# Hemangioma capilar lobular congénito da fossa nasal de um lactente - Caso clínico de uma rara apresentação

# Congenital lobular capillary hemangioma of the nasal cavity of an infant - A rare case report

Isabel Correia • Herédio Sousa • Inês Cunha • Vítor Sousa • Luísa Monteiro

RESUMO

Introdução: O hemangioma capilar lobular (HCL), originalmente denominado como granuloma piogénico, é uma lesão vascular benigna de crescimento rápido de etiologia desconhecida, com uma distinta arquitectura microscópica lobular, que afecta a pele e mucosa. Embora o HCL da cabeça e pescoço não seja incomum, a sua ocorrência na fossa nasal é pouco frequente, e ainda mais raro em crianças.

Material e métodos: Estudo retrospectivo de caso clínico de um lactente com uma massa na fossa nasal direita diagnosticada como HCL, utilizando o método descritivo.

Resultados: Esta é uma revisão da história de um lactente com 54 dias de idade referenciado ao nosso hospital por obstrução nasal severa causada por uma massa da fossa nasal direita. A criança apresentava dificuldade respiratória desde o nascimento, que se foi agravando progressivamente à medida que a massa aumentava de tamanho. Esta estava a ser responsável pelo abaulamento do lado direito da pirâmide nasal e septo e pelo aparecimento de edema periorbitário à direita. Após remoção da massa através de cirurgia endoscópica nasal, foi feito o diagnóstico histológico de HCL.

Conclusões: Os autores encontraram apenas a descrição de um caso na literatura de um HCL congénito da fossa nasal num lactente. Embora raro, o HCL deve ser incluído no diagnóstico diferencial de massas das fossas nasais na população pediátrica.

Palavras-chave: Hemangioma capilar lobular; granuloma piogénico; hemangioma; massa da fossa nasal; cirurgia endoscópica nasossinusal.

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*ABSTRACT*

*Introduction: Lobular capillary hemangioma (LCH), originally referred to as pyogenic granuloma, is a benign rapidly growing vascular lesion of unknown origin, with a microscopically distinctive lobular architecture that affects skin and mucosa. Although LCH of the head and neck is not uncommon, occurrence in the nasal cavity is rare, and even rarer in children.*

*Material and methods: A retrospective case review of an infant presenting at our pediatric hospital with a mass of the right nasal cavity diagnosed as lobular capillary hemangioma.*

*Results: This is a review of the history of a 54-days-old child with severe nasal obstruction caused by a mass of right nasal cavity. He had difficulty breathing since birth, which had been progressively worsening. The right nasal cavity mass had increased in size since birth and was responsible for bulging of the right side of nasal pyramid and septum and periorbital edema. After removal of the mass through endonasal surgery with endoscopic control, it was diagnosed as lobular capillary hemangioma.*

*Conclusions: The authors found in the literature only one case of congenital lobular capillary hemangioma of the nasal cavity. Although rare, lobular capillary hemangioma must be included in the differential diagnosis of nasal cavity masses in the infant population.*

*Keywords: Lobular capillary hemangioma; pyogenic granuloma; hemangioma; nasal cavity mass; endoscopic nasal surgery*

## INTRODUCTION:

Lobular capillary hemangioma (LCH), originally referred to as pyogenic granuloma, is a benign rapidly growing vascular lesion of unknown origin, with a microscopically distinctive lobular architecture that affects skin and mucosa.[1]

Most mucosal LCH of the head and neck arise in the oral cavity.[1] Although LCH of the head and neck is not uncommon, occurrence in the nasal cavity is rare[2], and even rarer in children. To the best of our knowledge, there are very few reports of this histological diagnosis in the nasal cavity in children,[1,3,4,5,6] and only one case of LCH presenting as a congenital nasal mass was reported in an infant.[7] We present a new congenital case of LCH of the nasal cavity.

## CASE REPORT

A 54-days-old boy was referred to our tertiary care pediatric hospital with severe nasal obstruction caused by a mass of right nasal cavity. He had a history of difficulty breathing since birth, which had been progressively worsening, causing stertor, suprasternal and intercostal indrawing and feeding problems. The right nasal cavity mass had increased in size since birth and was responsible for bulging of the right side of nasal pyramid and septum, proptosis and periorbital edema (figure 1).

There was no history of trauma and ante- and postnatal histories revealed no contributory factors.

Anterior rhinoscopy revealed a large pale, solid and non-pulsating mass, with elastic consistency, occluding the right nasal cavity and covered with serous secretions. It had a vessel on its anterior surface. The mass was bulging the nasal septum to the left, which was partially obstructing the left nasal cavity. The remainder of his head and neck examination was normal.

Magnetic resonance scan of the nasal cavities and brain confirmed the neoformation of about 30mm x 16mm x 22mm, which filled the right nasal cavity and caused bulging of nasal septum, of right side of soft palate and internal wall of right orbit, with no orbit invasion. Posteriorly, it extended into the nasopharynx. The lesion was hypointense on T1-weighted images

**FIGURE 1**

Lactent before surgery: bulging of the right side of nasal pyramid and septum, proptosis and periorbital edema

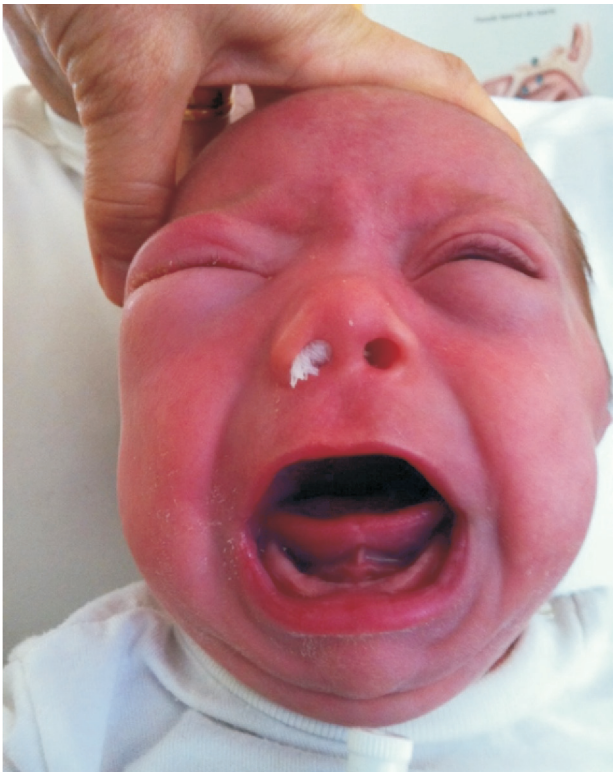


**FIGURE 2**

Magnetic resonance scan: T2 images showing a hyperintense mass; there is bulging of nasal septum and periorbital edema; coronal image shows the mass filling the right nasal cavity.

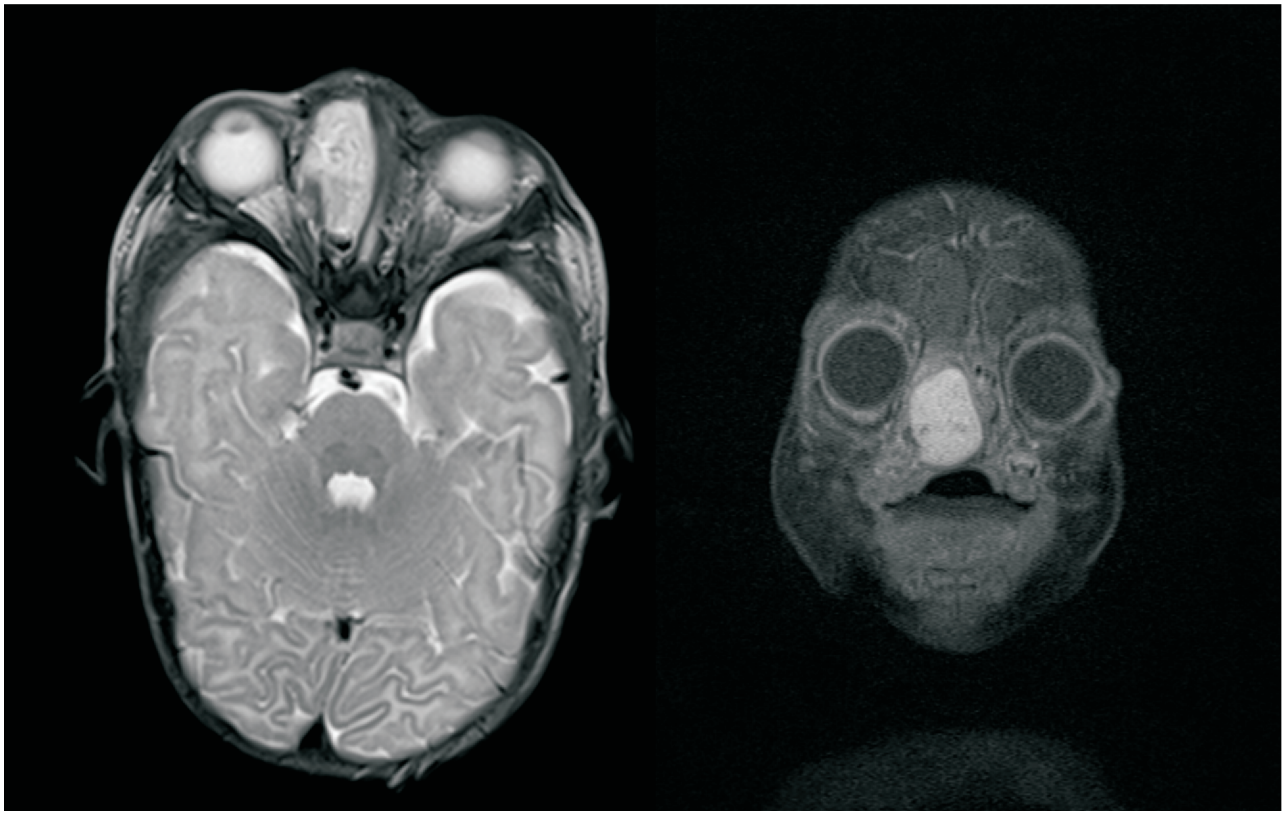
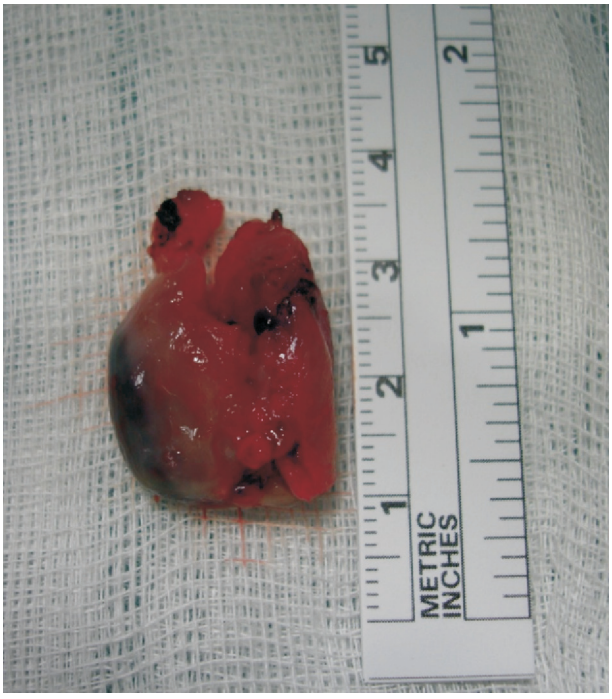
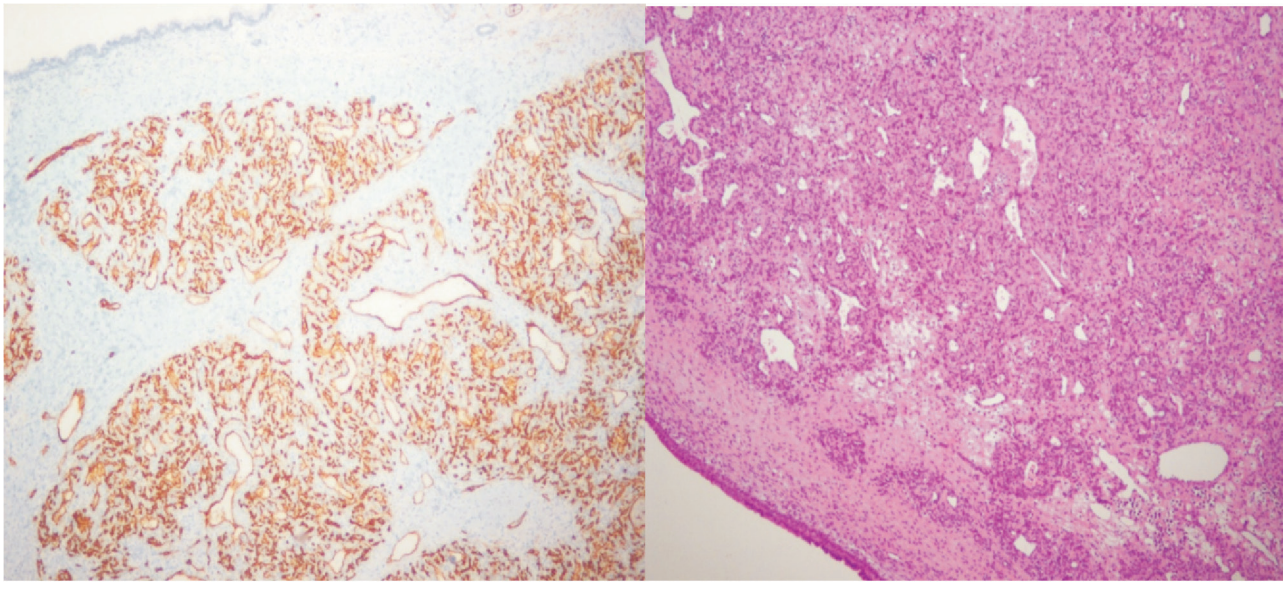


**FIGURE 3**
Macroscopic appearance of the mass after en bloc excision. There was no cleavage plane between the mass and inferior turbinate, so inferior turbinectomy was also performed.




and hyperintense on T2, and markedly enhanced with intravenous contrast gadolinium administration. There were no abnormalities on skull base (figure 2).
The infant was submitted to angiographic study of right external carotid artery, revealing a hypervascular mass, fed by the right sphenopalatine artery. An embolization with particle suspension was made.
The mass was excised under general anesthesia, through endonasal surgery with endoscopic control. The lesion was excised en bloc, using bipolar electrocoagulation. There was no cleavage plane between the mass and inferior turbinate, so inferior turbinectomy was also performed. All paranasal sinuses were free. Although its large size, it was extracted through the nostril (figure 3). Right nasal cavity was packed with surgicel. The estimated blood loss was 60ml. He remained mechanically ventilated through an endotracheal tube for 24 hours, being extubated without complications. He had improvement of symptoms and reinitiated breastfeeding with good tolerance.
Histological examination of the lesion revealed a vascular formation that contained small capillary vessels organized in a lobular and solid architecture (figure 4). The endothelial cells of the inner layer of blood vessels were positive for CD34, a typical endothelial cell

**FIGURE 4**
Histological preparations revealed anastomosing networks of capillary vessels organized in lobules in a fibrous stroma: The endothelial cells of the inner layer of blood vessels were positive for CD34, a typical endothelial cell immunohistochemical marker; Hematoxylin and eosin stain 40x.



immunohistochemical marker. The final histopathologic diagnosis was lobular capillary hemangioma.
Ten months later, the infant does not have nasal obstruction or other symptoms. Fiberoptic endoscopy shows no evidence of recurrence.

## DISCUSSION

LCH is a benign vascular tumor that is relatively common in the adult and pediatric population, in sites other than nasal cavity.[6] The most common sites of mucosal LCH are lips, tongue, gingiva and buccal mucosa.[2] They are rare in the nasal cavity of children, and extremely rare when congenital.

Classically, a hemangioma is either absent or flat at birth and then undergoes a period of rapid growth to present as a mass at around six weeks of age.[8] In our case, the infant presented at our hospital with 54-days-old (seven weeks and five days), and we know it was congenital because there was a history of difficulty breathing since birth, progressively worsening with the growing of the mass.

Newborns are generally obligate nasal breathers for the first few months of life and so nasal obstruction in this group can present as an acute respiratory emergency[8] in some cases.

LCH can occur at any age and usually presents as a papule or nodule that rapidly develops over a period of a few weeks and is prone to bleeding and ulceration.2 In the pediatric population, LCH has been reported more commonly in boys than in girls.[9]

The mechanism for the development of LCH is still obscure. Trauma, hormonal influences, viral oncogenes, underlying microscopic arteriovenous malformations, and the production of angiogenic growth factors have been postulated to play a role in the pathogenesis.[1]

The most common presenting symptoms of LCH of the nasal cavity include unilateral epistaxis and nasal obstruction.[1] In our case, the only symptom was nasal obstruction, progressively worsening since birth. The differential diagnosis of congenital nasal masses includes meningo-/encephaloceles, gliomas, hamartomas, chordomas and teratomas.[8]

The majority of LCH are usually small and tend to be localized in the anterior part of the septum or at the vestibule, but large lesions have a predilection for the lateral nasal wall and, in particular, for the inferior turbinate,[1] as in our case.

Magnetic resonance image is an essential imaging tool in the clinical evaluation of such lesions, to exclude intracranial connection or extension.[6] It was important in our case, to exclude other differential diagnosis of congenital masses, such as meningo-/encephaloceles and an eventual extension toward the skull base.

Definitive diagnosis and differentiation of LCH from other hypervascularized lesions is only possible after histological evaluation. It is characterized histologically by circumscribed anastomosing networks of capillaries arranged in one or more lobules in edematous and fibroblastic stroma.[9]

Diagnosis and definitive treatment of nasal LCH is accomplished by surgical excision.[2] Endoscopic excision is the preferred technique to ensure localization of the mucosal origin.[3] As with other sites of LCH, application of cautery at the base of the lesion is advocated in hopes of decreasing recurrence.[2]

The clinical course of LCH is usually benign following local excision of the lesion, although severe bleeding can occur and recurrences have been reported.[4] Endoscopic evaluation also allows excellent surveillance during follow-up in clinic.[2]

## CONCLUSION

LCH is a rare lesion in the nasal cavity that should be considered in the differential diagnosis of nasal masses in children and should be also considered when they are congenital, although exceptionally rare.

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