



Case Report

Giant Congenital Hemangioma: An Entity of Intermediate Behavior Between Rapidly Involuting Congenital Hemangioma and Non-Involuting Congenital Hemangioma. Presentation of A Case and Review of the Literature

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Abstract

Congenital hemangiomas are rare vascular anomalies differentiated from classical hemangiomas of infancy by their appearance, evolution, and lack of GLUT-1 immunoreactivity. Two classical types based on their clinical course have been described, rapidly involuting congenital hemangioma (RICH) and Non-Involuting Congenital Hemangioma (NICH). Recently, the term Partially Involuting Congenital Hemangioma has been proposed (PICH) to refer to another subtype of hemangioma with different evolution. We present a case of a large vascular tumour on the right flank, causing complications on evolution as cardiac failure. Initially treated with propranolol without success, afterwards, when the patient presented a secondary heart failure, an embolization was necessary, and finally surgical treatment resolved the tumour. The giant forms of congenital hemangiomas were associated with RICH subtype, and usually its present transitory thrombocytopenia and coagulation abnormalities. According to our case and the published cases of giant congenital hemangiomas, we think that all these forms of giant congenital hemangiomas might be included in PICH group.

Keywords: Congenital Hemangiomas; Giant Hemangiomas

Introduction

Congenital hemangiomas constitute a rare group of vascular anomalies separated from classic infantile hemangiomas based on the following: a different clinical aspect, a different evolution, they are fully developed at birth and do not have precursor lesions, and a different immunohistochemical pattern in the histopathological study, the lack of reactivity to GLUT-1, the marker of infantile hemangiomas. Initially, congenital hemangiomas were divided into two groups based on their clinical course, Rapidly Involuting Congenital Hemangiomas (RICH), which normally disappear spontaneously in the first year of life, and Non-Involuting Congenital Hemangiomas (NICH) which remain unchanged

over time. In 2004 Mulliken and Enjolras published a series of 15 patients within the congenital hemangiomas that call “missing links” type II [1]. These lesions that initially resemble a RICH, after a few months stop their involution and behave as NICH. In 2009 the term Partially Involuting Congenital Hemangioma (PICH) was introduced to denominate this group of lesions [2]. Histopathologically, these patients have characteristics of both poles, RICH and NICH.

Baselga et al. described a special group of RICH with large size that they call giant RICH [3]. The association of transient thrombocytopenia, coagulopathy and, in some cases, high output heart failure are the main features of giant RICH. We present a neonate patient with of a large vascular tumour on the right flank clinically compatible with giant RICH. Initially

was treated with propranolol without success, afterwards, when the patient presented a secondary heart failure, an embolization was necessary, and finally surgical treatment resolved the tumour. The tumour initially presented and behaved like RICH and after an initial involution, remained unchanging, adopting the clinical aspect and behaviour of another new category: Partially Involuting Congenital Hemangioma (PICH).

Case Report

We present the case of a new-born female infant, born at term, after an uneventful pregnancy, with a birth weight of 3,8 Kgr by vaginal delivery. Physical examination revealed a 10 x 10 cm, round-to-ovoid, soft and exophytic red-purple mass with prominent surface telangiectasias, surrounded by a pale halo, extended over the abdominal surface (Figure 1). The mass had a central ulceration covered by a scar, with well-defined limits. This lesion had not been detected by prenatal ultrasonography. Ultrasonography performed at birth showed a superficial mass, mostly and uniformly hypochoic, with some hyperechoic edges. Some cystic region could also be seen. Colour Doppler examination showed multiple high-flow large vessels in the tumour tissue. This diffuse vascular lesion, warm on palpation, with multiple vessels, was initially compatible with a congenital hemangioma.



Figure 1

Laboratory examinations revealed an initial platelet count after birth of $179 \times 103/\mu\text{L}$ (reference range $192\text{-}252 \times 103/\mu\text{L}$), which dropped on the second day (lowest level of $109 \times 103/\mu\text{L}$) but normalized during the first week with no additional therapy. In coagulation studies, prothrombin time was high (14,9 seconds; normal (age adjusted normal $8,5\text{-}12$ seconds), partial thromboplastin time was normal and fibrinogen was borderline low at $1,85\text{ g/L}$ and decreased until $1,19\text{ g/L}$ on the third day (reference range $1,67\text{-}3,09\text{ g/L}$). D-dimers were elevated ($10573\mu\text{gr/L}$; age adjusted normal $0\text{-}150\mu\text{gr/L}$). Our patient did not have petechiae or bleeding at any level. The liver function test was normal. Other laboratory studies were normal for age, and she did not present bleeding neither at birth nor during admission.

Histopathological findings of a lesional biopsy showed a vascular lesion, with small lobules of capillaries with plump endothelial cells separated by abundant fibrous tissue and dysplastic veins. Immunohistochemical staining with Glucose Transporter-1 (Glut-1) was negative. A diagnosis of congenital hemangioma was proposed. Magnetic resonance showed a subcutaneous T1 isointense and T2 signal hyperintense mass, with a rapid arterial phase enhancement that was consistent with a high-flow haemangioma. Echocardiography showed a structurally normal heart with poor right ventricular systolic dilation, so on the second day of life we started oral propranolol treatment at 3 mg/kg/day without any clear benefit. During the first days of life the patient started to develop clinical evidence of a secondary High-Output Cardiac Failure (HCF), and the chest X-ray demonstrated pulmonary oedema and cardiomegaly. In view of the results of the resonance and their clinical repercussion, an arterial embolization was mandatory.

The preliminary arteriogram using a 4Fr catheter (percutaneous femoral artery approach) demonstrated a well-defined mass that was nourished at the expense of the mammary intern, costal and lumbar arteries with important venous drainage. At 7 days of age, endovascular embolization using thrombogenic fibered platinum coils of the feeding arteries was performed to reduce flow and consequently the size of the tumour. There were no immediate complications associated with the procedure. After coil embolization, symptoms related to cardiac failure disappeared. Within days the tumour softened, decreased in volume and revealed a peripheral pallor with a central scar, suggesting a Rapidly Involuting Congenital Hemangioma (RICH). After 4 months of regression, it ceased remaining stable and leaving a telangiectatic pink plaque, with central scar and pale halo (Figure 2). Due to the size of the lesion and its aesthetic repercussions we decided to excise it when the patient was 12 months old. Surgery took place in only one stage, with no remarkable bleeding, using electrocautery for surgical dissection and haemostasis.



Figure 2

We approximated the skin with metal staples, keeping them

10 days after surgery, to avoid its dehiscence. Nevertheless, when they were removed, the wound partially opened, and secondary intention closure was needed. Finally, the aesthetic result was excellent (Figure 3). Histopathological examination of the full lesion revealed well-defined, large lobules of capillaries centred by large, a little bit stellate, draining channels (Figure 4A). The capillaries had a thin basement membrane, without inner elastic layer (the capillaries were negative for orcein stain). The endothelial cells had multifocal hobnail appearance (Figure 4B). Among the lobules there was an abnormal, scarring collagenous tissue, completely different to dermal collagenous, that contained large elongated and tortuous veins, compressed between the lobules, some of which were several millimeters in diameter (Figure 4C). Some of these dysplastic veins contained venous type valves (Figure 4D). Glucose Transporter-1 (GLUT-1) staining was negative. Because all of these findings, we thought that this tumour looked rather like a NICH. A lesion that begun as RICH and became as NICH must be named as PICH.

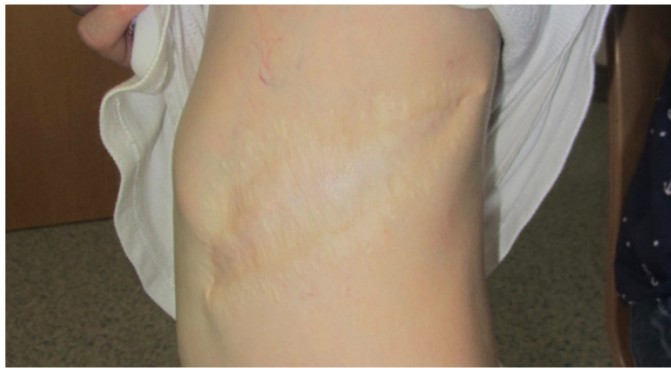


Figure 3

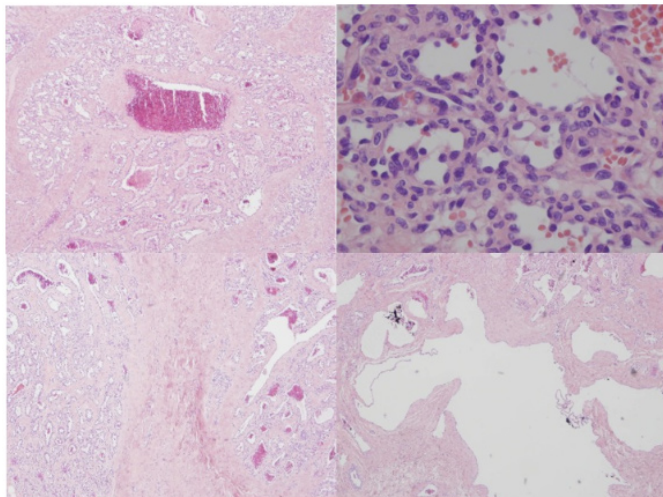


Figure 4 (A-D)

Discussion

In the two described polar forms of congenital hemangiomas, the RICH and the NICH evolution is totally different. The RICH involute spontaneously, without any treatment in a few months losing most of its mass. It is also characteristic that lipoatrophy occurs in the area on which they have settled, sometimes permanently. NICHs, on the other hand, remain stable over time and for their resolution it is usually necessary to perform surgical treatment. Before the description of these lesions performed in 1996, they were often interpreted as arteriovenous malformations [4]. But there are lesions that initially are diagnosed as RICH, with an initial involution similar to these, but over time, they stop their involution and clinically and histopathologically they are like an NICH [5]. This fact suggests that this polar departure proposed initially is not such. In fact, there is a clinical, radiological and histopathological overlap between the two polar forms of congenital hemangiomas RICH and NICH.

Congenital hemangiomas are generally warm but not pulsatile. When an arterial duplex is performed on them, high flow is usually found, with a mixed venous predominance vascularization and sometimes cystic areas or calcification. Radiologically, it is difficult to distinguish RICH, NICH, a high-flow infantile hemangioma or an arteriovenous malformation. From a histopathological point of view there are no specific findings of the two polar forms. Furthermore, in a recent study it is reported that in congenital hemangiomas there is an association with a somatic activating mutation in GNAQ and GNA, but the same in the RICHs as in the NICHs [6]. This fact has led to propose to some authors that there is a continuum between the 3 described types of congenital hemangiomas: RICH, PICH and NICH. Probably the three types are related and they constitute a lesional spectrum. In childhood hemangiomas, the growth rate, the final size reached and the rate of involution varies significantly between patients and others, and even in different lesions within the same patient.

Likewise, the origin of congenital hemangiomas seems likely to be the same, and the size, biological behavior, and rate and rate of involution varies according to local and individual factors of each patient, such as angiogenic response, healing mechanisms, capacity of inducing apoptosis in tissues, local tension, hypoxia, etc.. Baselga et al used the term RICH to describe a giant congenita hemangioma [3]. In these large lesions, complications such as thrombocytopenia, coagulation disorders (increased prothrombin time, decreased fibrinogen, increased D-dimers, etc.), high cardiac output, or bleeding are usually presents. The size of the congenital hemangioma is a determining factor for occurrence of complications. Deveza E et al report that these complications are more frequent in tumors large than 7 cm [7]. An association of tumor size and complications (heart failure, coagulopathy) are

commonly described for NICH and infantile hemangiomas.

In Kaposiform hemangioendothelioma, a limit size has also been described, from which Kassabach-Merritt syndrome occurs, although the mechanisms of coagulopathy in these lesions are different. Surgery or embolization is required when patients present with complications, and these lesions behave as PICH [2, 3, 5-23]. All of them have an intermediate behaviour, as a PICH.

Conclusion

Congenital hemangiomas are probably a lesional spectrum, and in their presentation and development host factors has relevant influence. The development of complications such as heart failure or bleeding disorders depends on the size as described in NICH, infantile hemangiomas or kaposiforme hemangioendothelioma. The term giant RICH should be reconsidered. Indeed, giant congenital hemangiomas generally behave as PICHs.

Declaration

The authors declare no conflict of interest present in the realization of this manuscript.

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