SCIENTIFIC REPORT

An ultrasound based classification of periocular haemangiomas

R J C Bowman, K K Nischal, K Patel, J I Harper

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Aims: To propose a classification system for periocular haemangiomas based on ultrasound evaluation.

Methods: Retrospective review of ultrasound images from children seen in the authors' unit with periocular haemangiomas. Static ultrasound images from 50 patients with periocular haemangiomas were reviewed as identified from a computerised database. Each haemangioma ultrasound image was classified into three categories: (1) preseptal only; (2) preseptal + extraconal; (3) preseptal + extraconal + intraconal. These were compared with the categories given to each patient at first presentation after dynamic scanning.

Results: Classification was possible from the static images in 44 (88%) cases. Of those classified 20 (45%) were preseptal only; 17 (39%) were preseptal + extraconal, and seven (16%) had an additional intraconal component. The classification in all 44 cases was the same as that given at the time of presentation. In the small number of cases which went to surgery or had neuroimaging, the ultrasound classification was confirmed.

Conclusions: Ultrasound classification was not difficult to perform and no child needed sedation or general anaesthesia for this exam. Ultrasound anatomical classification is an important first step in determining appropriate treatment of periocular haemangiomas. The authors present what they believe to be the first such classification.

Capillary haemangiomas are the most common orbital tumour of childhood.[1] They also occur elsewhere in the body and one study estimated that 10% of children under the age of 1 year have a visible haemangioma somewhere.[2] The tumours consist of anastamosing vascular channels with no true encapsulation. They have an unusual natural history usually presenting in the first few weeks of life (approximately 30% being visible at birth) and then undergoing a rapid proliferation and growth phase 3–6 months after diagnosis, followed by a slower phase of natural resolution such that approximately 30% of lesions may have resolved by the age of 3 years and 70% by the age of 7 years.[3] Despite this high rate of spontaneous resolution, periocular haemangiomas may cause permanent visual loss through amblyopia. The amblyopia may be anisometropic (caused by induced astigmatism), strabismic (caused by induced deviation of the eyeball), stimulus deprivational (caused by mechanical ptosis), or any combination of the above three. Such lesions may require treatment. Factors which influence the choice of treatment include the patient's age, visual status, and general health but also the anatomical extent and location in skin of the lesion. Previous anatomical classifications of these lesions have been based on clinical or surgical findings. We have found ultrasound helpful in determining the anatomical relations of the haemangioma, guiding treatment choice. In this paper we describe the ultrasound findings in a series of patients with periocular haemangioma and propose a classification system for these lesions based on ultrasound appearances.

METHODS

A retrospective study was conducted reviewing and classifying static ultrasound images from 50 patients with periocular haemangiomas obtained from the ultrasound database. In clinical practice, anatomical classification was made at the first consultation using dynamic scanning. Salient images were stored on the hard disc and copies made for the records. Ultrasonography was performed using an Acuson Sequoia 512 (Siemens AG, Munich, Germany), with a 15L8 linear probe held only in the horizontal and vertical planes with colour flow map overlay where necessary. The linear probe was found to be preferable to the vector design particularly for superficial preseptal lesions. The static images from the hard disc were reviewed, described, and classified by a single observer (KKN) according to their relation to two anatomical landmarks: the orbital septum and the cone of extraocular muscles. Three broad categories were therefore derived: (1) preseptal only; (2) preseptal + extraconal, and (3) preseptal + extraconal + intraconal. Category 1 therefore has no intra-orbital component whereas both 2 and 3 do involve the orbit. The orbital septum may be seen with ultrasound imaging (fig 1). The relation to the muscle cone was usually judged using Colour Flow Map overlay and the proximity of unusual high flow regions to the optic nerve.

In a small number of cases which underwent surgery and/or had neuroimaging, the ultrasound classification was compared to the anatomical findings at surgery.

RESULTS

Classification was possible from the static images in 44 (88%) cases. In the remainder not enough information was available on the captured images to make a definitive classification. Of those classified 20 (45%) were preseptal only, 17 (39%) were preseptal + extraconal, and seven (16%) had an additional intraconal component. Examples of each of these categories are shown in figures 2 to 4. Five of the 17 extraconal lesions were predominantly preseptal with only a small finger like projection through the septum (fig 1). The classification in all 44 cases was the same as that given at time of presentation when dynamic scanning was used to define extent of the lesion. In four cases surgical excision was performed and the ultrasound anatomical classification was confirmed by the surgical findings (two preseptal lesions which were completely excised and two preseptal + extraconal lesions which underwent subtotal excision). In one case neuroimaging was performed as well and this confirmed the ultrasound findings (fig 4).

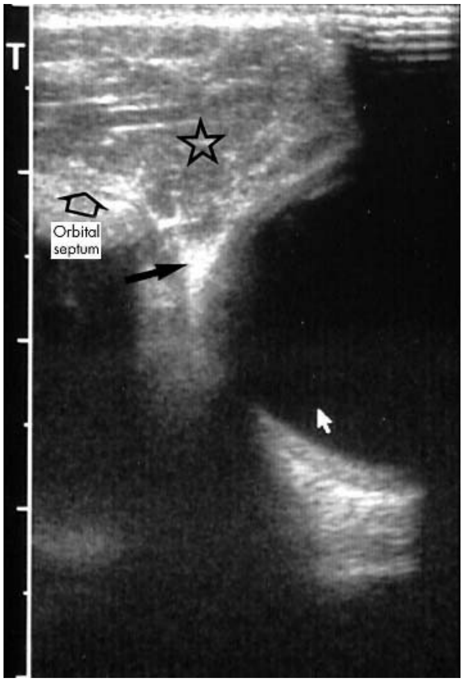

Figure 1 Ultrasound of a mainly preseptal haemangioma which has a finger like projection (black arrow) into the extraconal space of the orbit. The orbital septum is clearly seen (white arrow = vitreous cavity; star = haemangioma).

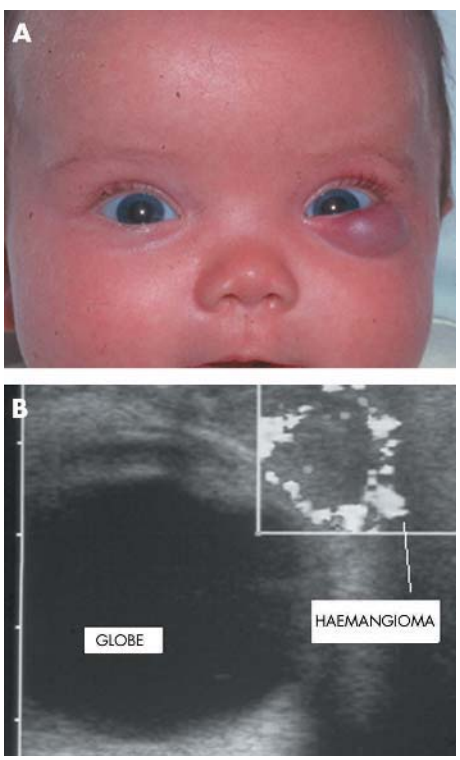

Figure 2 (A) Clinical photograph of a left lower lid deep periocular haemangioma. The term ''deep'' refers to the position of the haemangioma in the skin, hence its bluish appearance. (B) Ultrasound of case shown in (A). This is a vertical scan; the left of the picture is superior and the right is inferior. Colour Flow Map overlay confirms the presence of a haemangioma which is purely preseptal. (Reproduced with the parents' permission.)

Figure 3 (A) Clinical photograph of a deep periocular haemangioma. (B) A vertical ultrasound scan of the case shown (A). Again the left of the picture corresponds to the superior part and the right to the inferior part of the orbits. The haemangioma extends into the extraconal portion of the orbit. (C) Axial MRI of the case shown in (A). The haemangioma is seen to be preseptal and extraconal confirming the ultrasound findings. (Reproduced with the parents' permission.)

DISCUSSION

Previous classification of these tumours have included Haik's which divided them into superficial strawberry naevi (25%), bluish subcutaneous (68%), and deep orbital (7%).[4] Rootman described them slightly differently: superficial strawberry naevus (40%), combined subcutaneous and deep orbital (30%), and combined superficial strawberry and subcutaneous (30%).[5]

The ultrasound classification we present here is simple, easy to perform (even from static images, 88% in this series, and dynamic scanning is used in practice), and does not need sedation or a general anaesthetic unlike other imaging modalities such as CT or magnetic resonance imaging.

Both A and B scan ultrasound findings have previously been described as characterised by variable internal reflectivity.[6] Areas of low reflectivity are thought to correspond to solid hypercellular regions of endothelial proliferation, areas of moderate reflectivity to ectatic vascular channels, and areas of high reflectivity to fibrous septae separating tumour lobules. Computed tomography (CT imaging) often reveals a soft tissue mass in the anterior orbit or as an extraconal mass with finger like posterior projections such as those seen in five of our cases using ultrasound.[1] The CT tumour image enhances with iodinated contrast medium, and major feeding vessels supplying the tumour may be depicted. Using magnetic resonance (MR) imaging the tumour is of intermediate intensity on T1 weighted studies (hypointense to orbital fat and isointense to muscle).[7] The lesion is hyperintense on T2 weighted studies caused by slow blood

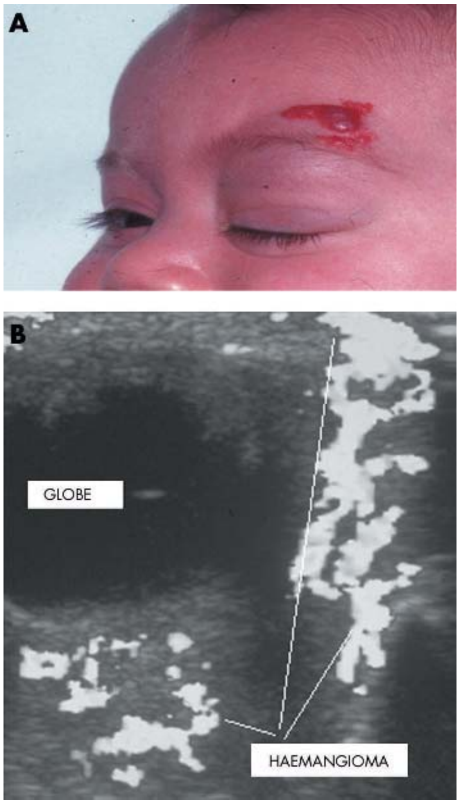

Figure 4 (A) Clinical photograph of a mixed haemangioma (superficial (bright red component) and deep (bluish component)). (B) Horizontal ultrasound scan of case shown in (A). The left of the picture is nasal and the right of the picture is temporal. The Colour Flow Map overlay confirms that the haemangioma has an extraconal and intraconal component. (Reproduced with the parents' permission.)

flow within the tumour mass; major feeding vessels appear as black serpigenous structures caused by the ''flow void'' phenomenon (rapidly moving blood in large vessels outpaces the stimulation-reception parameters of the MR sequence).[1] Although CT and MR imaging may both yield more detail in regard to the anatomical relations of the tumour, both would require a general anaesthetic or sedation in this age group whereas ultrasound does not because it is quick, painless, and requires minimal co-operation from the child. Surgical findings, although only available in four cases, confirmed the ultrasound classification in all four providing some support for its validity. In one case neuroimaging was performed as well and this confirmed the ultrasound findings (fig 3).

At our unit we use a management protocol for children with periocular haemangiomas which considers age, anatomical extent, location in the skin, and visual effect. The location of the haemangioma in the skin is defined as superficial (usually bright red), deep (usually bluish), or mixed (see figs 2–4). A deep skin lesion may be purely preseptal, whereas one with a superficial component may have orbital extension (see figs 2 and 4).

The ultrasound classification defines the anatomical extent of the lesion more accurately than previously described clinical classifications which tend to confuse orbital extent and location in skin.[4 5] Its importance lies in the fact that it may help define appropriate treatment—for example, a preseptal intralesional steroid injection is unlikely to be effective when the tumour is deep and intraconal.

We believe that ultrasound anatomical classification is an important first step in determining appropriate treatment of periocular haemangiomas. We present the first such classification to the best of our knowledge.

Authors' affiliations

R J C Bowman, K K Nischal, Department of Ophthalmology, Great Ormond Street Hospital for Children, London, UK
K Patel, Department of Radiology, Great Ormond Street Hospital for Children, London, UK
J I Harper, Department of Dermatology, Great Ormond Street Hospital for Children, London, UK

Correspondence to: Mr K K Nischal, Consultant Ophthalmologist, Great Ormond Street Hospital for Children, Great Ormond Street, London WC1N 3JH, UK; kkn@btinternet.com

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