CASE REPORT

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

Hemangiomas periorbitários: necessidade de conduta ativa - Relato de dois casos

Priscilla Maria Rodrigues Pereira[1]
Carlos Alberto Chirano Rodrigues[2]
Lívia Lima de Lima[3]
Adriana Valkira de Oliveira Mariano[1]
Sandra Adolfina Reyes Romero[1]

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

Resumo: Hemangioma é o tumor mais comum da infância, frequentemente situado na cabeça e pescoço. A órbita é frequentemente acometida e indica intervenção precoce e agressiva para evitar sérias complicações visuais. Reportam-se dois casos, nos quais há impedimento da visão no primeiro e, no segundo, um hemangioma profundo acomete áreas adjacentes, confirmado por exame radiológico. Demonstra-se sucesso terapêutico após corticoterapia sistêmica agressiva, evitando sequelas visuais permanentes, além do resultado estético satisfatório. O tratamento de escolha é o corticosteroide oral, devendo ser conduta individualizada e com bom seguimento clínico dos possíveis efeitos adversos.
Palavras-chave: Corticosteróides; Glucocorticóides; Hemangioma; Hemangioma capilar; Hemangioma cavernoso; Terapêutica

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. [2-5] 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-

Received on 10.12.2009.
Approved by the Advisory Board and accepted for publication on 17.05.2010.
* Study conducted at the Alfredo da Matta Foundation, Manaus, Amazonas, Brazil.
Conflict of interest: None / *Conflito de interesse: Nenhum*
Financial funding: None / *Suporte financeiro: Nenhum*

[1] Physician currently undergoing medical residency in Dermatology at the Getúlio Vargas Teaching Hospital of the Federal University of Amazonas, Manaus, Amazonas, Brazil.
[2] Clinical and surgical dermatologist, Alfredo da Matta Foundation; Preceptor in the Medical Residency Course in Dermatology, Alfredo da Matta Foundation, Manaus, Amazonas, Brazil.
[3] Physician currently undergoing medical residency, Alfredo da Matta Foundation, Manaus, Amazonas, Brazil.

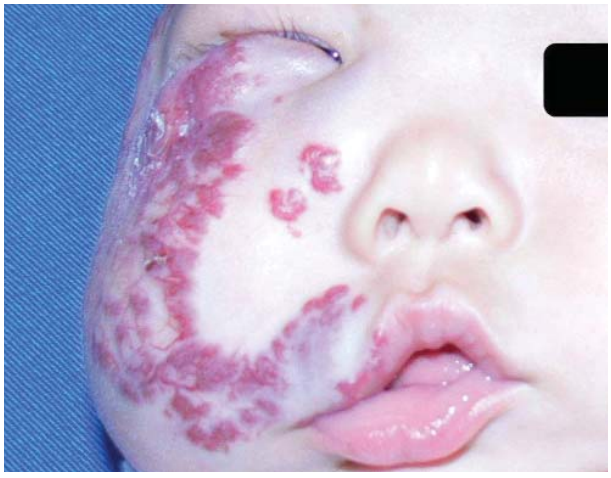

Figure 1: Case #1. Prior to treatment. A voluminous mixed hemangioma deforming the right hemiface and the lip, with a permanently open mouth and complete obstruction of the ipsilateral visual axis

rary immunosuppression and told to return to the dermatology clinic if they had any questions. Use of live and live attenuated vaccines was suspended during treatment. The baby gained weight and developed Cushingoid facies, which disappeared after discontinuation of therapy (Figure 2). The ophthalmologist detected no abnormalities and reported normal reflexes and normal sight. The child is currently healthy and undergoing three-monthly outpatient follow-up. Her growth and psychomotor development are compatible with her age (Figure 3).

Case 2: A one-year old baby girl, whose mother reported a purplish swelling on the right interpalpebral area that had been present since she was one month old. Examination showed the presence of a purplish, compressible, clearly outlined swelling on

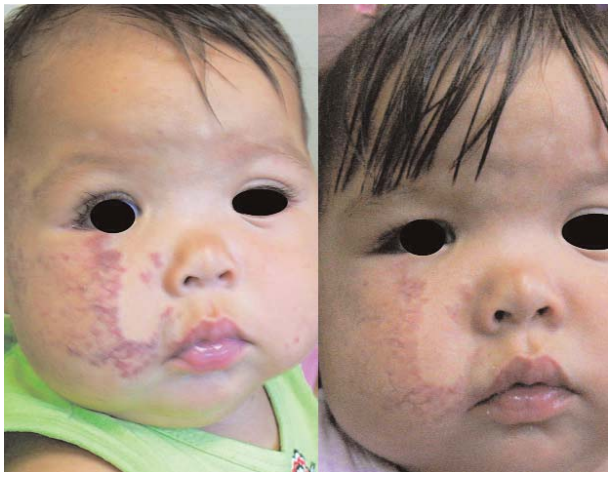

Figure 2: Case #1. Evolution of treatment. Left: significant clinical remission with a considerable improvement in the color of the lesion at 2 months of treatment. Right: Cushingoid facies at 4 months of corticosteroid therapy

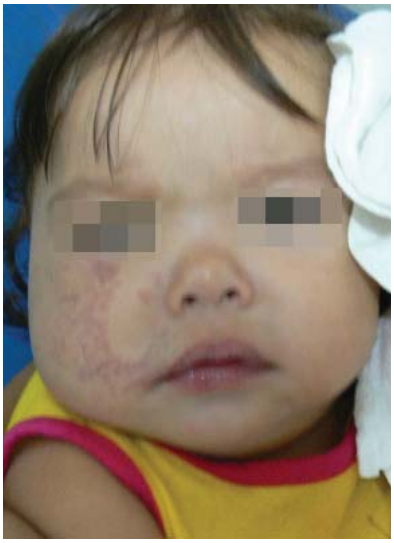

Figure 3: Case #1. Post-treatment. Child with residual telangiectasia on face six months after treatment

the lower right eyelid measuring 2.5 cm in diameter (Figure 4). No other skin lesions were found. Computed tomography of the orbit showed an expansive lesion with soft tissue density, multilobulated appearance and serpiginous areas, involving the lateral rectus and inferior rectus ipsilateral muscles, with medial displacement of the optic nerve, causing mild proptosis. In addition, the right orbital bone cavity had been widened (Figure 4). The contralateral orbital structure was normal and the possibility of extracranial involvement was excluded. The patient was diagnosed with a deep orbital hemangioma and referred to a pediatrician and ophthalmologist. No ophthalmological abnormalities were found. Treatment was initiated with prednisolone at a dose of 3 mg/kg/day, reducing the dose gradually over a 5-month period. This treatment resulted in remission, as shown clinically and by imaging exam (Figure 5). The child's family was informed regarding temporary immunosuppression, and the use of live virus and attenuated live virus vaccines was suspended during treatment. Post-treatment tomography showed complete remission of the vascular tumor, with no signs of involvement of the ocular globe, and normal extrinsic muscles. The thickness and trajectory of the optic nerve were normal, and the ocular globes were symmetrical (Figure 5). The child is currently healthy and is being followed up regularly, with no evidence of any vascular skin lesions.

DISCUSSION

The incidence of hemangiomas in newborn infants generally ranges from 1 to 2.6%, increasing, however, to over 10% in white infants. [3,6] Contrary to reports in the literature in which hemangiomas are found predominantly in white infants, both patients in the present case report are brown-skinned and, in fact, patient #1 is of native indian descent. Around half of all hemangiomas are present at birth, although it is unusual for them to be completely developed at

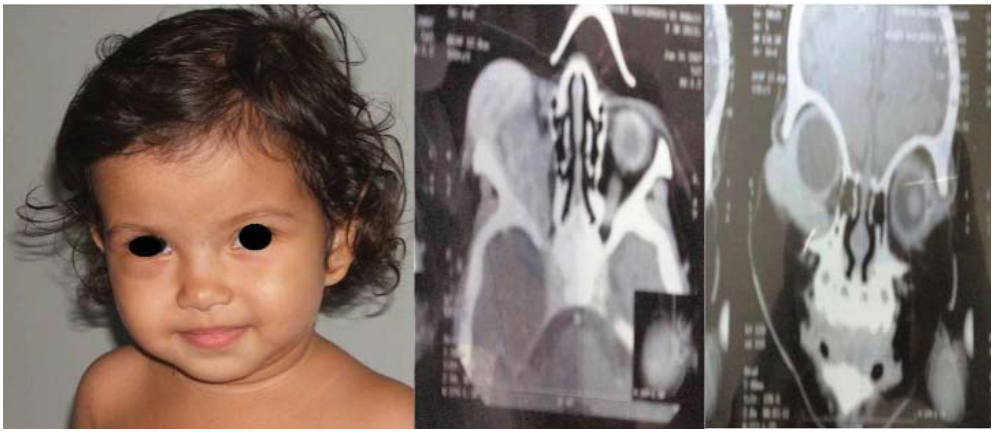

Figure 4: Case #2. Clinical and imaging findings prior to treatment. Deep interpalpebral hemangioma on the right side, with imaging exam showing proptosis of the ipsilateral ocular globe

this time. 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. [3] 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. [5] These are found principally in voluminous tumors of the face that result in disfigurement, [3] 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. [7] 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 circumscript, slowly growing, subcutaneous, bluish or purplish mass. [7] In general, proptosis is present, developing slowly, progressively and painlessly. In addition, there may also be visual impairment. [7] 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, [5] and their distribution appears to follow the embryologic fusion planes and the facial metameres. [3,4,8] The sites that involve potential health complications or risk of life are the cervicofacial, periorbital and anogenital regions, the auricle, subglottis, lumbosacral region and the parotid gland. [3,4] 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

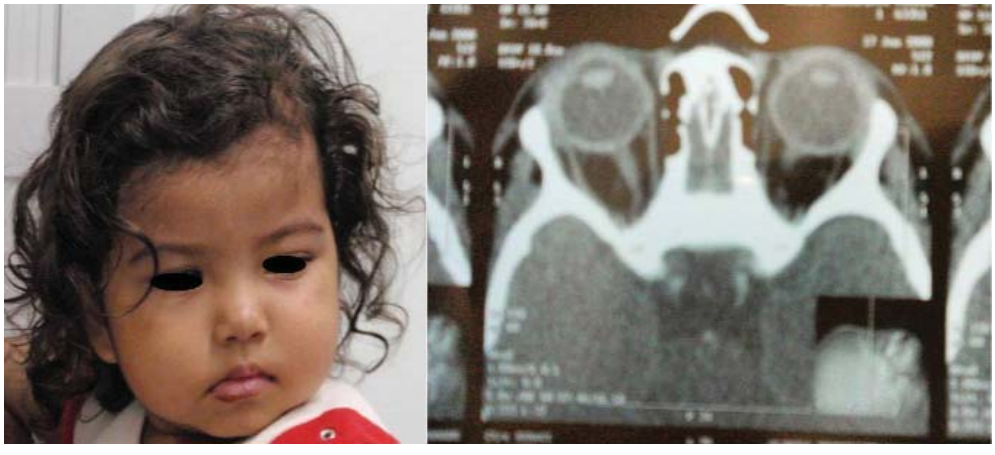

Figure 5: Case #2. Clinical and imaging findings following treatment. Clinical examination and imaging exam showing remission of the vascular lesion, with the ocular globe structures returning to normal, as shown in the supplementary test

vision by large lesions) and astigmatism due to direct compression of the ocular globe or to its retrobulbar expansion. [3,4] Other potential risks include: proptosis with corneal damage, strabismus, anisometropia, myopia and even blindness. [3,6] Immediate intervention is crucial, since sight may already be damaged after even as short a period as two weeks. [3,4,6,9,10] 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. [9] 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. [9.10] In the proliferative phase, the drug induces regression in 30-90% of cases. [3,4] Intralesional application should be avoided in periocular hemangiomas because of the risk of necrosis of the optic nerve, blindness and occlusion of the central retinal artery. [3] Safe alternatives include the oral and topical routes of administration, although the latter is considered relatively ineffective for reducing consequent astigmatism. [3] If corticosteroid therapy proves ineffective, interferon-alpha is a good option, although the high cost of this treatment and its higher toxicity (spastic diplegia) constitute limiting factors. [3,9] Surgical excision is reserved for cases in which pharmacological therapy has failed. [9] Radiotherapy may be effective; however, it is avoided because of the risk of inducing malignancies and because it interferes with bone growth. [9]

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. [3,10] 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. [2] Severe complications such as hypertension, cataract, glaucoma with irreversible blindness, aseptic necrosis of the hip and osteoporosis are rare in children. [2,3] 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. [2]

Immunosuppression due to a reduction in leukocyte migration should be taken into consideration and live and live attenuated vaccines should be contraindicated. [3] 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

1. Waner M, North PE, Scherer KA, Frieden IJ, Waner A, Mihm MC Jr. The Nonrandom Distribution of Facial Hemangiomas. Arch Dermatol. 2003;139:869-75.
2. George ME, Sharma V, Jacobson J, Simon S, Nopper AJ. Adverse Effects of Systemic Glucocorticosteroid Therapy in Infants With Hemangiomas corticoide. Arch Dermatol. 2004;140:963-9.
3. Metry DW, Hebert DW. Benign cutaneous vascular tumors of infancy: when to worry, what to do. Arch Dermatol. 2000;136:905-14.
4. Ersoy S, Mancini AJ. Hemifacial Infantile Hemangioma with Intracranial Extension: A Rare Entity. Pediatr Dermatol. 2005;22:309-13.
5. Gontijo B, Silva CMR, Pereira LB. Hemangioma da infância. An Bras Dermatol. 2003;78:651-73.
6. Dubois J, Milot J, Jaeger BI, McCuaig C, Rousseau E, Powell J. Orbit and Eyelid Hemangiomas: Is There a Relationship Between Location and Ocular Problems? J Am Acad Dermatol. 2006;55:614-9.
7. Sullivan TJ, Aylward GW, Wright JE, Moseley IF, Garner A. Bilateral multiple cavernous haemangiomas of the orbit. Br J Ophtalmol. 1992;76:627-9.
8. Chiller KG, Passaro D, Frieden IJ. Hemangiomas of Infancy: Clinical Characteristics, Morphologic Subtypes, and Their Relationship to Race, Ethnicity, and Sex. Arch Dermatol. 2002;138:1567-76.
9. Ranchod TM, Frieden IJ, Fredrick DR. Corticosteroid treatment of periorbital haemangioma of infancy: a review of the evidence. Br J Ophthalmol. 2005;89;1134-8.
10. Boyd MJ, Collin JRO. Capillary haemangiomas: an approach to their management. Br J Ophthalmol. 1991;75;298-300.

MAILING ADDRESS / ENDEREÇO PARA CORRESPONDÊNCIA:
Priscilla Maria Rodrigues Pereira
Av. Engenheiro Santana Junior, 1345 - Bloco A - Apto 301, Papicu
60175650 Fortaleza (CE) – Brasil
E-mail: pris17@hotmail.com

How to cite this article/*Como citar este artigo*: Pereira PMR, Chirano CAR, Lima LL, Mariano AVO, Romero SAR. Periorbital hemangiomas: the need for active management - a report of two cases. An Bras Dermatol. 2011;86(3):545-8.