

Presentation, progression and treatment of cutaneous hemangiomas – Experience of the Outpatients Clinic of Pediatric Dermatology - Hospital das Clínicas da Universidade de São Paulo*

*Apresentação, evolução e tratamento dos hemangiomas cutâneos – Experiência do Ambulatório de Dermatologia Infantil do Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo**

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Abstract: BACKGROUND - Hemangiomas are benign proliferations of endothelial tissue. They are the most common tumors of childhood and the majority has typical presentation and growth patterns.

OBJECTIVES - To report the experience of the Outpatients Clinic of Pediatric Dermatology of the Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo regarding presentation, progression and treatment of hemangiomas in childhood and to correlate them with literature data.

METHODS - We retrospectively evaluated 40 cases of hemangioma seen between March 1994 and November 2004, through analysis of medical and photographic records.

RESULTS - There was a predominance of females - 3:1 - and white patients (75%). Hemangiomas were present at birth in 77% of patients. In 85% of cases, the lesions were single and the majority was located in the head and neck (57%). The conservative management was adopted in most cases and the follow-up of patients showed involution of lesions started before 12 months of age in 65% of cases. The complete resolution occurred up to 3 years of age in 50% of patients, up to 6 years in 58%, up to 9 years in 83%, and up to 11 years in 100%.

CONCLUSION - Our cases are similar to those in the international literature in terms of gender and ethnicity, number and location of hemangiomas, although we have a higher percentage of lesions seen at birth.

Keywords: Clinical evolution; Epidemiologic studies; Hemangioma

Resumo: FUNDAMENTOS – Os hemangiomas, proliferações benignas do tecido endotelial, são os tumores mais comuns da infância, e a maioria possui apresentação e crescimento típicos.

OBJETIVOS – Relatar a experiência do Ambulatório de Dermatologia Infantil do Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo em relação à apresentação, evolução e ao tratamento dos hemangiomas da infância, correlacionando-os com dados da literatura.

MÉTODOS – Foram avaliados retrospectivamente, mediante análise de prontuários e registros fotográficos, 40 casos de hemangiomas atendidos no período de março de 1994 a novembro de 2004.

RESULTADOS – Houve predomínio do sexo feminino na razão de 3:1, e maior número de pacientes brancos (75%). O hemangioma estava presente ao nascimento em 77% dos pacientes. As lesões eram únicas em 85% dos casos, e a maior parte se localizava na região da cabeça e pescoço (57%). A conduta expectante foi a mais adotada, e o seguimento dos pacientes mostrou início da involução da lesão antes dos 12 meses de idade em 65% dos casos. A resolução total ocorreu até três anos de idade em 50% dos pacientes, até os seis anos em 58%, até os nove anos em 83% e até os 11 anos em 100%.

CONCLUSÕES – A casuística dos autores é superponível à da literatura mundial em relação às variáveis sexo e cor dos pacientes, número e localização dos hemangiomas, porém com maior porcentagem de lesões presentes ao nascimento.

Palavras-chave: Estudos epidemiológicos; Evolução clínica; Hemangioma

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Conflict of interests: None

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INTRODUCTION

For a long time there was some confusion in the terminology of vascular lesions. A better standardization started to be developed in 1982, when Mulliken and Glowacki proposed a classification in which the vascular anomalies were divided into hemangiomas and vascular malformations, based on their histopathological features, clinical and biological behavior.^{1,3} The International Society for the Study of Vascular Anomalies (ISSVA) modified this classification in 1996 and opted to divide the vascular lesions into vascular malformations (capillary, venous, lymphatic, arterial and combined) and vascular tumors. The latter have the marked characteristic of proliferation of endothelial cells and are divided into hemangioma, rapidly involuting congenital hemangioma, non-involuting congenital hemangioma, pyogenic granuloma, Kaposi-like hemangioendothelioma and tufted angioma.^{1,4,5}

Hemangioma is the most common benign tumor in childhood and it is estimated to affect 10-12% of children in their first year of life. It occurs more often in females and premature children, and it is observed in 23-30% of children who weigh less than 1,000 grams at birth.^{1,3,5,6} This condition can occur in individuals of any ethnicity, but there seems to be some a predominance in light-skinned individuals.^{3,6} The lesions are single in 80% of cases, and the most affected areas include the head and neck (60%), trunk (25%) and limbs (15%).^{1,5,7}

The clinical aspect of hemangiomas varies according with the depth of tumor involvement. Childhood hemangiomas are divided into superficial, deep and combined types. The superficial hemangiomas involve the superficial layer of the dermis and they are clinically seen as elevated, well-limited and bright red lesions surrounded by normal skin and poorly compressible upon palpation. The deep hemangiomas affect the deep dermal layers and the subcutaneous tissue, thus sparing the papillary dermis. They present as skin color or bluish, compressible nodules, and they can present telangiectasias in their surface and drainage vessels in the peripheral areas.^{1,2,5}

Most of the childhood hemangiomas present a typical growth pattern, and their main features include rapid proliferation and spontaneous involution. They are generally not apparent at childbirth, although a precursor lesion may exist in 30-50% of cases and manifests in different ways; telangiectasias surrounded by a pale halo, erythematous, anemic or ecchymotic maculae are the most common precursor lesions.^{1,5}

The natural evolution of childhood hemangiomas can be divided into three stages: proliferative, regressive and involuted.⁵ In general, hemangiomas

start their growth stage as early as in the first week of life, although some deeper lesions may start growing at a later stage. The initial proliferative stage of fast growth usually lasts between three to nine months and the majority does not go beyond 18 months. The duration of involution may vary greatly among children but it normally occurs gradually for a period of two to six years and it tends to be complete between 7 and 10 years of age.² In the natural history of hemangiomas, 30% of lesions involute up to 3 years of age, 50% up to 5 years of age and 90% up to nine years of age. In the stage of an already involuted hemangioma, several patients remain with residual lesions which may present clinically as redundant skin, atrophy, telangiectasias, scars and other symptoms.^{3,5}

The management of hemangiomas remains controversial, and each case must be carefully analyzed. Since they have a natural trend to recede spontaneously, watchful waiting is the most frequently adopted option. In selected cases, the currently available treatments include the use of systemic corticoid, intralesion corticoid, surgery, laser, cryotherapy, recombinant interferon alfa, chemotherapy and arterial embolization (restricted to those cases in which the hemangioma causes congestive heart failure and poses a risk to the patient's life).^{5,8-10}

This study was conducted with the purpose of reporting the experience of the Outpatients Clinic of Pediatric Dermatology of the *Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo* in regard to the presentation, progression and treatment of hemangiomas. The data obtained were compared with those from the international literature, contributing to reinforce the features of this vascular abnormality, thus helping physicians to choose the best management in each case.

PATIENTS AND METHODS

A retrospective study was carried out with a total of 40 patients evaluated at the Outpatients Clinic of Pediatric Dermatology of the *Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo*, between March 1994 and November 2004, who were diagnosed with hemangiomas.

Medical and photographic records of each patient were examined, and data related to gender and ethnicity; type, number and location of hemangiomas; their progression and treatment used were also obtained. The follow-up of progression of lesions was based on photographs, descriptions in medical records and data obtained by means of patient calling. Involution and age of total resolution of the hemangioma were also analyzed.

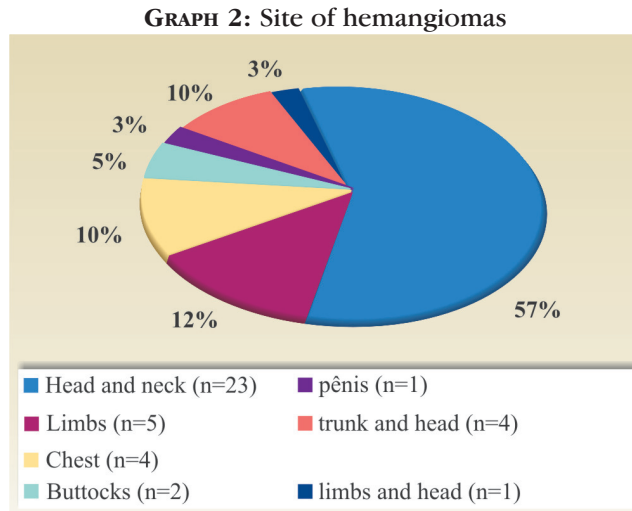
RESULTS

Out of 40 patients with hemangioma, 30 were female and 10 were male. The data show that 30 patients were white, 8 were mulattos and in 2 of them this information could not be obtained. In regard to the age of onset, most patients (77%) reported the presence of such lesions at birth (Graph 1). On physical examination, 34 patients (85%) presented one lesion, one patient (2.5%) presented two lesions and 5 patients (12.5%) presented three lesions. As to the location of the hemangiomas, there was some predominance in the head and neck (23 patients). Four patients presented lesions in the chest, 5 patients had lesions in the limbs, 2 patients presented lesions in the buttocks and one patient presented a lesion in the penis. Five patients showed involvement of more than one region, and in all these patients, one of the affected areas was the head and neck (Graph 2).

Watchful waiting was adopted in 30 out of 40 cases (75%) and it was based on the periodical inspection of the lesion (return visit every 6 months), photographic records and instructions to parents (Graph 3).

The patient's age at the first visit ranged from 2 months to 3 years (mean age of 8 months). Data about the beginning of the involution of hemangiomas were obtained in 26 of such patients, showing that in 17 cases (65%) the hemangioma involution started before 12 years of age and in 18 cases (69%) it took place before 18 months of age (Graph 4).

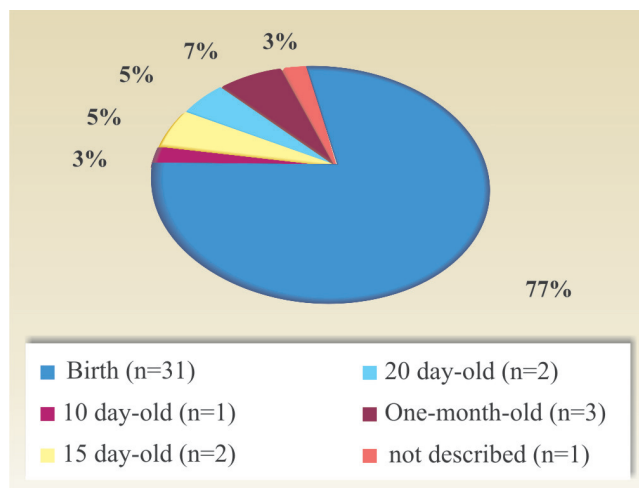
Total involution of hemangiomas was seen in 12 out of 40 cases. The age at which resolution occurred ranged from 9 months to 11 years (mean age of 5.4 years). In half of these patients, involution occurred up to the age of 3 years, in 58% it happened up to the age of 6 years, in 83% it happened up to the age of 9 years, and in 100% of the cases it occurred up to the age of 11 years (Graph 5). The remaining cases



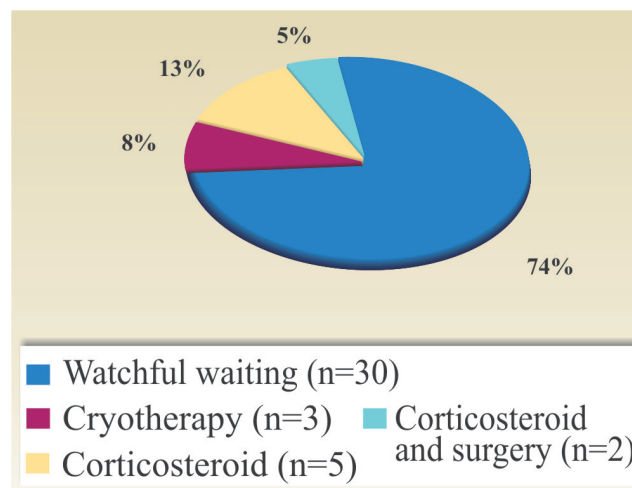
were still undergoing involution or they withdrew from the follow-up.

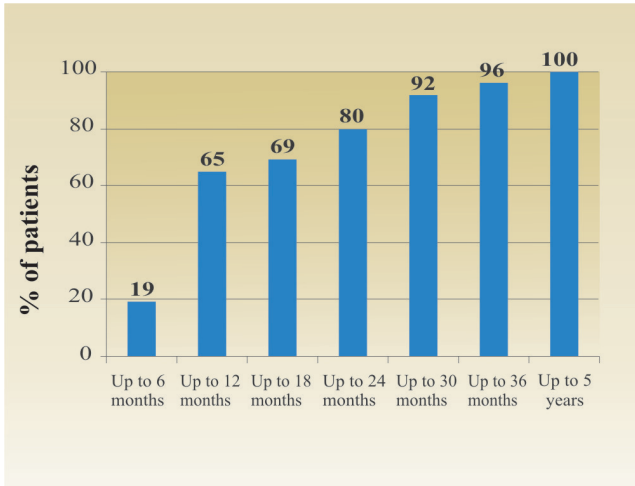
The patients were also divided into two groups depending on the features of the hemangiomas, which were classified as superficial (29 patients) or deep (11 patients). Watchful waiting was adopted in 23 patients with superficial hemangiomas and in 7 patients with deep hemangiomas. Total resolution of the lesion was seen in 8 out 23 patients with superficial hemangiomas in whom watchful waiting was adopted. The follow-up showed that half of these patients presented total resolution of the lesions up to the age of 3 years (mean age at resolution = 5.5 years). In 12 patients with superficial hemangiomas, the lesions were still undergoing involution at the time of data collection for this study and 3 patients dropped out.

GRAPH 1: Age at onset of hemangioma



GRAPH 3: Management adopted in cutaneous hemangiomas



GRAPH 4: Age at beginning of involution of hemangiomas

Watchful waiting was adopted in 7 out of 11 patients with deep hemangiomas, and 4 patients experienced total resolution. The patient's age at resolution varied from 9 months to 9 years (mean age of 5.2 years).

Ten patients received treatment, namely, cryotherapy or systemic corticotherapy. Two patients with deep hemangioma required the association of surgery for correction of the cicatricial lesions or entropion, and one patient was treated with Nd:YAG for the residual telangiectasias (Graph 3).

Three patients were treated with cryotherapy with varied responses. One patient reported relapse of the hemangioma after two sessions of cryotherapy. All patients were female and had two superficial hemangiomas (one in the supralabial area and another in the forehead) and a deep hemangioma (located in nasal dorsum).

Seven patients required systemic administration of corticoid drugs. The drug used was prednisone or prednisolone with a starting dose varying between 1 and 2 mg/kg/day. Four patients presented signs of involution of the hemangioma up to one month after the corticoid was introduced. One patient presented interruption of hemangioma growth although there were no signs of involution of the lesion even after 3 months of corticoid treatment. In two patients it was not possible to obtain sufficient data regarding the beginning of the involution process.

DISCUSSION

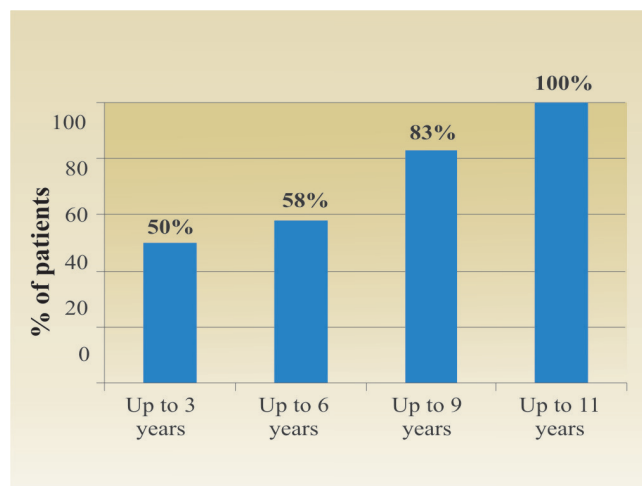
Data from the literature show that hemangiomas are seen more frequently among females in a ratio varying from 3:1 to 7:1 and light-skinned individuals seem to be more often affected. In our study there was also a predominance of females (3:1 ratio) and a larger number of white patients (75%). As to

age of onset of lesions, the incidence of hemangiomas at birth (77%) was higher than that described in the literature which varies between 30 and 50%. Just as described in the literature, the single lesions were more frequent (85%) and most hemangiomas were located in the head and neck (57%).^{1,5,7}

Although there is a great variation among patients, most of the lesions reach their maximum size by 9 to 12 months of age, and they start the involution process by 12 to 18 months.⁵ The evaluation of 26 patients revealed that, in 65% of cases, the onset of the involution process occurred before the age of 12 months and in 69% of those patients it occurred before the age of 18 months (Graph 4).

Data in the literature show that in the natural history of hemangiomas, 30% of all lesions undergo involution process until the age of 3 years, 50% until the age of 5 years and 90% until the age of 9 years. In our study, total resolution of hemangiomas occurred at a mean age of 5.4 years, with 50% of patients presenting resolution up to the age of 3 years, 58% up to the age of 6 years, 83% up to the age of 9 years and 100% up to the age of 11 years. There was no significant difference between the age of total resolution when deep and superficial hemangiomas were compared.

Watchful waiting was adopted in 74% of cases included in our study. Since the hemangiomas have a natural trend to resolve, in most cases the decision of not intervening and just having a periodical follow-up seems to be the best option. This approach was adopted preferably in cases of smaller hemangiomas located in areas that did not compromise the individual's development or function. It is important to point out that the follow-up involves not only the inspection of the lesion but it also includes its photographic

GRAPH 5: Age at total resolution of hemangiomas

records and continuous instructions to the parents.

The decision to introduce a specific treatment should be based on different factors such as the stage of progression at the moment of evaluation, growth rate, lesion size and location, risks and benefits of the proposed therapy and psychosocial implications of the tumor on the individual and his/her family.^{2,5,11}

Cryotherapy was used in three female patients. This can be the choice of treatment in cases of small lesions that may leave aesthetically poor scars. The possible formation of residual hypochromic maculae after cryotherapy always needs to be taken into account.

Seven patients were treated with prednisone or prednisolone. Therapy with systemic corticoid is indicated in patients with hemangiomas located in areas that may compromise vital structures or functions such as food intake, eyesight, hearing, intestinal discharge and breathing, or in patients with marked esthetical abnormalities. In this study, the medical indication in 4 cases was related with difficult opening of the eyes with a risk of amblyopia, and in 3 patients it was related with the extension of the hemangiomas

and their periorificial locations. Two patients with deep hemangiomas in the group treated with systemic corticoid therapy required the association of surgery for correction of the residual lesions.

CONCLUSION

This study reinforces the information about epidemiologic data and natural history of hemangiomas obtained in the literature and shows some differences as to age of onset of the hemangioma. This study with a follow-up of 10 years contributes with information about the follow-up of patients and evaluation of age at lesion involution; these data are rarely found in the Brazilian and Latin American medical literatures. Although the most appropriate management is still a subject of great discussion and it may vary among different institutions, the decision to just follow the involution continues to be the main approach adopted in the majority of hemangiomas due to their natural trend to spontaneous regression. However, in cases where a vital function may become compromised, early intervention should be adopted as soon as possible. □

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