neuritis, abducens and oculomotor nerve involvement have been noted with sphenoidal mucoceles.[2,3,6] A sphenoidal mucocele extending intracranially can be misdiagnosed as a malignancy.[1] Rarity of presentation, along with absence of positive indicators of infection (normal full blood count, ESR and C-reactive protein) and an inconclusive MRI appearance, led to a diagnostic dilemma in this case. In children with unilateral or bilateral optic neuropathy sinus infection should be suspected. CT scanning is the diagnostic imaging of choice in these patients.[7] The appropriate method of treating a sphenoidal mucopyelocele is transsphenoidal decompression and antibiotic therapy.[8]

The visual recovery depends on the acuity at the time of diagnosis.[4] Our case presented with no perception of light and did not improve beyond inaccurate projection despite surgical decompression of the mucopyelocele and antibiotic therapy. Therefore in children with optic neuropathy, ophthalmologists should be alert to the possibility of sphenoid sinus disease as an aetiological factor.

References

1. Oruckaptan HH, Akdemir P, Ozgen T. Isolated sphenoidal sinus abscess: clinical and radiological failure in preoperative diagnosis. Case report and review of the literature. Surg Neurol 2000;53:174–7.
2. Hiratsuka Y, Hotta Y, Yui A, Nakayasu L, Kanai A, Yamakawa T. Rhinogenic optic neuropathy caused bilateral loss of light perception [letter]. Br J Ophthalmol 1998;82:99–100.
3. Fujitani T, Takahashi T. Rhinogenic neuropathia of the optic nerve due to muco- and pyeloceles of the sphenoid sinus and posterior ethmoid cells [in German]. Laryngol Rhinol Otol 1984;63:361–3.
4. Lew D, Southwick FS, Montgomery WW, Weber AL, Baker AS. Sphenoid sinusitis: a review of 30 cases. N Engl J Med 1983;309:1149–54.
5. Newton N Jr, Baratham G, Sinniah R, Lim A. Bilateral compressive optic neuropathy secondary to bilateral sphenoethmoidal mucoceles. Ophthalmologica 1989;198:13–9.
6. Johnson LN, Hepler S, Yee RD, Batzdorf U. Sphenoid sinus mucocele (anterior clinoid variant) mimicking diabetic ophthalmoplegia and retrobulbar neuritis. Am J Ophthalmol 1986;102:111–5.
7. Hao SP. Mucocele of the sphenoid sinus with acute bilateral blindness: report of a case. J Formosan Med Assoc 1994;93:519–21.
8. Heller EM, Persky MS. Trans-septal sphenoidotomy for cystic lesions of the clivus. J Otolaryngol 1988;17:155–8.
9. Avery G, Tang RA, Close LG. Ophthalmic manifestations of mucoceles. Ann Ophthalmol 1983;15:734–7.

Mr R.K. Sekhri ✉
Department of Ophthalmology
Watford General Hospital
Vicarage Road
Watford WD18 0HB, UK

Sir,

Visual loss as the presenting feature of acute myeloid leukaemia

Isolated visual loss secondary to retinal pathology is a common referral to the ophthalmologist in the casualty setting. We present a patient who was found to have acute myeloid leukaemia, after an initial presentation of isolated decreased visual acuity in one eye. There have been very few reports in the literature of acute leukaemia presenting in this way, and it is important to be aware that this life-threatening blood disorder may initially present to the ophthalmologist.

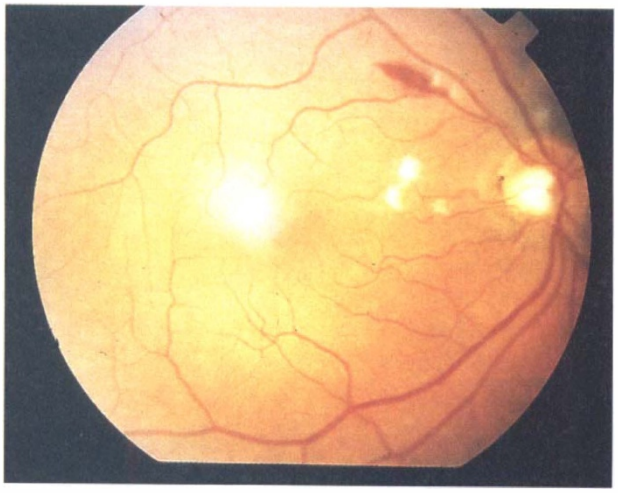

Fig. 1. *Fundus photograph of the right eye showing retinal haemorrhages and cotton wool spots near the optic disc.*

Case report

A 76-year-old Caucasian man presented to his optician with a 2 day history of blurred vision in the left eye. His visual acuity was recorded as 6/5 in the right eye and 6/24 in the left. Retinal haemorrhages at the left macula were observed and the patient was referred directly to the eye casualty.

On further enquiry, the patient stated that he had always enjoyed good health and normal vision. His past medical history was unremarkable except for one hospital admission for epistaxis and an operation for Dupuytren's contracture 15 years previously.

Ocular examination revealed marked tarsal conjunctival pallor, but otherwise healthy anterior segments. Fundoscopy demonstrated cotton wool spots and flame-shaped haemorrhages at both posterior poles (Fig. 1), with retinal and preretinal haemorrhage extending over the fovea in the left eye (Fig. 2). In both fundi, the arterial and venous calibre appeared normal, with no features indicative of hypertension.

Further investigation revealed a normal blood pressure (110/70 mmHg) and blood glucose (5.8 mmol/l). Serum electrolytes and liver function tests were within normal parameters. An urgent full blood count revealed a normocytic anaemia (Hb 6.0 g/dl), leucocytosis (WCC 38.6 $\times$ 10^9/l) and a profound thrombocytopenia (Plts 8 $\times$ 10^9/l). The blood film showed numerous blasts (36.3 $\times$ 10^9/l), with a variable nuclear:cytoplasmic ratio, some with cleft or budding nuclei, and single nucleolus. The bone marrow was hypercellular and replaced by blast cells. Cytogenetic analysis revealed monosomy 7 and additional material

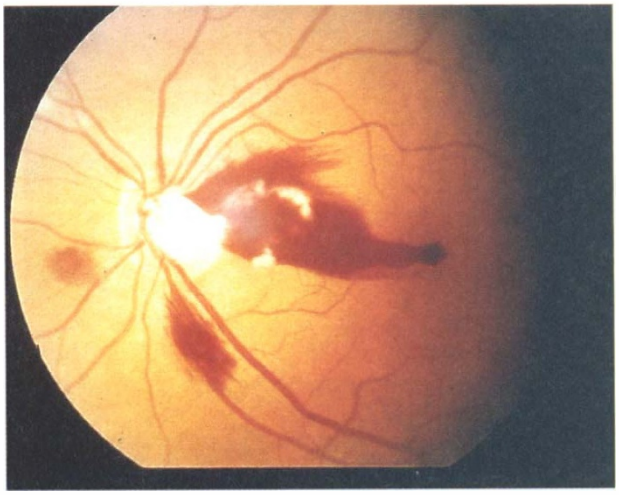

Fig. 2. *Fundus photograph of the left eye showing more extensive retinal and preretinal haemorrhage extending over the fovea and compromising visual acuity.*

on the arm of chromosome 16. The blast cell immunophenotype was positive for myeloid markers. A diagnosis of acute myeloid leukaemia was made.

During his stay in hospital, the patient developed severe haematemesis with progressive abdominal distention. He was treated with red cell and platelet transfusion, followed by broad spectrum intravenous antibiotics for pyrexia, but rapidly deteriorated in spite of treatment, dying 5 days after admission, presumably from massive intra-abdominal haemorrhage.

Comment

The differential diagnosis of retinal haemorrhages and cotton wool spots in the elderly population includes diabetes, hypertension and retinal occlusive disease. Blood dyscrasias are an uncommon cause, but if suspected may lead to early medical referral and appropriate treatment. In this case, the lack of evidence in the history and examination for hypertension, diabetes or arterial disease, and the presence of marked conjunctival pallor, led to immediate investigation and referral. It is interesting to note that this patient described no symptoms attributable to anaemia or thrombocytopenia despite his markedly abnormal blood count.

The incidence of adult acute myeloid leukaemia (AML) is approximately 3 per 100 000.[1] Symptoms may be sudden in onset with high fever and bleeding or be insidious with progressive weakness, pallor, low-grade fever, minor bleeding tendencies and recurrent infections. Ocular involvement in AML is frequent;[2] however, visual impairment as the presenting feature is rare and limited to a few case reports in the world literature.[3,4] The characteristic fundal signs of leukaemic retinopathy are those of an ischaemic vasculopathy and include retinal haemorrhages, typically at the posterior pole, cotton wool spots and infrequently retinal neovascularisation.

The extent to which retinal features correlate with the blood count and grade of leukaemia has been previously investigated. Guyer *et al.*[5] examined 117 consecutive patients with acute leukaemia, 66 of whom had acute non-lymphocytic leukaemia, in an attempt to correlate the fundus finding with the blood count. In their series, they reported an association between the presence of intraretinal haemorrhages and thrombocytopenia, and an association between white-centred haemorrhages and anaemia. However, the presence of cotton wool spots was independent of all haematological parameters. Karesh *et al.*[6] studied the fundal features of AML, and found that retinal signs were unrelated to age, sex, grade of leukaemia, leucocyte count, haematocrit and therapeutic response. They also noted that resolution of the ocular findings invariably followed if the induction phase of therapy was survived. In contrast, a study of ocular features in childhood acute leukaemia reported a relationship between ocular manifestations and survival.[7] However, ocular features recorded included not only retinopathy but also optic nerve, uveal and orbital infiltration as well as neuro-ophthalmic signs of central nervous system involvement. The survival rate of patients showing any ophthalmic features of leukaemia was found to be significantly lower than those who lacked such manifestations.

In summary, leukaemic retinopathy is a well-established clinical entity. This rare but life-threatening cause of isolated retinal signs should be considered in the differential diagnosis of any patient presenting with features of retinal ischaemia. Early referral may be life-saving.

References

1. Bishop JF. Adult acute myeloid leukemia: update on treatment. Med J Aust 1999;170:39–43.
2. Rosenthal AR. Ocular manifestations of leukemia: a review. Ophthalmology 1983;90:899–905.
3. Vangheluwe O, Ducasse A, Culioli B, *et al.* Macular manifestations of acute lymphoblastic leukemia: a case report. Bull Soc Ophtalmol Fr 1990;90:677–82.
4. Dutschke K, Siebenburger H, Schrader W. Acute lymphatic leukemia of the T-cell line: visual impairment as the initial symptom. Ophthalmologe 1998;95:831–4.
5. Guyer DR, Schachat AP, Vitale S, *et al.* Leukaemic retinopathy: relationship between fundus lesions and hematologic parameters at diagnosis. Ophthalmology 1989;96:860–4.
6. Karesh JW, Goldman EJ, Reck K, *et al.* A prospective ophthalmic evaluation of patients with acute myeloid leukemia: correlation of ocular and hematologic findings. J Clin Oncol 1989;7:1528–32.
7. Ohkoshi K, Tsiaras WG. Prognostic importance of ophthalmic manifestations in childhood leukaemia. Br J Ophthalmol 1992;76:651–5.

André Ismail
David Verity
Paul Brittain
Sussex Eye Hospital
Eastern Road
Brighton BN2 5BF, UK

Mr André Ismail ✉
10 Clifton Avenue
Finchley
London N3 1BN, UK