

Globe luxation in histiocytosis X

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SUMMARY A boy with histiocytosis X, first diagnosed at the age of 9 months, presented at the age of 5 years with left globe luxation. Under general anaesthesia the globe was reduced and a temporary paracentral tarsorrhaphy carried out. He was started on a course of oral steroids. Two years later he had normal visual acuity, full ocular movements, and healthy fundi.

Histiocytosis X, Langerhans cell histiocytosis, is an uncommon condition in which non-malignant histiocytes proliferate and invade various organs including skin, bone, liver, spleen, lung, and bone marrow. In most cases in children the condition is self-limiting, though a few have progressive disease and die from failure of infiltrated organs.^{1,2} Orbital involvement occurs in 20-25% of patients with histiocytosis X, and proptosis is the main ophthalmic problem.³⁻⁵

Globe luxation is rare, generally being precipitated by raised intraorbital pressure,⁶ lid manipulation,^{7,8} or trauma⁹ in patients with shallow orbits,^{10,11} lax orbital ligaments,¹² or anomalous extraocular muscles.¹³ Expanding orbital tumours may produce marked proptosis, but actual globe luxation is exceedingly rare. We describe a 5-year-old boy with orbital involvement by histiocytosis X who developed a luxated left globe.

Case history

At the age of 9 months this boy was diagnosed as having histiocytosis X when he presented with a cystic swelling behind his left ear, a follicular maculopapular rash, hepatosplenomegaly, miliary shadowing on chest x-ray, and a skull x-ray with multiple osteolytic lesions. Biopsy of both skin and the mass behind his left ear confirmed the diagnosis of histiocytosis X. Treatment was started with vinblastine, oral prednisone, and cyclophosphamide. On this regimen the histiocytosis X resolved, but over the subsequent three and a half years he had three further relapses involving the skin, left mastoid, and occipital region and was treated with courses of

vinblastine and prednisone and irradiation to the mastoid.

At the age of 3 he developed bilateral proptosis, more marked on the left. Investigations again revealed multiple bony lucencies in the skull. His proptosis was resistant to vinblastine and prednisone, and VP16 produced only minimal improvement. He therefore received radiotherapy (100 cGy) to both orbits at the age of 3 years 11 months. The proptosis did not completely respond, and at the age of 5 he was again put on prednisone 20 mg daily, and because of left lagophthalmos a lateral tarsorrhaphy was carried out. This produced a slow response, and the steroid dosage was reduced to 10 mg of prednisone on alternate days. A CT scan of his skull showed persistent bony defects of the middle cranial fossa, especially the lesser wing of the sphenoid bone and the petrous temporal bone.

At the age of 5 years 3 months he presented as an emergency with left globe luxation, which occurred while he was straining and which would not spontaneously reduce. His left visual acuity was counting fingers only, he had a hazy left cornea, and, while there was no definite afferent pupil defect, the left pupil showed a very sluggish reaction to light. A CT scan showed a much enlarged retroorbital mass (Fig. 1). Under an anaesthetic the luxated globe was reduced, a paracentral tarsorrhaphy performed, and the dose of prednisone increased to 60 mg daily. This produced a substantial reduction in the proptosis, and after three weeks the tarsorrhaphy was opened. During the subsequent 10 months the dose of steroids was gradually reduced to 15 mg on alternate days. Two years later the left eye shows no evidence of any permanent damage due to the luxation (Fig. 2). He had a visual acuity of 6/6 in both eyes, a full range of ocular movements, and healthy optic discs and fundi.

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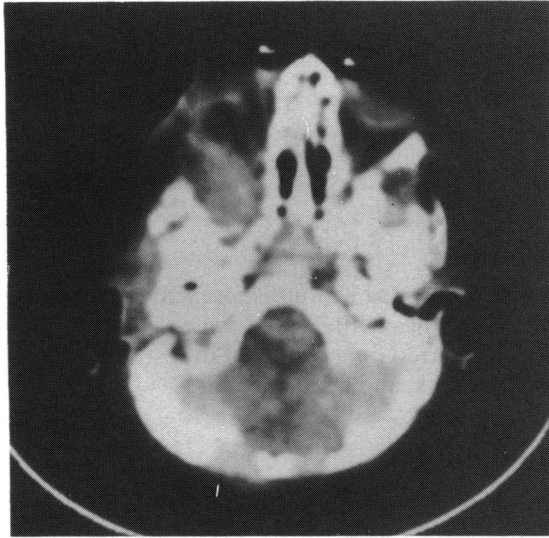


Fig. 1 CT scan of orbits showing luxated left globe with a large retroorbital mass.

Visual evoked responses to a pattern reversal stimulus with a 50-minute check at 95% and 50% were entirely within normal limits with respect to both latency and amplitude from each eye.

There have been no further episodes of globe luxation.

Discussion

Orbital involvement in histiocytosis X usually occurs in the chronic multisystem form of the condition which was previously called Hand-Schüller-Christian disease and rarely presents in the acute multisystem variety of Letterer-Siwe disease.³⁴ In one series with only soft tissue involvement the orbits were not affected, suggesting that they become so from adjacent bony involvement.³ The commonest signs of ophthalmic involvement are bilateral or unilateral

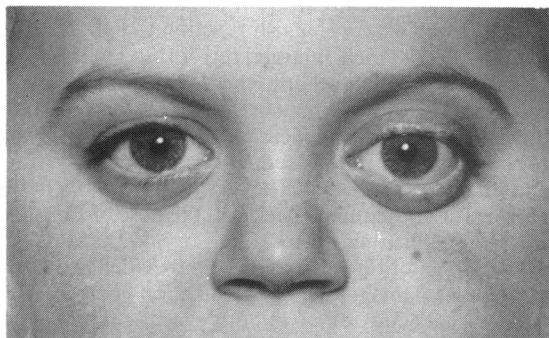


Fig. 2 Patient at 2½ years and after globe luxation.

proptosis and less frequently papilloedema with optic atrophy.^{3,5} When bilateral papilloedema is present it is often associated with central nervous system disease, and it may result in bilateral optic atrophy with visual impairment.³ Globe luxation is not described in several reviews of histiocytosis X.^{3,5}

In most cases of globe luxation due to either a shallow orbit or lax orbital ligaments reduction is easily achieved, and the patients have a good visual prognosis.¹⁴ In contrast luxation due to space occupying lesions is much more likely to cause loss of vision, and the management of the patient often involves orbital decompression,¹⁵ tumour excision, and very occasionally enucleation.¹⁶

As the majority of lesions in histiocytosis X are self-limiting, conservative treatment without jeopardising vision should be attempted. In the present case simple reduction supported by a temporary tarsorrhaphy and oral steroids produced a very satisfactory long-term result. The pre-existing lateral tarsorrhaphy which had been carried out to prevent exposure keratitis may have produced a tighter orbit, which would increase intraorbital pressure and increase the risk of luxation. Further, once the luxation had occurred, the smaller than normal palpebral aperture would have hindered reduction of the globe.

Radiotherapy,¹⁷ excision of the tumour mass, orbital decompression, or intralesional steroids are alternative approaches. Radiotherapy had already been administered in this case, and the other methods were not only more invasive than that adopted but also more likely to have an adverse effect on vision and ocular motility.^{18,19}

Severe proptosis in histiocytosis X should be carefully observed, and, if symptoms are progressive, therapy should be directed to preventing globe luxation. If this does occur, then conservative management may produce the best long-term visual prognosis. Lateral tarsorrhaphy should be avoided, for although it relieves corneal exposure it may increase the long-term risk of globe luxation.

References

- 1 Broadbent V, Pritchard J. Histiocytosis X—current controversies. *Arch Dis Child* 1985; **60**: 605–7.
- 2 Writing Group of the Histiocyte Society. Histiocytosis syndromes in children. *Lancet* 1987; i: 208–9.
- 3 Moore AT, Pritchard J, Taylor DSI. Histiocytosis X: an ophthalmological review. *Br J Ophthalmol* 1985; **69**: 7–14.
- 4 Lucaya JL. Histiocytosis X. *Am J Dis Child* 1971; **121**: 289–95.
- 5 Sims DG. Histiocytosis X. Follow-up of 43 cases. *Arch Dis Child* 1977; **52**: 433–40.
- 6 Duke-Elder S, MacFaul PA. *System of ophthalmology*. London: Kimpton, 1974; **13**: (2): 1233.
- 7 Carlson RE, Scheribel KW, Hering PJ, Wolin L. Exophthalmos, global luxation, rapid weight gain: differential diagnosis. *Ann Ophthalmol* 1982; **14**: 724–9.

- 8 Mertens PS. Subluxation of the eyeball. *Am J Ophthalmol* 1922; **5**: 290.
- 9 Chhabra HN, Kawuma AMS. Luxation of the eyeball. *Br J Ophthalmol* 1986; **70**: 150-1.
- 10 Fowler JG. Spontaneous luxation of the eyeball. *JAMA* 1941; **116**: 1206-8.
- 11 Mailer CM, Mawas E, Parizot H, Rceb J. Spontaneous luxation of the eyeballs. *Br J Ophthalmol* 1969; **53**: 846-53.
- 12 Offenbach B. Dislocation (luxation) of the eyeball. *N Engl J Med* 1954; **251**: 338-9.
- 13 Lyle DJ, McGavic JS. The cause of voluntary forward luxation of the eyeball. *Am J Ophthalmol* 1936; **19**: 316.
- 14 Walsh FB, Hoyt WF. *Clinical neuro-ophthalmology* 3rd ed. Baltimore: Williams and Williams, 1969; **1**: 702-4.
- 15 Wilson J, Von Haacke NP, Adams GGW, Dale BAB. Surgical decompression of the orbit. *J R Coll Surg Edinb* 1986; **31**: 22-5.
- 16 Takabatake M, Fujisaki H, Mori R. Two cases of orbital haemangioma. *Folia Ophthalmol Jpn* 1977; **28**: 217-20.
- 17 Ilzuka S, Hamada Y, Sakiyama T, et al. A case with metastatic orbital tumour of breast cancer. *Folia Ophthalmol Jpn* 1978; **24**: 842-4.
- 18 McCormick MS, Stell PM. Orbital decompression for progressive exophthalmos. *J R Coll Surg Edinb* 1986; **31**: 18-21.
- 19 Fells P. Orbital decompression for severe thyroid eye disease. *Br J Ophthalmol* 1987; **71**: 107-11.

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