CASE REPORT

Involution of a cystic hygroma of the face following local infection

Higroma cístico de face com involução após infecção local

Bárbara Nader Vasconcelos [1]
Marcela Duarte Villela Benez [2]
Aline Lopes Bressan [3]
Elisa Fontenelle de Oliveira [4]

Abstract: This report describes the case of a cystic hygroma on the face of a four-month old child. There was a history of congenital swelling of the right hemiface that decreased considerably following signs of infection. The cystic hygroma or lymphangioma is a rare congenital malformation of the lymphatic system that is present at birth in 50% of cases. It is usually located on the neck or face. It generally grows slowly and progressively and may compress and infiltrate adjacent structures. Its spontaneous regression occurs in only 6% of cases.
Keywords: Bleomycin; Infection; Lymphangioma, cystic; Lymphatic system

Resumo: Relatamos um caso de higroma cístico na face de uma criança de quatro meses de idade. Havia história de aumento de volume congênito na hemiface direita que involuiu consideravelmente após sinais de infecção. O higroma cístico ou linfangioma é uma malformação congênita rara do sistema linfático, presente ao nascimento em 50% dos casos. Localiza-se preferencialmente na região cervical e na face. Geralmente evolui com crescimento lento e progressivo, podendo comprimir e infiltrar estruturas adjacentes. Sua regressão espontânea ocorre em apenas 6% dos casos.
Palavras-chave: Bleomicina; Infecção; Linfangioma cístico; Sistema linfático

INTRODUCTION

The cystic hygroma or lymphangioma is a rare congenital malformation of the lymphatic system that is present at birth in 50% of cases.

CASE REPORT

A 4-month old, white female infant presented with congenital swelling on the right hemiface that developed suddenly in conjunction with a fever. At physical examination, the patient was found to be active and responsive, with a swelling on her right hemiface (Figure 1) that was soft and well defined, with a painful area of central hyperemia, extending into the submandibular region (Figure 2). No abnormalities were found in any other organ or system. Ultrasonography of the soft tissues was performed, showing an expansive, multiseptated, lobulated, cystic formation with some fine echoes in suspension, occupying the area of the right buccinator muscle. There was an apparent increase in echogenicity of the adjacent subcutaneous tissue, and no flow was detected using echo-Doppler, suggesting facial lymphangioma of the buccinator region. Computed tomography (CT) of the face and neck with venous contrast showed an expansive formation with a cystic, septated

Received on 10.10.2009.
Approved by the Advisory Board and accepted for publication on 11.05.2010.
* Study conducted at the Jesus Municipal Hospital and at the Pedro Ernesto Teaching Hospital, Rio de Janeiro, Brazil.
Conflict of interest: None / *Conflito de interesse: Nenhum*
Financial funding: None / *Suporte financeiro: Nenhum*

[1] Residency in Clinical Medicine at the Military Police Central Hospital. Postgraduate specialization in Dermatology at the Pedro Ernesto Teaching Hospital, State University of Rio de Janeiro (UERJ), Rio de Janeiro, Brazil.
[2] Residency in Clinical Medicine at the Armed Forces Central Hospital. Postgraduate specialization in Dermatology at the Pedro Ernesto Teaching Hospital, State University of Rio de Janeiro (UERJ), Rio de Janeiro, Brazil.
[3] Residency in Clinical Medicine at the Galeão Air Force Hospital. Postgraduate specialization in Dermatology at the Pedro Ernesto Teaching Hospital, State University of Rio de Janeiro (UERJ), Rio de Janeiro, Brazil.
[4] Substitute Professor of Dermatology, Pedro Ernesto Teaching Hospital, State University of Rio de Janeiro (UERJ), Rio de Janeiro, Brazil. Dermatologist at the Jesus Municipal Hospital. Head of the pediatric dermatology outpatient clinic at the Casa de Misericórdia, Rio de Janeiro, Brazil.

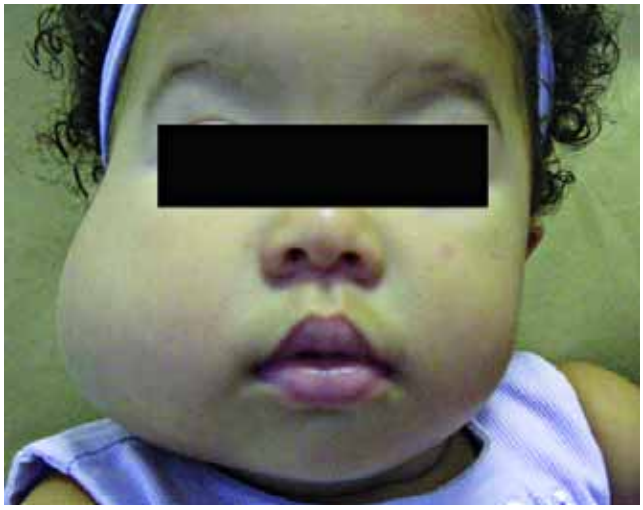

Figure 1: Cystic hygroma of the face. Swelling on the right hemiface

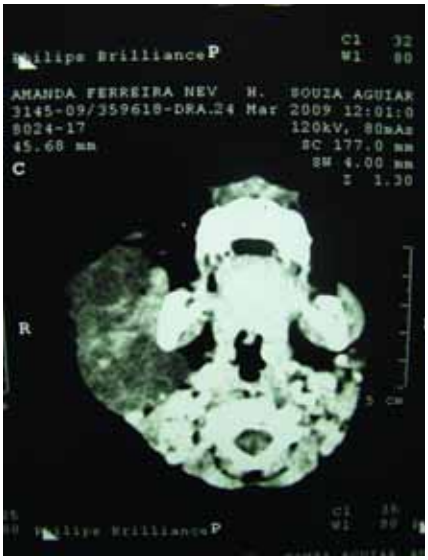

Figure 3: Computed tomography of the face with venous contrast. Expansive, cystic, septated formation measuring 5.8 x 4.8 x 4.0 cm on the right parotid gland

appearance, measuring 5.8 x 4.8 x 4.0 cm, on the right parotid gland (Figure 3), which was indistinguishable from the lesion. The CT also revealed enlarged lymph nodes located in the right submandibular chain and bilaterally in the internal jugular chains, in addition to which the maxillary sinus and the ethmoidal cells on the right side were sealed. The patient also had leukocytosis without left shift and an increase in erythrocyte sedimentation rate (ESR). Oxacillin was administered and treatment with intralesional bleomycin was proposed. However, one month after an infection of the expanding mass, the lesion began to involute without any other therapeutic intervention (Figure 4). The patient is currently being followed up clinically.

DISCUSSION

Cystic hygroma or lymphangioma is a rare congenital malformation of the lymphatic system diagnosed in children under two years of age. It is present at birth in 50% of cases. It is generally located on the neck or face, but may affect the axillas, lateral chest or mediastinum. [1] It probably occurs due to a fault in the formation of the communication between the lymphatic system and the jugular vein on the fourteenth day of pregnancy, leading to lymph stasis and the formation of cysts. [2] It consists of lymphatic cysts covered by an endothelial layer. They are classified in accordance with their size as macrocystic (cystic hygroma), microcystic (cavernous and capillary) or intermediary forms. [3]

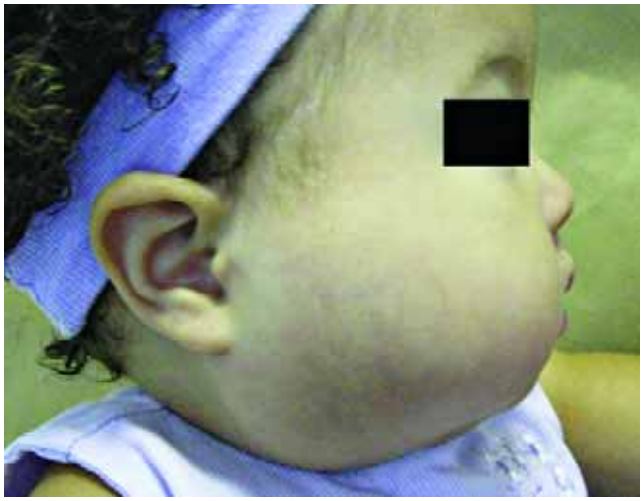

Figure 2: Profile of the lesion, showing the expanse of the mass, which extended into the submandibular region

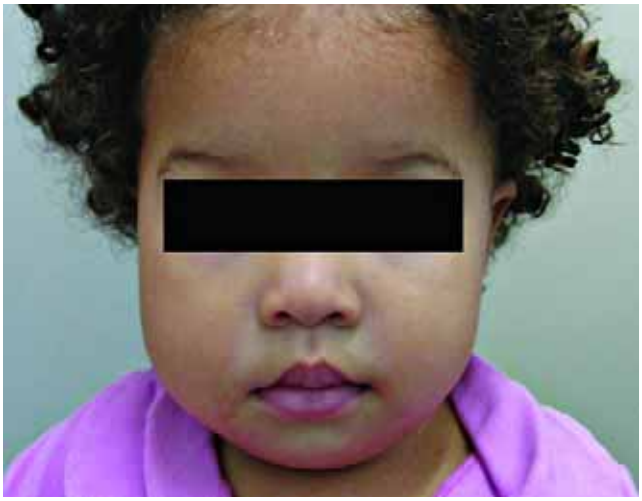

Figure 4: After six months, significant involution following local infection

The malformation may be detected by ultrasonography prior to birth, from the fourth month of pregnancy onwards. [2] Some cases diagnosed prenatally are associated with karyotypical abnormalities or malformation syndromes (Down, Turner's or Noonan syndromes) and possibly with teratogenic agents. Clinical diagnosis is simple: a soft, cystic mass under normal-looking skin, which is generally asymptomatic; however, hemorrhage may occur, making the lesion edematous, painful and violaceous. Its natural history is characterized by slow, progressive growth with compression and infiltration of adjacent structures, and its clinical status depends on its location. Diagnosis may be confirmed by imaging methods and also by puncture followed by cytological examination of the fluid. Spontaneous regression may occur in 6% of cases, generally following an infection, as in the patient in the present case report.

Treatment of lymphangioma depends on its clinical presentation and its risks of complication. The most commonly used therapy is surgical and complications include: damage to adjacent structures, formation of fistulae, infection and suture dehiscence. Mortality ranges from 2% to 6%. Recurrence of the lesions has been described in up to 27% of cases. Another treatment alternative is the application of sclerosing agents such as bleomycin and hypotonic saline solutions that provoke inflammation of the vascular endothelium, leading to total or partial regression of the lymphangioma. Diffusion of these substances through the wall of the cysts to the adjacent tissues may provoke an inflammatory reaction and retraction of the scar tissue, resulting in unsightly scars and making any subsequent surgery more complicated. A new agent, OK-432, produced from *Streptococcus pyogenes* (Group A streptococcus), has been used with good results and a low recurrence rate. [4,5] In this case, we opted for clinical follow-up since there were already signs of involution of the condition. ❑

REFERENCES

1. Enjolras O. Vascular Malformations. In: Bolognia JL, Jorizzo JL, Rapini RP, editors. Spain: Mosby Elsevier. 2008. p. 1581-95.
2. Oak SN, Redkar RG, Kapur VK. Posterior midline cervical fetal cystic hygroma. J Postgrad Med. 1992;38:93-6.
3. Sampaio SAP, Rivitti EA. Dermatologia. 3 ed. São Paulo: Artes Médicas; 2007. p.1197-1225.
4. Ruiz Jr. E, Valera ET, Veríssimo F, Tone LG. Uso de OK-432 em crianças com linfangioma. J Pediatr. 2004;80:154-8.
5. Rautio R, Keski-Nisula L, Laranne J, Laasonen E. Treatment of Lymphangiomas with OK-432 (Picibanil). Cardiovasc Intervent Radiol. 2003;26:31-6.

Mailing address / Endereço para correspondência:
Bárbara Nader Vasconcelos
Rua Marques de Abrantes,178. Aptº 1805 Flamengo
22.230-060 Rio de Janeiro - RJ, Brazil
E-mail: bnvasconcelos@gmail.com

Como citar este artigo/*How to cite this article*: Vasconcelos BN, Benez MDV, Bressan AL, Oliveira EF. Involution of a cystic hygroma of the face following local infection. An Bras Dermatol. 2011;86(1):135-7.