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Hairy Polyps: A New Case Presentation and a Pathogenetic Hypothesis

Jonathan C. Simmonds, MD; Jad Jabbour, MD, MPH; Jennifer A. Vaughn, MD; Vera A. Paulson, MD, PhD; Dennis S. Poe, MD, PhD; Reza Rahbar, DMD, MD

Hairy polyps are benign lesions found in the oropharynx or nasopharynx that are thought to be present at birth and can lead to upper airway obstruction in infants. Also known as naso-oropharyngeal choristoma, they are increasingly viewed as aggregates of bigeminal tissue, likely from the first or second branchial arches, found in aberrant locations. They are benign lesions that are usually successfully treated by surgical excision. Here we present a rare case of a hairy polyp originating in the eustachian tube of a 7-week-old male, discuss our management of the patient, and put forth a new hypothesis as to the origin of these lesions.

Key Words: Hairy polyps, naso-oropharyngeal choristoma, dermoids, eustachian tube, magnetic resonance imaging, embryology, middle ear.

INTRODUCTION

Hairy polyps are masses derived from ectodermal and mesodermal germ layers that are commonly found in the oronasopharynx. They appear as a sausage-like pedunculated mass with a white/gray keratinized epithelium interspersed with hair-bearing areas. In the literature, the terms dermoids, hairy polyps, and naso-oropharyngeal choristoma (a choristoma being an aggregation of polygeminal tissue in an aberrant location) are often used interchangeably. They are considered the most common congenital tumor of the naso-oropharynx, with an incidence of 1 in 40,000.

The primary purpose of this study was to provide a guide to evaluating and managing patients with hairy polyps, based on a review of the literature on this condition, and describe a case of a hairy polyp emanating from the left eustachian tube of a 7-week-old male. We also propose a mechanism by which hairy polyps could develop from sequestrated first pharyngeal pouch tissue during the development of the middle ear.

MATERIALS AND METHODS

A retrospective chart review was conducted and information from hospital documentation was utilized. A systematic search in Ovid MEDLINE, PubMed, and Google Scholar was conducted. The Medical Subject Headings term “hairy polyps” as well as relevant synonyms such as “dermoids” and “choristoma” were used to compile an initial list of articles. The search was narrowed to articles on nasopharyngeal or oropharyngeal neoplasms. Relevant articles were isolated after reviewing abstracts. Articles were excluded if they were not written in English or if they pertained to monogeminal lesions, such as dermoid cysts, or trigeminal lesions (teratomas). The search was not limited to pediatric patients. The bibliographies were manually reviewed and cross-referenced with our initial search to obtain additional citations. Hospital documentation was reviewed at our institution for similar cases and was combined with the information gathered from our literature review to suggest a possible mechanism for the development of hairy polyps.

RESULTS

Case Presentation

A 7-week-old male was transferred to our institution from an outside hospital for evaluation and treatment of a left-sided nasopharyngeal mass. The patient’s gestation and delivery at 37 weeks were uneventful. He required high-flow nasal cannula briefly after delivery due to transient tachypnea of the newborn, but was discharged home in stable condition. He was subsequently readmitted at...
6 weeks of age after developing retractions and respiratory distress requiring continuous positive airway pressure. He underwent direct laryngoscopy, bronchoscopy, and flexible nasopharyngoscopy, which revealed a pedunculated nasopharyngeal mass with keratinized squamous epithelium and multiple dark hairs. Distal tracheomalacia was also noted and felt to be an incidental finding. An esophagram ruled out a vascular ring. Computed tomography (CT) showed a 1.5 × 3.2–cm lobulated, well-circumscribed, heterogeneous soft tissue mass with rim enhancement that was located anterior and inferior to the internal carotid artery and extended from the nasopharynx into the left middle ear with expansion of the bony portion of the eustachian tube (Fig. 1A). On magnetic resonance imaging (MRI), the mass had mixed signal intensity on the fast spin-echo T2-weighted sequences, foci of high signal on the T1 sequences, and slightly heterogeneous enhancement with areas of fat suppression consistent with a teratoma, lipomatous tumor, or hairy polyp (Fig. 1B–D). A CT-guided biopsy of the lesion showed benign squamous epithelium. A tympanostomy tube was placed to relieve a left-sided mucoid effusion prior to transfer to our institution.

After evaluation by our team, the patient was consented for excision of the mass and taken to the operating room. After repeating a direct laryngoscopy and bronchoscopy, a Crowe-Davis mouth gag was placed to expose the oropharynx and the patient was suspended from Draffin rods. The palate was retracted with a suction catheter and under direct visualization with a mirror, the inferior portion of the mass was grasped with angled Blakesley forceps and a portion excised with monopolar cautery. At this point, the rest of the mass was noted to be mobile, and with gentle manipulation, the mass was delivered from the surrounding tissue with virtually no bleeding. Nasopharyngoscopy with a 2.7-mm rigid endoscope confirmed that no residual mass was present.

Postoperatively the patient was taken to the neonatal intensive care unit. A CT angiogram obtained 2 days after the procedure showed a nonspecific small rounded intermediate density opacity in the hypotympanum with differential considerations including postoperative blood products, granulation tissue, or residual polyp (Fig. 2). There was no residual mass within the eustachian tube and no compromise to the surrounding vasculature.

On macroscopic examination, the lesion was composed of a pedunculated, soft-tissue mass measuring 2.5 × 1.5 × 0.5 cm and covered by tan-white, hair-bearing skin. Microscopic examination of the bisected mass demonstrated a polypoid lesion comprised of a mesenchymal core of adipose tissue and elastic cartilage lined by keratinizing squamous epithelium, with accompanying hair follicles and hyperplastic sebaceous glands, and transitioning to respiratory mucosa in one small focus at the apex, features consistent with a congenital hairy polyp (Fig. 3). At the patient’s 1-year follow-up visit, the patient was noted to be doing well. He had no middle ear effusion on exam and he had no evidence of residual disease, so no additional imaging was obtained.

**DISCUSSION**

**Presentation**

Hairy polyps typically present within the first month after birth with obstructive symptoms such as respiratory distress or feeding difficulties. They can also obstruct the eustachian tube orifice leading to a unilateral middle ear effusion or form between palatal shelves causing a cleft palate and velopharyngeal insufficiency. Almost 60% are found in the nasopharynx, most commonly from the lateral nasopharyngeal wall or the eustachian tube, 25% are found on the tonsil or tonsillar pillars, and the rest are found on the palate. They

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**Fig. 1.** (A) Preoperative contrast-enhanced CT in bone windows shows asymmetric smooth widening of the osseous left eustachian tube and opacification of the left middle ear cavity. Preoperative contrast-enhanced MRI shows a well circumscribed polypoid soft tissue mass centered in, and widening, the left eustachian tube with protrusion into the nasopharynx. The mass demonstrates a sausage like morphology and increased signal on the axial T2-weighted imaging (B) with fat signal components on the axial T1 precontrast imaging (C) and an enhancing soft tissue core on the axial, postcontrast, fat-saturated imaging (D). CT = computed tomography; MRI = magnetic resonance imaging.
are 3.5 to 5 times more common in females and are 6.5 times more likely to occur on the left side.4,5 Unlike teratomas, they have no malignant potential, and only one case of recurrence after surgical excision has been documented.8

Evaluation

In newborn patients where an obstructing mass is suspected, nasal and transoral endoscopy are the gold standard for establishing a diagnosis. This is especially important, as polyps <3 cm are commonly missed on physical exam, and therefore pose a greater chance of leading to obstruction.11 Fine-needle aspiration has been advocated as a means to establish a diagnosis in these lesions.12 However, in our experience, we found MRI to be useful for evaluating these lesions.

Imaging

Imaging is useful in the diagnosis of hairy polyps, which have characteristic radiologic features. On CT they appear as well-circumscribed masses containing fat and soft tissue–density components. CT is useful to demonstrate the widening of the osseous eustachian tube as well as for assessing the bony integrity of the carotid canal. On MRI, hairy polyps typically demonstrate heterogeneous high signal on the T2-weighted sequences. On T1-weighted sequences, there are areas of high signal corresponding to the fatty components, with suppression of the signal on fatsaturated imaging. Enhancing components of the polyps can be demonstrated on both modalities.

Pathology

As mentioned above, hairy polyps contain cartilage, smooth muscle, striated muscle (mesoderm), as well as skin with sebaceous glands, eccrine sweat glands, and hair follicles (ectoderm). Sinonasal/respiratory mucosa can also be seen. The differential for these lesions include teratomas, dermoid cysts, which are more cystic than polypoid, cholesteatoma, especially those arising in the middle ear, and keratin debris with foreign body giant cells.

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**Fig. 2.** Postoperative CT angiogram demonstrates interval resection of the mass in the patulous left eustachian tube. There is small nonspecific rounded opacity in the hypotympanum (arrow) with intermediate density, which may reflect postsurgical blood products, granulation tissue, or residual polyp. The left internal carotid artery is intact. CT = computed tomography.

**Fig. 3.** Macroscopic (A) and corresponding microscopic (B, H&E) scanning power images of the bisected mass demonstrate a polypoid lesion comprised of a mesenchymal core of adipose tissue and elastic cartilage (*) lined predominantly by skin (=), with a small focus of respiratory mucosa at the apex (—). (C, H&E) Higher-power (16x) magnification of the lesion’s apex highlights the transition between respiratory mucosa (—) and skin (=), with its accompanying hair follicles (●) and hyperplastic sebaceous glands (●). H&E = hematoxylin and eosin.
Pathogenesis

Traditionally, hairy polyps or dermoids are considered one of the four germ-layer lesions proposed by Arnold in 1870; hairy polyps being the only bigeminal lesion, with ectodermal and mesodermal tissue, whereas teratoids, teratomas, and epignathi representing trigeminal lesions at varying levels of differentiation. Hairy polyps are not definitively associated with any particular syndrome, though they have been noted to occur more frequently in patients with first and second branchial arch anomalies such as those with branchio-oto-renal syndrome and aural atresia. This has led some to speculate that they are displaced first pharyngeal pouch or second branchial arch tissue. The residual opacification in the hypotympanum on the postoperative CT could signify the origin of the mass. This raised similarities to a previous case report from our institution, in which a patient with branchio-oto-renal syndrome was found to have a middle ear hairy polyp that originated from the border of the eustachian tube adjacent to the glenoid fossa and hypotympanum.

The origin of these lesions in the hypotympanum raises an interesting question regarding the process by which they form. Until recently, hairy polyps were thought to be strictly localized to the naso-oropharynx; however, an increasing number are recognized to originate from the eustachian tube. Modern estimates suggest 20% to 45% originate within the eustachian tube or middle ear. Along with the recognition that a significant number of patients also had branchial arch abnormalities, this argues that hairy polyps represent displaced tissue either from the first pharyngeal pouch or second branchial arch. As Heffner et al. and Burns et al. argued, almost all hairy polyps are found on structures developed from the first pharyngeal pouch or second branchial arch. Dutta et al. elaborated on this, arguing that these lesions arise during the development of the endodermal tubotympanic recess as it forms the middle ear cavity and eustachian tube. Although this theory has gained popularity, hairy polyps do not contain endodermal tissue, and no mechanism has been proposed to explain the presence of the squamous epithelial tissue found in hairy polyps.

Contemporary understanding of the development of the middle ear reveals a plausible mechanism for their formation. The middle ear cavity is lined with epithelium that, until recently, was thought to be from the endoderm of the first pharyngeal pouch. However, half of the middle ear cavity is actually lined with epithelium derived from mesenchymal cells of neural crest origin. The middle ear cavity develops as the endoderm from the first pharyngeal pouch extends from the nasopharynx toward the developing middle ear ossicles and the mesenchymal neural crest cells surrounding them. This bubble of endoderm breaks down as it approaches the otic capsule, allowing a flood of mesenchymal cells into the developing cavity.

Fig. 4. In the development of hairy polyps, the endoderm of the first pharyngeal pouch (red) expands into the middle ear cavity surrounded by mesenchymal cells from the neural crest (light blue). The auditory bullae then stimulates the mesenchymal cells to transform into the epithelium that lines the superior half of the middle ear (dark blue), and the mesoderm expands and lines the inferior half (red). If the organization or differentiation of these mesenchymal cells is interrupted, they would have the potential to develop into hairy polyps and could attach and form in the middle ear, eustachian tube, or along any part of the first pharyngeal pouch. Images adapted from Tucker.
Development of the auditory bullae, the bony encasement of the middle ear, then stimulates the mesenchymal cells from the neural crest to retract and differentiate into the epithelium that lines the superior half of the middle ear cavity.\textsuperscript{19} It is likely that hairy polyps develop when these mesenchymal cells fail to organize and differentiate into the epithelium that lines the middle ear. This primordial conglomerate of mesenchyme in the process of undergoing mesenchymal–epithelial transition located at the breakdown of the first pharyngeal pouch could develop into a hairy polyp at any location or on any structure that develops from the first pharyngeal pouch or second branchial arch (Fig. 4). This would support Dutta et al.’s assertion that hairy polyps may not be bigeminal in origin but rather originate solely from the neuroectoderm.\textsuperscript{5} It would follow, then, that any disruption to the development of the auditory bullae and their ability to stimulate mesenchymal cells from the neural crest would promote the formation of hairy polyps and, indeed, failure in the formation of these auditory bullae has been noted in EYA1 inactive mice, the gene associated with branchio-oto-renal syndrome and other branchial arch anomalies that are commonly seen in patients with hairy polyps.\textsuperscript{20,21}

The true incidence of branchial anomalies in patients with hairy polyps has not been studied; however, if future research confirms this association, then the diagnosis of hairy polyps may warrant an evaluation for branchial anomalies. It has also been argued by Dutta et al. that the regulation of epithelial–mesenchymal interactions by Hox and sonic hedgehog (shh) genes, which determine left–right symmetry during morphogenesis, may also explain the predominance of left-sided lesions.\textsuperscript{5}

**Management**

Because the standard of management for these lesions is excision, hairy polyps originating from the eustachian tube present a dilemma to the otolaryngologist because, as in our case study, they appear to arise alarmingly close to the carotid artery, raising the possibility that the lesion may be vascular in nature. MRI was useful in this regard as it narrowed our differential to hamartomas, teratomas, or dermoids.\textsuperscript{22} However, this did not negate the risk of excision. Hairy polyps have been associated with carotid artery atresia and have caused neurological sequelae from vascular compression ischaemia, so the concern for bleeding is prudent.\textsuperscript{5,8} Nevertheless, the ease in which the mass was delivered from the surrounding tissue was a surprise. It has been argued that torsional forces on the pedicle of the lesion can cause necrosis in the stalk, which would make excision of these lesions easier than one would expect.\textsuperscript{23} Excision using a snare transorally often results in complete excision without any bleeding.\textsuperscript{24}

**CONCLUSION**

Hairy polyps are bigeminal masses derived from ectodermal and mesodermal germ layers. They are usually found on structures derived from the first pharyngeal pouch or second branchial arch and are commonly associated with branchial anomalies. Here we present a case of a hairy polyp originating from the left eustachian tube along with a discussion of the workup and management of these patients. We also suggest a new hypothesis for the embryologic basis for their development, based on recent understanding of the development of the eustachian tube and middle ear.