

Prevention of stimulus deprivation amblyopia in children with eyelid capillary haemangioma

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Abstract

Aim: To present a case of eyelid capillary haemangioma with complete eye closure in an infant.

Methods: A case is discussed of a 5-week-old baby with eyelid capillary haemangioma. The ophthalmic and orthoptic findings are documented, with photographs. Investigation and treatment are described to illustrate the measures taken to prevent stimulus deprivation amblyopia of the affected eye.

Results: Due to rapid growth of the haemangioma there was complete eye closure with an inability to open the eye. Full-time occlusion of the non-affected eye was commenced and combined with steroid treatment of the haemangioma. The haemangioma reduced in size significantly. At the age of 53 weeks, visual acuity appears equal and binocular vision responses are demonstrable.

Conclusion: Total occlusion of the good eye to prevent stimulus deprivation amblyopia of the affected eye is an option when methods of keeping the affected eye open have failed.

Key words: Haemangioma, Occlusion, Stimulus deprivation amblyopia

Introduction

Haemangioma can affect about 1 in 10 babies, 60% of whom are affected in the head and neck area. Orbital haemangiomas are of particular concern as they can either obstruct the visual axis or exert pressure on the globe, depending on their location.¹ This in turn can result in a range of visual problems including stimulus deprivation amblyopia, induced astigmatism, anisotropic amblyopia or squint.² The following case study illustrates an alternative measure taken to prevent stimulus deprivation amblyopia in an infant who presented with a large capillary eyelid haemangioma.

Case report

Five weeks of age

A 5-week-old baby presented with a left upper lid capillary haemangioma. At the time of presentation the

haemangioma did not impinge on the visual axis, vision both eyes open was age-appropriate and there was no obvious manifest deviation. Binocular single vision tests were not possible due to the young age of the baby. On examination, the haemangioma appeared to be limited to the upper eyelid, sitting on the superior margin of the left pupil and being entirely preseptal (no image available). The patient was found to have healthy fundus and media with no significant refractive error for age.

Immediate treatment was not deemed necessary. However, an intralesional steroid injection was scheduled for 6 weeks ahead in anticipation of growth of the haemangioma to a significant size. The parents were informed of the importance of a clear visual axis and instructed to contact the department if there was any significant change.

Eight weeks of age

Three weeks later, the parents reported a sudden rapid increase in the size of the haemangioma, which left the infant unable to open the left eye (Fig. 1). The haemangioma extended into the orbit and the cornea could not be visualised even with mechanical retraction with Desmarres retractors. The left upper eyelid could not be taped up.

The child was immediately admitted to the ward for intravenous pulse methylprednisolone for 3 days to stop progression of the haemangioma. This was followed by a 2 mg/kg oral dose of the steroid prednisolone, which was tapered over 15 weeks (Table 1). Further refractions at this point were not possible due to the size of the haemangioma.

At this time, the decision was taken to occlude the right eye to prevent stimulus deprivation amblyopia of the left eye. The parents were advised to occlude the right eye for all waking hours minus one (Fig. 2); they were not given an occlusion chart. Babies of this age sleep on average for 16 hours and are awake for 8 hours. This child had a similar sleep pattern and was therefore on occlusion for approximately 7 hours a day.

Nine to 14 weeks of age

At 9 weeks, a good response was noted to the steroid treatment as the eye was beginning to open. It was noted that the oral steroids were containing the growth of the haemangioma but not resolving it, so at 14 weeks of age

Table 1. Treatment table

Age (weeks)	State of haemangioma	Steroid treatment	Occlusion treatment
8	Complete left lid closure. Cornea could not be visualised even with mechanical retraction	Intravenous pulse methylprednisolone for 3 days, followed by a 2 mg/kg dose of oral prednisolone	TORE full-time except 1 hour (6–7 hours)
10	LE much improved – beginning to open	Oral prednisolone reduced to 1.5 mg/kg daily	No change
11	LE no real change	Oral prednisolone reduced to 1 mg/kg daily	No change
14	LE opens when RE is occluded	Intralesional steroid injection of 20 mg triamcinolone and 2 mg dexamethasone combined with oral prednisolone	No change
17	Left lid obscures upper half of pupil	Prednisolone 5 mg reducing by 1 mg every 2 weeks	TORE 5–6 hours daily
22	No change	Steroid treatment stopped	TORE 4 hours daily
27	Left lid covering half of pupil	Prednisolone 3 mg reducing by 1 mg every 2 weeks	TORE 2 hours daily for next 7 weeks
34	Slight increase in size of haemangioma. Visual axis clear	No steroid treatment for last week. Restarted: prednisolone 1 mg OD for 1 month then stop	Occlusion stopped

LE, left eye; RE, right eye; TORE, total occlusion of the right eye.

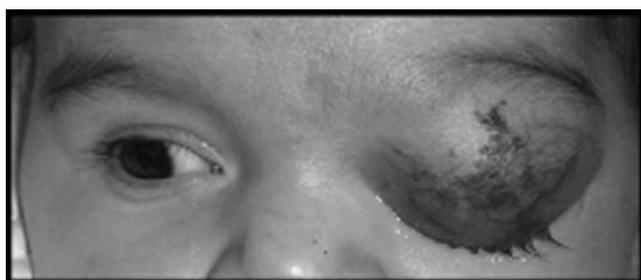


Fig. 1. Rapid growth of the haemangioma within 2 weeks of initial presentation.



Fig. 3. The patient aged 43 weeks showing the residual bulk of the haemangioma following steroids.



Fig. 2. The patient on occlusion therapy.

an intralesional steroid injection of 20 mg triamcinolone and 2 mg dexamethasone was administered combined with the oral steroid (prednisolone).

During this period total occlusion of the right eye continued (Table 1). The left eye was now beginning to open, allowing visual acuity testing of this eye. The vision of the child was recorded as right 1.6 cy/cm and left 0.64 cy/cm with Teller acuity cards.

Seventeen to 22 weeks of age

At 17 weeks of age there was a significant reduction in the size of the haemangioma so that only half the pupil was obscured. Occlusion was reduced to 5–6 hours a day and the dose of oral steroid was reduced. The vision was right and left eyes 2.4 cy/cm, although the left eye response was slightly slower.

At 22 weeks occlusion was reduced to 4 hours a day, and vision was right: 6.5 cy/cm, left: 4.8 cy/cm. Oral steroids were stopped.

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Twenty-seven weeks of age

By 27 weeks of age the haemangioma had increased slightly in size. Oral steroids were restarted with the view to gradually tapering off. Visual acuity was equal with no squint detected; therefore, occlusion was reduced to 2 hours a day.

Forty-three weeks of age

At 43 weeks of age a residual bulk of the haemangioma remained which did not impinge on the visual axis and all treatment was stopped (Fig. 3). Refraction revealed right +2.50 DS, left +1.50 DS/+2.00 DC × 90 and glasses were prescribed for full-time wear.

Fifty-eight weeks of age

At the most recent assessment the child was 58 weeks old. Visual acuity with glasses was right and left eyes 4.8 cy/cm, and 6.5 cy/cm both eyes open. On cover test there was no manifest deviation with or without glasses. The response to a 20 dioptre base-out prism was normal and the patient achieved 550" of arc on the Lang stereotest.

Discussion

The infant described presented with a capillary haemangioma of the left eyelid, which increased rapidly in size to cause complete eye closure, an inability to open the eye and the risk of stimulus deprivation amblyopia. Schwartz *et al.*³ reported that haemangiomas of the eyelids and orbit measuring less than 1 cm in the greatest dimension did not have any associated amblyogenic

factors. In contrast, 53% of patients whose lesion measured greater than 1 cm became amblyopic in the affected eye.

If possible, haemangiomas are left to resolve without treatment because of the risks involved with steroid injections and surgery. Any resulting visual problems are then treated with glasses and occlusion therapy as necessary.^{4,5} However, where there is a risk of visual impairment, as in this case, early intervention is necessary.

The current literature concentrates on whether steroids or surgical intervention is the better option in order to avoid problems. Levi *et al.*⁶ advocate the use of surgery, especially before the age of 13 months, to reduce astigmatism. This is supported by Slaughter *et al.*,⁷ who reported surgery to be effective with very few complications. Morrell and Willshaw⁸ used steroid injections and reported a reduction in haemangioma of at least 25% in 81% of patients studied, with a marked reduction in the amount of astigmatism.

Steroids are generally the preferred choice of treatment even though they give a slower rate of reduction in size of the haemangioma compared with surgery. Generally steroids can affect appetite, weight gain and immunisations. Although this child was well whilst on the steroids, care had to be taken with vaccinations. Current recommendations regarding immunisations are to continue with all killed vaccines (a vaccine prepared from dead micro-organisms) whilst on steroid treatment (although these may have to be repeated), and to avoid live vaccines.

Treatment with propranolol has recently been found to be effective with fewer side-effects.⁹ However, there is insufficient research on its use to enable a comparison with steroids.

There is sparse literature on the preservation of good vision in the affected eye whilst a patient is awaiting treatment for a haemangioma or whilst a haemangioma is resolving. Taping up the upper eyelid to clear the visual axis and ensuring that the eyes are regularly lubricated has been discussed.¹⁰ Scleral immersion shells such as those used for A-scans have also been described to keep the eye open but require lubrication every 5 minutes and therefore are not practical.¹¹ This report illustrates an alternative method to prevent stimulus deprivation amblyopia in the affected eye.

The frequency of stimulus deprivation amblyopia in general is reported to be approximately 3%. Due to the severity of this type of amblyopia, it has been found to have a poor prognosis despite intensive treatment.^{12,13} In this case, vision was compromised at 8 weeks of age and the decision was made to occlude the non-deprived eye immediately, even though a uniocular measure of visual acuity had not been obtained. The vision and growth of the haemangioma were carefully monitored, with the amount of occlusion and dose of oral steroids adjusted as necessary. At the most recent review this child had equal visual acuity, measured with preferential looking, and no manifest strabismus.

At present, it is difficult to determine what the long-term prognosis is for good visual acuity in each eye and binocular single vision (BSV) in this child. Although preferential looking tests have been found to be useful in monitoring interacuity difference in young babies,¹⁴ these can overestimate vision. For this reason the aim is to continue monitoring visual acuity and BSV responses.

Conclusion

Total occlusion of the good eye to prevent stimulus deprivation amblyopia of the affected eye is an option when methods of keeping the affected eye open have failed. The authors intend to provide an update on the visual outcome for this child at age 3 to 4 years.

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