OCULAR RELAPSE IN A PATIENT WITH ACUTE LYMPHOBLASTIC LEUKEMIA: A CASE REPORT

CHEONG SOON KENG
TAN POOI LIEW

SUMMARY

A female Malay patient with acute lymphoblastic leukemia in remission for one year developed bilateral visual loss. The ophthalmoscopic appearance showed infiltration of optic nerves and retinae by leukemic cells. At that time, her blood, bone marrow and cerebral spinal fluid remained normal. She had received prophylactic cranial irradiation and intrathecal methotrexate as part of the treatment programme. It was likely therefore she had a leukemic relapse from a pharmacologic sanctuary in the eyes. She responded to local irradiation but did not regain her sight. Three months later, she had a bone marrow relapse.

INTRODUCTION

Visual loss in patients with leukemia is not uncommon and usually associated with meningeal leukemia or systemic disease.[1] Ocular leukemia occurring in patient in remission is most unusual. We report a patient with ocular leukemia preceding bone marrow relapse.

Cheong Soon Keng
MBBS (Mal), DCP (Lond), MRCP (UK)
Lecturer

Tan Pooi Liew
MBBS (Lucknow), FRCS (Edin), DO (Lond)
Formerly Associate Professor

Faculty of Medicine
National University of Malaysia
50300 Kuala Lumpur, Malaysia

CASE REPORT

A 23-year-old female Malay was referred in November 1982 from the Tanjung Karang Hospital with a history of lethargy, anorexia and weight loss for two months. Two weeks before admission, she experienced generalised body-ache and recurring fever, despite medication. Passing of black, tarry stools prompted the patient to the hospital. Abnormal blood cells were seen in the blood film and she was referred to the Kuala Lumpur General Hospital for further treatment.

On admission, she was found to have high temperature (39°C), marked pallor, tachycardia (104 beats per minute), cervical lymphadenopathy, and hepatosplenomegaly. Laboratory investigations confirmed that she had acute lymphoblastic leukemia by morphology and cytochemistry.

She was induced with prednisolone, vincristine, daunorubicin and L-asparaginase. She went into clinical and haematological remission about one month after beginning treatment. A course of cranial prophylactic irradiation (2400 rads) was also given. Following induction, she continued to receive maintenance therapy comprising oral

methotrexate, oral 6-mercaptopurine, and subcutaneous cytosine arabinoside intermittently. Every three months, she also received a course of reinduction with prednisolone and vincristine. During this period she was treated on an outpatient basis and led a normal life.

Approximately one year after remission, she complained of loss of vision in her left eye. She was immediately referred to the ophthalmologist who detected leukemic infiltration of the retina of both eyes and complete loss of vision in the left eye. Fundal changes were as seen in fundal photographs (Figs. 1, 2, 3). At that time physical examination other than the eyes was essentially normal. Laboratory investigations including a bone marrow biopsy and a lumbar puncture were within normal limits. A course of local irradiation (500 rads) was given to the left eye. However she refused treatment for her right eye. She continued her maintenance therapy without fail.

On follow-up, she had progressive loss of vision in her right eye though her left eye remained very much the same. About three months later, she had a haematological relapse with blast cells found in both bone marrow and peripheral blood. Her untreated eye had then shown changes similar to that of the treated one before irradiation. She was readmitted to the hospital and was reinduced with the same regime. Meanwhile a course of local irradiation (500 rads) was given to the right eye. A repeat lumbar puncture then was within normal limits and no malignant cell was seen. She achieved a second clinical and haematological

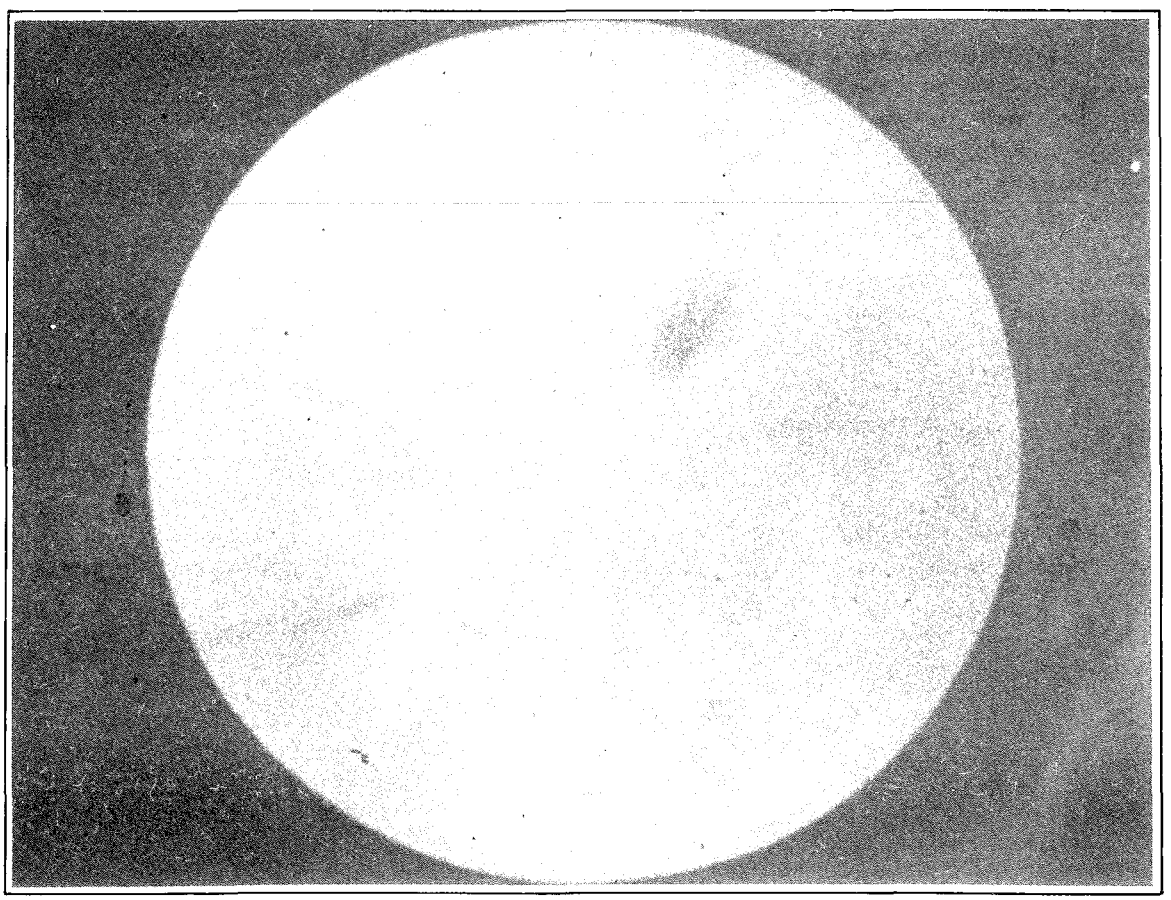

Fig. 1 Fundal photograph showing a choked disc and retinal haemorrhages in the left eye.

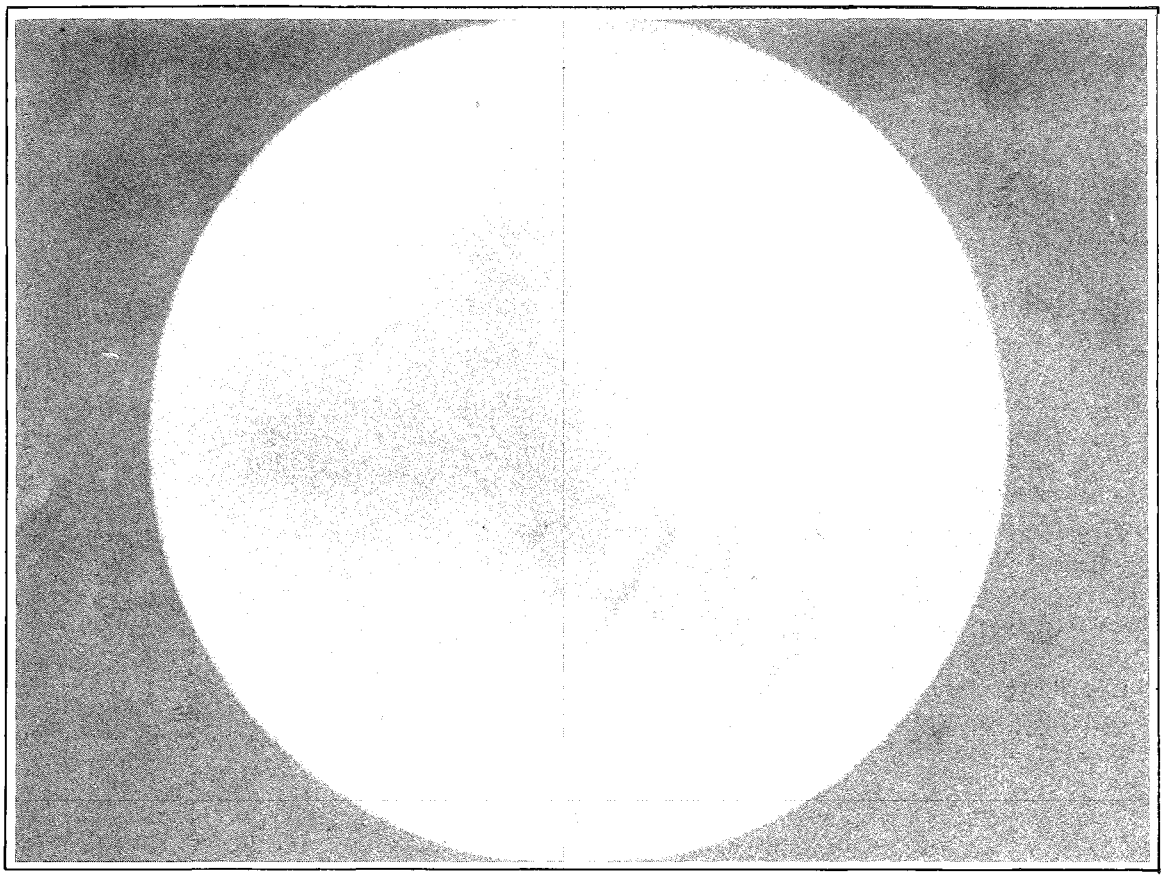

Fig. 2 Fundal photograph showing a choked disc and retinal pressure lines in the right eye.

remission. At present, she is alive and remains in remission while still on maintenance chemotherapy. She never regained her eye-sight.

DISCUSSION

The appearance of choked disc or papilloedema is not uncommon in patients with leukemia.[2] It may be due to the following conditions: increased intracranial pressure, prolonged corticoid therapy, leukemic infiltrates of the orbit or optic nerve or both, perivascular infiltrate resulting in venous engorgement, and leukemic infiltrate of the nerve head. The last three causes may or may not be associated with increased intracranial pressure.

In the absence of systemic or meningeal leukemia and increased intracranial pressure, the ophthalmoscopic appearance of this case most likely is due to the last three mechanisms.

This patient had received prophylactic cranial irradiation to prevent meningeal leukemia. But it has not prevented her from getting an isolated extramedullary relapse in the eyes. This may be due to the fact that prophylactic cranial irradiation given to her excluded the eyes. She also received four doses of prophylactic intrathecal methotrexate. It could not reach the eye because the subarachnoid space ends just posterior to the globe. Thus the isolated ocular relapse and the subsequent bone marrow relapse probably originates from this pharmacologic sanctuary.

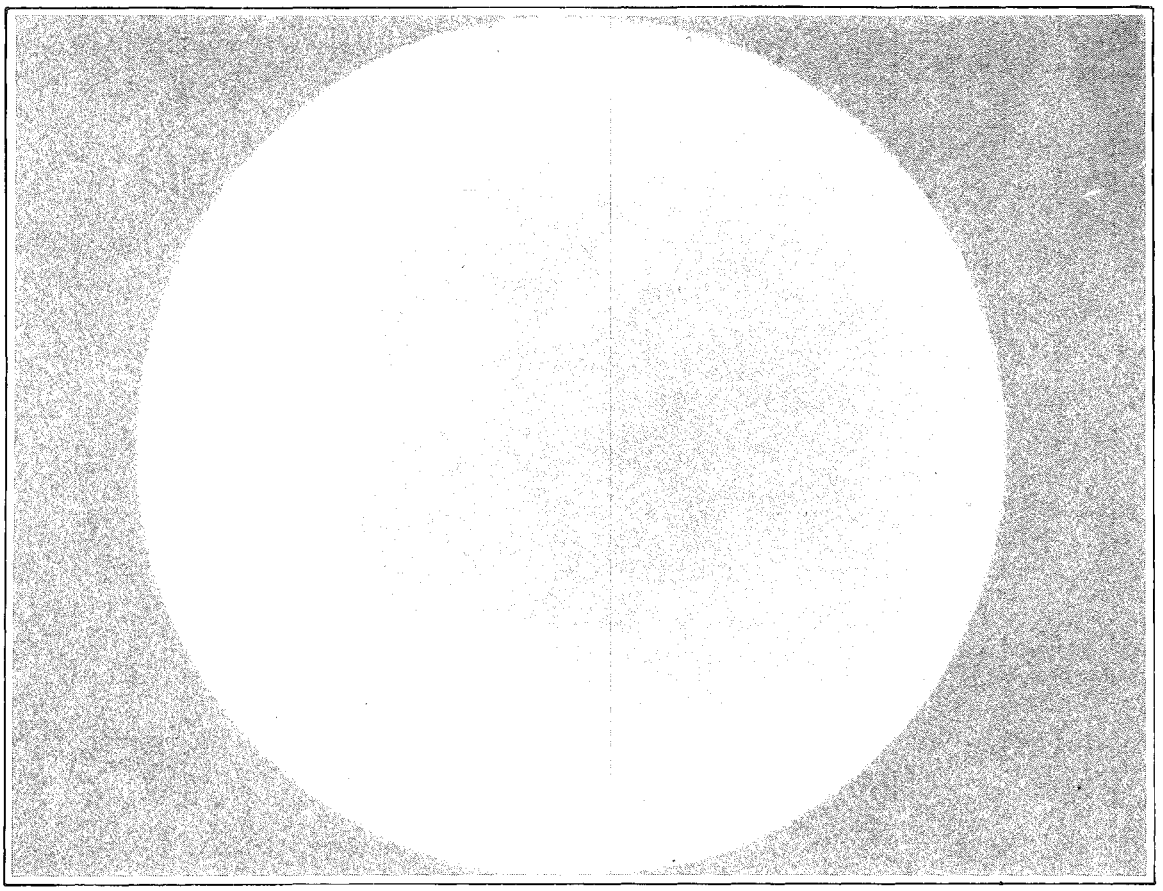

Fig. 3 Fundal photograph showing whitish retinal infiltrates of leukemic cells.

As ocular relapse is rare, prophylactic irradiation to the eyes should not be recommended because the attendant complications far outweigh the benefit. However, if ocular leukemia is present together with systemic and meningeal leukemia, there should be no hesitation to recommend irradiation to both eyes in conjunction with craniospinal irradiation and systemic/intrathecal chemotherapy.

In this case, the ocular relapse preceded bone marrow relapse. As there was no evidence of a systemic or meningeal disease, the patient was continued on the same maintenance chemotherapy. In retrospect, in addition to local irradiation to the eye, a change of chemotherapy regime should have been done. This could have prevented her bone marrow relapse.

REFERENCES

1 Murray K H, Paulino F, Goldman J M, Galton D A G, Grindle C F J. Ocular involvement in leukemia. *Lancet* 1977; i: 829–831.

2 Ellis W, Little H L. Leukemic infiltration of the optic nerve head. *Am J Ophth* 1973; 75: 867–871.