Specular microscopic findings of corneal deposits in patients with Bietti's crystalline corneal retinal dystrophy

WADA YUKO, NAKAZAWA MITSURU, ABE TOSHIAKI, SHIONO TAKASHI, TAMAI MAKOTO

Journal or Publication Title: British Journal of Ophthalmology
Volume: 83
Number: 9
Page Range: 1095-1095
Year: 1999
URL: http://hdl.handle.net/10097/51506

doi: 10.1136/bjo.83.9.1088k

<table>
<thead>
<tr>
<th>著者</th>
<th>三代恒雄, 仲澤智也, 安藤俊明, 輔野達昭, 田上真樹</th>
</tr>
</thead>
<tbody>
<tr>
<td>タイトル</td>
<td>Specular microscopic findings of corneal deposits in patients with Bietti's crystalline corneal retinal dystrophy</td>
</tr>
<tr>
<td>依頼者</td>
<td>British Journal of Ophthalmology</td>
</tr>
<tr>
<td>年</td>
<td>1999</td>
</tr>
<tr>
<td>月</td>
<td>9</td>
</tr>
<tr>
<td>ページ</td>
<td>1095-1095</td>
</tr>
<tr>
<td>URL</td>
<td><a href="http://hdl.handle.net/10097/51506">http://hdl.handle.net/10097/51506</a></td>
</tr>
<tr>
<td>doi</td>
<td>10.1136/bjo.83.9.1088k</td>
</tr>
</tbody>
</table>
LETTERS TO THE EDITOR

Surgically removed submacular nematode

Editor,—Intraocular filariasis is an uncommon disease in civilised countries. In only a few cases has the worm been surgically removed from the posterior portion of the eye and identified. In this report we describe a patient who had a filaria-like worm in an epimacular and/or submacular lesion, which was successfully removed surgically.

CASE REPORT
A 38 year old woman complained of decreased visual acuity in her left eye of 5 days’ duration. Her medical history was unremarkable. The patient had two pet dogs, and she had not travelled to foreign countries in recent years. On examination, her corrected visual acuity was 20/20 in the right eye and 20/100 in the left. No inflammation was present in the anterior segment or vitreous cavity bilaterally. Ophthalmoscopic examination disclosed a white worm, approximately 3 disc diameters in length, equivalent to about 4.5 mm, moving slowly in the macula at the epi retina. Epiretinal and intraretinal haemorrhages were observed in and around the macular region. The body of the worm was tapered at one end and slightly rounded at the other (Fig 1). A round, preretinal haemorrhage was observed on a branch of the superonasal retinal artery, which could have been the route of entry into the eye. Numerous subretinal hypopigmented tracks with small haemorrhages were noted in the superior retina, and perivascular haemorrhages were observed around the inferior branch of the central retinal vein (Fig 2). Fluorescein angiography showed numerous hyperfluorescent tracks without dye leakage. The patient’s blood test revealed a slightly increased white blood cell count (10 200 x10/3; normal <9000) and elevated IgE in the serum (680 U/ml; normal <250).

On the following day pars plana vitrectomy was performed and the worm was found partially moved into the subretinal space of the macula. The worm was aspirated successfully through a 20 gauge silicone tipped needle and submitted for parasitological study; however, it was lost during transportation to a different laboratory. Parasitological study of the patient’s serum detected an antibody to Dirofilaria. Six months after surgery, the patient’s visual acuity was still 20/50. Ophthalmoscopically the hypopigmented tracks had faded, and mildly irregular pigment was observed at the deep retina in the macular lesion. Cone and rod electroretinograms to full field stimuli were normal bilaterally.

COMMENT
Our patient owned two dogs, and her ocular findings could be differentiated from toxocariasis, in which only larvae of Toxocara canis can infect humans. The length of the Toxocara larva is about 400 µm, and grows no longer. Although no direct microbiological evidence of Dirofilaria infection was obtained in our patient, a positive antibody to Dirofilaria in the serum indicated its infection. There have been many reports of intraocular filariasis; however, the filariae were removed and identified in only six of 56 cases reviewed by Beaver in 1989. Direct parasitological microscopic examination is necessary for accurate diagnosis, but serological study can be helpful, as shown in the present case.

Various types of management for intraocular parasites have been reported. Direct photocoagulation to the worm body has been successfully reported in cases with filaria-like worms and in one case with insect parasites. It has been suggested that photocoagulation denatures the parasite proteins and mitigates the immune reaction. If the parasite is located in the posterior pole of the retina, however, photocoagulation may cause permanent visual impairment, and surgical removal should be selected. Furthermore, photocoagulation would make parasitological identification impossible. Periretinal or subretinal parasites were retrieved successfully by pars plana vitrectomy in several reported cases. In the present case, the visual acuity was still 20/50 6 months postoperatively. The migrating worm may have caused considerable damage to macular function, therefore, we believe that intraocular parasites should be removed as soon as possible.

The authors are grateful to Dr Keizo Yamaguchi for parasitological examination and Mr Shingo Yamazaki for fundus photographs.

SHUICHI YAMAMOTO
MASANORI HAYASHI
SHINOBU TAKEUCHI
Department of Ophthalmology,
Toho University Sakura Hospital, Japan

Correspondence to: Shuichi Yamamoto, MD, Department of Ophthalmology, Toho University Sakura Hospital, 564-1 Shimoshrui, Sakura, Chiba 288-8741, Japan.

Accepted for publication 19 July 1999


Acanthamoeba keratitis occurring with daily disposable contact lens wear

Editor,—Up to 92% of cases of acanthamoeba keratitis occur in contact lens users of all types. Failure to disinfect soft contact lenses and the use of chlorine disinfection systems are major factors accounting for the increase in cases of acanthamoeba keratitis. It is thought that “daily disposable” contact lenses, which require no disinfection, will have a much lower risk for the development of all forms of infectious keratitis. We report a case of acanthamoeba keratitis occurring in a daily disposable contact lens wearer. We believe this is the first such reported case.

CASE REPORT
A healthy 21 year old woman, who had been wearing daily disposable contact lenses for 1 year, wore her lenses for 4 hours on 1 day and then stored the lenses in preserved saline solution overnight in a new contact lens case. She re wore the lenses for a further 3 hours the following day. She reports that this was the first time that she had done this. She then developed a painful left eye. This was initially treated as conjunctivitis by her general practitioner and local accident and emergency department. After 2 weeks her local eye unit suspected acanthamoeba keratitis and performed an epithelial scrape that “revealed amoeba species”. She was then referred to our unit for opinion.

On examination her visual acuity was reduced to 6/18 in the left eye. There was conjunctival injection and a mild scleritis. The cornea showed diffuse punctate staining with linear epithelial infiltrates.

Figure 1 The cornea showing diffuse punctate staining with linear epithelial infiltrates.
Corneal infection can occur. Disinfection and contaminated lens cases were introduced in 1995. When used properly the lens is discarded after 1 day’s wear only, though some corneal infiltrates remained.

**COMMENT**

Acanthamoeba are free living protozoa commonly found in soil and water, including unrelated sources, such as exposure to peripapillary capillaries. Indocyanine green (ICG) angiography showed slight hypofluorescence of the macular area at a late phase. Analysis of the mean deviation in HVF over the clinical course corresponded with the visual acuity (Fig 1, bottom). In addition, m-ERG (Veris III, Tomey, Nagoya, Japan) was analysed during the clinical course. In this examination, the fundus was divided into four foci and the sum of amplitudes in each group was measured (Fig 2, top). In the left eye, the sum of amplitudes was altered individually but all of them were affected during the clinical course. Only in the inferior temporal area did it correspond with HVF. The values in the right eye were about 5000 μV in each focus. This is almost the same as normal volunteers in our clinic (data not shown).

**Clinical course of acute zonal occult outer retinopathy in visual field and multifocal electroretinogram**

Edmonton.—Patients with acute zonal occult outer retinopathy (AZOOR) may present with a normal fundus examination and almost normal fluorescein angiography (FA), despite severe loss of visual field and electroretinogram (ERG) abnormalities. The lesion defined zones of the retinal receptor cells; however, the course of the disease remain unclear. There is also no established conclusion about progression of visual field loss.

**CASE REPORT**

A healthy 26 year old woman presented to our outpatient clinic complaining of acute onset of visual disturbance in her left eye. Her corrected visual acuity was 20/20 in her right eye, and 20/100 in her left. The pupils were equal and reactive normally. Slit lamp and fundus examination, computed tomography, magnetic resonance imaging scan, and general examination were normal. HVF 30–2 demonstrated blind spot enlargement breaking out to the inferotemporal periphery in the left eye (Fig 1, top). Full field ERG showed grossly reduced A and B waves in the left eye. The FA showed slight leakage from peripapillary capillaries.

**Figure 1** (Top) Raw images of Humphrey 30–2 visual fields in the left eye. (Bottom) The relation between clinical course and mean deviation (MD) of Humphrey 30–2 visual field and visual acuity in the left eye. Asterisks indicate the day in which multifocal ERG was analysed. The roman numerals correspond with raw images in the upper part of the figure.
The findings of AZOOR that we observed in our patient suggest that the retinal recovery assessed by m-ERG was different for the lesion (main focus was related to the infero-temporal retina) and was delayed compared with visual acuity and HVF. Perhaps this delayed retinal recovery reflects a subtle microcirculatory disturbance that cannot be clearly detected by FA or ICG. Slight leakage in FA and slight hypofluorescence on the late phase in ICG would suggest such a microcirculatory disturbance.

When better understanding of the aetiology and pathophysiology of AZOOR is available, the clinical response to appropriate therapy may perhaps best be followed by m-ERG.

KANAKO YASUDA
MASAHIKO SHIMURA
MICHIRU NORO
MITSURU NAKAZAWA
MAKOTO TAMAI
Department of Ophthalmology, School of Medicine, Tohoku University, Sendai 980-8574, Japan

Correspondence to: Masahiko Shimura, MD, Department of Ophthalmology, School of Medicine, Tohoku University, Sendai 980-8574, Japan.

Accepted for publication 26 March 1999

Hypotonic maculopathy following pneumatic retinopexy: a UBM study

KANAKO YASUDA
MASAHIKO SHIMURA
MICHIRU NORO
MITSURU NAKAZAWA
MAKOTO TAMAI
Department of Ophthalmology, School of Medicine, Tohoku University, Sendai 980-8574, Japan

Accepted for publication 26 March 1999

Figure 2 (Top left) In the multifocal ERG, the fundus was divided into four foci. (Top right) Sum of the amplitudes in each foci was altered during the clinical course. (Bottom) The three dimensional topography and sum of the amplitudes in each of four foci of the m-ERG were indicated.

Figure 1 UBM picture of right eye showing the external part of the dehiscent cataract wound, as a narrow slit, before repair (area between arrows). There is a full blown hypotonous maculopathy in this eye, with VA 20/200.

Below October 1997 and November 1997 he was presented to our unit on three occasions complaining of further deterioration of vision to 20/200, IOPs between 3–5 mm Hg, and a full blown hypotonous maculopathy. After 3 months of hypotony of “unknown origin”, a diagnosis was made by a glaucoma specialist using indentation gonioscopy revealing a dehiscence of the cataract wound. A preoperative UBM study confirmed the presence of an internal wound gap, behind the limbus, superotemporally, 3 mm in circumference. Ultrasonically the wound dehiscence was depicted as a narrow slit (Fig 1) with a flat inadvertent bleb above which was not apparent clinically. A surgical repair of the wound was decided upon. Intraoperatively no definite dehiscence could be clinically identified. Balanced salt solution through a 30 gauge needle was repeatedly injected under pressure to the anterior chamber but this failed to localise any suspicious area. At this point, based on the ultrasonic study, two 10-0 nylon interrupted sutures were placed through the sclera parallel to the limbus in the suspicious area. These bites were moderately deep in an attempt to engage the internal flap of the cataract wound.

Two weeks postoperatively, the pressure normalised (IOP 12 mm Hg), maculopathy was reversed, and visual acuity improved to 20/50.

Hypotonic maculopathy is an unusual and, to our knowledge, the first reported complication of this kind after pneumatic retinopexy. We hypothesise that the original cataract wound did not heal properly and the additional cryoprobe manipulation caused the

CASE REPORT

A 63 year old white man, complaining of a shadow in his right visual field, was referred to our department in April 1997 for further management. Ocular history was notable for bilateral extracapsular cataract extraction with posterior chamber implant. Ultrasound biomicroscopy (UBM) proved to be a valuable adjunct in both the diagnosis and management of this complicated case.

Between October 1997 and November 1997 he was presented to our unit on three occasions complaining of further deterioration of vision to 20/200, IOPs between 3–5 mm Hg, and a full blown hypotonous maculopathy. After 3 months of hypotony of “unknown origin”, a diagnosis was made by a glaucoma specialist using indentation gonioscopy revealing a dehiscence of the cataract wound. A preoperative UBM study confirmed the presence of an internal wound gap, behind the limbus, superotemporally, 3 mm in circumference. Ultrasonically the wound dehiscence was depicted as a narrow slit (Fig 1) with a flat inadvertent bleb above which was not apparent clinically. A surgical repair of the wound was decided upon. Intraoperatively no definite dehiscence could be clinically identified. Balanced salt solution through a 30 gauge needle was repeatedly injected under pressure to the anterior chamber but this failed to localise any suspicious area. At this point, based on the ultrasonic study, two 10-0 nylon interrupted sutures were placed through the sclera parallel to the limbus in the suspicious area. These bites were moderately deep in an attempt to engage the internal flap of the cataract wound.

Two weeks postoperatively, the pressure normalised (IOP 12 mm Hg), maculopathy was reversed, and visual acuity improved to 20/50.

Hypotonic maculopathy is an unusual and, to our knowledge, the first reported complication of this kind after pneumatic retinopexy. We hypothesise that the original cataract wound did not heal properly and the additional cryoprobe manipulation caused the...
wound to leak. The UBM study provided us with an interesting insight into how the scleral suture repair may have worked to correct the wound leak as shown in Figure 2. The fact that hypotony resolved after suturing the wound, indicated that the external part of the wound was secure (no slit is apparent) despite the fact that the internal part of the wound was gaping even more postoperatively. This finding implies, therefore, that only minimal overall alteration of the wound architecture postoperatively, sufficient to rectify the leak. We believe that UBM is a valuable adjunct in the management of similar cases by clearly identifying both the presence and exact location of leak. Finally, pneumatic retinopexy should be performed with caution, especially in cases of previously operated eyes with large incision wounds.

**CASE REPORT**

A 26 year old man complained of gradual and painless diminution of vision in both eyes, right more than left, for the past 2–3 years. There was a history of an overhead high tension electric transmission cable accidentally falling on the patient’s head 4–5 years earlier. This had resulted in immediate burn to the scalp. The patient was visually asymptomatic till about 1 year after the mishap, when he began to notice the gradual fall in vision that had progressed to its present state.

Corrected visual acuity was 20/200 right eye and 20/60 left eye. A 15 × 2.5 cm linear, sagittal scar extending from the frontal to the occipital region of the head was noticed. The lids, conjunctiva, cornea, and pupils showed no abnormality in either eye. Fundus examination was unremarkable. Slit lamp examination revealed multiple, mid-peripheral snowflake-like anterior subcapsular lens opacities in both eyes, right greater than left. In the right eye some of these opacities were seen encroaching into the visual axis and additionally a few posterior subcapsular opacities were noticed (Fig 1).

In review of the history of electrical injury and classic location and typical appearance of the lens opacities, a diagnosis of bilateral electric cataract was made. Extracapsular cataract extraction (ECCE) with posterior chamber intraocular lens implantation was undertaken in both eyes, right eye first followed 3 months later in left eye. The intraoperative and postoperative course were uneventful and the patient has achieved corrected visual acuity of 20/20 in both eyes.

**COMMENT**

Involvement of the lens exclusively, sparing other ocular structures is rare. This case documents such a possibility and also highlights the salient features involving electric trauma to the lens. The scalp burn in this case represents the entrance wound for the electrical energy but the lack of an exit wound makes this case particularly peculiar. Both entry and exit sites for the electric current have been reported by all previous authors.

The excellent surgical results noted in both eyes of this patient are in keeping with the similar result reported by Portellos et al. This observation should encourage the ophthalmologist to undertake surgery for electric cataract, where necessary, without any undue concern.

**Figure 1** The characteristic anterior subcapsular lens opacities. (A) Right eye, (B) left eye.

**Figure 2** Cross section through temporal artery showing disrupted internal elastic lamina (arrow).

**Letters**

**Temporal artery biopsy specimens**

**EDITOR,—**Temporal artery biopsies are performed routinely on patients suspected of having giant cell arteritis. Of 131 pathology specimens examined at University of Illinois at Chicago Eye Center from 1975 to 1998, the most common diagnosis was atherosclerosis with myointimal fibrosis (63%) followed by giant cell arteritis (13%). In about 6% of cases we encountered calcific sclerosis confined to the tunica media which was associated with mild tissue disorganisation surrounding the calcific plaque and disruption of the internal elastic lamina (Fig 1).

Monckeberg’s sclerosis as seen in these specimens was first described by Monckeberg in 1903. It commonly affects medium size muscular arteries and is described in femoral, tibial, radial, coronary, cerebral, and visceral arteries. However, its association with the temporal artery is uncommon. The infrequent occurrence of this condition in the temporal artery and the presence of a fragmented inter-

**Monckeberg’s sclerosis in temporal artery biopsy specimens**

**EDITOR,—**Temporal artery biopsies are performed routinely on patients suspected of having giant cell arteritis. Of 131 pathology specimens examined at University of Illinois at Chicago Eye Center from 1975 to 1998, the most common diagnosis was atherosclerosis with myointimal fibrosis (63%) followed by giant cell arteritis (13%). In about 6% of cases we encountered calcific sclerosis confined to the tunica media which was associated with mild tissue disorganisation surrounding the calcific plaque and disruption of the internal elastic lamina (Fig 1).

Monckeberg’s sclerosis as seen in these specimens was first described by Monckeberg in 1903. It commonly affects medium size muscular arteries and is described in femoral, tibial, radial, coronary, cerebral, and visceral arteries. However, its association with the temporal artery is uncommon. The infrequent occurrence of this condition in the temporal artery and the presence of a fragmented inter-

**Figure 1** Cross section through temporal artery showing disrupted internal elastic lamina (arrow) and large calcific plaques (C) in the tunica media (haematoxylin and eosin; original magnification ×40).
nal elastic lamina should not be erroneously interpreted as sequelae of previous arterial inflammation. The pathophysiology of Monckeberg’s arteriosclerosis is still unclear, but it can be induced in animal models by injecting adrenalin, nicotine, parathyroid hormone, and vitamin D. In addition, lumbar sympathectomy has been shown to promote occurrence of Monckeberg’s arteriosclerosis of the lower extremities in humans. Automatic dysfunction from diabetic neuropathy is thought to be responsible for the occurrence of Monckeberg’s in diabetic patients. Unlike atherosclerosis, Monckeberg’s arteriosclerosis is a benign condition and does not cause vascular thrombosis. In conclusion, Monckeberg’s arteriosclerosis of the temporal artery may be seen occasionally in the temporal artery. It is an interesting histological diagnosis that has little clinical significance but can be recognized in temporal biopsy specimens.

BIENVENIDO V CASTILLO JR
ELISE TORCZYNSKI
DEEPAK P EDWARD
Department of Ophthalmology, University of Illinois at Chicago, IL, USA

Correspondence to: Dr Deepak P Edward, Department of Ophthalmology, University of Illinois at Chicago, 1855 W Taylor Street, Chicago, IL 60612, USA.

Accepted for publication 9 April 1999


Retinal vascular abnormality in Poland’s syndrome

EDITOR,—Poland’s syndrome is a congenital anomaly first described in 1841 consisting of unilateral hypoplasia or aplasia of the pectoralis major muscle and ipsilateral upper extremity abnormalities which often include ipsilateral syndactyly. Since then a number of associated anomalies have been reported. These include absence of the pectoral minor muscle, absence or atrophy of ipsilateral ribs two to five, aplasia of the ipsilateral breast or nipple, and simian crease of the a finger. Though we are not aware of direct evidence supporting this hypothesis.

To our knowledge, this is the first reported case of coexistence of juxtafoveal telangiectasis, renal agenesis, and Poland’s syndrome. Although we are not aware of direct evidence reported in the literature indicating any retinal involvement linked to this syndrome, it is plausible that the three anomalies found in this patient may have common original vascular causative factors. Therefore, we recommend a careful eye fundus examination of patients presenting with this syndrome.

CASE REPORT

We examined a 39 year old man previously diagnosed with Poland’s syndrome who came to our clinic because he had experienced blurred vision in the right eye for 2 months. Computed axial tomography, arteriography, and abdominal echography revealed absence of the left kidney. There was no history of diabetes or hypertension. The ophthalmological examination of the right eye revealed a visual acuity of 20/50 and a paracentral relative scotoma. The right eye fundus showed pericentral capillary telangiectasis, retinal vascular distortion, moderate perimacular hard exudates, and retinal swelling, all of them more prominent in the temporal perimacular area. Fluorescein angiography clearly showed the vascular abnormalities (Figure 1). The left eye fundus was normal. A diagnosis of juxtafoveal retinal telangiectasis was made.

Figure 1 Early venous phase fluorescein angiogram of the right eye, macular area. Note the easily visible telangiectatic retinal capillaries (arrows). These abnormal vessels showed significant leakage of fluorescein in the late phase of the angiogram.

This work was partially supported by grants XUGA-20082897 and DGESIC PB97-0521.

FRANCISCO GOMEZ-ULLA
Servicio de Oftalmología, Facultad de Medicina, Complejo Hospitalario Universitario de Santiago, E-15705 Santiago de Compostela, Spain

FRANCISCO GONZALEZ
Servicio de Oftalmología y Departamento de Fisiología, Facultad de Medicina, Complejo Hospitalario Universitario de Santiago, E-15705 Santiago de Compostela, Spain

Correspondence to: Dr Francisco Gomez-Ulla or Dr Francisco Gonzalez, Servicio de Oftalmología,

Compleso Hospitalario Universitario de Santiago, c/Caballos nº E-15705 Santiago de Compostela, Spain.

Accepted for publication 9 April 1999

Latanoprost is a recent addition to the medical management of raised intraocular pressure in chronic open angle glaucoma and ocular hypertension. It is a potent ocular hypotensive agent with few ocular or systemic side effects.

We report a case of bilateral optic disc oedema developing soon after commencing treatment with latanoprost which resolved once therapy was stopped.

CASE REPORT

An asymptomatic 64 year old woman presented with raised intraocular pressure. She maintained good general health, had no significant JNB, Weaver or family history, and was not on any medication. Snellen visual acuities were 6/5 both eyes. The intraocular pressures were 28 mm Hg right eye and 26 mm Hg left eye. Ocular examination was otherwise unremarkable with open angles, normal optic nerves, and full Humphrey 24-2 visual fields. She was thus diagnosed as having ocular hypertension and consented to enter a prospective double masked trial comparing some of the intraocular pressure lowering drugs. Therapy was commenced with one of the drugs involved in the study and at a 1 month review she reported no problems with the drops. The intraocular pressures had lowered to 16 mm Hg in both eyes and the examination was otherwise unchanged. At her third visit 2 months later, she was again asymptomatic with visual acuities of 6/5 in both eyes and intraocular pressures of 15 mm Hg. However, examination of the optic nerves revealed bilateral oedema which was more prominent in the left eye. There were no signs of uveitis in either eye, pupillary reflexes were normal, colour vision and Amsler testing were not affected, and the visual fields were full. At this point the code for the trial drug was broken and it was seen that she had been using latanoprost 0.005% eye drops at night to both eyes over the 3 month period. A neurological consultation failed to find any neurological abnormality and all haematological and biochemical analyses were normal. A computed tomographic scan with contrast showed no abnormality and she was discharged from neurological review. Follow up in the eye clinic revealed no change after 72 hours. The latanoprost was stopped and the disc swelling had largely resolved at 1 week. By 10 weeks


both optic nerves looked normal. Visual acu-
ties were still 6/5 in both eyes and there was no 
loss of colour vision or visual field. The 
intracocular pressures had increased to 22 mm 
Hg in both eyes.

COMMENTS

Latanoprost is a prostaglandin F2 
alanogue which acts by increasing uveoscleral outflow. 
Side effects include increased iris 
pigmentation, hypertrophia and increased 
eyelash pigmentation, anterior uveitis in 
patients with complicated glaucoma or in 
those having had previous incisional 
surgery, and cystoid macular oedema occur-
ring soon after beginning latanoprost in pseudo-
diaphakic or aphakic eyes. Ocular hypotony 
with choroidal effusions and facial rash have 
also been attributed to latanoprost. To the 
best of our knowledge, optic disc oedema 
associated with latanoprost has not previously 
been described. The mechanism behind this 
association is unclear. One may not be 
surprised to see optic nerve swelling in associ-
ation with signs of posterior uveitis or 
hypotony but in this case it occurred without 
any sign of ocular inflammation and the lowest 
recorded intraocular pressure was 15 mm Hg. 
It may be feasible that the perfusion to the optic 
nerve heads via the short posterior 
ciliary arteries was compromised by a 
prostaglandin-like action manifesting as disc 
oedema and that latanoprost acid and prostaglandin F2 
at high concentrations could 
cause vasoconstriction of bovine ciliary 
arteries and a similar action cannot be 
discounted in this case. The rapid resolution 
of the swelling with seemingly no long term 
sequela once latanoprost was stopped would 
perhaps support this hypothesis.

Owen Stewart
Louise Walsh
Milind Pande
Department of Ophthalmology, 
Hull Royal Infirmary, Hull

Correspondence to: Mr Owen Stewart, 
Department of Ophthalmology, 
St James’s University Hospital, 
Leeds LS9 7TF.

Accepted for publication 12 April 1999

Somatostatin scan positive gastrinoma 
ocular metastasis

Editor,—Symptomatic ocular metastases are 
uncommon despite the 4% prevalence in 
patients dying of all types of malignancy in 
postmortem series.1 We report a case of ocular 
metastasis from a gastrinoma, which was part 
of the Wermer’s syndrome (multiple endo-
crine neoplasia (MEN) type 1), diagnosed by 
indium labelled octreotide scanning.

CASE REPORT

A 57 year old man presented with a 1 week 
history of blurring in his peripheral vision in 
his right eye and severe loss of visual acuity, 
worse early morning. He had been diagnosed 
with MEN type 1, 8 years previously after two 
perforated jejunal ulcers (1978, 1990) led to a 
diagnosis of Zollinger–Ellison syndrome, and 
a hyperplastic parathyroid gland had been 
removed for hypercalcaemia (1990). His 
mother had MEN type 1.

Ophthalmic examination revealed 6/12 acu-
ity in the right eye and 6/5 in the left. His 
other segments were unremarkable. His 
right fundus showed a solid amelanotic lesion 
about one disc diameter above the right disc. 
Ultrasoundography demonstrated a base of 13 
mm and a height of 8 mm. He also had bilateral 
inferior retinoschisis. One month later the 
tumour base measured 14·5 mm and the 
thickness measured 7·9 mm. The posterior 
edge now practically abutted the optic disc 
(Fig 1). There was subretinal fluid accumula-
tion.

A liver ultrasound scan showed multiple 
lesions and a tumour biopsy was composed of 
small solid islands of polygonal cells with 
granular cytoplasm, diagnostic of metastatic 
neuroendocrine carcinoma (immunostaining 
positive for chromogranin, neuron specific 
enolase, and NCAM, negative for S-100 and 
HMB45 (melanoma markers)). His urinary 
5HIAA was marginally raised at 133 pmol/24 
hours (normal up to 75), and a fasting intesti-
nal peptide screen, showed a highly elevated 
gastrin level (on omeprazole 40 mg per day) of 
343 pmol/l (normal range 0–40 pmol/l) but 
normal levels of other polypeptides.

No primary tumour or further metastases 
were seen using body computed tomograph 
and magnetic resonance imaging scans and 
the I-123 MIBG scan (meta-iodobenzyl gua-
nidine) was also negative. An indium (In-111) 
labelled octreotide scan at 1 and 4 hours 
+SPET showed focal areas of increased 
uptake in the right orbit (Fig 2), the nasal 
region, mediastinum, multiple sites in the 
liver, and possibly other abdomen sites. How-
ever, a positive octreotide scan is not exclu-
sively seen with neuroendocrine tumours, 
since other tissues have somatostatin recep-
tors. These include high grade lymphoma, 
some small cell lung cancers, occasional 
tumours of the breast, and in chronic inflam-
matory conditions where there is T cell activa-
tion including endocrine ophthalmopathy with 
orbital involvement. Thus, although an 
absolute positive diagnosis of metastatic gas-
trinoma cannot be made definitively in the 
absence of histology, in the context of this 
case it is highly probable that the choroidal 
tumour is due to ocular metastasis from 
gastrinoma.

COMMENTS

Gastrinoma may occur sporadically or as part 
of multiple endocrine neoplasia. MEN type 1 
is a rare disorder, usually inherited in an auto-
osomal dominant fashion with high penetrance 
which affects multiple endocrine glands (hy-
perparathyroid hyperplasia in 80–90%, pan-
creatic islet tumours in 50%, and pituitary 
adrenomas in 40–50%).

Ocular metastasis from gastrinoma has not 
previously been reported, although it has been 
reported in other neuroendocrine tumours— 
for example, carcinoid. Multiple or metastatic 
gastrinomas (especially if actively secreting) 
are best localised by a new radioisotope scan 
using radioactively labelled somatostatin 
analogues.5 Somatostatin is a peptide elabo-
rated by the delta cells of the islets of Lang-
han and the hypothalamus. It inhibits the 
secretion of gastrin by gastric mucosa and 
many other hormones such as insulin, thyro-
tropin, and corticotropin. Somatostatin recep-
tors are found on neuroendocrine tumours 
(80% of gastrinomas) and indium labelled 
occteotide visualises all somatostatin receptor 
positive gastrinomas.6 Our scans revealed 
somatostatin avid liver metastases. Interest-
ingly, the scan also very clearly confirmed the 
ocular/orbital disease. Orbital radiotherapy 
and chemotherapy were recommended.7

KEIR E LEWIS
PAUL B ROGERS
Department of Clinical Oncology

JOHN HUNGERFORD
Department of Ocular Oncology

KEITH E BRITTON
Department of Nuclear Medicine

P NICHOLAS PLOWMAN
Department of Clinical Oncology

Correspondence to: Dr Plowman.
Accepted for publication 12 April 1999


Figure 1 Retinal photograph showing amelanotic neuroendocrine tumour.

Figure 2 Image from indium (In-111) labelled octreotide scan taken 4 hours after injection showing very avid focal uptake in the right orbit.
Retinal neovascularisation in Goltz syndrome (focal dermal hypoplasia)

EDITOR,—This is the first reported case of Goltz syndrome with documented peripheral retinal non-perfusion and subsequent retinal neovascularisation and vitreous haemorrhage. In the eye this represents solely a mesodermal disturbance compared with the more common cases which present with both mesodermal and neuroectodermal disturbances, such as coloboma or microphthalmia.

CASE REPORT
Goltz syndrome was diagnosed shortly after birth in a girl with linear lesions of atrophic skin following Blaschko’s lines on the trunk and symmetrical syndactyly of the third to fourth fingers and second to third toes. Initial ophthalmic screening revealed no ocular anomalies. Dental screening revealed the congenital absence of one deciduous incisor. Anomalies, in particular syndactyly, polydactyly, or adactyly as well as skeletal anomalies; in particular synostosis, kyphosis, spina bifida occulta, rib and scapula anomalies; and dental anomalies, especially hypodontia.

COMMENT
Focal dermal hypoplasia is a rare disorder of ectodermal and mesodermal dysplasia originally described by Goltz et al.1 It is characterised by congenital atrophic skin changes often associated with herniation of the subcutaneous fat; skeletal anomalies, in particular syndactyly, polydactyly, or adactyly as well as scoliosis, kyphosis, spina bifida occulta, rib and scapula anomalies, and dental anomalies, especially hypodontia.

Ocular anomalies occur in 40% of cases.2 Coloboma have been reported in one third of cases, then less frequently microphthalmia, strabismus, nystagmus, and ectopia lentis.3 Other reported ocular anomalies include anophthalmia, corneal clouding, aniridia, heterochromia, and optic atrophy. Rarely ectropion and ptosis may occur as well as lid margin or conjunctival papillomatous lesions (histologically angioblastomas).4,5 Only one case of cloudy vitreous has been reported. This was in association with microphthalmia, aniridia, and lens subluxation.6 Retinal sclerosis or hypopigmentation was reported in Goltz’s original case review.7 No attempt was made to explain these findings. In our case, there was peripheral retinal non-perfusion and temporal retinal telangiectasia with subsequent neovascularisation and vitreous haemorrhage.

The differential diagnosis of retinal vascular anomalies associated with skin lesions includes incontinentia pigmenti (IP) and Cockayne’s syndrome. Although focal skin atrophy may occur in IP, the initial skin lesions are vesicles and bullae which may later become pigmented macules. The skin lesions in Cockayne’s syndrome are pigmented scars due to light sensitivity and trauma. Neither syndrome is associated with digital anomalies.

The genetic anomaly in Goltz syndrome remains to be determined. Most cases are sporadic. It is generally thought to be X-linked dominant with lethality in males, like IP; however, 9% of cases are male. These are proposed to be the result of half chromatid mosaicism.8 They are deletions in the region of the chromosome Xp22.9 These are a suggested site, though these must be differentiated from the deletions seen in microphthalmia with linear skin defects (MLS) and that of microphthalmia, dermal aplasia, and sclerocornea (MIDAS), which are now considered to be distinct entities.9 The wide variation in severity of expression is thought to be due to mosaicism.

A.A.S. DUNLOP
Medicinal Retinal Unit, Moorfields Eye Hospital, City Road, London

J. HARPER
Department of Dermatology, Great Ormond Street Hospital for Children, London

A.M.P. HAMILTON
Medicinal Retinal Unit, Moorfields Eye Hospital, City Road, London

Correspondence to: Dr A A S Dunlop, 66 King Street, Newcastle, NSW 2300, Australia

Accepted for publication 19 April 1999


Retinitis sclopetaria associated with airbag inflation

EDITOR,—Chorioretinitis sclopetaria is a severe form of blunt trauma, caused by a high velocity object grazing the globe but not penetrating it. It is a concussion injury, which usually manifests as severe choroidal and retinal rupture associated with haemorrhage but an intact sclera.1 We report a case of chorioretinitis sclopetaria resulting from deployment of an airbag in a stationary motor vehicle.

CASE REPORT
A 32 year old pregnant woman was the driver in a car accident in which the airbag failed to deploy at the time of impact but inflated when she returned to sit in the driver’s seat approximately 5 minutes after the accident occurred. The patient was not wearing spectacles and there was no evidence of injury from other sources.

At presentation the visual acuity was hand movements in the left eye and 6/6 in the right eye. Examination revealed evidence of swollen eyelids and marked chemosis and subconjunctival haemorrhages on the left, with no bony injury and a full range of eye movements. Anterior segment examination revealed a clear left cornea and a quiet anterior chamber with a microhyphaema. The lens was clear and there was no evidence of vitreous haemorrhage. There was no angle recession and the intraocular

Figure 1 Colour fundus photograph of right temporal retina showing perfused retina (photo right) to peripheral non-perfusion (photo left) with fibrotic vessels and neovascular complex in the transition zone. (Inferior dislocation artefact.)

Figure 2 Fluorescein angiogram of area showing peripheral non-perfusion and haemorrhage arising from the neovascular complex. The avascularity of the retinal vessels appears normal in the transitional zone.
pressure was recorded at less than 4 mm Hg. Fundal examination revealed evidence of a retinal tear although details were obscured by a diffuse vitreous haemorrhage. An ultrasound examination showed a vitreous haemorrhage and large retinal tear but no evidence of a scleral perforation.

An examination under anaesthesia performed the following day confirmed that there was no scleral rupture and indirect ophthalmoscopy confirmed the findings of diffuse vitreous haemorrhage and a retinal tear.

At the 2 week postoperative clinic visit, visual acuity had improved to 6/18. There was 2+ cells in the anterior chamber and the intraocular pressure was 24 mm Hg. Posterior segment findings were an intragel haemorrhage and a retinal tear. Observation under anaesthesia performed 4 months later confirmed the findings of diascytosis, and choroidal ruptures have been reported in the literature but this is the first report of retinitis sclopetaria resulting from airbag deployment.

COMMENT

Airbags are designed to protect the driver from direct impact from the steering wheel, dashboard, and windscreen. They are designed to inflate in 10 ms in response to sudden deceleration and during deployment, the airbag is propelled out of its storage compartment at speeds of more than 100 mph. Following inflation the airbag deflates slowly within seconds.

Facial and ocular injuries associated with airbags have been reported in the literature. Skin abrasions, burns, and eyelid ecchymoses are the most common facial injuries. Reported ocular injuries include orbital fractures, keratitis, corneal abrasions, hyphaemmas, angle recession, and lens subluxation. In the posterior segment, vitreous and retinal haemorrhages, commotio retinae, retinal tears and dialyses, and choroidal ruptures have been reported. 4 To our knowledge, this is the first reported case of retinitis sclopetaria secondary to airbag inflation.

Although airbags have clearly been shown to reduce serious morbidity and mortality associated with road traffic accidents, they are associated with a number of injuries directly attributable to their inflation. Some of these are serious ocular injuries and it is important for ophthalmologists and others involved with trauma cases to be aware of these complications. A full ophthalmic assessment is mandatory in all cases and this should include indentation ophthalmoscopy. A variety of posterior segment injuries have been reported in the literature but this is the first report of retinitis sclopetaria resulting from airbag deployment.

Figure 1 Airbag injury with retinitis sclopetaria, an intragel haemorrhage, and attached retina.

Figure 1 Specular microscopic findings. (A) Many crystalline deposits are observed at the limbus. (B) Specular microscopic findings re-examined after 1 year. The change of the locations and forms of crystalline deposit from the same vessels can be seen.

Specular microscopic findings of corneal deposits in patients with Bietti's crystalline corneal retinal dystrophy

Editor,—In 1937, Bietti1 first described three cases of tapetoretinal degeneration characterised by yellowish glistening retinal crystals, tapetoretinal degeneration with choroidal sclerosis, and marginal crystalline deposits of the cornea. Although more than 100 cases of crystalline retinopathy have been reported, crystalline deposits of the corneal limbus have been observed in only four out of 52 Japanese patients with crystalline retinopathy. Recently, observation using specular microscopy2,3 has been reported to be useful in detecting crystalline deposits at the limbus of patients with crystalline corneal retinal dystrophy. Therefore, in this study, we examined four patients with crystalline retinopathy using specular microscopy under a “con-surface” mode, which is used for the observation of the corneal surface, and we detected the deposits at the limbus.

Twelve months after the initial specular microscopic examination, we reinspected the crystalline deposits of two cases. Interestingly, the changes in the locations and forms of the crystalline deposits in the corneal limbus were exposed over time (Fig 1). More crystalline deposits were found in the patients with more advanced retinopathy. It is supposed that corneal deposits and fundus deposits are essentially the same and it is suggested that crystalline retinopathy is caused by systemic abnormality. Although the exact pathogenesis of crystalline deposits is still uncertain, it is possible that destroyed fibroblasts appear to glitter or fibroblasts with crystalline-like deposits look glittering during breakdown. Further biochemical or cellular biological studies are needed to clarify these possibilities.

YUKO WADA Department of Ophthalmology, Tohoku University School of Medicine, Sendai Japan

MITSURU NAKAZAWA Department of Ophthalmology, Hirosaki University School of Medicine, Hirosaki, Aomori, Japan

TOSHIKI ABE TAKASHI SHIONO MAKOTO TAMAI Department of Ophthalmology, Tohoku University School of Medicine, Sendai Japan

COMMENT

Airbags are designed to protect the driver from direct impact from the steering wheel, dashboard, and windscreen. They are designed to inflate in 10 ms in response to sudden deceleration and during deployment, the airbag is propelled out of its storage compartment at speeds of more than 100 mph. Following inflation the airbag deflates slowly within seconds.

Facial and ocular injuries associated with airbags have been reported in the literature. Skin abrasions, burns, and eyelid ecchymoses are the most common facial injuries. Reported ocular injuries include orbital fractures, keratitis, corneal abrasions, hyphaemmas, angle recession, and lens subluxation. In the posterior segment, vitreous and retinal haemorrhages, commotio retinae, retinal tears and dialyses, and choroidal ruptures have been reported. 4 To our knowledge, this is the first reported case of retinitis sclopetaria secondary to airbag inflation.

Although airbags have clearly been shown to reduce serious morbidity and mortality associated with road traffic accidents, they are associated with a number of injuries directly attributable to their inflation. Some of these are serious ocular injuries and it is important for ophthalmologists and others involved with trauma cases to be aware of these complications. A full ophthalmic assessment is mandatory in all cases and this should include indentation ophthalmoscopy. A variety of posterior segment injuries have been reported in the literature but this is the first report of retinitis sclopetaria resulting from airbag deployment.

Figure 1 Airbag injury with retinitis sclopetaria, an intragel haemorrhage, and attached retina.
Specular microscopic findings of corneal deposits in patients with Bietti's crystalline corneal retinal dystrophy

YUKO WADA, MITSURU NAKAZAWA, TOSHIAKI ABE, et al.

Br J Ophthalmol 1999 83: 1088
doi: 10.1136/bjo.83.9.1088k

Updated information and services can be found at:
http://bjo.bmj.com/content/83/9/1088.4.full.html

These include:

References
This article cites 3 articles
http://bjo.bmj.com/content/83/9/1088.4.full.html#ref-list-1

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/