Respiratory Epithelial Adenomatoid Hamartomas (REAH)

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INTRODUCTION

Hamartoma is a malformation that presents as a benign non-neoplastic overgrowth or mass of disorganized tissue indigenous to the area of its occurrence. Sinonasal hamartomas are extremely rare and most often of the pure epithelial type.

Respiratory epithelial adenomatoid hamartoma (REAH) was first described by Wenig and Heffner in 1995 and was the largest series to date at that time, with 31 cases occurring in the nasal cavity, paranasal sinuses, and nasopharynx. REAH is a benign entity characterized by abnormal glandular proliferation of the surface ciliated respiratory epithelium surrounded by thick eosinophilic basement membrane with no atypia or metaplastic squamous change. Though commonly an incidental finding on pathology reports, correctly diagnosing REAH is essential to prevent more aggressive surgical resections than necessary as well as to prevent undertreatment of more malignant pathology.

METHODS AND MATERIALS

Pathology specimens were reviewed from all endoscopic sinus surgery cases performed at UCLA Medical Center between January 2000 and May 2011. Of the 3,120 sinus surgeries performed during this time, there were 54 patients who were diagnosed with REAH over this 10-year period. All specimens were reviewed by the senior head and neck pathologist (S.B.). A retrospective patient chart review was then conducted to determine age, sex, location, associated findings, radiographic features, and recurrences among these identified patients.

RESULTS

54 patients with REAH were identified over a 10-year period. No biopsies done prior to definitive surgery revealed REAH in the specimen. One case had evidence of squamous metaplasia of the submucosal glands during definitive surgery. There were no distinguishing symptoms on review of systems that would suggest the diagnosis of REAH. Similarly, there were no characteristic features on endoscopic examination consistent with the diagnosis of REAH.

While the majority of cases had findings of REAH within the sinuses, 8 (15%) were present as isolated masses within the nasal cavity. Of these eight cases, 5 (63%) had REAH as the only pathology within the specimen. 2 (25%) cases were centered on the posterior nasal septum, 3 (37.5%) were centered on the middle turbinate, and 3 (37.5%) were centered on the inferior turbinate.

24 (44%) were associated with an allergic type of chronic sinusitis, and 9 (17%) were associated with nasal polyposis. 44 (81%) cases of REAH were present within a background of chronic sinusitis or chronic allergic sinusitis, and were the dominant pathology within the specimen.

There were 2 (3.7%) recurrences during this 10-year period, with no recurrences following repeat surgery. The average follow-up was 3.8 years.

CONCLUSIONS

REAH is a benign entity characterized by abnormal glandular proliferation of the surface ciliated respiratory epithelium, admixed with goblet cells with no atypia or metaplastic change, rimmed by a thick basement membrane. REAH may be confused with other pathology, such as inflammatory polyps, inverted papillomas, and low grade adenocarcinoma can lead to unnecessarily more extensive surgical interventions. In contrast, mistaking other more aggressive pathology for REAH may lead to incomplete resections without clear margins. Therefore, correctly diagnosing each of these pathologic diagnoses is paramount.

REFERENCES