**Respiratory Epithelial Adenomatoid Hamartoma,**

**a different presentation**

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Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

**FUNDING AND CONFLICT OF INTERESTS**

Competing interests: None. | Sponsorships: None. | Funding source: None.

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*REAH, a different presentation*

**RESPIRATORY EPITHELIAL ADENOMATOID HAMARTOMA,**

**A DIFFERENT PRESENTATION**

**Introduction:** Respiratory Epithelial Adenomatoid Hemartoma (REAH) has been included in the World Health Organization’s classification of tumours since 2005. It results from non-neoplastic overgrowth of glandular tissue. It is rarely located in the upper airway and in 70% of these cases it’s present in the nasal septum. Within the described locations, it has never been described as attached to the nasal floor.

**Objective:** Presentation of a clinical case with REAH located in the nasal floor.

**Material and methods:** We present the case of a 56-year-old male patient with progressive right nasal obstruction for 2 years. A nasal endoscopy showed a whitish, lobulated and large lesion with well-defined contours and with solid and polypoidal components. The incisional biopsy was inconclusive and the computed tomography (CT) of the paranasal sinuses showed obliteration of the right nostril and of the upper nasopharyngeal and oropharyngeal airways by a lobulated lesion. We performed endoscopic excision of the 9 cm lesion attached to the nasal floor. Histopathologic study revealed REAH.

**Results:** Of uncertain etiology, REAH appears to correlate with chronic inflammation. It presents with non-specific symptoms, which may mimic other nasal lesions. Despite not being described in the literature, our case presented REAH located in the nasal floor. Only excisional biopsy determines the diagnosis. Treatment involves complete endoscopic excision.

**Conclusion:** REAH are underdiagnosed and identification is important to avoid more aggressive treatments. In this work we present a case of location in the nasal floor, not described in the literature to this date.

**Keywords:** Respiratory Epithelial Adenomatoid Hamartoma, Nasal floor, Endoscopic Sinus Surgery

**Sort summary**

In this study we present a clinical case of Respiratory Epithelial Adenomatoid Hamartoma with insertion on nasal floor, never described before.

**Introduction**

Respiratory Adenomatoid Hamartoma (HAER) is a rare tumor that has been included in the World Health Organization’s (WHO) classification of tumours since 2005.1 It results from non-neoplastic overgrowth of glandular tissue, a local malformation of multiple aberrant cell types.2 This tumor is rarely located in the upper airway and in 70% of these cases it’s present in the nasal septum. Within the described locations, it has never been described as attached to the nasal floor.3 It usually occurs unilaterally and surgical resection is curative.

During clinical evaluation, this pathology can mimic a sinonasal carcinoma, with therapies that are associated with significant morbidity.

**Case Report**

#### We present the case of a 56-year-old male patient, non-smoker, with a personal history of exposure to dust in construction work and without a medical history of rhinosinusitis and allergies or family history of nasal pathologies. He showed progressive right nasal obstruction for 2 years associated with hyposmia and posterior rhinorrhea. He denied experiencing otologic and cervical symptoms.

#### A nasal endoscopy showed a whitish, lobulated and large lesion with well-defined contours and with solid and polypoidal components. It occupied the entire right nasal cavity, extending to the rhinopharynx through the right choanae. There was no contact bleeding. The rest of the physical exam findings were unremarkable.

#### An incisional biopsy was performed inthe practice. The histopathologic study revealed “mucosa lined by respiratory-type epithelium with reactive epithelial changes, expanded chorion at the expense of fibrosis and mild chronic inflammatory infiltrate, with no malignant neoplasm tissue observed”.

#### The Computed Tomography (CT) of the paranasal sinuses showed “obliteration of the posterior region of the right nasal cavity and of the upper nasopharyngeal and oropharyngeal airways by a lobulated lesion marginal well delimited with heterogeneous density after iodinated contrast enhancement”, with no other alterations and no exclusion of malignancy. (Fig. 1 and 2)

#### The patient was scheduled to undergo an endoscopic sinus surgery under general anesthesia with priority. Intraoperatively, a 9 cm lesion attached to the nasal floor was observed (Fig. 3), which was entirely excised with electrocautery, aspiration and “Blakesley”. There was minimal bleeding. The histopathology study revealed Respiratory Epithelial Adenomatoid Hamartoma (REAH).

#### After surgery, the patient denied experiencing nasal obstruction, hyposmia and rhinorrhea. He was discharged from the hospital on the following day.

#### Because there was a delay making the Nuclear Magnetic Resonance (NMR) in our centre, the patient ended up being operated before the exam. Nonetheless, it confirmed the complete excision of the lesion. (Fig. 4 and 5).

**Discussion**

Of uncertain etiology, REAH appears to correlate with chronic inflammation, in which mast cells play an important role. It presents with non-specific symptoms such as nasal obstruction, chronic sinusitis, deviated septum, frontal headache rhinorrhoea, hyposmia/anosmia or epistaxis.3

It can be observed in two forms: isolated REAH or in association with another inflammatory process such as nasal polyposis.3 It can mimic nasal polyposis, inverted papilloma or adenocarcinoma, squamous cell carcinoma, olfactory neuroblastoma and lymphoma.2

Aggressive surgical intervention and adjuvant therapies for squamous cell, adenocarcinoma, adenoid cystic carcinoma and mucosal melanoma are associated with significant morbidity. Therefore, the correct diagnosis is imperative to avoid aggressive therapy.2

Most HAERs occur in adult males between third and nineth decades, with predominance of the fifth decade. 2,3

REAH was highly associated with a long duration of nasal polyposis, asthma and history of repeated sinus surgery.

This lesion is a complex diagnosis that often cannot be based on clinical findings nor radiological evidence alone.2

Previous reports have identified REAH arising from the nasal septum (especially the posterior aspect), lateral and posterior nasal walls. However, we describe the first case arising from the nasal floor.2,3

The most frequent finding on CT paranasal sinus is opacification of the affected paranasal sinus extending to the nasal septum. *Vira et al* concluded that no distinguishing features could differentiate it from any other sinonasal lesion. But *Lima et al* demonstrated that enlargement of the olfactory cleft greater that 10 mm is characteristic of REAH.3,4,5 However, these features were not found in our patient since our case presented REAH attached to the nasal floor.

Incisional biopsy is not recommended due to the heterogeneity of the lesion. Thus, only excisional biopsy determines the diagnosis.2 In literature, the histhopathologic analysis is characterized by a prominent glandular proliferation, with small to medium-sized, round to oval glands that are widely separated by stromal tissues. Glands are lined by multi-layered ciliated respiratory epithelium with goblet cells and have a thick, eosinophilic basement membrane. They don’t present atypia or metaplastic change.2

Treatment of this pathology involves complete endoscopic excision. The prognosis is good with risk of recurrence with a incomplete excision.1

**Conclusion**

REAHs are underdiagnosed and proper identification is important to avoid more aggressive treatments such as mutilating surgeries and adjuvant therapies. In this article, we present a case of location in the nasal floor, not described in literature to this date.

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**Figure legends**

**Fig 1 and 2:** CT-scan paranasal sinus (soft tissue window constrasted) – Coronal (1) and Axial (2) views with obliteration of the posterior region of the right nasal cavity and of the upper nasopharyngeal and oropharyngeal airways by a lobulated lesion marginal well delimited with heterogeneous density after iodinated contrast enhancement

**Fig. 3:** REAH - 9m, spherical, well-circumscribed polypoidal tumor

**Fig. 4, 5.** MR post-operative image – Coronal (4) and Axial (5) views showing complete excision of the lesion