B-ENT, 2019, **15**, 375-378

Hairy polyp of the oropharynx: a report of two cases and literature review

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Key-words. Hairy polyp; naso-oropharyngeal choristoma; dermoid; embryogenesis.

Abstract. Hairy polyps are rare bigerminal masses that arise most often in the naso-oropharynx. With an incidence of 1 in 40,000 live births, this entity is generally not well known by ENT-specialists, neonatologists, or paediatricians that care for newborns. Due to its location, a hairy polyp can have serious consequences. This diagnosis must be considered in newborns with respiratory obstructions or feeding difficulties. A thorough flexible nasopharyngoscopy examination must be performed. We describe two cases of hairy polyps in the oropharynx and review the literature.

Introduction

Hairy polyps are rare. They arise most often in the naso-oropharynx, and they comprise both ectodermal and mesodermal elements. This entity was first described by Brown-Kelly in 1918.[1] Patients typically present with respiratory obstructions or distress and/or feeding or swallowing difficulties. Most often, the complaints arise within the first days after birth, although occurrences in adults have been described.[1] We describe two recent cases that were diagnosed in newborns in our department.

Case reports

The first case was a preterm boy, born at nearly 36 weeks postmenstrual age. The first clinical examination was normal. On day 3, the newborn presented with episodes of obstructive apnoeas, with deep desaturation and stridor. He was transported to a secondary centre with neonatal intensive care. Flexible nasopharyngoscopy was performed by an ENT, Head, and Neck-specialist. A white mobile mass was found pediculated on the left nasopharyngeal side of the soft palate. A CT scan confirmed that a 15 ´ 8 ´ 7 mm, fatty, polypoid mass, arose from the soft palate. The patient was transported to a tertiary centre for resection and further management. An additional MRI showed a heterogeneous mass, with a high signal intensity on T1-weighted sequences and little enhancement after contrast administration (Fig. 1a-c). The mass was resected with a combined approach under general anaesthesia. Endoscopic control was maintained

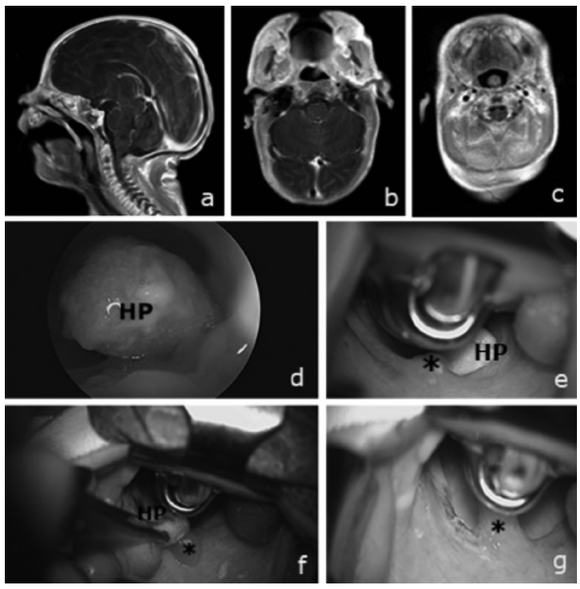

Figure 1

(a) Sagittal and (b, c) axial T1-weighted MRIs show the hairy polyp arising from the left side of the soft palate. (d) Endoscopic view of the hairy polyp (HP). (e) Oral view of the hairy polyp (HP); the uvula is marked with an asterisk. (f) Transoral resection of the polyp while applying traction with forceps; the attachment on the left side becomes clear. (g) Transoral view at the end of surgery.

Poster Presentation: Belgian ENT Society, Spring Symposium, 23 February 2019, Palace of the Academies, Brussels, Belgium

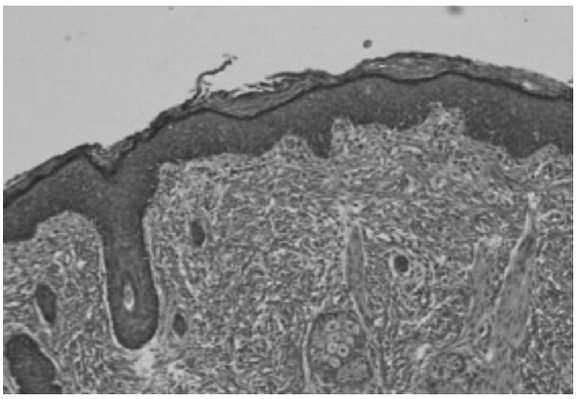

Figure 2

Polypoid specimen lined with stratified keratinizing squamous epithelium, supported by a core of fat tissue. (H&E stain, ´50 magnification).

through the nose. The pedicle of the mass was cut through the mouth with forceps and unipolar cautery (Fig. 1d-g). The pathology revealed a polypoid structure lined with stratified keratinizing squamous epithelium, a centre of immature, lobulated adipose tissue, and a fibrous stroma with adnexal structures. The patient recovered rapidly and was discharged from the hospital one day postoperatively. No specific postoperative care was necessary. A check-up performed 3 weeks after surgery showed complete regression of respiratory symptoms. No further follow-up was indicated.

The second case was a two-day-old baby girl, born at term. She experienced an acute cyanotic episode during breastfeeding. At that moment, the mother noticed a mass emerging from the mouth, which disappeared a few seconds later. A clinical examination of the mouth showed a mass that arose from the nasopharyngeal side of the palatum molle on the left side. This structure was confirmed with flexible endoscopy. A CT scan showed a large, fatty mass that arose from the left oropharynx. It protruded through the hypopharynx and oesophagus, over a distance of 3.3 cm. The patient was transported to a tertiary centre for resection. Upon close inspection, the mass arose from the left tonsillar pillar and was covered with hair. Like the previous case, the mass was resected under general anaesthesia with unipolar cautery. The pedicle was cut with nasendoscopic control to ensure complete resection. A pathological investigation showed a polypoid specimen lined with stratified keratinizing squamous epithelium, and normal adnexal structures, consistent with a hairy polyp (Fig. 2).

Discussion

The histopathological definition of a hairy polyp is a mature, fibrofatty, mesodermal core surrounded by keratinizing squamous epithelium with normal skin appendages. Most hairy polyps originate in the oronasopharynx, as observed in our two cases. Other locations include the oral cavity, the mastoid, and the middle ear. Hairy polyps arise 6.5 times more often on the left side than on the right side. To date, there is no adequate explanation for this preference. A female preponderance has been generally accepted, but this might be less pronounced than previously thought. In fact, a recent literature overview by Dutta et al stated that the ratio of female:male occurrences was 3.5:1, instead of 6:1, as reported previously by many authors.[1]

Although hairy polyps are rare, with an incidence of <1 in 40,000 live births, it is the most common congenital nasopharyngeal mass.[1,2] The differential diagnosis includes lesions of intracranial origin, including craniopharyngioma, Rathke's cleft cyst, pharyngeal hypophysis, glioma, neuroblastoma, and (meningo)encephalocele. However, in contrast to the hairy polyp, all of these lesions lack a fat density upon imaging. The lack of a fat density upon imaging also occurs with haemangiomas and thymic/thyroglossal/lingual cysts. Finally, a hairy polyp cannot be differentiated from a hamartoma, oral teratoma (epignathus), or dermoid cyst with imaging.[3]

On CT scans, the fatty core is visualized as a low-density mass, which might be enhanced with contrast administration, due to the fibrous elements. MRI T1-weighted sequences show high signal intensity, which is lost after fat suppression.[4]

When faced with neonatal respiratory distress, non-pulmonary lesions should be considered. Inspiratory stridor is often associated with upper airway obstructions.[5] Other symptoms associated with hairy polyps include feeding or swallowing difficulties, obstructive sleep apnoea, snoring, and ear problems.[1]

Due to their location, these lesions can cause serious problems by producing upper airway obstructions. Some case studies reported that a hairy polyp caused serious cerebral hypoxia, following an episode of acute respiratory distress.[6,7] Koike et al reviewed the literature to identify risk factors associated with respiratory distress. They found

that tumours smaller than 3 cm were more likely to cause respiratory distress than large tumours. This might be explained by the fact that a small lesion is easy to miss in routine clinical and endoscopic investigations.[7]

When an upper airway disease is suspected, an ENT-specialist should be consulted and a flexible nasopharyngoscopy should always be performed.[5,6] A CT or MRI scan can aid in confirming the diagnosis of an upper airway disease.[6] No guideline has emphasized that imaging is absolutely necessary, but it is performed in almost every case report, as reported in the present study.

Based on previous studies, a combined transoral and nasendoscopic approach appears to be the treatment of choice for these lesions.[1] Several advantages are associated with this technique. The greatest advantage is the improved visualization of the pedicle. This advantage increases the chances of a safe, complete resection and the rapid control of potential bleeding.[9]

No local recurrence has been described following a complete resection of a hairy polyp. However, a local recurrence after an incomplete resection was described by Chang et al. In that case, progressive growth of the lesion caused symptoms at 6 years after the first resection.[10]

It is difficult to devise a descriptive classification system, because the embryogenesis of hairy polyps is not known definitively. Consequently, hairy polyps have been called teratomas, dermoid cysts, or hamartomas. True teratomas include tissue from all three germ layers, which is not the case with a hairy polyp.[11] Some authors classify hairy polyps as dermoid cysts, inspired by Arnold's classification of the complex germ-layer lesions in the nasopharynx.[1] However, this classification requires caution, because Arnold never referred to hairy polyps as dermoid cysts, which are true cysts that derive from a single germ layer and contain desquamated epithelial products. Instead, Arnold referred to hairy polyps as 'dermoid', because they included bigerminal tissue.[1,2] Hamartomas are an abnormal overgrowth of normal tissue in an anatomic area that the tissue is normally found. However, this classification is not suitable for hairy polyps, because there is no hairy skin in the pharynx.[3] In contrast, a choristoma is normal polygerminal tissue found in an abnormal anatomical location. This classification seems to be the closest fit for hairy polyps.[11] However, this purely descriptive term does not give an embryological description of the hairy polyp.

Although different theories have been put forward to explain the origin of hairy polyps, no definitive explanation has been accepted. Earlier authors often considered that the hairy polyp was a limited form of teratoma; therefore, it was thought to be a neoplastic entity that arose from pluripotent cells. This theory could explain the occurrence of hairy polyps in adults; however, a strong argument against this theory rests on the fact that, unlike teratomas, a hairy polyp has never been associated with a malignant transformation or recurrence after a complete resection. Currently, most authors believe that the hairy polyp is a developmental malformation. One specific theory that seems to be gaining support is the association between the hairy polyp and branchial arch abnormalities.[1] One recent article by Simmonds et al supported this association. Indeed, they put forward a detailed hypothesis of how the hairy polyp develops. They suggested that hairy polyps originated as mesenchymal cells from the neural crest that failed to differentiate into epithelium in the superior half of the middle ear cavity.[12]

Conclusion

Hairy polyps are rare bigerminal masses that comprise both ectodermal and mesodermal elements. Due to their location, they can lead to serious consequences and life threatening situations. Therefore, this diagnosis should always be kept in mind when assessing newborns with breathing or feeding difficulties. The embryological classification remains an issue of discussion, although a growing number of authors tend to support an association between hairy polyps and branchial arch abnormalities.

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