a lower respiratory tract infection. She had a history of dry eyes, treated with hypromellose eye drops. On examination proptosis measured 23 mm on the right and 21 mm on the left. She had marked restriction of extraocular movement in all directions of gaze in both eyes (Fig 1). Visual acuity with pinhole was 6/9–1 right and 6/12+3 left. Often there are no features of active inflammation, and there may be aggressive local destruction of bone with invasion of adjacent structures including brain, meninges, and sinuses. Reliable differentiation from lymphoma is only possible with immunohistochemical studies. Management is difficult because of poor response to both steroids and radiotherapy. Kennerdell recommends early, aggressive treatment with surgery, steroids, radiotherapy, or a combination. Results of the use of systemic chemotherapeutic agents such as azathioprine and cyclophosphamide, in addition to the above modalities, have been reported by Rootman et al.

Figure 1 Restriction of ocular movement in nine positions of gaze.

Figure 2 Computed tomography showing (A) calcification in the centre of the right orbit, and (B) bilateral calcification in the lacrimal fossae.
orbit. It also demonstrates and confirms orbital calcification as part of the disease process, and we recommend that idiopathic sclerosing inflammation should be considered in the differential diagnosis of calcification within the orbit.

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Therapy of subhyaloidal haemorrhage by intravitreal application of rtPA and SF gas

EDITOR—Subhyaloidal haemorrhage in the premacular space may cause a sudden loss of central vision in eyes, where macular function was good before the incidence. It can be caused by different disorders such as vitreoretinal traction of different origins, trauma, diabetic retinopathy, or occur spontaneously—for example, following partial detachment of the posterior hyaloid mem- brane. Different therapeutic approaches have been adopted for treatment of this situation. Spontaneous resorption of the haemorrhage can be awaited, which may be limited by the frequently slow course of resolution. Nd:YAG laser photodisruption of the posterior hyaloid membrane has been described to achieve dis- tribution of the haemorrhage in the vitreous, which resulted in accelerated clearing and visual improvement.1 Pars plana vitrectomy can be performed for complete surgical sepa- ration of the posterior hyaloid membrane and removal of the whole haemorrhage.

We report on a case of acute premacular subhyaloidal haemorrhage, which was treated successfully by subsequent injection of recombinant tissue plasminogen activator (rtPA) and sulphur hexafluoride gas (SF6).

CASE REPORT
A 55 year old healthy woman (apart from medically controlled arterial hypertension) presented with a 1 day history of acute decrease of central vision to 20/200 in her right eye. Visual acuity in the left eye was 20/20, and there was no history of other or previous ocular disorders. Funduscopy re- vealed a subhyaloidal haemorrhage in the right eye which extended between the tempo- ral vascular arcades (Fig 1). A small retinal area with intraretinal haemorrhages, retinal oedema, and epiretinal fibrovascular prolifera- tion following occlusion of a small venous branch above the temporal superior arcade could be identified as the origin of the haem- orrhage. Scatter laser photocoagulation was performed in the area of venous occlusion immediately. After 2 days, the surgical proce- dure was performed similar to the pneumatic displacement therapy of subretinal haemor- rhages: after peribulbar anaesthesia, ocu- lopression was applied twice for 10 minutes to reduce intraocular pressure. Then, 25 µg of rtPA (Actilyse, Boehringer-Ingelheim, Ger- many) were injected in the central vitreous cavity via pars plana. Following another two courses of oculopression, 0.3 ml of SF6, were injected in the vitreous cavity after 30 minutes. For further reduction of intraocular pressure, a limbal paracentesis was carried out and aqueous humour was released.

On the first postoperative day, detachment of the superior half of the posterior hyaloid membrane could be observed with diffuse intravitreal blood, and visual acuity had increased to 20/25. After 2 weeks, the fundus image was almost clear and the patient experi- enced no further visual impairment. During a 4 month follow up, visual acuity returned to 20/20. No increase in intraocular pressure was noted during the whole follow up period. At 4 months, fundoscopy showed regular findings except for the small area of venous occlusion (Fig 2), the crystalline lens showed no increase in opacification compared with the preoperative findings and to the other eye.

COMMENT
In many cases, subhyaloidal premacular haemorrhage demands therapeutic interven- tion. Although the finding theoretically can be awaited, which may be limited by the potential risks for further damage to ocular structures. To induce distribution of the premacular blood in the vitreous cavity and consequently accelerate clearing, Nd:YAG laser photodisruption of the posterior hyaloid membrane has been described.1 However, this form of treatment may result in damage to the underlying retinal tissue, especially in cases of thin subhyaloidal blood layers, of increased vitreous opacification, and of reduced patient compliance. Certainly, visual function can almost instantly be restored by pars plana vitre- cectomy with surgical separation of the poste- rior hyaloid membrane and evacuation of all blood. However, vitrectomy—even though a routine procedure—has numerous risks and side effects. The progression of lens nuclear sclerosis even after uneventful vitrectomy is a well known complication, which occurs in almost all cases. Intraoperative retinal breaks and postoperative proliferative vitreoretinopha- thy may result in Retinal detachment and severe loss of visual function. The intravitreal injection of rtPA and SF gas has recently been reported by different authors to induce pneu- matic displacement of subretinal haemorrhage in cases of age related macular degeneration.1

A significant reduction of central scotoma size with this comparably minimally invasive proce- dure has been pointed out, especially considering the minor side effects compared with the potential complications after vitrec- tomy with subretinal surgery.

To our knowledge this is the first report on the application of this surgical technique for the indication of central subhyaloidal haemor- rhage. The intravitreal injection of fibrinolytic agents such as urokinase to induce resolution of vitreous haemorrhage of different origins has already been described in the previtec- tomy era and has been investigated experimental-ly.1 More recently, induction of posterior vitreous separation by injection of rtPA has been shown both experimentally1 and clinically. Plasmin formed from vitreal plasminogen by rtPA breaks up extracellular matrix proteins of the hyaloid membrane, thus inducing separation. Additionally, these breaks may allow blood to pass the membrane even before further detachment, as we were able to see in our case, where diffuse distribu- tion of blood in the vitreous cavity had already.
ocurred 30 minutes after injection of rTPA. Vitreous separation and further distribution of blood is then promoted by the injected gas bubble, which constantly rolls across the posterior pole if prone positioning is maintained by the patient in the early postoperative period.

The low risk profile of the procedure has been pointed out for its use in subretinal haemorrhage. \(^1\) Retinal break formation after intravitreal gas injection is a well known complication. Therefore, before gas injection a thorough examination of the peripheral retina should be carried out to detect any pre-existing breaks or degenerations. The relatively small gas volume injected to achieve coverage of the posterior pole for this indication should further lower the risk of secondary break formation. Compared with vitrectomy, cataract formation or progression does not occur after intravitreal injection of fluid or gas as we know from retinal detachment surgery.

This case demonstrates the effective treatment of a dense central subhyaloidal haemorrhage by the injection of rTPA and SF.

The minimally invasive procedure resulted in restoration of useful visual function within a day after surgery and in recovery to full visual acuity within 2 weeks. No side effects could be attributed to the procedure compared with the potential risks of vitrectomy.

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Sebaceous gland carcinoma of the eyelid and palpebral conjunctiva in a patient with Muir-Torre syndrome

EDITOR—Sebaceous carcinoma (SC) of the eyelid and palpebral conjunctiva is rare. \(^1\) We present a patient with this entity and Muir-Torre syndrome (MTS).

MTS is characterised by the concurrent or sequential development of at least a single sebaceous gland tumour (adenoma, epithelioma, or carcinoma) with or without keratoacanthomas and a minimum of one internal malignancy. A recent review revealed that 163 cases fulfilling the diagnostic criteria for MTS have been reported since 1913, with 318 internal malignancies. \(^2\) Colorectal (47%), urogenital (21%) malignancies predominate, and nearly half the patients have two or more internal cancers. Our case is remarkable and was previously reported 7 years ago for presenting the highest number of malignancies described in this syndrome (eleven) and a prolonged survival of 26 years. \(^3\)

CASE REPORT

A 62 year old man came to our clinic with a diagnosis of MTS. \(^4\) In 1972 he had undergone a right hemicolectomy for undifferentiated Dukes' B adenocarcinoma plus adenomatous polyp; 16 months later, a colonoscopy showed metachronous carcinoma and a total colectomy was performed with end ileostomy for adenocarcinoma in a tubulovillous adenoma. \(^5\) In 1983 a sebaceous epithelioma had been resected from the left thigh. Between 1986 and 1987 four transurethral resections at the right ureter were performed for low grade tumours: partial excision of the right renal pelvis, a segmental left ureterectomy and two transurethral resections at the right ureteric meatuses.

In 1978 a segmental right retiurectomy was performed for transitional cell carcinoma (T1 No Mo). In 1983 a sebaceous epithelioma had been resected from the left thigh. Between 1986 and 1987 four transurethral resections at the right ureteric meatuses were performed for low grade tumours: partial excision of the right renal pelvis, a segmental left ureterectomy and two transurethral resections at the right ureteric meatuses.

In 1987 a radical right nephroureterectomy was performed for invasive carcinoma (T3a No Mo). In 1989 he was diagnosed with rectal carcinoma and an abdominopelvinic resection was performed (Dukes' B adenocarcinoma plus adenomatous polyp); 16 months later, a colonoscopy showed metachronous carcinoma and a total colectomy was performed with end ileostomy for adenocarcinoma in a tubulovillous adenoma. \(^6\) In 1993 a left upper lid. Physical examination disclosed a whitish, gelatinous, and vascularised papillomatous lesion on the tarsal conjunctiva located on the inner part of the left upper eyelid. It protruded as the eyelid was turned up, did not affect the free edge and was 0.5 cm long, with a base width of 0.2 cm (Fig 1). The patient also presented with more than 40 facial lesions (face and neck), each about 0.4 cm wide, and which had appeared in the previous year; they were rounded, well delimited, and adherent to deep tissues. One of them was located in the mid part of the lower eyelid, and did not affect the free edge. Both eyelid tumours were resected by excisional biopsy. The genetic study showed mutations in the hMSH2 gene (mut exon 14 ms/h2; del at 2239; codon 747).

COMMENT

MTS is a rare autosomal dominant disorder characterised by the association of sebaceous gland tumours with internal malignancies. This syndrome is now considered a subtype of the more common hereditary non-polyposis colorectal cancer syndrome (HNPPC). \(^7\) This last condition has been ascribed to mutations in the hMSH2 gene (mut exon 14 ms/h2; del at 2239; codon 747).

patients. \(^7\) The genetic study showed mutations in the hMSH2 gene in our patient. This case reveals the indolent course of multiple neoplasms in patients with MTS and the prolonged survival that is possible after surgical treatment.

The skin lesions may be the first sign in 41% of these patients, and in some cases the cutaneous tumours may precede the appearance of the internal disease by as many as 25 years, although more often they follow the diagnosis of at least the first visceral malignancy. \(^8\) Our case presented a sebaceous epithelioma in the left thigh in 1983, 11 years after the first malignancy and did not present SC on the eyelid, face and palpebral conjunctiva until 1998, 15 years later.

The mean age for the appearance of the skin tumours is 53 years (range 27–90 years) and the mean age for detection of the initial visceral neoplasm is 50 years; our case was diagnosed at ages 36 and 47 years old. The male:female ratio is 2:1.

Sebaceous carcinomas of the eyelid comprise 1–5% of all malignancies in this anatomic location and have been noted for their frequent metastatic potential. In contrast, extraocular sebaceous carcinoma is exceedingly rare, with fewer than 100 reported cases; metastatic spread is infrequent. \(^9\) We have found only one case of differentiated sebaceous carcinoma from the palpebral conjunctiva, which was described as uncommon. \(^9\) Therefore, this would be the second case reported in this location. There are 37 MTS patients with documented sebaceous gland carcinomas.

Histopathological diagnosis in our case was SC well differentiated in the lesion of the skin and SC moderately differentiated in the conjunctival lesion (Fig 2).

The treatment of sebaceous neoplasm is surgical excision. Complete removal of the tumour virtually eliminates the chance of recurrence in extraocular locations. Lesions of

Figure 1 Polyoid mass located in the temporal aspect of the tarsal conjunctiva without free edge affection.

Figure 2 (A) The tumour was composed of lobular or sheets of cells separated by a fibrovascular stroma. Focally the lesion showed a pagetoid spread of neoplastic cells through the metastatic conjunctiva (haematoxylin and cosin x100). (B) Neoplastic cells showed variable sebaceous differentiation, with finely vacuolated or foamy cytoplasm. Nuclei were large, with prominent nucleoli. Scattered mitoses were present (haematoxylin and cosin x400).

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the eyelid may have metastatic recurrence even with adequate initial excision.\textsuperscript{a} Patients with an MTS associated cutaneous lesion should have a complete evaluation for gastrointestinal or genitourinary cancers. Although the penetrance of this disease is variable, its autosomal dominant inheritance suggests that relatives should be screened for sebaceous gland tumours and internal malignancy and followed on a regular basis.\textsuperscript{a}
COMMENT

Two children underwent combined liver/kidney transplantation at 15 and 18 months of age for primary oxalosis (PH 1). Profound alteration of the retinal pigment epithelium was evident before transplantation. There was no progression of retinal oxalosis after transplantation. No recent visual acuity was 20/50 in each eye of the first patient and 20/40 and 20/100 in the second patient. No strabismus was noted. The prominent feature in both children was the bilateral symmetrical submacular RPE changes with extensive fibrosis (“white geographic maculopathy”). Neither child demonstrated any peripheral crystalline retinopathy or optic atrophy.

Primary hyperoxaluria type 1 (PH 1) is caused by a deficiency of the hepatic peroxisomal enzyme alanine:glyoxylate aminotransferase (AGT). This enzyme is encoded by the AGXT gene on chromosome 2q37.3. In the absence of AGT, glyoxylate is not adequately converted to less toxic metabolites and is instead metabolized to oxalate and glycolate. AGT has pyridoxal phosphate as its cofactor. In the majority of patients the disease results from a fucosidation of the normal gene product, but in one third of the patients there is a misrouting of the enzyme to the mitochondria instead of the peroxisomes.

Primary hyperoxaluria type 2 (PH 2) is due to a missense mutation of the mitochondrial enzyme glyoxylate reductase. The biochemical criteria for diagnosis include hyperoxaluria and t-glyceric aciduria.

Oxalosis has classically been included in the differential diagnosis of crystalline retinopathy. This differential includes Bietti’s crystalline dystrophy and cystinosis, though both of these also manifest corneal crystals. Additionally, talc and canthanthrene retinopathy should be included as well as methoxyhexanoyl toxemia and tamoxifen retinopathy. Intranretinal crystals have also been described in advanced stages of herporthromatocytoma, gyrate atrophy, and Stjoren-Larsson syndrome.

These children represent the product of treatment of the underlying metabolic problems by liver/kidney transplantation for the most severe form of infantile primary hyperoxalosis. Despite early intervention, these children developed white geographic maculopathy and poor vision. Previously, 15 ocular cases have been reported in the English literature citing the typical funduscopic picture of crystalline retinopathy, black geographic ringlet maculopathy, and optic atrophy.

Those previous reports indicated the maculopathy caused only mild, if any, visual impairment, whereas the worst vision was in those patients with optic atrophy. Unlike the previous reports, in our children their maculopathy was associated with poor vision in all but one eye, despite combined liver/kidney transplantation at an early age. It may be that our patients were younger and with a more severe disease. We postulate that the visual decrement is a result of subfoveal oxalosis. This one eye with good vision shows less subretinal fibrosis than the fellow eye with poor vision.

Successful hepatorenal transplantation has been followed by progressive and ultimately complete mobilisation of the oxalate deposits with resolution of the manifestations of systemic oxalosis including cardiomyopathy, cardiac dysrhythmias, and osteodystrophy. It may be that if there was not significant foveal involvement or optic nerve damage before transplantation these children could have never described an excellence in vision. Given that these two patients underwent liver/kidney transplantation by age 18 months old and yet have poor vision, it is imperative that treatment be initiated at the earliest possible stage.

The patients described above represent early involvement of retinal oxalosis with infantile PH 1, yet neither child demonstrated any crystals. It is important to the ophthalmologist to be cognizant of the “non-crystalline” retinopathy which is the result of calcium oxalate deposition and subsequent RPE reaction.

As more children will be diagnosed appropriately with newer molecular testing and with the increased success of combined liver and kidney transplantation, the ophthalmologist will be more involved with these patients and must be aware of the wide spectrum of ocular manifestations of oxalosis.

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References


Retinal telangiectasis and angiod streaks

EDITOR—As the name implies, angiod streaks may resemble blood vessels. Initially described by Doyne in 1889 as a case presentation to the Ophthalmological Society of the United Kingdom, there was consensus that these streaks were vascular in nature, and thus aptly named. It remained for Kofer in 1916 to correctly delineate the level of the streaks at Bruch’s membrane. Chloroidal neovascularization (CNV) has been reported as a complication of angiod streaks to occur in 70% to 86% of cases. Retinal telangiectasis, however, has never been described in association with angiod streaks.

Figure 1

CASE REPORT

A 66 year old white woman presented with gradual visual loss in her right eye for 2 months. Her best corrected visual acuity was right eye 20/40 and left eye 20/20. Slit lamp biomicroscopy of the anterior segment was normal. Fundus examination showed macular oedema in the right eye and prominent peripapillary atrophy with irregularly radiating streaks in the left eye (Fig 1). Fluorescein angiography disclosed bilateral peripapillary angiod streaks and juxtapfoveal retinal telangiectasis in the right eye. There was no evidence of CNV (Fig 2).

COMMENT

The development of CNV is a common finding in angiod streaks. Histopathological studies demonstrated linear breaks in Bruch’s membrane in addition to extensive calcification. These cracks in Bruch’s membrane may be bridged by a thin hypopigmented layer of retinal pigment epithelium, thus predisposing to the ingrowth of fibrovascular tissue from the choroid into the subpigment epithelial space in at least three out of four patients, usually occurring during the third to fifth decade of life.1

Despite the age of 66 years, the patient presented here did not show any evidence of CNV or related scaring. However, mild peripapillary streaks were present in both eyes. It has been reported previously that streaks with this appearance are not associated with fundus abnormalities or macular lesions typically observed in patients with pseudoxanthoma elasticum, such as reticular pigmented changes or peau d’orange appearance.2 Regarding the different clinical appearance and the benign course, these mild peripapillary streaks were considered a separate entity and have also been termed senile atrophic lines or pseudostreaks.3 Characteristically, in senile streaks, peripapillary helicoidal choroidal atrophy is often much more prominent than the streaks themselves (Figs 1 and 2).

References


Figure 1 Fundus photographic views of the right eye (A) shows macular oedema and biomicroscopically visible telangiectasis (arrow). In the left eye, prominent peripapillary atrophy and mild irregularly radiating streaks (arrows) are present (B).
The macular oedema of this 66 year old woman, however, resulted from leakage of juxtafoveal telangiectasis. A classification of idiopathic juxtafoveal retinal telangiectasis was proposed by Gass and coworkers in 1982, and updated in 1993. According to the biomicroscopic and fluorescein angiographic findings, three distinct groups of patients at risk were categorised—unilateral, non-familial, biomicroscopically visible telangiectasis with intraretinal exudation; bilateral, occult telangiectasis with minimal exudation and superficial retinal crystalline deposits; bilateral, biomicroscopically visible telangiectasis with minimal exudation and capillary occlusion, associated with systemic disease.

The aetiology of angiod streaks as well as of retinal telangiectasis remains unknown. Although hundreds of eyes with angiod streaks have been observed clinically and some of them have been studied histopathologically, the reason for calcification and for the development of cracks of Bruch's membrane remained unclear. Abnormal calcification of elastic tissue, a component of Bruch's membrane, is seen in other parts of the body in pseudoxanthoma elasticum and in Paget's disease. These two entities are the most common systemic association with angiod streaks, bilateral telangiectasis and angiod streaks may theoretically present together.

In our case, unilateral juxtafoveal telangiectasis caused macular oedema. There was no evidence of systemic disease or vascular occlusion. Therefore, regarding the various histopathological features and systemic diseases associated with angiod streaks and retinal telangiectasis, we assume that both clinical entities occurred in a coincidental way. It is important to note, however, that retinal telangiectasis can occur in an eye with angiod streaks, and it should not be confused with CNV. In elderly patients beyond the sixth decade of life, there is a senile form of angiod streaks which is unlikely to cause CNV, but may be associated with retinal telangiectasis.

**CASE REPORT**

A 33 year old white woman presented with a painless red swelling of the medial left lower lid (Fig 1A) that had been present for several weeks. There was an obstruction of both lower and upper canalicus and also mild oedema of the upper lid. On palpation, the swelling was presented as a remarkably firm subcutaneous mass extending deep into the medial canthal area. Otherwise, all ocular findings were normal. The patient's previous ocular history was unremarkable apart from a left sided refractory dacryocystitis at age 3 that had been investigated and treated at another university eye department. The parents had documented refractory dacryocystitis at age 3 that had been investigated and treated at another university eye department. The parents had documented refractory dacryocystitis at age 3 that had been investigated and treated at another university eye department. The parents had documented refractory dacryocystitis at age 3 that had been investigated and treated at another university eye department.

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Wound healing was without any complications, and the cosmetic result was excellent. On repeat probing, the canalicus was now patent, and no recurrence was seen up to 2 years after the surgery. In contrast with our histopathological expectations, a biochemical analysis of the excised tissue using infrared, mass, and nuclear magnetic resonance spectroscopy...
This patient presented with a lipid-rich lesion similar to the so-called paraffin granuloma which represents an inflammatory reaction to exogenous lipid. Usually, exogenous lipid gets access into the tissue in the form of ointments used in or in close vicinity to the eye. In contrast with paraffin, however, the triglyceride esters were analysed in our patient are not usually present in ophthalmic ointments. Triglycerides are rather a constituent of naturally occurring lipids and can be expected—for example, in fat necrosis after trauma. Our patient did not exhibit any features of a pre-existing lipomatous lesion such as, for example, a lipomerdor; moreover, there was no history of trauma or mechanical irritation. Thus, the most likely explanation for the presence of a lipogranuloma remains a “complication” from the treatment of her dacryocystitis 30 years ago. Various lipid based substances have been used for rinsing of, and instillation into, the canaliculus or lacrimal sac. These ointments, however, are usually also based on paraffin or Vaseline. Other lipid based materials have been employed as contrast material for viewing the lacrimal passage. One of the substances that has been commonly used for contrast dacryocystography is Lipiodol, an iodised poppy seed oil which is a characteristic mixture of glyceric esters of various fatty acids including mainly oleic, linoleic, linolenic, palmitic, and stearic acid (information from Byk Konstanz). Thus, the lipid composition of Lipiodol corresponds remarkably well to the mixture that was analysed in our specimen. The iodine present in the original substance can be expected to have been removed and transferred to the thyroid, and, with endogenous fat and ointments exhibiting somewhat different components, there is convincing evidence that Lipiodol can indeed be regarded as the initiating agent. Similar problems after instillation of other lipid based substances into the lacrimal drainage system and one case of a granulomatous inflammation initiated by the application of a lipid based contrast medium to the orbit have been reported but the time between the original “insult” and the development of an inflammatory reaction was always much shorter. This suggests that, in our case, a minor injury to the canaliculus or lacrimal sac might have occurred, allowing only a very small amount of lipid based material to reach the surrounding tissues and cause a self-propagating inflammatory process. As triglycerides are much more similar to human body fat than paraffin, one could speculate that this might further help to explain the unusually long time lapse between the primary application and the clinically relevant granulomatous reaction seen in our patient.

We are very grateful to Dr S Moss, Novartis, Switzerland, for performing the biochemical analysis.

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Figure 1 (A) Clinical appearance at first presentation, showing a red subcutaneous swelling (indicated by arrows) in the left medial canthus extending into the lower lid. The left upper eyelid also appears somewhat oedematous. (B) Intraoperative appearance of the lesion, revealing yellowish lipid-like tissue of firm consistency. Note that the tumour is infiltrating the surrounding tissues without evidence of a capsule or pseudocapsule. (C) Histology shows connective tissue with numerous lipid vacuoles of different sizes that are mostly surrounded by multinucleate giant cells. Note also the dense chronic inflammatory cell infiltrate. No genuine orbital fat is seen in this section. (Paraffin section, haematoxylin and eosin, x170).
subhyaloid membrane owing to the poorly applied contact lens and cataract. Therefore, we decided to perform intravitreal tPA and C$_3$F$_8$ injection under topical anaesthesia. Twenty minutes after injecting intravitreal 0.1 ml 0.1 ml tPA (total dose of 25 µg), 0.5 ml of 100% C$_3$F$_8$ was injected into the vitreous cavity. A paracentesis was done to decrease the intraocular pressure. The patient was told to maintain the face down position for 2 weeks. Three days after the injection, the subhyaloid haemorrhage was displaced by the gas bubble out of the macular region. The haemorrhage slowly decreased in size over 2 weeks, and then markedly decreased. After 2 months, the subhyaloid haemorrhage had completely cleared (Fig 2). Her vision in the right eye increased to 20/70 on her last visit.

COMMENT
Although the subhyaloid haemorrhage was somewhat old and very thick, it was rapidly displaced out of the macular region within 3 days. We suggest that tPA worked to lyse the blood clot. The vision of 20/70 may be attributed to cataract and retinal damage caused by the subhyaloid haemorrhage.

Tissue plasminogen activator and C$_3$F$_8$ injection seems to be an alternative way to clear the subhyaloid haemorrhage especially in retinal and retinal pigment epithelium (RPE) function is not clear. In addition, factors that determine the progression of the vitelliform foveal lesion leading to impairment of visual acuity in patients with BVMD are not understood. We present a case in which blunt trauma was associated with deterioration of visual acuity and macular scar formation in a patient with BVMD.

CASE REPORT
A 14 year old male presented to our clinic after being hit in his right eye by a fist 40 days earlier. He complained of reduced visual acuity in the right eye increased to 20/70 on her last visit.

Blunt trauma in Best's vitelliform macular dystrophy

EDITOR,—Recently, the association of Best's vitelliform macular dystrophy (BVMD) with mutations in chromosome 11 has been reported, and candidate genes that may be affected by these mutations have been identified. However, the role of these genes in retinal and retinal pigment epithelium (RPE) function is not clear. In addition, factors that determine the progression of the vitelliform foveal lesion leading to impairment of visual acuity in patients with BVMD are not understood. We present a case in which blunt trauma was associated with deterioration of visual acuity and macular scar formation in a patient with BVMD.

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orescence in the area where the subretinal material was previously present (Fig 2A). In the left eye, early hypofluorescence and late staining of the foveal vitelliform lesion were seen (Fig 2B).

COMMENT
Submacular haemorrhage has been previously reported in several cases of BVMD during the natural course of the disease. In addition, Benson et al reported in 1975 a single case of a child with BVMD who had blunt trauma complicated by rupture of the vitelliform lesion, subretinal haemorrhage, scarring of the fovea, and severe reduction of visual acuity. In an attempt to explain the occurrence of such haemorrhages in BVMD, it was suggested that a metabolic abnormality causes the retinal pigment epithelium and Bruch’s membrane in these patients to be especially vulnerable to rupture, with resultant subretinal bleeding from the choriocapillaris. Interestingly, one of the recently identified candidate genes for BVMD is indeed expressed in the RPE.

Our patient had the typical clinical and electrophysiological findings of BVMD. His visual acuity deteriorated in one eye after he suffered blunt trauma that resulted in damage to the retinal pigment epithelium and Bruch’s membrane, as evidenced by subretinal bleeding and scarring of the fovea. The yellowish subretinal material that was later absorbed may also represent remnants of dehaemoglobinised subretinal haemorrhage (the patient was first examined 40 days after the trauma). Alternatively, this yellowish material may have originated from the vitelliform lesion ruptured by the trauma. The visual acuity deteriorated following this rupture with formation of a fibrous foveal scar.

The present case and the case reported by Benson et al demonstrate that the consequences of blunt trauma in BVMD can be devastating. Vitelliform lesion disruption, haemorrhage and, perhaps, choroidal ruptures may have otherwise persisted for many years. In some cases, visual acuity may have originated from the vitelliform lesion ruptured by the trauma. The visual acuity deteriorated following this rupture with formation of a fibrous foveal scar.

Epidermoid carcinoma arising in an ocular leishmaniasis lesion

Emerston.—American tegumentary leishmaniasis is an endemic disease occurring in Latin America, especially in Brazil. The agent is the protozoan Leishmania, transmitted by sand flies. Leishmania v braziliensis causes the mucocutaneous form of the disease, in which systemic dissemination to mucous membranes follows the primary ulcerative skin lesion.

We report a patient with nasal and conjunctival mucous leishmaniasis who developed an epidermoid carcinoma in the orbit.

CASE REPORT
A 58 year old man presented with a painless conjunctival mass of 2 months’ duration. He reported nasal and mouth wounds and nasal flattening for 3 years and had a depressed scar in his right leg caused by an ulcer that developed 6 years before. He had been treated with N-methylglucamine antimonials 2 years before and underwent nasal reconstruction 1 year later. The patient had been a smoker and an alcoholic for 30 years and lived in an endemic region for tegumental leishmaniasis.

Slit lamp examination revealed a lower left conjunctival mass eroding the walls of the left maxillary sinus, zygomatic arch, and orbital floor (Fig 1D). Computed tomography revealed a solid mass eroding the walls of the left maxillary sinus, zygomatic arch, and orbital floor (Fig 1D). The patient underwent left maxillectomy and orbit exenteration. One year after surgery, the patient presented with partial dehiscence of the frontal flap, a local biopsy showing carcinoma recurrence. Face magnetic resonance did not show signs of tumour. The patient did not return to the hospital.

COMMENT
The upper airway is the most affected site in mucocutaneous leishmaniasis (MCL), and
extensive destruction of nasal mucous membranes and cartilage, invasion of the face sinuses, oral cavity, larynx, and pharynx may occur. Diagnosis is confirmed by the demonstration of intracellular leishmania amastigotes in Giemsa stained slit skin smears, although the parasite may be difficult to find in chronic lesions. Immunohistochemistry for leishmania antigens is an important diagnostic tool. Histopathological analysis discloses features ranging from inflammatory infiltration of mononuclear cells and neutrophils to a granulomatous reaction. Pentavalent antimony is the drug of choice for the treatment of all types of leishmaniasis. In resistant cases, amphotericin B or pentamidine isothionate is indicated.

Cutaneous leishmaniasis of the eyelid is the most common ophthalmological finding in oriental and occidental cutaneous leishmaniasis, and conjunctival involvement may appear as an associated or isolated finding. Other ocular features in MCL include interstitial keratitis, iridocyclitis, and chronic dacryocystitis.

In the case reported here, the malignant tumour may have been present in the initial condition, but the diagnosis of MCL and the favourable response to treatment meant that a more extensive search for an additional diagnosis was not carried out. The development of a neoplasm at the site of a previous dermal scar is a well recognised phenomenon. Basal cell carcinomas have been reported to arise from a previous cutaneous leishmaniasis lesion. To our knowledge, this is the first case of development of a squamous cell carcinoma from a previous mucous leishmania lesion with major involvement of the eye.

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