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Naso-oropharyngeal Choristoma in an Adult

Re-living the Controversy

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The Prologue

"Not one of them is without meaning; not one that might not become the beginning of excellent knowledge, if only could we answer the question – why is this rare? or, being rare, why did 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.

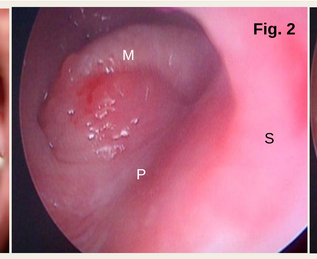
METHODOLOGY: Case report with review of literature.

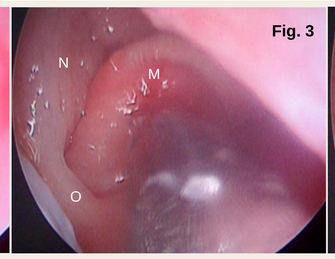
RESULTS: A 42-year-old woman presented with sleep apnea and gradually-progressive dysphagia for 4 years, with right-sided nasal obstruction for 2 years. On examination, a large, pedunculated smooth-walled, non-tender mass, free on all sides except superiorly, was seen to occupy the entire oropharynx, 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 bigerminal teratoma with no evidence of dysplasia, leading to the diagnosis of a naso-oropharyngeal choristoma. The patient recuperated well and was disease-free on 8-month follow-up.

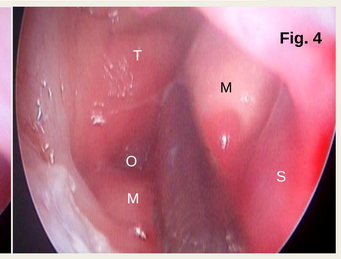
CONCLUSIONS: The naso-oropharynx is the most common site for teratoid lesions in the head and neck region, and these mostly are seen in the neonates and in early infancy producing symptoms related to mass effect. However, though this group of non-neoplastic lesions presenting as choristoma in this region 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.

Case Report

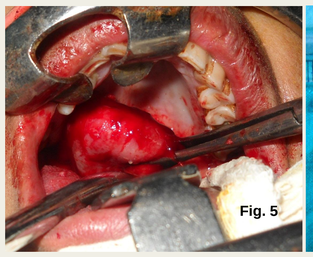
A fleshy pedunculated mass, free on all sides except superiorly, seen hanging from the nasopharynx.



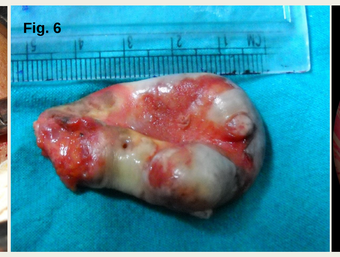




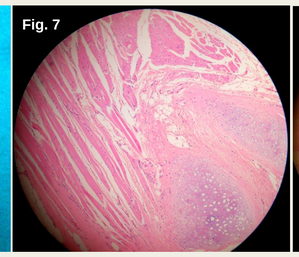
The mass had its origin from the right lateral nasopharyngeal wall, precisely from the Eustachian tube orifice, extending into the adjacent soft palate. (S = septum, P = soft palate, M = mass, N = lateral nasopharyngeal wall, O = Eustachian tube orifice, T = torus)

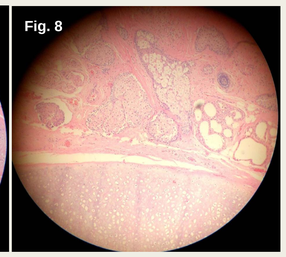


The mass was removed by combined endoscopic and trans-oral approach.



The lesion following excision: solid, firm, fleshy, heterogeneous, bosselated in appearance.





Histopathology showed mature tissue derivatives from both ectoderm and mesoderm (stratified squamous epithelium, sebaceous glands, hair follicle, fibroadipose tissue, cartilage, and muscle fibers). [H & E; x 400]

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 bigerminal composed of ectodermal and mesodermal derivatives. Though characteristically bigerminal, there are several reports of HPs where authors have referred them as teratoma, teratoid, or more specifically, bigerminal "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 "dysontogenetic tumors of the nasopharynx", they have often been referred to as "tumors", 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[1]. In fact, HPs are occasionally associated with congenital anomalies (cleft palate, uvular agenesis, ankyloglossia, facial hemihypertrophy, osteopetrosis, hypospadias, left carotid artery atresia, agenesis of external auricle, bifurcation of tongue and branchial arch sinuses), lending support to the theory of developmental error.

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

No.	Citation	Location	Age	Sex	Presentation
1.	Present case	Pharyngeal end of Eustachian tube right	42 yrs.	F	Difficulty in swallowing, intermittent respiratory obstruction
2.	Tariq MU, Din NU, Bashir MR. Hairy polyp, a clinicopathological study of four cases. Head Neck Pathol **2013**; March 15. Epub ahead of print. DOI: 10.1007/s12105-013-0433-4	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.			
3.	Green VS, Pearl GS. A 24-year-old woman with a nasopharyngeal mass. Arch Pathol Lab Med **2006**; 130: e33-4	Left lateral nasopharyngeal wall	24 yrs.	F	Earache, hearing loss, feeding difficulty, respiratory distress
4.	Cerezal L, Morales C, Abascal F, Canga A, Gómez J, Bustamante M. Magnetic resonance features of nasopharyngeal teratoma (hairy polyp) in adult. Ann Otol Rhinol Laryngol **1998**; 107: 987-90	Left lateral nasopharyngeal wall	50 yrs.	M	Recurrent epistaxis
5.	Franco V, Florena AM, Lombardo F, Restivo S. Bilateral hairy polyp of the oropharynx. J Laryngol Otol **1996**; 110: 288-90	Between the palatine arches (bilateral)	58 yrs.	F	Feeding difficulty, respiratory distress

***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 bigerminal choristomas in the head-neck are linked to the development of the 1st and 2nd pharyngeal arches[1,2]. During the 4th week of development, dorsal part of the 1st pharyngeal arch endoderm (the pharyngeal pouch) along with that of the 2nd join 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[3] may explain the left-sided preponderance noted during involvement of the lateral nasopharyngeal wall.

***The expanding domain of the "teratoma family"** - should neoplasia be considered to explain hairy polyps in adults?*

Though the origin of these bigerminal lesions can be explained by the theory of developmental malformation, this seems inadequate to implement in adults where they arise in areas that were so long unaffected in their life. Our patient did not have any mass 4 years back when she was completely asymptomatic. This may lead us to speculate whether bigerminal choristomas in the naso-oropharynx could be neoplastic in origin – a proposition supported by earlier researchers, and by the ongoing controversy related to the nosology of the complex germ-layer lesions itself. Green and Pearl (Table I) in their report have stated them as "neoplastic", while others have put them under the family of "benign teratoma". In fact, the definition of teratoma is experiencing a paradigm shift from them being referred to as the conventional "trigerminal" (Arnold, 1880) to a mass composed of any two germ layers (Tariq et al; Table I). The complex germ-cell lesions seem to belong to a larger "teratoma family". HP has therefore been referred to as a "primitive form of teratoma" by Karabekmez et al[4]. Weaver et al have even defined teratoma as a tumor of multiple tissues not indigenous to their site of origin, emphasizing on the aberrant location rather than on its composition.

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

A growing group of researchers are now saying that these bigerminal lesions in the naso-oropharynx should better be called "choristomas" – the heterotopic cell-rests (*choristo* = separated)[2,5]. Owing to an inciting factor (trauma), pluripotent cells during development get released from the local governing influences that would have otherwise led them to the pre-destined tissue morphogenesis, or gets misdirected or trapped during migration so that they cannot reach the targeted organ (the "missed target hypothesis") forming heterotopic tissues. Though a choristoma is essentially non-neoplastic, this alternative classification system might address the existing controversies related to the genesis of the complex germ-layer lesions.

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Conflict of interest/ financial disclosure None declared.