

BRITISH JOURNAL OF OPHTHALMOLOGY

Editorial

Ophthalmic manifestations of childhood leukaemia

The treatment of childhood leukaemia stands out as one of the great achievements of modern haematological practice: remission and cure are now commonplace for a disease which was, at one time, uniformly fatal. Unfortunately, many patients still suffer relapses and in these individuals the promise of a permanent cure is less certain. In this issue Ohkoshi and Tsiaras highlight the prognostic importance of ophthalmic involvement in childhood leukaemia. In a 15 year study period 28 of 131 patients treated developed ocular complications and, of these, regrettably only one patient survived. The remaining 27 patients all died within 28 months from the onset of the ophthalmic involvement. All patients with ophthalmic manifestations had either bone marrow relapse or central nervous system involvement and the authors attribute the poor prognosis to these associated factors. Ocular involvement would, therefore, appear to be the harbinger of a relapse rather than its progenitor.

Whilst the authors must be congratulated for a detailed and informative study, some caution must be exercised in interpreting their results. Patient recruitment began in 1972 and it is probable that recent treatment regimens will provide a greater prospect of remission or cure; particularly in cases of relapse following initial therapy. Indeed, repeated modifications of therapeutic regimens used in the treatment of acute lymphoblastic leukaemia have resulted in progressive improvements in patient survival.¹ Recent studies have also reported long lasting second remissions in about one third of children suffering from this condition.²

In addition to its primary aim, this article serves to remind the ophthalmologist of the diverse spectrum of the ocular involvement in childhood leukaemia which can, broadly speaking, be divided into three groups: neuro-ophthalmic features associated with central nervous system involvement, vascular abnormalities reflecting changes in haematological status, and direct infiltration of the ocular tissues.

Neuro-ophthalmic signs of central nervous system disease include papilloedema, secondary to raised intracranial pressure, and isolated cranial nerve palsies. Vascular abnormalities typically affect the retina and include intraretinal haemorrhages, cotton wool spots, and venous occlusions.³ Vitreous haemorrhage may also occur. Infiltration typically affects the optic nerve, anterior segment, and orbit. Whilst optic nerve infiltration is usually related to central nervous system involvement, anterior segment infiltration frequently occurs in the absence of central nervous system disease. Novakovic *et al*⁴ recently reported eight cases of anterior segment involvement in acute childhood leukaemia, only two of whom had concurrent central nervous system involvement or bone marrow relapse. This is in sharp contrast to the results of the study reported in this issue. Anterior segment involvement is characterised by either diffuse whitish thickening of the iris or a pseudohypopyon. Any child with leukaemia who develops 'iritis' should be regarded as having a relapse of the leukaemia until proved otherwise⁴ and, in such cases, anterior chamber paracentesis or iris biopsy is essential.

Although the ophthalmologist has a secondary role in the treatment of childhood leukaemia, a prompt recognition of the ocular manifestations and their importance as a sign of possible extramedullary disease is crucial if appropriate therapy is to be initiated.

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- 1 The Medical Research Council UKALL trials 1972-84. Improvement in the treatment for children with acute lymphoblastic leukaemia. *Lancet* 1986; i: 408-11.
- 2 Henze G, Fengler R, Hartmann R, Kornhuber B, Janka-Schaub G, Neithammer D, *et al*. Six-year experience with a comprehensive approach to the treatment of recurrent childhood acute lymphoblastic leukemia (ALL-REZ BFM 85). A relapse study of the BFM group. *Blood* 1991; 78: 1166-72.
- 3 Schachat AP, Markowitz JA, Guyer DR, Burke PJ, Karp JE, Graham ML. Ophthalmic manifestations of leukemia. *Arch Ophthalmol* 1989; 107: 697-700.
- 4 Novakovic P, Kellie SJ, Taylor D. Childhood leukaemia: relapse in the anterior segment of the eye. *Br J Ophthalmol* 1989; 73: 354-9.



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Br J Ophthalmol 1992 76: 641
doi: 10.1136/bjo.76.11.641

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