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A review of 58 patients with periorbital haemangiomas to determine appropriate cases for intervention☆

Russell J. Bramhall [a], **Awf Quaba** [b,*]

[a] *Glasgow Royal Infirmary, 84 Castle Street, Glasgow G4 0SF, UK*
[b] *Department of Plastic Surgery, St John's Hospital at Howden, Howden Road West, Livingston EH54 6PP, UK*

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Summary *Background:* Periorbital haemangiomas cause severe effects on visual development. They can be treated with some success, but there is currently no literature on the selection of patients for intervention. Our study aims to review all periorbital haemangioma cases in Edinburgh over the past 20 years, to set more stringent guidelines regarding the appropriate selection of patients for treatment and determine prognostic indicators of visual outcome.
Methods: A review of 58 patients since 1980 with significant lesions, seen by ophthalmologists and/or plastic surgeons.
Results: 82% of those presenting were female. The mean age of presentation was 4 months and 2 months in more serious cases. Mean follow up was for 5.6 (range 0—15) years. Loss of visual field was the most sensitive prediction of the development of visual complications (67%), but least specific (76%). Pressure on the globe and pupillary obstruction were more specific (77—100%), but less sensitive (33—50%).
Conclusions: Loss of visual field, partial/complete pupillary obstruction and pressure exerted on the globe are all valuable prognostic indicators. The size and position of the lesion are also relevant, but are probably due to these factors. Based on these results we recommend specific guidelines for the selection of patients for intervention.

Haemangiomas are the most common tumour of infancy. The incidence in the general population has been reported by different studies as 1.0—2.6%[1,2] and 5—12%.[3,4] They are more common in girls with a female:male ratio usually reported in the region of 3:1.[6—8] Haemangiomas can present as a superficial strawberry naevus, dark blue/purple subcutaneous

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* Corresponding author. Tel.: +44 (0) 1506 419666.
E-mail address: marion.mikkelsen@wlt.scot.nhs.uk (A. Quaba).

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lesion, deep orbital tumours or any combination of these.[9] Diagnostic factors include an increase in size on crying and the presence of other non-periocular marks.[8] Less than 30% are present at birth, but they undergo a rapid proliferative phase and 90% are apparent within the first 4 weeks.[6] However, most slowly regress with 50% having involuted by age 5 years, 75% by 7 years and 90–100% by 10 years.[2,10,11]

Current guidelines on recommended treatment have proved difficult to establish with variations in practice existing between units. However, the current treatment of choice is usually intralesional steroid injections. Since these lesions tend to regress naturally, a 'wait-and-see' policy is often preferred. There have been numerous publications concerning the advantages and disadvantages of treatment modalities, but the question of whether to treat at all has attracted much less attention. Whilst there are a number of prognostic indicators which suggest a greater threat to the development of normal vision, decisions remain exceedingly speculative concerning the initiation of active treatment. Massive lesions or those resulting in significant pupillary obstruction are invariably treated. However, guidelines are much less clear about the treatment of smaller haemangiomas, which may only reduce the visual fields slightly, not obstructing the pupil. This is particularly uncertain with lesions of the lower lid and glabellar, since these are less likely to present a visual obstruction. The primary issue appears to be judging whether the haemangioma is causing enough pressure on the globe to result in a refractive error. This also causes amblyopia, but due to anisometropia rather than deprivation. Patients with periorbital haemangiomas are usually referred to ophthalmology for an assessment of predicted visual disturbance if the growth is causing either obstruction or pressure on the globe.

This study aims to collate all appropriate information regarding the periorbital haemangioma cases seen in this unit over the past 20 years. With this we aim to set more stringent guidelines regarding the appropriate selection of patients requiring treatment. The use of obstruction to visual field and pressure on the globe as determinants for active intervention will be assessed and any alternative factors reported.

Patients and methods

Patients

This study collected information from the case notes of the 62 patients with periorbital haemangiomas who have attended The Royal Hospital for Sick Children (Edinburgh) or St John's Hospital (Livingston) between 1980 and 2005. Of these, four patients have absent notes concerning treatment received elsewhere. All relevant information for the remaining 58 patients in this report has been retrieved regarding attendance to other plastics and ophthalmology units.

Data

The details extracted were those of their initial presentation, management, follow up and a description of their condition. Every patient had photographs taken at each attendance in the plastics unit.

This information was used to compare the visual complications evident in different groups of patients. The effect of size, location, the level of obstruction to vision, and pressure subjected on the globe were investigated. The locations compared were those affecting the upper lid only, upper lid/eyebrow, lower lid, glabellar, and those that constituted massive/multiple facial lesions. The lower lid category includes larger haemangiomas of the cheek that extended superiorly to include the eyelid.

Measurements

As measurements were rarely recorded and only relevant in relation to the size of the patient, size was measured from photographs to be the maximum diameter of the deep component in relation to the width of the palpebral fissure. The distance from the estimated centre of the pupil to the centre of the deep component was also measured to assess the relevance of proximity to the orbit (see Fig. 1). These measurements were recorded with the software package Image Tool 3.0. The average of three recordings of each measurement was used. Fifty patients had photographs of the lesion at its peak severity.

Analysis

The validity of using the lesions' size and proximity to the eye, obstruction to visual field and pressure on the globe as prognostic indicators in the decision to treat was investigated by calculating their sensitivity and specificity for predicting visual complications. The consistency, reasoning and general trends in treatment decisions were also investigated by comparing prognostic factors and treatment undertaken. Any patients who had never been referred to an ophthalmologist were assumed to have no visual complications for the purposes of this study, unless contrary evidence was available.

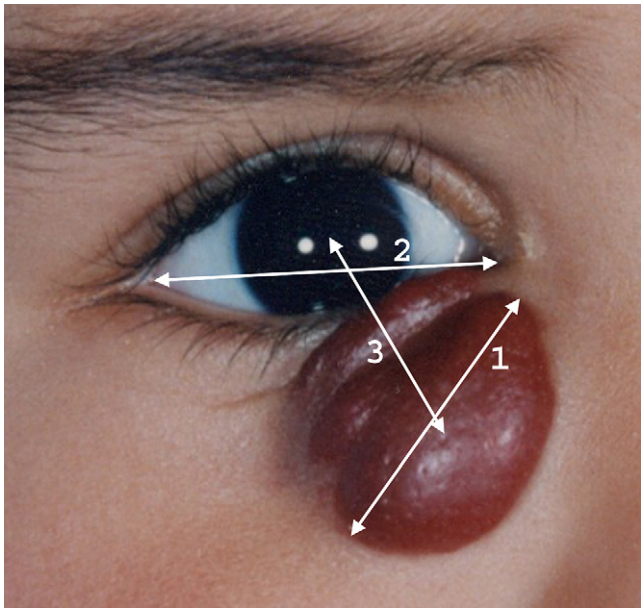

Figure 1 Size measurements. Size of lesion (1), width of palpebral fissure (2) and proximity of lesion to globe (3).