Periorbital hemangiomas: the need for active management - Report of two cases

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Abstract: Hemangioma is the most common tumor of childhood and is commonly located on the head or neck. The orbit is often affected and early and aggressive intervention is required to prevent serious visual complications. This paper reports on two cases. In the first case, the patient’s vision was impaired, while in the second case a deep hemangioma affecting adjacent areas was confirmed radiologically. Treatment with aggressive systemic corticotherapy was successful, thus avoiding permanent damage to the patients’ vision. Furthermore, esthetic outcome was satisfactory. The treatment of choice is oral corticosteroids and management should be individualized and should include careful follow-up to monitor possible adverse effects.

Keywords: Adrenal cortex hormones; Glucocorticoids; Hemangioma; Hemangioma, capillary; Hemangioma, cavernous; therapeutics

INTRODUCTION

Hemangiomas are the most common tumors in childhood, affecting more female than male children in a proportion that ranges from 2:1 to 5:1.1 They affect 10-12% of children; however, this proportion increases to over 20% in premature infants, principally those weighing under 1000 grams at birth.2,3,5

The most common sites are the head and neck, with the orbit being the site of superficial hemangiomas in 4% of biopsies performed on children.1,3–5

CASE REPORT

Case #1: RN, a 4-month old baby girl, was brought to the dermatology clinic by her mother because of a voluminous congenital hemangioma on her face that had been preventing her right eye from opening since birth. The mother complained that her daughter’s mouth was permanently open, resulting in salivary incontinence, with her tongue continuously protruding from her mouth (Figure 1). Examination showed a superficial, voluminous, low-flow, reddish vascular tumor with well-defined borders and paler central areas of regression, completely occluding the child’s right eye, occupying much of the right hemiface and deforming her lip. Diagnosis of a voluminous, superficial periorbital hemangioma was confirmed by tomography, which excluded the possibility of intracranial involvement. Treatment was implemented with methylprednisolone at a dose of 1 mg/day, reducing the dose gradually over a 6-month period. With treatment, the lesion gradually lightened in color until complete clinical remission was achieved, leaving only residual surface telangiectasia. Photographs were taken monthly to record clinical response. Treatment was multidisciplinary and included periodic evaluations by a pediatrician and ophthalmologist. The patient’s family was instructed with respect to tempo-
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546 Pereira PMR, Chirano CAR, Lima LL, Mariano AVO, Romero SAR


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In the remaining cases, they appear in the first month of life, as occurred in the case of patient #2 whose mother noticed only a purplish stain at the site of the lesion in the first month of her baby’s life. The phase of progressive growth of the tumor is from 6 to 12 months of age, hence the importance of the early implementation of active therapy in those patients who require treatment, since this is the phase in which response to treatment is best. Generally, growth ceases by 18 months of age and this phase is followed by a period of slow involution until 10 years of age. The first sign of involution is a central greyish discoloration that is seen more easily in superficial hemangiomas such as that in case #1, which gradually grew lighter in color. Residual skin alterations such as scarring, atrophy, redundant discolored skin and telangiectasia remain in over 40% of children. These are found principally in voluminous tumors of the face that result in disfigurement, as occurred in case #1 in which telangiectasia persisted after treatment. In this case, however, the family was satisfied, since they had initially been more concerned with esthetics and with the child’s constant salivation, because the mother had been wrongly informed at a healthcare clinic that she should wait until the child was two years of age before seeking treatment.

Deep hemangioma has been referred to as the most common form of orbital tumor. Its diagnosis is generally made by tomography or magnetic resonance imaging, which shows a hypointense mass on T1 and a hyperintense mass on T2, thus determining the extent and depth of the lesion. Clinically, it presents as a circumscribed, slowly growing subcutaneous, bluish or purplish mass. In general, proptosis is present, developing slowly, progressively and painlessly. In addition, there may also be visual impairment. In the infant in case #2, although the clinical appearance of the condition was mild, it was compatible with descriptions in the studies reviewed; however, the child had no visual abnormalities despite proptosis. More than half of all hemangiomas occur on the head or neck, and their distribution appears to follow the embryologic fusion planes and the facial metames- res. The sites that involve potential health complications or risk of life are the cervicofacial, periorbital and anogenital regions, the auricle, subglottis, lumbo-sacral region and the parotid gland. Hemangiomas at these sites should be treated as soon as diagnosed. When the tumor is located on the upper eyelid, the risk of visual complications increases, principally amblyopia (unilateral or bilateral loss of vision with no other apparent cause that results from occlusion of
vision by large lesions) and astigmatism due to direct compression of the ocular globe or to its retrobulbar expansion. 

Other potential risks include: proptosis with corneal damage, strabismus, anisometropia, myopia and even blindness. 

Immediate intervention is crucial, since sight may already be damaged after even as short a period as two weeks.

To our surprise, the child who had visual occlusion at 4 months had no visual sequelae. Early ophthalmological examination is mandatory prior to, during and 6 months after discontinuation of therapy.

Unfortunately, in these two cases, interdisciplinary collaboration within the public healthcare network proved difficult and there were delays. Some ophthalmologists refused to accompany the cases because the patients concerned were children, making examination more difficult because the child, particularly the newborn infant, would be unable to remain still.

Systemic corticosteroids are the first line of therapy. In the proliferative phase, the drug induces regression in 30-90% of cases. IntraleSIONal application should be avoided in periorcular hemangiomas because of the risk of necrosis of the optic nerve, blindness and occlusion of the central retinal artery. Safe alternatives include the oral and topical routes of administration, although the latter is considered relatively ineffective for reducing consequent astigmatism.

If corticosteroid therapy proves ineffective, interventional therapy and its higher toxicity (spastic diplegia) constitute limiting factors. Surgical excision is reserved for cases in which pharmacological therapy has failed. Radiotherapy may be effective; however, it is avoided because of the risk of inducing malignancies and because it interferes with bone growth.

The use of corticoids for six months or more in children with hemangiomas often results in transitory adverse effects such as: Cushingoid facies, particularly in the first 1-2 months of therapy; personality alterations such as depression, agitation and insomnia in one-third of cases; delays in bone growth up to two years, although this is restored in the majority of children; and reversible gastric dyspepsia in 20% of cases. Hypothalamic-pituitary-adrenal axis suppression may develop after 2-4 weeks of supra-physiologic doses of glucocorticoid, increasing in frequency when drugs with a long half-life are used (intramuscular or intraleSIONal), when administration is divided into two or more doses per day or in the case of a single dose given at a time of the day other than the morning. Severe complications such as hypertension, cataract, glaucoma with irreversible blindness, aseptic necrosis of the hip and osteoporosis are rare in children. The latter is difficult to measure, since there are no standard measurements of bone mineral density for children; therefore, physicians should be attentive to any bone symptom or evidence of calcium loss.

Immunosuppression due to a reduction in lymphocyte migration should be taken into consideration and live and live attenuated vaccines should be contraindicated. This recommendation is fundamental, as shown in case #1 in which there was a problem of discordance between the advice given by the pediatrician, who advised the family not to treat the child because of the risk of severe preventable diseases as a result of not vaccinating the infant, and that given by ourselves. Here, the family was informed regarding the risks associated with the child’s lack of use of her right eye and they were assured that their baby’s health would be taken care of. The guidance provided by this service persuaded the parents to agree to treatment.

Despite the natural benign course of the majority of hemangiomas, periorbital lesions require early intervention in order to preserve the child’s sight. Management should be individualized and multidisciplinary.

REFERENCES


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