A Giant Congenital Lobular Capillary Haemangioma

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Abstract

Lobular capillary haemangioma is a relatively common benign mucocutaneous lesion in paediatric age.

The etiopathogenesis is not very clear, but it seems that lesions originate de novo from a process of lobular vascular proliferation that appears to be neoplastic.

Congenital onset is usually very rare; it happens only in 1.1% in literature.

We report a case of a newborn with a giant congenital lobular capillary haemangioma of the head (left occipital region) which increased rapidly with superficial ulcers in the early days of life.

The baby was operated on by full-thickness excision using the linear closure technique; this treatment allows histological examination, ensures the most efficacy aesthetic results and gives the lowest recurrence rate.

Keywords: Neonatal; Giant mucocutaneous lesion; Surgical excision

Introduction

Angiomas are the most common cutaneous defect at the birth and they include haemangiomas and vascular malformations.

Haemangiomas, the most common paediatric tumours, are benign tumours of capillary endothelium. They are caused by local defects of angioblastic tissue proliferation during the final stage of cutaneous capillary system differentiation which occurs in the perinatal period when it is still disorganized [1].

Correct classification differentiates haemangiomas into superficial, deep or sub-cutaneous and mixed haemangiomas [2].

Besides, haemangiomas can be evident at birth or may appear later.

Lobular capillary haemangioma (LCH) is a relatively common benign mucocutaneous lesion in children and young adults [3-8].

The modern definition of LCH is widely accepted and reflects the opinion that the underlying process consists in a lobular vascular proliferation probably of neoplastic origin [3,5,9,10].

Case Report

We report the case of a Caucasian male born at term by normal delivery with birth-weight of 2990 grams.

The pregnancy was regular and the gynaecologic examinations showed normal pre-natal growth parameters.

At birth the baby had a big elastic-parenchymatous mass (ultrasound dimensions: 3 cm × 1.7 cm) on the left occipital side with normal dominant skin and red-wine superficial capillaries in the central area.

Initially, manual compression did not produce a fovea sign or ischemization of superficial vascularization.

Figure 1: Ultrasound shows the intense arterial and venous vascularization of the lesion and the slight vascular pedunculation with possible intra-cranial connection.

On first day of life, power-Doppler ultrasound showed a left occipital mass with homogeneous structure without liquid or calcium elements; the lesion showed intense vascularization, both arterious and venous.
venous, with slight vascular pedunculation. These vessels seem to have a connection with intra-cranial theca (Figure 1).

Successively, cranial radiography confirmed bone cranial integrity and ruled out evident calcium elements inside the lesion. The differential diagnosis, in fact, includes teratoma (easily excludible with imaging exams), basal cell carcinoma, squamous cell carcinoma, granulation tissue, haemangioma and angiosarcoma [3].

On the fourth day of life, the mass was enlarged and more intensely vascularized (dimensions: 4 cm × 4.5 cm: Figure 2), the consistence became softer and more sinkable at manual compression; superficial micro ulceration events are probably caused by rubbing of occipital region on the pillow (Figure 3).

The post-operative course was regular and the baby was discharged on the second day.

The histological examination confirmed the initial suspicion of congenital LCH.

The baby is well, both before and after the operation, with normal reactivity and is feeding exclusively on mother’s milk.

**Discussion**

Lobular capillary haemangioma is a common benign mucocutaneous lesion in children and young adults [1,5-8]. This lesion was formerly known as pyogenic granuloma because it was thought to be a reactive lesion secondary to trauma and pyogenic infection [5-11]. Lobular capillary haemangioma is most commonly located in the head and neck area, followed, in order of decreasing frequency by the trunk, upper and lower extremities. The preponderance occurs on the skin, while the remaining ones involve the mucous membranes (particularly the oral cavity and conjunctivae) [1,3,12]; the sub-cutaneous [13] and intravenous [5,8] forms are most rare.

Usually LCH presents as a solitary, red-purple nodule. The etiology is poorly understood and is usually thought to represent an overgrowth of granulation tissue after trauma or a foreign body reaction such as an insect bite, infections, increased levels of female sex hormones, chronic irritation and aberrant microscopic arteriovenous anastomoses [3,4]. However, most probably, they originate de novo from a process of lobular vascular proliferation that appears to be neoplastic [5,9,10].

Typically, LCH grows rapidly for a few weeks and the stabilizes at a size of about 2 cm in diameter, although larger swellings (giant LCH) are known to occur [14,15]

In children, the mean age of onset is 6.7 years; 42% of cases occur by 5 years of age, 12% occur before 1 year of age and only in 1.1% of cases the lesion is present at birth. Big size and congenital appearance made the peculiarity of the lesion.

Giant LCHs (larger than 4 cm) of the skin, although infrequent, were reported in literature [6,14,16], but never at birth, to our knowledge.

Early LCH is histologically identical to granulation tissue, appearing as highly vascularized connective tissue with capillaries and venules into an edematous matrix. At the mature stage a fibromyxoid stroma separates the lesions into lobules containing aggregates of capillaries and venules with plump endothelial cells. The edema has now disappeared. The epidermis shows inward growth at the lesion base, forming a so-called epidermal collarette and determining slight vascular pedunculation. The arrival of extensive fibrosis indicates the final stage of regression [1,3,7].

Histological findings comprise a proliferation of capillaries with a dense infiltrate of neutrophils, prominent endothelial cells and a normal number of mast cells in an edematous fibrous matrix in contrast to increased mast cells characteristic of proliferative phase hemangiomas [1,5,9].

Various treatment modalities have been used to remove LCH including surgical and medical options. Effective means include full-thickness excision, shave excision, laser surgery, sclerotherapy, curettage, electro-coagulation or combination of methods [1,3,4].
Generally, the choice of treatment depends on the localization, size and recurrence.

Full-thickness excision with linear closure offers the lowest recurrence rate, allows histological examination and ensures the most effective aesthetic results [3,17].

Shave excision followed by electrocoagulation is an effective therapeutic alternative that minimizes scar formation while preserving the ability to confirm the diagnosis with histological examination. Unfortunately, this method has a high recurrence rate (43.5% like those lesions treated exclusively with electrocoagulation) [4,17,18].

Others methods, more conservative, such as pulsed-dye laser surgery, sclerotherapy and exclusive electrocoagulation do not allow a histological examination which is fundamental to avoid incorrect diagnosis (in one series, 18% of LCH were incorrectly diagnosed) [1,17-20] and they may be reserved to small lesions.

Conclusion

Lobular capillary hemangioma is a benign tumor that can be localized on the skin, mainly in the head and neck area, as well as on the mucous membranes. In our case, the giant dimensions and the newborn appearance give the peculiarity to the lesion. Its congenital onset is very rare [1] and the standard treatments include invasive and non-invasive methods. The standard treatment among invasive techniques is the full-thickness excision which is to be reserved to big lesions [18]; furthermore although it requires general anesthesia, it provides the lowest recurrence rate and allows the histological examination for avoiding misdiagnoses.

References