Brit. J. Ophthal. (1966) **50**, 92

LEUKAEMIC GLAUCOMA*

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A 54-year-old man presenting for an eye examination was found to have open-angle glaucoma in both eyes, with normal fundi. Subsequent events confirmed that the glaucoma was the initial clinical manifestation of chronic lymphatic leukaemia. Because, to the authors' knowledge, glaucoma has not been reported as the presenting sign of leukaemia, this case is here presented.

Case Report

A 54-year-old white male was first seen in October, 1962, with the chief complaint of swelling and itching about the eyes of three months' duration. Except for a left facial paresis three months previously, which cleared in one week, and a slight loss of hearing in the right ear, he had always enjoyed good health. There were no other complaints. Family history revealed that two maternal aunts had glaucoma.

On examination, the uncorrected visual acuity was 20/20 in each eye. External examination revealed only slight thickening of the lids, without any redness or pitting oedema. There was no proptosis and motility was normal. The corneae were clear and the anterior chambers were of good depth. Slight injection of the limbus was seen in both eyes, but biomicroscopy was otherwise normal. Ophthalmoscopy revealed normal discs and no abnormalities. The visual field was full to 3/330 white in each eye. However, on routine tonometry, the intra-ocular tension was 50 mm. Hg Schiötz in each eye.

The patient was first given 2 per cent. pilocarpine solution 5 times daily with 250 mg. acetazolamide twice daily. Four days later, at re-examination, there had been no change in the intra-ocular tension at all. Because he failed to respond to therapy he was admitted to hospital for further study.

General physical examination revealed a few petechiae on the buccal mucosa. Several enlarged lymph nodes were palpated in the neck, axillae, and inguinal areas. The spleen was moderately enlarged but not tender. No other abnormalities were noted.

Radiographs of the skull and paranasal sinuses were normal. A flat plate of the abdomen confirmed the splenomegaly. A routine haemogram, however, revealed a white blood count of 120,000. Several repeat examinations during the next two weeks revealed a consistent leucocytosis ranging up to 230,000. Because of this finding a bone-marrow aspiration was performed which revealed a hypercellular marrow with marked proliferation of small lymphocytes. A diagnosis of chronic lymphatic leukaemia was made. In addition to pilocarpine, epitrate, and Diamox, the patient received systemic steroids, Leukeran, and irradiation to the liver and spleen. On this therapy the intra-ocular tension fluctuated widely between 20 and 50 mm. Hg Schiötz.

* Received for publication February 2, 1965.

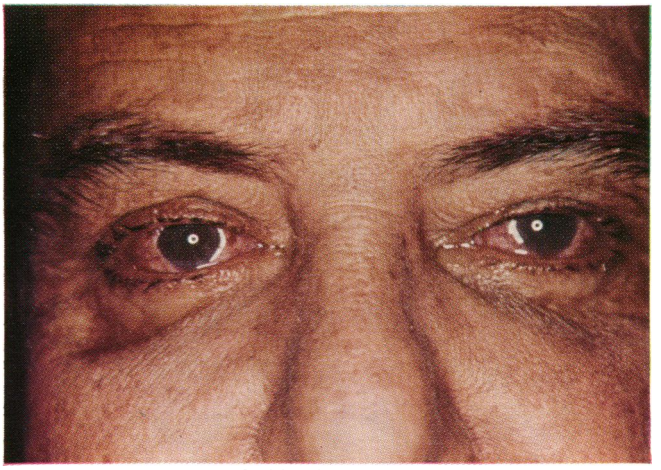

FIG. 1.—Appearance of lids, bulbar chemosis at limbus, and thickening of malar skin.

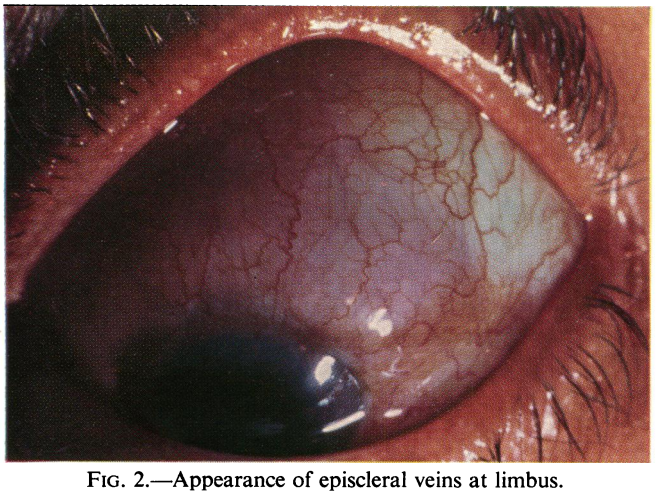

FIG. 2.—Appearance of episcleral veins at limbus.

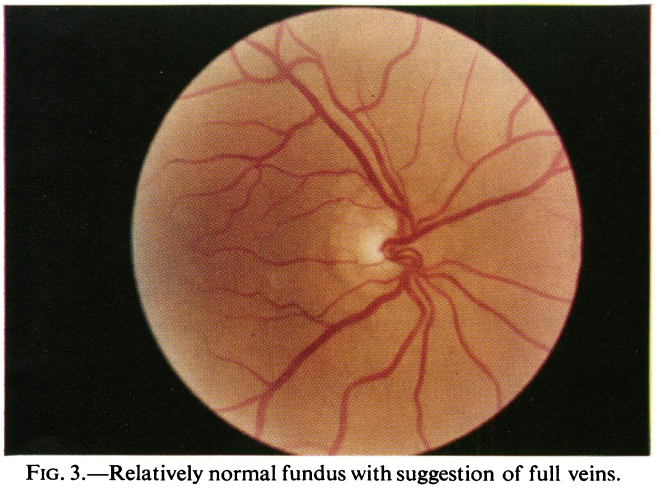

FIG. 3.—Relatively normal fundus with suggestion of full veins.

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During the next four months the patient was followed up as an out-patient. It was extremely difficult to maintain the intra-ocular tension at a reasonable level with topical therapy. By March, 1963, a malar flush was apparent and the episcleral veins now appeared congested. A purplish infiltration around the limbus could now be seen in each eye. Ophthalmoscopy revealed for the first time a few superficial retinal haemorrhages and tortuous vessels. (Figs 1–3.)

With an intra-ocular pressure of about 50 mm. Hg on medical therapy, surgery was contemplated. As it appeared that the glaucoma was due to leukaemic infiltration of the limbal tissues in each eye, however, it was decided to first treat the globes with irradiation. On March 15, 1963, the patient was given 400 roentgens/air through a 5×5 cm. port to each eye. Thereafter a transient elevation of intra-ocular pressure occurred for a few days, but shortly after pressure declined. Within a few weeks it fell to the low twenties and remained normal thereafter despite discontinuance of all glaucoma therapy. The patient died in July, 1963, and a post-mortem examination was not made.

Discussion

Leber reported a case in 1878 in which ocular signs preceded the detection of leukaemia. His patient had infiltration of all four lids as well as exophthalmos for a year before succumbing to the disease. Saradarian (1940) reported a case in which the bulbar conjunctivae were raised by an extensive salmon-coloured or reddish thickening, reminiscent of the case here considered. Biopsy of this case revealed numerous undifferentiated blast cells. Section of a submental lymph node revealed atypical lymphatic leukaemia. O'Rourke and O'Connor (1957) presented an excellent review of ocular involvement in leukaemia up to 1953. Reese (1951) recorded puffiness of the lids as being present in 2·5 per cent. of a series of 149 definitely established cases of leukaemia. Allen and Straatsma (1961) have reviewed 76 cases of leukaemia, finding the most frequent change to be a thickening of the conjunctiva at the limbus.

In none of the above series, however, was the presence of elevated intra-ocular pressure recorded as a presenting sign of leukaemia. The case here reported would certainly appear to be due to limbal infiltration blocking the aqueous outflow channels, for there was no significant response to miotics. Violaceous tissue could later be seen in this area, and irradiation of the globes caused prompt control of the glaucoma. The presence of normal fundi in this case is considered of interest. Most ophthalmologists consider leukaemia in the differential diagnosis only when they encounter the presence of tortuous retinal vessels or diffuse haemorrhagic retinopathy. In the case here reported, when the patient was first seen he had white eyes with no proptosis and no fundus changes at all, and the condition was identical with the clinical appearance of chronic open-angle glaucoma in both eyes. Only the failure to respond to miotic therapy prompted his admission to hospital, and a routine haemogram of nearly 100,000 white blood cells led to the correct diagnosis.

Summary

A 54-year-old man presented with open-angle glaucoma and normal fundi at routine eye examination. Except for intra-ocular pressures of 50 mm. Hg (Schiötz) in both eyes, no ocular abnormalities were present. The fact that the glaucoma was

the presenting sign of chronic lymphatic leukaemia in this case is of interest. A total failure to respond to miotic therapy, contrasted with a prompt fall in intraocular pressure after irradiation, implicated the limbal veins in the glaucoma. A routine blood count should be considered in the patient with an unusual or refractory glaucoma.

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