Rapidly involuting congenital hemangioma*

Susana Brás[1]
Cristina Amaro[2]
Pedro Mendes-Bastos[2]

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Abstract: Rapidly involuting congenital hemangioma is a rare vascular tumor that generally has a good prognosis. The authors describe a case of a newborn girl with a left cervical vascular lesion. Image exams were performed, and the lesion slowly decreased, leaving redundant skin. Considering all of the findings, a final diagnosis of a rapidly involuting congenital hemangiomas was suspected.

Keywords: Congenital abnormalities; Congenital, hereditary, and neonatal diseases and abnormalities; Hemangioma; Infant, newborn; Infant, newborn, Diseases; Vascular neoplasms

INTRODUCTION

Vascular anomalies are categorized by the International Society for the study Group of Vascular Anomalies into two types of lesion: vascular tumors and vascular malformations.[1]

Congenital hemangiomas are benign vascular tumors that have grown to their maximum size at birth and do not exhibit accelerated postnatal growth.[2] They can be subdivided into three subgroups: rapidly involuting congenital hemangioma (RICH), noninvoluting congenital hemangioma (NICH), and partially involuting congenital hemangioma (PICH). [1,3,4]

The authors describe a case and review the topic in order to clarify the RICH characteristics and to distinguish it from other congenital vascular tumors.

CASE REPORT

The authors describe a case of a neonate girl, born with a left cervical mass, following an uncomplicated pregnancy with normal prenatal scans. Examination on the 14th day of age revealed a soft pink-purple, without fremitus, compressible mass of 6x4 cm surrounded by a pale halo (Figure 1). The infant was hemodynamically stable, and her platelet count was unremarkable.

Ultrasound demonstrated a 40x44x11 mm left cervical and occipital lesion with heterogeneous vessels and a high blood flow (Figure 2). Upon magnetic resonance imaging (MRI), these lesion presented high intensity on T2-weighted images and intense contrast captation without intracranial extension or deformities of the underlying bone. Angio-MRI showed an irregular branch of the left occipital artery to the lesion and some transcranial vessels to the sigmoid sinus (Figure 2). These images were compatible with a vascular anomaly. Considering the clinical findings, a congenital hemangioma was suspected. By the 12th month of age, the lesion had slowly decreased, leaving slightly redundant skin (Figure 1). These findings were compatible with the diagnosis of a RICH.

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1 Dermatology Service, Centro Clínico da GNR, Lisboa, Portugal.
2 Dermatology Center, Hospital CUF Descobertas, Lisboa, Portugal.

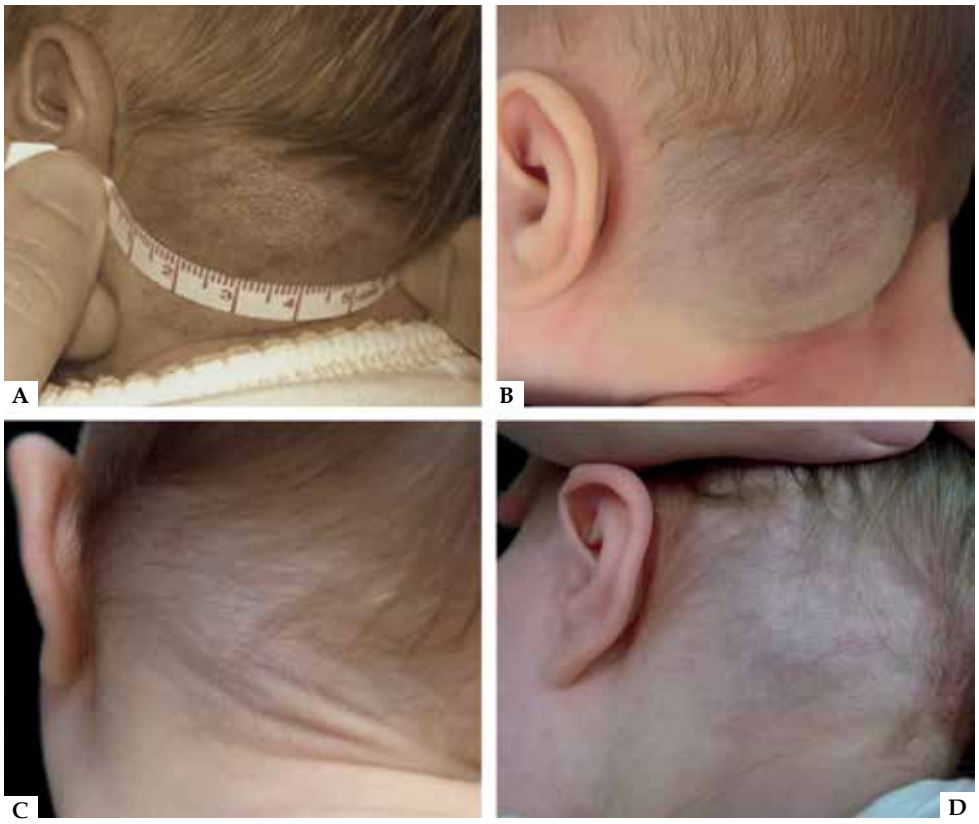

Figure 1: Clinical picture of congenital hemangiona: **A -** with 14 days of evolution**, B -** with one month of evolution; **C -** with 8 months of evolution; **D -** complete regression at twelfth month

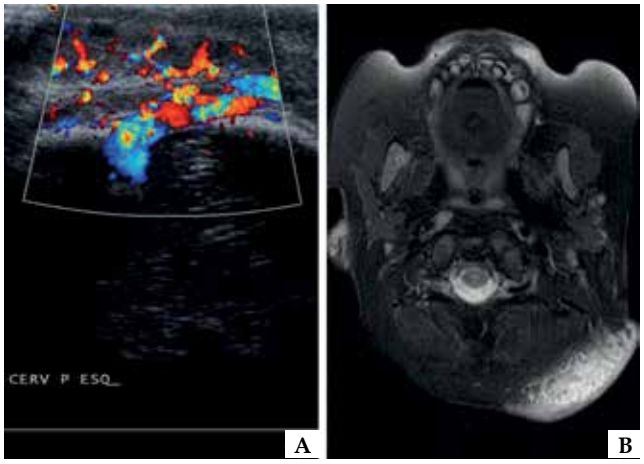

Figure 2: A - Ultrasound image showing heterogeneous high blood vessels; **B -** magnetic resonance imaging showing high intensity of the lesion on T2-weighted images

DISCUSSION

Congenital hemangiomas have an almost equal distribution among the sexes, are usually solitary, and are mainly located in the head or near a joint in the limbs.[2,3] They have a slightly variable morphology; however, common features include red-purple color with multiple tiny or coarse telangiectasias, often a surrounding pale halo, and sometimes a central ulceration, linear scar, or central nodularity.[2,3]

RICH involutes in the first 6-14 months of life, leaving a residual patch of thin skin with prominent veins and a normal blood flow, with little, if any, subcutaneous fat.[2]

In a smaller proportion of patients, involution may be incomplete, leaving a vascular plaque with coarse telangiectasia on the surface and a peripheral bluish white border, resulting in a lesion that is indistinguishable from NICH. These congenital hemangiomas, which have a distinct behavior, with rapid involution during the first 12 to 30 months of life (typical of RICH) before stabilizing in size and appearance (NICH-like lesions), are now defined as PICH.[3]

Some authors hypothesize that NICH could be considered a later stage of PICH because of the similarities in appearance, persisting fast flow, and similar histopathological features. On the other hand, other authors, based on the different clinical behavior and reports, or RICH with fetal involution, hypothesize that NICH could be a PICH with partial regression occurring in utero.[3,5]

Congenital hemangiomas can be detected in utero by antenatal ultrasound, at the end of the first trimester and, more commonly, at the beginning of the second trimester. [2,6] If diagnosed antenatally, the lesion can be followed by ultrasound or MRI to define the tumor characteristics and monitor growth.[7] A distinction between RICH and NICH lesions cannot be made on prenatal ultrasound.[6]

On Doppler ultrasound imaging, all tumors initially exhibit fast-flow, are usually confined to the subcutaneous fat, and are more likely to be heterogeneous and contain calcifications.[8,9] RICH initially has a high blood flow and switches to low-flow when involuting.

NICH and PICH remain as fast-flow lesions.[2]

The presence of large and tortuous vessels in RICH and NICH can simulate arteriavenous malformations, but these present low-resistance arterial flow and arteriorized venous flow signals.[8]

In an MRI, congenital hemangiomas exhibit high intensity flow voids on T2-weighted images and iso-intensity on T1-weighted images; prior to involution they enhance avidly and homozygously.[2] RICH angiographic findings include inhomogeneous parenchymal staining, large and irregular feeding arteries in a disorganized pattern, multiple various sized aneurysms, direct arteriovenous shunts, and intravascular thrombi.[9]

Histophatologically RICH and NICH demonstrate overlapping features and particularities which may help to distinguish them. RICH is characterized by well-defined, variably sized lobules containing small capillaries and prominent draining vessels in the dermis and subcutaneous tissue. There are often fewer lobules in the center of the lesion, as it is the first area to involute. The blood vessels possess a thin basement membrane and the endothelial is moderately plump in the early stages. Later, the basement membrane appears thickened and lobules are surrounded by fibrous tissue, dystrophic calcification, thrombosis, and hemosiderin deposition; extramedulary hematopoiesis may also be observed.[2,3,10] NICH consists of well-defined, large lobules of small vessels and draining channels. The blood vessels have a thin basement membrane and endothelial cells can be hobnailed. Interlobular areas contain fibrous tissue with large arteries, dilated veins, and arteriovenous microfistulae. Unlike infantile hemangiomas, all congenital hemangiomas are glucose transporter-1 (GLUT1) negative.[2,3]

The management of RICH lesions with a typical clinical presentation and characteristic features on ultrasound or magnetic resonance imaging consists of observation during the involution phase. In many patients, involution may begin within the first week of life. Prevention of ulceration may be accomplished by applying petrolatum ointment to the surface of the lesion several times daily.[10]

Lesions that are firm or show no involution may be difficult to differentiate, even following imaging, from more serious tumors, such as congenital fibrossarcoma or rhabdomyosarcoma. Such lesions require early biopsy and GLUT1 staining.[10]

Surgical excision or embolization are indicated for persistent ulceration, hemodynamic instability, thrombocytopenia, or bleeding that does not respond to medical therapy.[5] NICH and PICH can also be treated with excision after 18 months without signs of regression. Transient thrombocytopenia is generally limited to the first two weeks of life and is generlly not complicated by bleeding problems.

Rapidly involuting congenital hemangioma is a rare vascular tumor that generally presents a good prognosis. Image exams associated with rapid involution of the lesion help to distinguish it from other vascular anomalies. The accurate diagnosis allows for an adequate treatment and an appropriate parent counseling. ❑

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Mailing address:
Susana Brás
Alameda de Santo António dos Capuchos,
1169-050 Lisboa, Portugal.
E-mail: bras.susana@gmail.com

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