Aetiology of childhood proptosis

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Objective: To retrospectively analyse causes of childhood proptosis and their investigations and treatment.

Methodology: The records of children under 15 years of age presenting with proptosis to The Children’s Hospital, Camperdown, Sydney, were reviewed for the period 1983–92 inclusive.

Results: Fifty-seven cases of proptosis were found. In order of frequency the causes were: orbital cellulitis 22, thyroid eye disease 8, optic nerve ± optic chiasm glioma 8, orbital rhabdomyosarcoma 7, metastatic neuroblastoma 4, orbital neurofibroma 3, orbital haemangioma 2, metastatic Ewing’s sarcoma 2 and orbital dermoid cyst 1.

Treatment depended on the cause (antibiotics, antithyroid drugs, chemotherapy, radiotherapy, surgery) and was multidisciplinary.

Conclusions: The most common cause of proptosis in children presenting to The Children’s Hospital, Camperdown, Sydney, was infective orbital cellulitis. The most useful initial investigation was an orbital computed tomography scan. Treatment depended on the cause of the proptosis and was multidisciplinary.

Key words: Exophthalmos; orbit; orbital cellulitis; proptosis.
The clinical approach to the management of a child with proptosis largely depends on the history of presentation. By far the most common cause in our series was infective orbital cellulitis. The typical presentation was with pain, eyelid erythema, chemosis and proptosis, with or without fever and malaise. Although all cases were secondary to sinusitis many cases gave no definite preceding history of sinusitis. The differential diagnosis of this inflammatory presentation is delineated in Table 2.

Children with Grave’s disease may have other ocular and systemic signs and symptoms of thyrotoxicosis. Optic nerve gliomas associated with neurofibromatosis (type I) in this group notably had a worse course than those independent of systemic disease. This contrasts with traditional teaching that gliomas associated with neurofibromatosis have a more quiescent course, but may be accounted for by the small numbers of this subgroup within this study.

No cases of the following lesions causing proptosis were seen: lymphoma, haemangioma, orbital spread of retinoblastoma, Langhan’s cell histiocytosis, pseudotumour and tumours of adipose, osseous, fibro-osseous and cartilaginous tissues.

The child underwent a subtotal surgical excision and at present the visual acuity is normal with no sign of recurrence.

DISCUSSION

The age of onset of orbital rhabdomyosarcoma varied from 3 to 12 years. One case was Stage 2, four were Stage 3 and two were Stage 4. One case later developed acute lymphoblastic leukaemia. All cases were treated with chemotherapy and radiotherapy, with one case having a bone marrow transplant in addition. The visual outcome of the involved orbit varied from normal to blind, with one case dying.

The age of onset of orbital metastatic neuroblastoma varied from 9 months to 7 years. Treatment included surgery, chemotherapy, radiotherapy and in one case bone marrow transplant. The visual acuities ranged from normal (this case died) to blind.

The age of presentation of the 2 cases with orbital haemangioma was at birth and at 7 months. There was no record of visual acuity or treatment for the first case. The second case had initial acuities of 6/30 in each eye (as assessed by Teller visual acuity cards) and had systemic interferon with some success in tumour regression at 4 months follow-up after initial failure with steroid therapy.

The 2 cases of Ewing’s sarcoma presented at 11 and 13 years. Both had radiotherapy, chemotherapy and surgery. One child died and the other had a visual acuity of 6/60 in the involved eye.

The one case of orbital dermoid cyst presented at 3 years. The child underwent a subtotal surgical excision and at present the visual acuity is normal with no sign of recurrence.
To distinguish tumours, such as haemangiomas, as being likely to be malignant in nature. In our experience, this is most frequently required when a rhabdomyosarcoma is suspected.

For optic nerve and chiasm lesions. It is important to liaise with the pathologist. Frozen sections are useful to ensure that the correct tissue has been biopsied.

To date, the duration of a haematoma (the state of and location of the haemoglobin and its breakdown and, in the case of an excision biopsy, that the margins are clear of tumour. Specimens should be provided fresh or in liquid nitrogen for immunohistochemistry, in formalin for light microscopy and in glutaraldehyde for electron microscopy.

Treatment of proptosis in children depends on the cause and is often multidisciplinary. Children with orbital tumours require management with input from a number of paediatric subspecialities, including ophthalmology, oncology, radiotherapy and neurosurgery. More than any other group in our series, they illustrate the need for a multidisciplinary approach to the diagnosis, treatment and total management of a child presenting with proptosis.

Other investigations that need to be considered, depending on the clinical presentation are presented in Table 3.

The principal indication for orbital biopsy is to obtain tissue diagnosis when proptosis is due to a mass lesion, which is likely to be malignant in nature. In our experience, this is most frequently required when a rhabdomyosarcoma is suspected.

### REFERENCES


