Hairy polyp faríngeo causando de dificuldade respiratória num recém-nascido – caso clínico / Pharyngeal hairy polyp causing respiratory distress in neonate – a case report

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Pharyngeal hairy polyp

Casos clínicos

**Resumo**

Os *hairy polyps* faríngeos são lesões raras com origem na nasofaringe ou orofaringe. Ocorrem predominantemente no sexo feminino, com predilecção para o lado esquerdo. A sua etiologia continua a ser mal compreendida. Apresenta-se tipicamente no período neo-natal com dificuldade respiratória e dificuldade na alimentação. Os métodos de imagem são fundamentais para identificar o conteúdo de gordura da lesão. A ressecção cirúrgica é a terapêutica de escolha. Apresentamos um caso clínico de um *hairy polyp* nasofaríngeo, causando dificuldade respiratória desde o nascimento.

**Palavras-chave**

Pescoço; Nasofaringe; Orofaringe; Doenças e alterações congénitas, hereditárias e neonatais; Neoplasias faríngeas

**Abstract**

Pharyngeal hairy polyps are rare lesions that arise from the nasopharynx or oropharynx. It occurs predominately in females, with predilection for the left side. Its etiology remains poorly understood. It typically presents in the neonatal period with respiratory distress and feeding difficulties. Imaging is fundamental to identify the high fat content of the lesion. Surgical resection is the treatment of choice. We report a case of a nasopharyngeal hairy polyp causing respiratory distress since birth.

**Key-words**

Neck; Nasopharynx; Oropharynx; Congenital, Hereditary, and Neonatal Diseases and Abnormalities; Pharyngeal Neoplasms

**Introduction**

Despite being globally rare, hairy polyps are the most common congenital tumors of the naso-oropharynx. Imaging is fundamental in the differential diagnosis of neonatal pharyngeal masses. In the case of hairy polyps, the high fat content is the key.

We report the case of a 1-day-old girl with respiratory distress diagnosed with a nasopharyngeal hairy polyp after performing ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI).

English language texts of the last 6 years (January 2011 to January 2017) were collected from PubMed/MEDLINE database using hairy polyp as key-word. Of 35 records, 29 full-text articles (mostly case reports/series) were selected, incorporating clinical data from 38 patients.

**Case report**

A new-born girl was delivered by caesarean section at 39 weeks and 6 days of gestation due to acute foetal distress. At birth, she presented with respiratory distress. APGAR scores at 1, 5 and 10 minutes were 6, 7 and 7, respectively. She was intubated at 5 minutes of life and extubated 5 minutes later. New intubation had to be performed and after several difficult attempts of extubation, steroid therapy was initiated, given the possibility of edema of the glottis. On the 4th day, a left, rounded, supra-glottic mass was identified during laryngoscopy.

US showed a well-defined, ovoid, pharyngeal lesion with mixed echogenicity, adjacent to the endotracheal tube. No vascularity was seen on colour Doppler (Fig. 1a and 1b). A contrast-enhanced CT was performed, showing a 35-mm, large lesion occupying the nasopharynx, displacing the endotracheal tube forward and to the left, and enveloping and displacing the naso-gastric tube to the left, extending from the base of the skull to the supra-glottis. It was hypodense, with a median density of 3 Hounsfiled units (HU) and a minimum density of - 80 HU). No bony changes were identified (Fig. 2).

MR confirmed the previous findings, depicting a cystic nasopharyngeal lesion, insinuating anteriorly into the choanas, and extending inferiorly to the level of the valleculae. It was hyperintense on T2-weighted images (WI). At the lower part of the lesion, spontaneous hyperintense content was found on T1WI, loosing signal intensity with fat suppression. No intracranial extension was identified (Fig 3a, 3b, 3c and 3d).

Complete surgical resection was performed. The postoperative period was complicated by reopening of the ductus arteriosus. Favourable evolution with closure of the ductus arteriosus was achieved after conservative therapy.

Macroscopically, a pink, pedunculated, polypoid lesion was documented. Histological examination showed a polypoid lesion covered by keratinizing stratified squamous epithelium, with hair follicles, sebaceous glands and a central core with adipose tissue, mature muscle tissue and cartilage. The distal margin of the tubular fragment was lined by respiratory epithelium, consistent with complete excision. These findings confirmed the diagnosis of hairy polyp.

**Discussion**

Hairy polyps are the most common congenital tumors of the naso-oropharynx. It derives from two germ layers, ectoderm and mesoderm. Ectoderm-derived tissues include keratinizing stratified squamous epithelium and skin appendages. Mesoderm component may include fibro-adipose tissue, muscle or cartilage. They are benign masses, with limited growth potential. Malignant transformation has never been reported. The incidence of hairy polyp is 1/40000 live births. It occurs predominately in females, as seen in our case. Among cases in which there is reference to the laterality, a predilection by the left side was found1-4.

It remains debatable whether hairy polyps are congenital defects like development malformations or primitive teratomas, or belong to the spectrum of neoplastic disorders1. Several theories about its origin have been proposed. Burns et al5 suggested a second branchial arch origin, while Vaughn et al6 described a case associated with first branchial arch sinus. Interestingly, hairy polyps have also been described as accessory auricles7,8. Exceptionally, they can occur in adults1,9.

In 2012, Yilmaz et al10 reviewed 35 cases corresponding to a period of 17 years. We performed a recent and larger review, including 38 cases from January of 2011 to January of 2017, a period of 7 years. We speculate that hairy polyps are not as rare as previously thought. More advanced methods of investigation, including imaging methods, may have permitted more diagnoses, mainly in less symptomatic patients.

Hairy polyps have been reported to arise from oropharynx, nasopharynx, soft palate, hard palate, tonsils, Eustachian tube, middle ear cavity, external auditory canal, and tongue11. In recent years, other locations have been described for the first time, such as the nasal vestibule, nasal septum, and lower lower lip11-14. They are usually solitary lesions, but there are some records of bilateral or two unilateral simultaneous lesions9,15-17.

Hairy polyps do not represent any syndromic disorder nor have familiar predisposition, but are occasionally associated with cleft palate, uvular agenesis, ankyloglossia, facial hemihypertrophy, low-set ears, osteopetrosis, osteporosis, hypospadias, left carotid artery atresia, agenesis of external auricle, bifurcation of tongue, branchial arch sinuses, and congenital hypothalamic hamartoma1,11,18,19.

Clinical presentation depends on the size, location, shape and level of the hairy polyp, but it usually presents with respiratory distress and feeding difficulties in the neonatal period1. It can also be identified as a clinically detectable oral mass4,20-22. Occasionally hairy polyp presents as masses with ischemic or hemorrhagic changes, presumably due to torsion15,22-24.

Respiratory difficulties owing to blockage of upper respiratory tract can cause respiratory distress, cyanosis, stridor and acute airway emergency. Feeding problems often occurs in cases when it impinges the esophagus, thus leading to drooling, vomiting, coughing and gagging attacks22,25. When the Eustachian tube is involved, chronic ear drainage, recurrent otitis media and hearing loss may occur3,26,27. Neurological complications may occur secondary to vascular compressions and ischemia2,10,20,28.Moreover, it is believed that the obstruction in fetal swallowing mechanisms may lead to polyhydramnios21,22,25,29.

Koike et al30 stated that polyps smaller than 3 cm in diameter have a higher risk of respiratory distress than do those bigger than 3 cm. Small hairy polyps are more difficult to diagnose and may be lethal because of delayed diagnosis30.

In the workup of oropharynx or nasopharynx masses, imaging is essential to assess the origin and extent of the lesion, determine possible intra-cranial extension, exclude neurological and otorhynolaryngological complications, and plan surgery26.

In newborns, US remains the first-line examination for the study of cervical masses. It is a safe and fast method of imaging that does not require sedation or anesthesia. However, US is operator-dependent and does not allow accurate visualization of deep structures. Hairy polyps typically show predominant high echogenicity due to the high fat content of the lesion. However, it often shows central low echogenicity, corresponding to its fibrous core, which may reveal vascularity on color Doppler31.

CT has the advantage of allowing for rapid acquisition times, thus avoiding deep sedation. Its main disadvantage is the use of ionizing radiation. Characteristic imaging features on CT include a smooth polypoid lesion with fat attenuation and a central linear soft tissue attenuation, that corresponds to the fibro-vascular stalk26,31. It can also show bony changes20-22,32-35.

MRI provides excellent depiction of soft tissues and does not use ionizing radiation36. The main disadvantage is the need of sedation or anesthesia. On MRI, a hairy polyp usually presents as a well-defined, non-enhancing mass with a relatively homogeneous matrix, surrounded by a smooth capsule3,35. It is predominantly hyperintense on T1WI and T2WI, with loss of signal intensity with fat suppression techniques. The fibro-vascular stalk appears as a linear structure with intermediate signal on T2WI, which can enhance after intravenous contrast administration26. MRI is useful to delineate soft tissue extension and exclude intra-cranial involvement33,35.

Typical imaging features can help narrow the differential diagnosis (table 1). The high fat content is useful to exclude lesions such as neuroblastoma, meningo-encephaloceles, vascular anomalies, and embryonic cysts22-24,31,34,37. At the same time, MRI narrows the differential diagnosis of a neonatal oropharyngeal or nasopharyngeal mass to hairy polyps, hamartomas, teratomas – which tend to be more heterogeneous –, and lipomas and their variants – which very infrequently occur not only in the pharynx but also in the neonate26.

Cases of auto-amputation have rarely been reported21,22-25. After ensuring the airway safety, the treatment of choice is surgical excision at the base of the stalk10,25,34. The recurrence of hairy polyps is not usual. A case of progressive growth due to incomplete resection 6 years before was described38.

**Conclusion**

Although rare, hairy polyps should be considered in cases of respiratory distress at birth. In the workup of oropharynx or nasopharynx masses, imaging is essential to assess the origin and extent of the lesion, narrow the differential diagnosis, determine intra-cranial extension, exclude complications, and plan surgery. We believe that MRI is an excellent imaging method for characterizing nasopharyngeal lesions in neonates and assessing for their potential complications. After an initial US, which remains the first-line method in the study of cervical masses, MRI is preferable over CT due to lack of ionizing radiation and better depiction of soft tissues.

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**Table 1.** Characteristic imaging features of hairy polyp.

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| **Lesion** | **Hairy polyp** |
| **US/Doppler** | Hyperechogenic lesion, often with central low echogenicity. The central fibrous core may have flow signals on color Doppler. |
| **CT** | Smooth polypoid lesion with fat attenuation and a central linear soft tissue attenuation. |
| **MR** | Well-defined, homogeneous, non-enhancing mass, surrounded by a smooth capsule. Hyperintense on T1WI and T2WI, with loss of signal intensity with fat suppression.  A linear central structure, with intermediate signal on T2WI, may enhance after intravenous contrast administration. |

**Figure 1.** (a, b) Transverse, oblique US image of the neck using a 13-5 MHz linear probe shows a well-defined ovoid lesion with mixed echogenicity within the pharynx (arrows), adjacent to the endotracheal tube (white asterisk). Mylo-hyoid muscles (white points); Tongue (red asterisks); Vertebral body (red circle).

**Figure 2.** Axial constrast-enhanced CT image shows a lesion occupying the nasopharynx (arrows), displacing the endotracheal tube forward and to the left, and enveloping and displacing the naso-gastric tube to the left.

**Figure 3.** (a,b) Sagittal and axial T2WI show a hyperintense lesion with epicenter in the nasopharynx (arrows), insinuating anteriorly through the choanas and extending inferiorly to the level of the valleculae. (c,d) Axial T1WI without and with fat-suppression shows hyperintense fat content within the lesion, which looses signal intensity after fat suppression.