Naso-oropharyngeal Choristoma in an Adult
Re-living the Controversy

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The Prologue

“The prologue is not in order, it cannot be one of them, it must not be made of a beginning of excellent knowledge, if only could we answer the question — why is this rare? or, being rare, why didn’t it in this instance happen?”
Sir James Paget, 1882

Abstract

OBJECTIVE: We present a rare case of naso-oropharyngeal choristoma in an adult, and discuss its origin from an embryological perspective.

STUDY DESIGN: Case report.

METHODOLOGY: Case report with review of literature.

RESULTS: A 42-year-old woman presented with sleep apnea and gradually-progressive dysphagia for 4 years, with right-sided nasal obstruction for 2 years. On examination, a large, pedunculated smooth-walled, non-tender mass, free on all sides except superiorly, was seen to occupy the entire oropharynx, more towards the right. Nasal endoscopy revealed the lesion to be attached to the right Eustachian tube orifice and the adjacent epipharyngeal surface of the soft palate. Imaging was non-contributory. The mass was excised by a combined naso-endoscopic and trans-oral approach. Histopathology suggested bigeminal teratoma with no evidence of dysplasia, leading to the diagnosis of a naso-oropharyngeal choristoma. The patient recovered well and was disease-free on 8-month follow-up.

CONCLUSIONS: The naso-oropharynx is the most common site for teratoid lesions in the head and neck region, and these mostly are seen in the neonates and in early infancy producing symptoms related to mass effect. However, though this group of non-neoplastic lesions presenting as choristoma in this region in adults is rare, this has questioned the genesis of the dermoid/teratoma complex in terms of origin, nomenclature, and histology. This report deals with the embryology of these lesions and the related controversies through the presentation of a rare yet representative case in an adult.

Table I: Bigeminal choristomas of the naso-oropharynx reported in adults from 1988 to March 2013 in English language indexed literature

<table>
<thead>
<tr>
<th>No.</th>
<th>Citation</th>
<th>Location</th>
<th>Age</th>
<th>Sex</th>
<th>Presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Present case</td>
<td>Pharyngeal end of Eustachian tube right</td>
<td>42 yrs.</td>
<td>F</td>
<td>Difficulty in swallowing, intermittent respiratory obstruction</td>
</tr>
<tr>
<td>2.</td>
<td>Tariq MU, Din NU, Bashir MR. Hairy polyp, a clinicopathological study of four cases. Head Neck Pathol 2013; 15: 35. Epub ahead of print. DOI: 10.1007/s11155-013-0434-3</td>
<td>Reported a series of 4 patients, of which 2 were in their late teens (aged 17 and 18 years). The lesions were present in the nasopharynx in 2 cases, and 1 each in lower lip and palate. Of the 4 patients, 3 were male, and 1 female.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3.</td>
<td>Green VS, Pearl DS. A 4-year-old woman with a nasopharyngeal mass. Arch Pathol Lab Med 2006; 130: e33-4</td>
<td>Left lateral nasopharyngeal wall</td>
<td>24 yrs.</td>
<td>F</td>
<td>Earache, hearing loss, feeding difficulty, respiratory distress</td>
</tr>
</tbody>
</table>

Fig. 1
A fleshy pedunculated mass, free on all sides except superiorly, seen hanging from the nasopharynx.

Fig. 2
The mass had its origin from the right lateral nasopharyngeal wall, precisely from the Eustachian tube orifice, extending into the adjacent soft palate. (S = septum, P = soft palate, M = mass, N = lateral nasopharyngeal wall, O = Eustachian tube orifice, T = tonsils)

Fig. 3
Histopathology showed mature tissue derivatives from both ectodermal and mesodermal (cartilaginous epipharynx, salivary glands, hair follicles, fat, fibrous tissue, cartilage, and muscle fibres). (H & E x 400)

Fig. 4
The lateral nasopharyngeal wall is the commonest subsite in the naso-oropharynx where HPs have been reported; the Eustachian tube constitutes ~40% of them. Following widespread use of diagnostic naso-endoscopy, there is growing evidence that Eustachian tube might be the predominant site of origin. This, along with reports of HPs in the middle ear cleft, suggests that bigeminal choristomas in the head-neck are linked to the development of the 1st and 2nd pharyngeal arches. During the 4th week of development, dorsal part of the 1st pharyngeal arch endoderm (the pharyngeal pouch) along with that of the 2nd to form the tubotympanic recess which forms the middle ear cavity and Eustachian tube. The speculative role of the sonic hedgehog gene products on the expression of Hox gene transported during the population of the pharyngeal arches by migrating neural crest cells may explain the left-sided preponderance noted involvement of the lateral nasopharyngeal wall.

Fig. 5
The lesion biotrofically excision: solid, firm, fleshy, heterogeneous, bosselated in appearance.

Fig. 6
The mass was removed by combined endoscopic and trans-oral approach.

Discussion

Hairy polyps (HP), the commonest congenital tumor of the naso-oropharynx, and described by Brown-Kelly in 1918, are most commonly noted in the female neonates. They are extremely rare in adults with only 5 reported cases in the last 25 years, our patient being the 6th one (Table I). The reason is not known, probably because of our incomplete understanding of the embryogenesis of the complex germ-cell lesions.

A developmental malformation, or a "primitive teratoma"?

HPs are typically bigeminal composed of ectodermal and mesodermal derivatives. Though characteristically bigeminal, there are several reports of HPs where authors have referred them as teratoma, teratoid, or more specifically, bigeminal "teratomas". In contrast, they might actually have their origin in a single germ-cell lineage, the neuroectoderm that has the major contribution to the head-neck mesenchyme (the ectomesenchyme). Thus, in spite of the fact that HPs more closely resemble dermoids according to Arnold's classification of the so-called "dysembryogenic tumors of the nasopharynx", they have often been referred to as a "teratoma".

A "teratomas", suggesting their association with teratoma, a true neoplasia. However, unlike teratoma, growth potential of HP is slow with no malignant potential. Also, cartilaginous tissues present in such lesions are in the form of curved plates of characteristic thickness that resemble fetal pinna – quite unlike the orientation seen in teratomas. These led one group of authors to comment that they should not be considered as primitive teratoma, but strictly a developmental malformation. In fact, HPs are occasionally associated with congenital anomalies (cleft palate, urologic anomalies, ankyloglossia, facial hemihypertrophy, osteotropism, hypospadias, left cardiac artery atresia, agenesis of external auditory meatus, bifurcation of tongue and branchial arch sinuses), lending support to the theory of developmental error.

Understanding the origin – development of the pharyngeal arches and its molecular control

The lateral nasopharyngeal wall is the commonest subsite in the naso-oropharynx where HPs have been reported; the Eustachian tube constitutes ~40% of them. Following widespread use of diagnostic naso-endoscopy, there is growing evidence that Eustachian tube might be the predominant site of origin. This, along with reports of HPs in the middle ear cleft, suggests that bigeminal choristomas in the head-neck are linked to the development of the 1st and 2nd pharyngeal arches.

The pathogenesis of heterotopic cell rests – should this be called a choristoma?

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