

Figure 3 Anterior segment of the left eye. Microcornea, 6 mm in diameter, with a small sharply demarcated central opacity and an anterior synechia except at 4-5 o'clock (arrow) are noted.

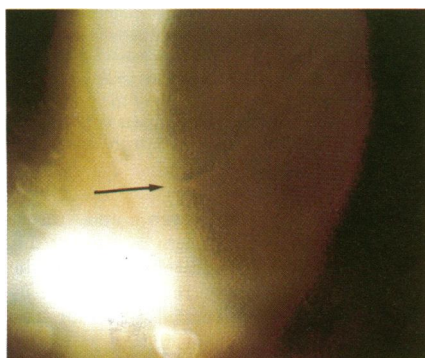


Figure 4 Gonioscopic finding with gonioprism of the right eye. There is an iris strand to the temporal posterior embryotoxon (arrow).

was shallow, and the central iris was attached to the periphery of the corneal opacity.

We diagnosed Peter's anomaly. Haematological analysis and chromosomal analysis with G-banding were all within the normal range. A paediatrician found no developmental retardation or systemic malformation. Hypertelorism was excluded. Ultrasonography showed a normal posterior segment.

Intraocular pressure was 12 mm Hg (RE) and 13 mm Hg (LE) with a portable applanation tonometer under general anaesthesia. The corneal diameters were 9 mm (RE) and 8 mm (LE) (that is, microcorneas). In the right eye, anterior synechia were present at 9 o'clock and 11 o'clock near the margin of the central corneal opacity (Figs 1A and B). In the left eye, an anterior synechia was present at the margin of the central corneal opacity except at 4-5 o'clock (Fig 3). Gonioscopic examination with a prism revealed several iris strands attached to the temporal and upper portions of a prominent Schwarbe's line, posterior embryotoxon in the right eye (Fig 4). In the left anterior chamber angle, we identified posterior embryotoxon but no iris strand was identified. Ophthalmoscopy showed a normal right optic disc and intact retina. The ocular fundus of the left eye was hardly visible because of corneal opacity. Ophthalmic examinations of his parents revealed no abnormal findings. There was no evidence of chromosomal abnormality of them. At 15 months, the ocular tension had been within the normal range.

COMMENT

Most cases of Peter's anomaly are sporadic, although reports of parental consanguinity and more than one affected sibling support an autosomal recessive or irregularly dominant mode of inheritance in some cases. Our

patient is sporadic. He has several iris strands attached to the posterior embryotoxon in the temporal and upper parts of the anterior chamber of the right eye. This patient also has a mild degree of Axenfeld's anomaly without glaucoma.

Mesenchymal cells which differentiate into corneal endothelium, stroma, iris, and aqueous outflow structure have been proved to be derived from neural crest cells by histochemical method.^{5,6} Bahn *et al*⁷ suggested that corneal endothelial disorders, Peter's anomaly, congenital glaucoma, posterior embryotoxon, Axenfeld's anomaly, Rieger's anomaly, and sclerocornea result from abnormal neural crest cell migration. Thus, in most cases of Peter's anomaly and of Axenfeld's anomaly the pathogenesis is related to abnormal migration of neural crest cells. It is speculated that the pathogenesis in our patient with Peter's and Axenfeld's anomalies is related to abnormal migration of the neural crest cells. If this hypothesis is correct, it would seem that the already reported cases of Peter's anomaly with Axenfeld's anomaly are disproportionately scarce. The reason for this disproportion may be that: (1) the wide corneal opacity conceals the anterior chamber; (2) most patients with this anomaly are babies or children, so gonioscopic examination is very difficult without general anaesthesia; (3) in patients with synechia between the central iris and the margin of the corneal opacity, the anterior chamber angle is hard to visualise, because of the central iris obstruction, with a gonioscopic mirror without a gonioscopic prism.

Careful gonioscopic examination with a prism under general anaesthesia of patients with Peter's anomaly would presumably reveal latent Axenfeld's anomaly.

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Subretinal hypopyon in polylmphocytic leukaemia

EDITOR.—It is well known that leukaemias can manifest themselves in ocular tissues. Most clinical and pathological studies suggest an incidence of at least 50% among leukaemic patients.¹ Ocular involvement is

markedly more frequent in acute than in chronic disease. All ocular tissues may become involved, mostly, however, choroid and retina. Retinal manifestations are haemorrhages, exudates, congested tortuous vessels, perivascular sheathing, and exudative or haemorrhagic retinal detachment which are summarised under the term leukaemic retinopathy. We report on a clinical sign of leukaemic retinopathy which has not been described previously, a subretinal hypopyon.

CASE REPORT

A 59-year-old male patient suffered from polylmphocytic leukaemia for 6 years. After splenectomy which had been performed 1 year after onset the leukaemic state was stable with a white cell count between 100 and 150×10⁹/l, no anaemia, and no thrombocytopenia. For 9 months the disease progressed showing an increasing white cell count. Chemotherapy had to be administered. When the patient was referred to our department he complained of blurred vision and ocular pain. At that time white cell count was 400×10⁹/l, haemoglobin 8.0 mg/100 ml, and platelets 200×10⁹/l. Visual acuity was 0.8

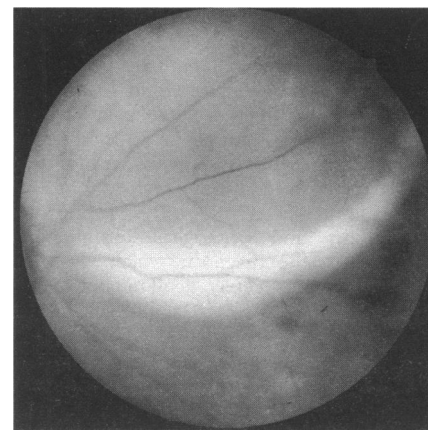


Figure 1 Subretinal aggregation of leukaemic cells at the bottom of serous retinal detachment in leukaemic retinopathy.

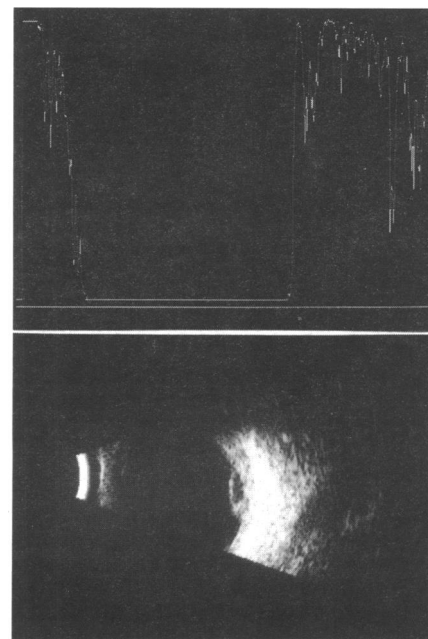


Figure 2 A- and B-scan in upright position: aggregation of cellular elements at the bottom of subretinal serous fluid (for further explanation see text).

right eye and 1-0 left eye. Slit-lamp examination showed cells and flare in the right anterior chamber with retrocorneal precipitates and cellular infiltration of the posterior vitreous. Funduscopy revealed bilateral disseminated intraretinal haemorrhages. The right fundus showed exudative retinal detachment of the upper nasal quadrant with an underlying whitish mass forming a horizontal level which changed according to the patient's position (Fig 1).

Ultrasound A-scan in upright position of the patient showed a 100% high spike due to retinal detachment and medium to highly reflective subretinal spikes at the bottom of the subretinal space. The latter were the results of irregular acoustic interfaces² caused by cellular (leucocytic) sedimentation (Fig 2). Ultrasound B-scan in the upright position confirmed the findings of A-scan investigation (Fig 2). A gravity dependent behaviour of subretinal cellular elements was clearly demonstrated as the higher reflective elements moved upwards and finally mixed with the lower reflective elements while the patient's position was gradually changed horizontally.

A few weeks after diagnosis of leukaemic retinopathy platelets dropped to $80 \times 10^9/l$. The patient deteriorated suffering from incurable fever and died.

COMMENT

The above described ophthalmic findings represent ocular involvement in leukaemia. As pointed out by Rosenthal³ and Decker *et al*⁴ the iritis-like signs are based on iris infiltration of leukaemic cells which invade the aqueous humour and may precipitate on the endothelium or even form a hypopyon. Paracentesis and cytological examination can confirm the diagnosis which was not possible in our severely debilitated patient. The fundus changes correspond well to the signs of leukaemic retinopathy.^{5,6} Guyer *et al* found an association between the presence of intraretinal haemorrhages and thrombocytopenia.⁷ In our patient, however, the manifestation of intraretinal haemorrhages preceded the decrease of platelets. As Fischer *et al* stated leukaemic cells can obstruct small vessels if the white cell count exceeds $150 \times 10^9/l$. Leukostasis leads to ischaemia and rupture of small vessels.⁸ This mechanism might explain intraretinal haemorrhage without thrombocytopenia. Serous retinal detachment has been found as part of leukaemic retinopathy.⁹ A subretinal hypopyon-like aggregation of cellular elements as shown on ultrasound, however, is rare and has not been reported in the literature to the best of our knowledge.

As there was no consent to postmortem examination the ophthalmic diagnosis could not be proved. As ocular signs and symptoms did not worsen but slightly improved after chemotherapy bacterial or fungal intraocular infection, which had to be considered, did not seem probable. The rapid fatal outcome after onset of ocular involvement in our patient confirms the statement of Rosenthal that leukaemic retinal infiltration combined with a high leucocyte count is 'an ominous prognostic sign'.³

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Pseudomonas keratitis associated with biofilm formation on a disposable soft contact lens

EDITOR,—A 26-year-old woman attended the casualty department at Moorfields Eye Hospital with a 1 week history of a sore left eye. The patient had been wearing Acuvue contact lenses (Vistakon, Johnson & Johnson, Berks) for myopic correction for 5 months. Lenses had not been disposed of according to the manufacturer's recommendations, and the same pair of contact lenses had been used for the past 5 months. Lenses were worn on an extended wear basis for 7 days, followed by disinfection using Oxsept 1 and 2 (Allergan, High Wycombe, Bucks). No surfactant cleaner was used before disinfection.

On admission, her left visual acuity was hand movements. She had a large corneal abscess with a 3 mm hypopyon (Fig 1). Microbiological analysis of the corneal scrape identified the causative organism as *Pseudomonas aeruginosa*. Part of the worn lens was macerated using a tissue homogeniser and culture of the homogenate recovered *Pseudomonas aeruginosa*, with similar antibiotic sensitivities as the isolate from the cornea. A second part was fixed in 2.5% glutaraldehyde with 0.05% ruthenium red for electron microscopy. Scanning electron microscopy of the posterior surface of the contact lens revealed widespread bacterial colonisation, with rod-shaped organisms embedded within an extensive extracellular matrix (Fig 2). Microbiological analysis of the contact lens storage case revealed a mixed confluent culture of *Pseudomonas aeruginosa* and *Serratia marcescens*. Culture of the disinfection and spray saline solution revealed no microbial growth. Sampling of body sites

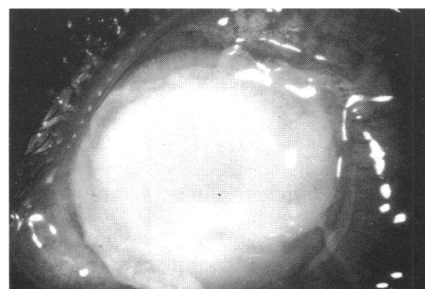


Figure 1 Large corneal abscess with dense corneal infiltrate and 3 mm hypopyon in the left eye.

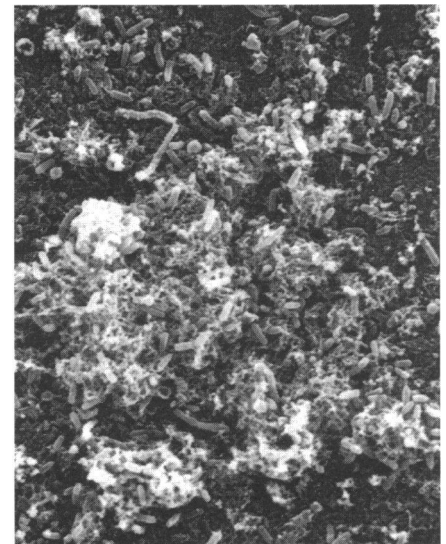


Figure 2 Scanning electron micrograph showing widespread colonisation of the posterior lens surface by rod shaped organisms which are embedded within a sponge-like extracellular matrix (magnification $\times 1180$).

including fingers, toe web, and throat revealed no persistent patient carriage of this organism. Microbiological analysis of the domestic water supply revealed no easily identifiable environmental source for this organism.

Initial treatment comprised intensive unpreserved ticarcillin drops (1%) every 30 minutes and systemic ciprofloxacin for 12 days. One week after admission, the patient was discharged and continued with ticarcillin drops hourly and gentamicin ointment at night. After a further week, there was healing of the corneal epithelial defect and the therapy was modified to ticarcillin drops 2 hourly, dexamethasone drops four times a day, and gentamicin ointment at night. Two weeks later, the corneal epithelialisation was complete and the topical treatment was slowly reduced. Five months later, the patient underwent successful penetrating keratoplasty, with a resultant visual acuity of 6/9.

COMMENT

Extended wear of contact lenses is a major predisposing factor for microbial keratitis and has a greater risk of disease compared with other modes of contact lens wear.¹ In this case, involving a disposable contact lens wearer, non-compliance with both the surfactant cleaning regime and contact lens disposal may have been additional associated risk factors.²

Bacterial colonisation of the contact lens by organisms similar to those recovered from the cornea, suggests the potential role of the contact lens as a vector for pathogenic organisms. Such colonisation may prolong the retention time of organisms at the ocular surface and production of an extracellular matrix (glycocalyx) may protect adherent organisms from host defence mechanisms and from the antimicrobial effects of contact lens disinfection systems. Bacteria within a biofilm on contact lens storage cases have been shown to be significantly more resistant to contact lens disinfection regimes compared with planktonic organisms.³

The role of the bacterial biofilm in the pathogenesis of contact lens related keratitis is not well understood. Prevention of persistent colonisation of contact lenses either by establishing the source of organisms, by modifying