

A Case of Respiratory Epithelial Adenomatoid Hamartoma

L.G. Marin, V. Trombitas, S. Albu

IInd Department of Otolaryngology, University of Medicine and Pharmacy Cluj-Napoca, Cluj-Napoca, Romania

Rezumat

Un caz de hamartom adenomatoid epitelial respirator

În acest studiu descriem un pacient în vârstă de 50 de ani, de sex masculin, internat pentru o formațiune tumorală nazală unilaterală. Diagnosticul histopatologic a fost de hamartom adenomatoid epitelial respirator (HAER). HAER este o entitate histopatologică recent descrisă care se manifestă clinic prin obstrucție nazală, rinoree, epistaxis, hiposmie și cefalee. Este o leziune rară nazală sau a sinusurilor paranasale, dar este necesară menționarea în diagnosticul diferențial al formațiunilor tumorale nazale deoarece este de natură benignă și poate fi rezecată complet chirurgical.

Cuvinte cheie: hamartom adenomatoid, obstrucție nazală, tumori nazale, diagnostic histopatologic

Abstract

We report a case of a 50-year-old man diagnosed with a unilateral nasal mass found to be a respiratory epithelial adenomatoid hamartoma (REAH) upon pathologic examination. REAH is a recently described pathologic entity that can present with nasal obstruction, congestion, rhinorrhoea, epistaxis, hyposmia, and headaches. It is a rare lesion of nasal

and paranasal sinuses, but should be considered in the differential diagnosis because it is a benign lesion and complete surgical resection is curative.

Key words: adenomatoid hamartoma, nasal obstruction, nasal tumors, histopathologic diagnosis

Introduction

Hamartomas are benign malformations or inborn errors of tissue development. The term "hamartoma" was first described by Albrect in 1904. A hamartoma is composed of disorganized tissue indigenous to the particular site, but with an excess of one or more of the tissue types. Hamartomas can occur in any area of the body, with a predilection for the lung, kidney, and intestine (1). Hamartomas of the head and neck region, in particular the nasal cavity and paranasal sinuses, are very rare (1). One particular type of hamartoma, first described by Wenig and Heffner in 1995 (2), is the respiratory epithelial adenomatoid hamartoma (REAH). Among sinonasal hamartomas, the REAH subgroup of hamartomas represents a rare entity that has been described in only a handful of case reports.

Case report

A 50-year-old male patient presented with a 6-month history of progressive nasal obstruction, unilateral purulent nasal discharge, posterior rhinorrhoea, headache and halitosis. The patient underwent previous allergy tests, with negative results, was a non-smoker and had no significant medical history.

Corresponding author: Silviu Albu, MD
Professor, IInd Department of Otolaryngology
University of Medicine and Pharmacy Cluj-Napoca
Republicii Street, no. 18, 400015, Romania
E-mail: silviualbu63@gmail.com

The endoscopic examination revealed an S-shaped nasal septum deviation, congestion of the nasal mucosa, right mucopurulent nasal discharge and multiple white, oedematous polypoid masses located in the right sphenothmoid recess, connected to the posterior nasal septum (Fig. 1).

The CT scan showed the presence of soft tissue masses in the posterior aspect of the nasal fossa, opacification of the right ethmoid sinus and heterogeneous opacification of the right sphenoid sinus (Fig. 2).

The patient underwent complete endoscopic surgical excision of the polypoid masses and right sphenoidectomy. The microscopic examination of the excised tissue revealed numerous tubular structures lined by simple cuboidal epithelium or ciliated columnar epithelium, separated by

stromal tissue (Fig. 3). The stroma had an oedematous aspect and contained a rich inflammatory cell infiltrate, composed of diffusely spread lymphocytes, plasma cells and eosinophils (Fig. 4). A diagnosis of REAH was made.

Discussion

Hamartomas are benign lesions defined as aberrant differentiation which may produce a mass of disorganized, but mature specialized cells or tissue indigenous to the site of growth (1). In 1995 Wenig and Heffner published a series of 31 cases, describing a type of hamartoma characterized by prominent glandular proliferations lined with ciliated respiratory epithelium originating from the surface epithelium, called

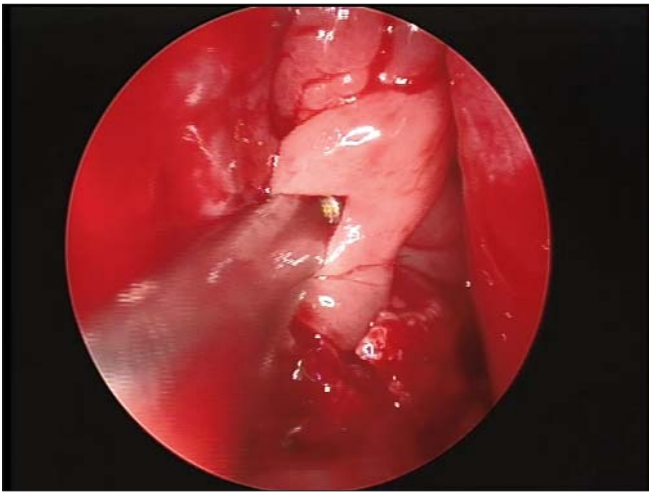


Figure 1. Endoscopic appearance of an hamartoma

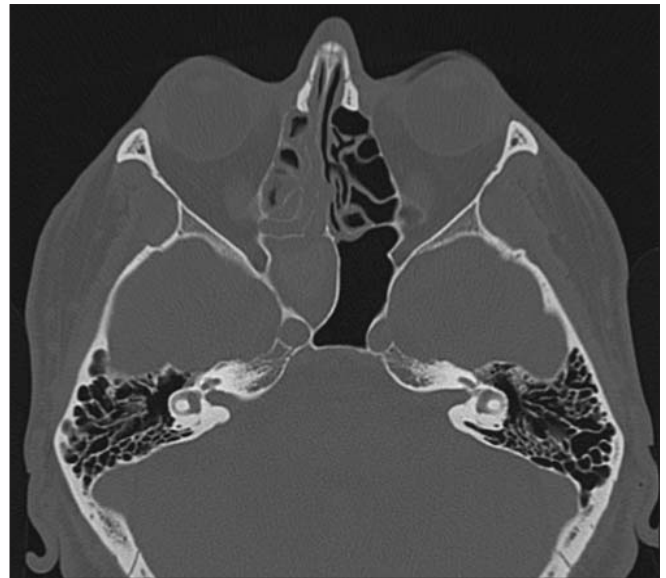


Figure 2. Axial CT scan demonstrating opacification of the right ethmoid and sphenoid sinus

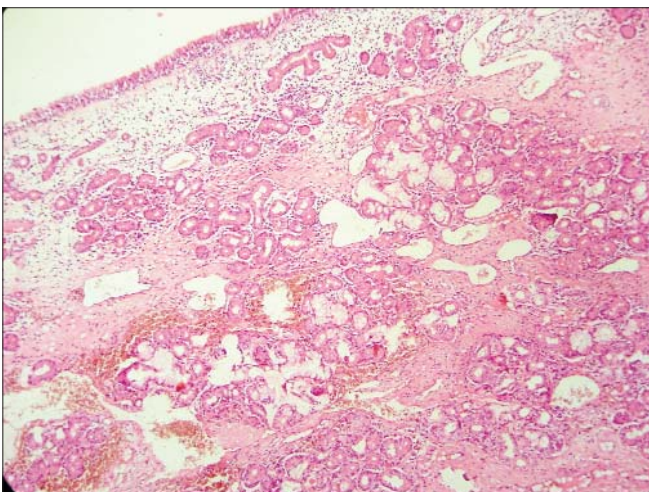


Figure 3. Hematoxylin and eosin stain demonstrating glandular proliferation and glands lined with ciliated respiratory epithelium

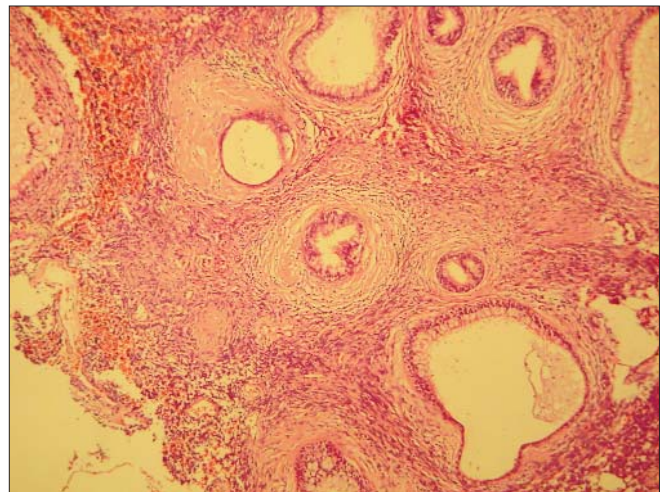


Figure 4. The proliferated glands of various sizes are separated by stromal tissue and filled with amorphous debris

Respiratory epithelial adenomatoid hamartoma (REAH) (2). Later, a related subtype was defined, COREAH (Chondro-Osseous Respiratory Epithelial Hamartoma), differing from REAH by the fact that it contains islands of cartilage interspersed throughout the lesion (3).

Respiratory epithelial adenomatoid hamartomas occur in the nasal cavity, paranasal sinuses and nasopharynx, with predilection for the nasal cavity, especially the posterior septum (1,4,5). The lesion often involves both nasal cavities and is commonly associated with chronic rhinosinusitis (5,6). In fact, it seems that the inflammatory process may be the etiologic precursor of REAH (3). In addition, REAH is frequently associated with nasal polyps.

REAH is a benign lesion predominantly affecting men in their third to ninth decades of life, with a median age of 58 years. Usually the presenting symptoms are nasal obstruction and stuffiness, rhinorrhoea, epistaxis, facial pain, proptosis and hyposmia, extending on a period of months and even years.

Hamartomas are characterized by self-limited proliferation and slow growth, a finding that is radiologically demonstrated by bony expansion, rather than erosion. Also, REAH is associated with enlargement of the olfactory clefts on CT scan, the difference being significant in individuals with bilateral disease (1,7). Radiologically, the most common finding is an opacification of the affected sinus and some connection to the nasal septum (1-5).

On gross evaluation, REAHs appear as shiny, edematous polypoid masses, with various sizes, that tend to be fleshy to firm, yellow to white in color. Microscopically, the lesions are characterized by glandular proliferation, which tends to be submucosal (1). The glands are lined with a single layer of ciliated respiratory epithelium originating from the surface respiratory epithelium. They are round to oval in shape, small to medium in size with prominent dilation. The stroma is usually oedematous and contains a mixture of inflammatory cells including eosinophils. Complex glandular growth and cribriform architecture is absent (1).

Although REAH is considered a non-neoplastic entity, molecular genetics findings, as presented in Ozolek and Hunt's study, suggest that REAH may in fact be a benign neoplasm and not a hamartoma, as it was originally believed (1,7).

REAH has no tendency to regress spontaneously and complete surgical excision is curative. It is important to differentiate REAH from other sinonasal lesions, such as inverted papilloma or adenocarcinoma, to avoid an overly aggressive treatment. REAH can be misdiagnosed as an inflammatory polyp because of the clinical and morphological similarities. Second in the differential diagnosis of REAH, inverted papillomas tend to be more aggressive lesions, having the capacity to destroy bone, extend along mucosal surfaces and invade adjacent vital structures. Histologically, inverted papillomas are characterized by invagination of the surface

epithelium in the underlying stroma, consisting of papillary fronds with a delicate fibrovascular core, covered by multiple layers of epithelial cells (6,7). Sinonasal adenocarcinomas originate from the glandular epithelium and are characterized by a complex glandular growth pattern with a back-to-back or cribriform pattern lacking intervening connective tissue. A few criteria used to differentiate REAHs from low grade adenocarcinomas are individual glands surrounded by eosinophilic basement membranes, lined by ciliated respiratory epithelium, the absence of cribriform architecture, lack of nuclear stratification, lack of dysplasia or increased mitotic rate (8). Immunohistochemically adenocarcinoma demonstrates higher reactivity for MIB-1 staining than REAH (8). Furthermore, adenocarcinoma shows behavior of bone destruction and invasion into the surrounding soft tissue on imaging. The treatment requires radical aggressive surgery and radiotherapy (9).

While REAH occurs mostly in the nasal cavity, paranasal sinus hamartomas have been described. Because hamartomas do not have malignant potential complete removal via conservative surgery is curative. With endoscopic approaches exploring the basement of the mass it is necessary to ensure en bloc resection. A misdiagnosis of REAH as either inverted papilloma or adenocarcinoma would likely result in far more aggressive surgical intervention than is needed for the REAH.

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