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Respiratory Epithelial Adenomatoid Hamartoma-A Case Report

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Abstract: *Respiratory epithelial adenomatoid hamartomas (REAHs) are rare, benign glandular proliferations of the sinonasal cavity and nasopharynx, originating in the schneiderian epithelium. Recognition and awareness of this benign lesion is necessary to distinguish it from inverted papilloma and adenocarcinoma, to avoid aggressive surgery. We report a case of REAH diagnosed on histopathology.*

Keywords*: Respiratory epithelial adenomatoid hamartoma, nasal obstruction, sinonasal cavity, inverted papilloma and sinonasal adenocarcinoma.*

I. Introduction

Hamartomas are benign, non-neoplastic lesions that occur secondary to tissue-development anomalies and they are composed of overgrowth of mature cells and tissues that normally occur in the affected location.[1]They are common in the lung, kidney, liver, spleen, and intestine but are extremely rare in the upper aerodigestive tract.[2]

Hamartomas can be further classified into mesenchymal and epithelial subtypes according to predominant element on histopathologic examination. A particular subset of epithelial subtype known as respiratory epithelial adenomatoid hamartoma (REAH) was first described in 1995 by Wenig and Heffner in a series of 31 cases from the files of the Otolaryngic Tumor Registry at the Armed Forces Institute of Pathology.[3]

They are commonly reported in the nasal cavity and nasopharynx.[1] In the nasal cavity, REAHs are mostly associated with the posterior nasal septum, although lesion arising from the lateral nasal wall has also been reported. The involvement of the maxillary sinus is extremely rare, although occasional occurrence of REAHs in areas surrounding nasal cavity, such as the ethmoid sinus,frontal sinus and nasopharynx has been described.[4] It is a benign condition but the histopathological features can mimic inverted papilloma or well differentiated adenocarcinoma leading to misdiagnosis.[5] The challenge of this lesion is to not overdiagnose it as malignant because it can be treated with a simple excision as opposed to radical surgery.[6] We present a case report of a 48-year-old male with REAH localized in the right nasal cavity.

II. Case Report

A 48 year male presented to the ENT OPD with complaints of nasal obstruction and hyposmia for the past one year. There was no history of facial pain, epistaxis or eye problems. He was otherwise healthy with no known co-morbidities. On physical examination there was a polypoidal mass occupying the right nasal cavity. He underwent endoscopic biopsy.

2.1 Gross-: Specimen consisted of three bits of grey-white to grey-brown tissue largest measuring 0.6 cm×0.3 cm×0.2 cm.

2.2 Microscopy-: Sections studied showed tissue bit lined by pseudostratified ciliated epithelium with squamous metaplasia. Subepithelium showed multiple glands composed of multilayered respiratory epithelium with mucin secreting cells. Thick hyalinised basement membrane with areas of fibrosis were seen. Chronic inflammatory cell infiltrates were also seen along with few seromucinous glands. Features were suggestive of respiratory epithelial adenomatoid hamartoma.

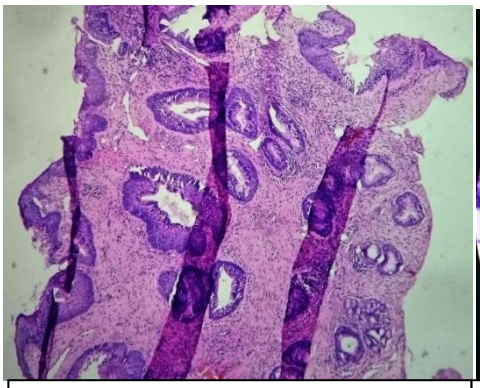

Figure 1- Microscopy shows pseudostratified ciliated epithelium with subepithelium showing multiple glands. H&E 10X10.

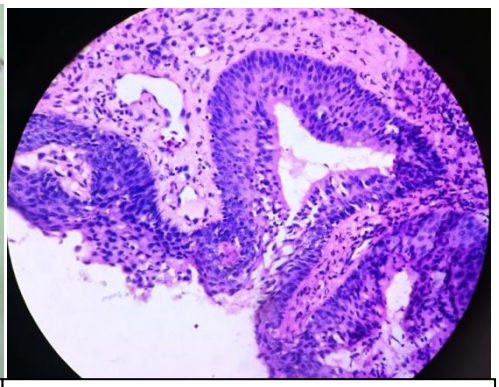

Figure 2- Microscopy shows pseudostratified ciliated epithelium with squamous metaplasia. H&E 40X10.

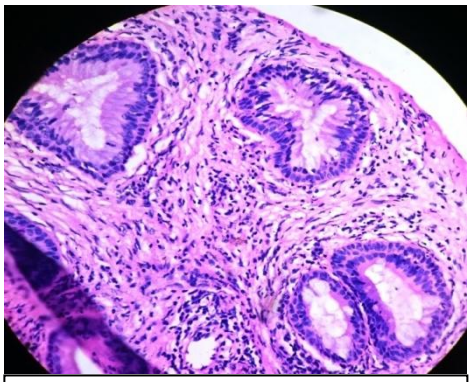

Figure 3- Microscopy shows mucin filled glands composed of multilayered respiratory epithelium along with stroma showing chronic inflammatory cell infilterates. H&E 40X10.

III. Discussion

Hamartomas may be viewed as malformations composed of an excessive proliferation of a specific cellular component endogenous to a given tissue. In contrast to neoplasms, hamartomatous proliferations do not have the capability of continuous unimpeded growth and are, therefore, self-limiting.[7]

REAH is a distinct and rare type of hamartoma occurring in the sinonasal tract. The etiology is unclear but is thought to involve an inflammatory process. This hypothesis arises from the observed occurrence of REAH in the setting of rhinosinusitis, inflammatory polyposis, and sinus surgery.[8] Since Wenig's seminal series, literature regarding this lesion has been primarily limited to case reports and histopathological analyses.[9]

Over 80% of the patients with REAH are males, age ranging from the third to the ninth decade of life, with a median age in the 6th decade.[2] Commonly described presenting complaints are nasal obstruction, nasal stuffiness, epistaxis, rhinorrhea, chronic recurrent sinusitis, facial pain, proptosis and hyposmia.[3]

Radiographically the most common finding of REAH is an opacification of the affected sinus and some connection to the nasal septum.[5] Calcification may be visualized within the lesion but bone erosion and intracranial involvement are uncommon.[3]

One study that investigated the molecular genetic changes in REAH revealed a higher loss of heterozygosity (LOH) at loci located on chromosome 9p and 18q than expected for a benign lesion. The 9p region is known to code for two structurally distant tumor suppressor proteins, while LOH at 18q has been shown to be common in recurrent or metastatic squamous cell carcinoma of the head and neck. Despite these reported molecular alterations, REAH are completely benign lesions.[10]

Macroscopically, REAHs appeared as edematous, yellow–pink masses with a glistening surface similar to inflammatory polyps but were generally darker, with a more indurated, rubbery consistency.[9] Histologically, the lesions are characterized by a prominent tubular glandular proliferation lined by ciliated respiratory epithelium, originating from the surface epithelium, stromal hyalinization which envelops the adenomatous proliferation, and mucous cell metaplasia. The basement membrane is thickened. No destructive growth is

noted.[11] The nuclear features of REAH are bland. Prominent nucleoli are rare, as are mitoses. These findings support the benign nature of the lesion.[6]

The exact differential diagnosis is made by histopathologic examination.[1] The differential diagnosis includes inflammatory polyps (fewer glands) and inverted schneiderian papillomas (which have a hyperplastic squamous epithelium with only a few goblet cells and a thin basement membrane). More important is the differential diagnosis of sinonasal low grade adenocarcinoma, which usually demonstrates a back to back glandular pattern with nuclear atypia, prominent mitotic activity, desmoplastic stroma and perineural invasion.[11] Despite characteristic histological features that distinguish these tumours, the differential diagnosis can be challenging in cases of small or fragmented endoscopic biopsies.[4]

Although considered benign, REAHs have been shown to be locally aggressive. Potential for intracranial and orbital extension can occur if left untreated, particularly in the setting of frontal sinus involvement. Successful treatment of REAHs involves complete surgical resection, frequently accomplished using a transnasal endoscopic procedure. [9] There are no reports of recurrent, persistent, or progressive disease, in the literature.[2]

IV. Conclusion

Respiratory epithelial adenomatoid hamartoma truly is a lesion in its infancy. It mimics inverted papilloma and sinonasal adenocarcinoma. Diagnostic misinterpretation is a serious issue regarding this lesion. Pathologists must be aware of this entity in order to avoid overdiagnosis and excessive surgical procedures for the patient.

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