

## Respiratory epithelial adenomatoid hamartoma of the maxillary sinus: A case report

**Gerry Raymond Joviolo, Kartono Sudarman, Agus Surono**

### ABSTRACT

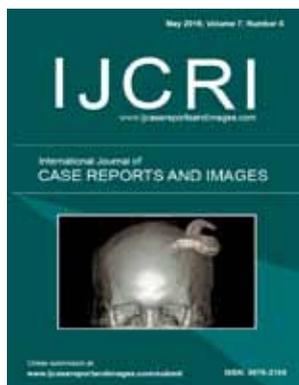
**Introduction:** Hamartoma is non-neoplastic tumor characterized by excessive proliferation of tissue in parts of the human body. Hamartoma is a very rare in head and neck regions.

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**Conclusion:** Even though REAH is very rare, otolaryngologist should be aware of the pathologic entity of this disease to differentiate REAH with inverted papilloma, adenocarcinoma or other paranasal sinuses inflammation. Misinterpretation of REAH as a chronic sinus inflammation may lead to inadequate treatment.



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# Respiratory epithelial adenomatoid hamartoma of the maxillary sinus: A case report

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## ABSTRACT

**Introduction:** Hamartoma is non-neoplastic tumor characterized by excessive proliferation of tissue in parts of the human body. Hamartoma is a very rare in head and neck regions. **Case Report:** A case of 33-year-old woman who was initially diagnosed as a chronic sinus inflammation. After Caldwell-Luc approach for surgery, histopathological examination showed respiratory epithelial adenomatoid hamartoma (REAH) of the left maxillary sinus. It was an atypical localization because the most common site for REAH was in the nasal cavity. **Conclusion:** Even though REAH is very rare, otolaryngologist should be aware of the pathologic entity of this disease to differentiate REAH with inverted papilloma, adenocarcinoma or other paranasal sinuses inflammation. Misinterpretation of REAH as a chronic sinus inflammation may lead to inadequate treatment.

**Keywords:** Neoplasm, Paranasal sinus, Respiratory epithelial adenomatoid hamartoma, Surgery

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Gerry Raymond Joviolo<sup>1</sup>, Kartono Sudarman<sup>1</sup>, Agus Surono<sup>1</sup>  
**Affiliations:** <sup>1</sup>Department of Otorhinolaryngology-Head and Neck Surgery, Faculty of Medicine, Universitas Gadjah Mada, Sekip Utara, Yogyakarta, Indonesia.  
**Corresponding Author:** Agus Surono MD ORL-HNS PhD, Department of Otorhinolaryngology-Head and Neck Surgery, Faculty of Medicine, Universitas Gadjah Mada, Sekip Utara, Yogyakarta, Indonesia; Email: agus\_srn@yahoo.com

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## INTRODUCTION

Hamartoma term was firstly introduced by Albrecht [1] as the primary non-neoplastic malformations of tissue composed of excessive proliferation of cells and tissues indigenous to particular site of the body. Hamartoma may occur in any part of the body such as the surface of epithelium, seromucous glands, fibrous stroma, and vessels [2]. They are common in the spleen, lung, liver, and kidney and intestinal, but rarely found in the head and neck regions [3].

Hamartoma of the head and neck was initially described by Wenig and Heffner [4], a subgroup of hamartoma more often involving upper aerodigestive tract, which was the respiratory epithelial adenomatoid hamartoma (REAH). They found 31 cases with lesion occurred in the nasal cavity, paranasal sinuses, and nasopharynx associated with tumor originating from surface epithelium with proliferation and accumulation of glands and ducts, lined by ciliated respiratory epithelium, and surrounded by edematous or inflammatory background and sometimes invagination of the respiratory epithelium is observed. The proliferation is not derived from seromucous glands that usually found in the region [3–5]. The pathogenesis of REAH is still speculative. Congenital and prolonged inflammatory are often considered to be predisposing factors. The REAH mostly affect men with male to female ratio of 7:1, ranging from third to ninth decades of life with a median age in the sixth decade of life [3, 6].

## CASE REPORT

A 33-year-old female presented with more than five months history of nasal blockage with posterior

rhinorrhea. The patient also complained toothache since five years ago. The patient had taken antibiotic treatment but the symptoms were still present. There was no other significant medical history. The physical examination demonstrated swollen of left inferior turbinate, posterior rhinorrhea and upper left 1st molar caries without any mass found. Computed tomography (CT) scan revealed hypodense mass at the left maxillary sinus suggesting a mucocele of the left maxillary sinus (Figure 1). The patient underwent Caldwell-Luc approach of surgery under general anesthesia to evacuate the presence of mass in the left maxillary sinus. Mass of the left maxillary sinus was shown like a sac filled with serous fluid.

Histopathological examination revealed that the lesion was composed of tumor with glandular proliferation covered with pseudostratified ciliated respiratory epithelial cells. The lesion surface was lined in direct continuity with the ciliated respiratory epithelium creating a papillary appearance with elongated invagination to the submucosa. Other abnormal features including hyalinization with eosinophilic basement membrane covering the glands, stromal edema with chronic inflammatory proliferation was also noted (Figure 2). The patient was well recovered and without recurrence of symptoms at first year follow-up.

## DISCUSSION

Hamartoma has been described as a mass that developed with abnormal tissue growth. Unlike neoplasms, hamartoma has no capacity to grow continuously, therefore their proliferation is self-limiting. Respiratory epithelial adenomatoid hamartoma (REAH) is a rare type of hamartoma and found in the nasal cavity, the paranasal sinuses and nasopharynx. Wenig and Heffner [4] performed the largest study of REAH in 1995 by identifying 31 cases from Tumor Registry at the Armed Forces Institute of Pathology which consisted of 27 men and 4 women, aged from 27–81 years old, with mean age of 58 years, and the youngest reported was nine years old. In this case report, the patient was woman with age of 33 years old showing an incidence of REAH at the 4<sup>th</sup> decade of life.

The REAH has been commonly identified at the nasal cavity (70%), most often at the posterior nasal septum. The other sites are the ethmoid and frontal sinuses, nasopharynx and rarely occurred in the maxillary sinus. Di Carlo et al. [6] reported 15 cases of REAH found in the anterior half of the olfactory clefts bilaterally. Only limited literatures of REAH were published, and mostly were case reports. The previous reports of REAH have described at least  $\pm$  60 cases confirming rare entity of this disease. This makes a challenge for this lesion not to be misdiagnosis as malignant because it can be treated with simple excision rather than radical surgery. In this case, REAH developed in the left maxillary sinus without any mass in the nasal cavity. Only three cases of REAH in the maxillary sinus had been published in literature.

The presenting symptoms of REAH have been shown as nasal obstruction, nasal stuffiness, rhinorrhea, epistaxis, anosmia/hyposmia and chronic sinusitis [3]. Kessler [7] reported a case of maxillary sinus of REAH presenting as a periapical radiolucency of the first molar without any sinonasal symptom. Symptoms usually occur for a few months up to eight years. Our case showed the symptoms of nasal blockage and posterior rhinorrhea. Opacification of the affected sinus and some connection to the nasal septum are the most common finding of REAH radiologically. The lesion tends to grow slowly and be able to cause bone expansion rather than bone erosion. Lima et al. [8] found that REAH significantly enlarged the width of the olfactory cleft.

Pathological examination showed that REAH tends to have a polypoid appearance, fleshy to firm, dark brown to white masses with varying size and area of induration



Figure 1: Coronal view of computed tomography scan showed the mass in left maxillary sinus.

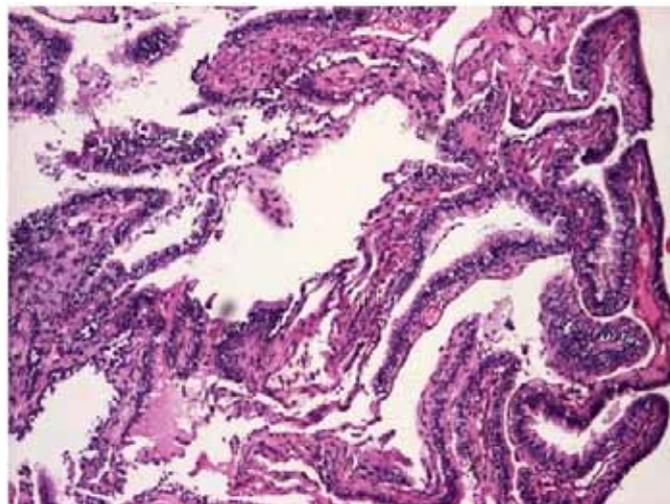


Figure 2: Microscopic appearance of respiratory epithelial adenomatoid hamartoma (REAH). The tissue showed glandular proliferation with invagination of surface epithelium to the submucosa.

noted when the tumor is cut. The histologic features are dominated by the presence of glandular proliferation with the gland covered by the ciliated respiratory epithelium originating from the surface respiratory epithelium with proliferation tends to be submucosal. Other features are like stromal hyalinization and thickening of basement membrane, stromal edema, seromucous gland proliferation, chronic inflammatory cell, and no dysplastic or neoplastic changes seen [9, 10].

The pathogenesis of REAH is unknown. As chronic inflammatory cell often found in REAH histologic view, inflammation is hypothesized may induced REAH [3]. In this case, we found the patient who had teeth caries of upper left 1st molar, could be considered as a risk factor for sinusitis. Prolonged inflammation of the maxillary sinus may develop into REAH [3, 6]. It is a challenge for otolaryngologist to make a distinction of REAH with inflammatory disorders in maxillary sinus because small number of reported cases of REAH in maxillary sinus. The treatment of REAH is complete local excision and there has not been any report of recurrence or progressivity of the disease in literature. Respiratory epithelial adenomatoid hamartoma (REAH) in paranasal sinuses have often been differentially diagnosed with inverted papilloma, adenocarcinoma and nasal polyp [4]. In this case, CT examination suggested mucocele of maxillary sinus prior to pathologic result.

The REAH can be differentiated with nasal polyp from the gross examination of indurations and histological findings of extensive glandular proliferation and stromal hyalinization in REAH. These findings cannot be found in nasal polyp. When differentiating of inverted papilloma with REAH, examination should carefully be performed. The treatments between both diseases are slightly different. The REAH needs local excision while inverted papilloma needs more extensive surgical excision. Inverted papilloma clinically has the ability to destroy bone and invade adjacent vital structures. Histologically, epithelial proliferation with marked thickening, intra epithelial mucous cysts and the presence of inflammatory cells in the epithelium are specific finding of inverted papilloma. The REAH tends to be a single cell layer ciliated respiratory epithelium and inverted papilloma lined by multiple layers of squamous, ciliated, columnar or transitional epithelium. The REAH is also difficult to be differentiated against low grade adenocarcinoma. Stroma identified between the ciliated glands in the case of REAH is the best way to differentiate it from low grade adenocarcinoma. Immunohistochemistry examination using antibody MIB-1 (KI-67) shows higher immunoreactivity of adenocarcinoma compared to hamartoma [3, 9, 11].

## CONCLUSION

A case of respiratory epithelial adenomatoid hamartoma (REAH) has been reported. Despite REAH is

very rare, it is important for otolaryngologist to be aware of this pathological entity and to differentiate REAH with inverted papilloma or adenocarcinoma in order to avoid unnecessary aggressive surgery. Misinterpretation of REAH as any sinus inflammatory may result in inadequate treatment.

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## Author Contributions

Gerry Raymond Joviolo – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Kartono Sudarman – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Agus Surono – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

## Guarantor

The corresponding author is the guarantor of submission.

## Conflict of Interest

Authors declare no conflict of interest.

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## ABOUT THE AUTHORS

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**Gerry Raymond Joviolo** is Faculty of Medicine in Department of Otorhinolaryngology -Head and Neck Surgery, Universitas Gadjah Mada, Sekip Utara, Yogyakarta, Indonesia.



**Kartono Sudarman** is Faculty of Medicine in Department of Otorhinolaryngology -Head and Neck Surgery, Universitas Gadjah Mada, Sekip Utara, Yogyakarta, Indonesia.



**Agus Surono** is Faculty of Medicine in Department of Faculty of Medicine -Head and Neck Surgery, Universitas Gadjah Mada, Sekip Utara, Yogyakarta, Indonesia. His area of interest include head and neck cancer and bronchoesophagology.  
E-mail: agus\_srn@yahoo.com

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