Sir,
**Anterior chamber infiltrate in 3-month-old:
rare presentation of myeloid sarcoma**

Myeloid sarcoma (MS) is an extramedullary tumour of immature myeloblasts associated with leukaemias.¹ MS commonly presents after diagnosis of active leukaemia, during a recurrence, or as a blastic transformation of myelodysplastic syndrome. Rarely, MS arises *de novo*.^{2,3} Here, we describe a case of MS presenting *de novo* with a white anterior chamber infiltrate.

Case report

A 3-month-old infant presented to an outside ophthalmologist with a unilateral 'discoloured iris.' He was referred for retinoblastoma *vs* endophthalmitis. Examination showed a fluffy, white anterior chamber infiltrate and scarce vitreous cell in the right eye. The rest of the ocular exam and peripheral blood studies were unremarkable.

Figure 1 Presentation at first exam under anaesthesia showing bilateral proptosis, lacrimal gland swelling, and bilateral anterior chamber infiltrates.

Figure 2 MRI demonstrates prominent lacrimal gland swelling.

Social reasons delayed the exam under anaesthesia and MRI for 12 days. Unexpectedly, he developed bilateral proptosis, lacrimal gland swelling, a larger right anterior chamber infiltrate, and new left anterior chamber infiltrate (Figure 1). MRI of the brain showed infiltrative bone marrow processes with involvement of bilateral globes, lacrimal glands, extraocular muscles, and paranasal sinuses (Figure 2).

Lacrimal gland biopsy revealed MS. The H&E stain showed sheets of tumour cells and the lysozyme stain was positive (Figures 3a and b). Work-up of blood and bone marrow showed no evidence of malignancy.

After two courses of 10-day cycles of cytarabine, daunorubicin, and etoposide, CNS involvement was seen and additional intrathecal therapy was started. His lesions regressed and anterior segment infiltrates disappeared (Figure 4).

Before his fifth course, rare leukaemic blasts were noted in the CSF. These cleared with intrathecal therapy; however, during recovery, leptomeningeal enhancing lesions were noted. He was started on palliative oral VP-16 (etoposide). After multiple relapses despite external radiation and chemotherapy, the decision was

Figure 3 (a) H&E reveals sheets of tumour cells with a high mitotic rate. (b) Lysozyme staining is positive. Photos courtesy of Dr. Hans Grossniklaus.

Figure 4 Significant resolution of infiltrates and proptosis after two cycles of chemotherapy.

made to start palliative whole brain radiation. He passed away 4 months later.

Comment

MS is rare and often misdiagnosed.^{2,3} The incidence of a single MS is 2% in AML,¹ whereas orbital MS accounts for 1 of 250 cases.⁴ An anterior chamber infiltrate might be misdiagnosed as retinoblastoma or hypopyon, resulting in delayed or improper treatment. We present this case to emphasize that MS should be considered in patients with white anterior chamber infiltrates even if previously healthy.

Conflict of interest

The authors declare no conflict of interest.

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Sir,
Reply to Spiteri Cornish and Reddy

We read with interest the paper by Spiteri Cornish and Reddy entitled 'The use of propranolol in the management of periocular capillary haemangioma – a systematic review.'¹ The authors provide an excellent review regarding the changing paradigms for the management of patients with infantile haemangiomas (IHs) and provide clear evidence as to the effectiveness of propranolol. The authors were unable to find an RCT comparing propranolol with a recognised standard or a placebo and concluded that an RCT would be beneficial but difficult to perform. The primary reason being that clinical equipoise cannot be met; given the weight of recent evidence the majority of clinicians would now be reluctant to revert to previous treatments for IHs, and therefore cannot ethically choose at random which treatment to provide.² An RCT in this situation will have inherent flaws with confounding factors and bias.

Glasziou *et al*³ ask whether an RCT in this situation is actually required, illustrating similar historic milestones in medical practice where a new intervention has provided equally dramatic results. Occasionally, an RCT is not performed despite the intervention becoming mainstream practice, with examples including insulin to treat diabetes, defibrillation for ventricular fibrillation, and sutures to repair large wounds. They go on to describe the 'rate ratio' between the signal or intervention (propranolol) and the background noise (previous treatments). This is calculated as the rate of improvement of the lesion during the new treatment divided by the rate with previous treatments. Ideally, this ratio should be over 20, that is treatment with propranolol should increase the rate of improvement by over a factor of 20 when compared with a control that allows for bias and other confounding factors.

Clearly, the efficacy of propranolol is not disputed and issues regarding adverse effects should now be monitored using long-term data collection in the form of either a prospective database or registry. Such observational data will highlight rarer treatment complications and long-term effects. The optimal mode, dose, and duration of treatment of propranolol are yet to be defined and well-designed RCTs in this context would certainly be of benefit.

References

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