

Respiratory epithelial adenomatoid hamartoma of the maxillary sinus: case report

Amartoma adenomatoide epiteliale respiratorio del seno mascellare: caso clinico

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Key words

Maxillary sinus • Paranasal sinus neoplasm • Chronic sinusitis • Surgical treatment • Epithelial adenomatoid hamartoma

Parole chiave

Seno mascellare • Neoplasie dei seni paranasali • Sinusiti croniche • Terapia chirurgica • Amartoma adenomatoide epiteliale

Summary

The case is described of a male patient with respiratory epithelial adenomatoid hamartoma of the left maxillary sinus that initially presented as a chronic sinus inflammation. This benign lesion is characterized by glandular proliferation originating from the surface of the respiratory epithelium. Maxillary sinus localisation is very rare but is very important to be able to distinguish hamartomas from schneiderian papillomas of the inverted type and adenocarcinomas, potentially requiring aggressive surgical treatment. Moreover, misinterpretation of the respiratory epithelial adenomatoid hamartoma as chronic sinus inflammation may result in inadequate treatment. The clinical and pathological features of this lesion are discussed.

Riassunto

Si descrive un caso di amartoma adenomatoide epiteliale respiratorio del seno mascellare esordito come una sinusite cronica. L'amartoma adenomatoide epiteliale respiratorio è una rara lesione benigna caratterizzata dalla proliferazione ghiandolare dell'epitelio respiratorio. La localizzazione al seno mascellare è del tutto eccezionale, ma è importante riconoscerla e differenziarla dai papillomi invertiti e dagli adenocarcinomi che necessitano di trattamenti chirurgici più aggressivi e dalle sinusiti ipertrofiche che necessitano, per contro, di un trattamento più conservativo. Vengono discussi gli aspetti clinici e anatomopatologici di questa patologia.

The term "hamartoma" was first used in 1904 as tumour-like, but primary, non-neoplastic malformations or inborn errors of tissue development¹.

Hamartomas can occur in any part of the body, for example, the surface epithelium, seromucous glands, fibrous stroma, and vessels².

They are common in the lung, kidney, liver, spleen, and intestine but are extremely rare in the upper aerodigestive tract³.

In 1995, Wenig and Heffner found a subgroup of hamartoma more often involving the nasal cavity or nasopharynx and named it respiratory epithelial adenomatoid hamartoma (REAH)⁴.

This rare benign lesion originates in the schneiderian epithelium, with glandular elements arising from this epithelium but not from the seromucous glands.

The case is described of REAH of the maxillary sinus and a review is made of reports concerning adenomatoid hamartomas in the sinuses and nasal cavity.

Case report

A 62-year-old male presented with a > 3-year history of cosmesia with posterior rhinorrhea. Symptoms were not relieved by antibiotic treatment. The patient also had a

history of neurosurgical treatment of the left trigeminal neuralgia with middle facial anaesthesia.

Chest X-ray, electrocardiogram (EKG) and routine blood tests were all within normal limits. Physical examination showed polyps occupying the left middle meatus. Computed tomography (CT) revealed complete opacification of the left maxillary sinus with bone thickening (Fig. 1). The patient underwent functional endoscopic sinus surgery under general anaesthesia to remove polyps and the lower portion of the uncinate process, followed by the creation of a middle meatus antrostomy to restore sinus ventilation and normal function.

The maxillary sinus was occupied by a purulent fluid and the mucosa was hyperaemic, hypertrophic and very bloody. On the basis of intra-operative findings biopsy was required and a histological diagnosis of REAH was made.

Given the diagnosis of the REAH of the maxillary sinus, the tumour was resected using the Caldwell-Luc procedure. The maxillary sinus was covered with a circumscribed and polypoid lesion with a smooth surface, of rubbery consistency and a red-brown appearance (Fig. 2). Histological examination revealed that the lesion was composed of well-formed, branching glands of various

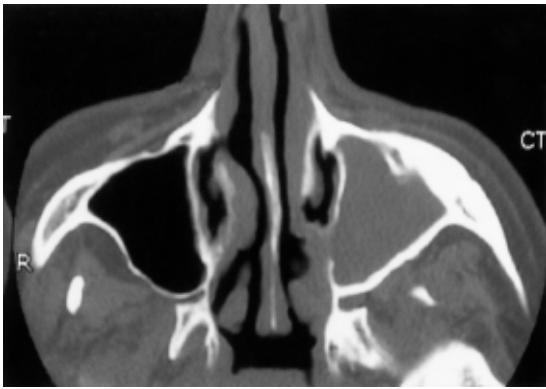


Fig. 1. Axial view of CT scan: complete opacification of left maxillary sinus.

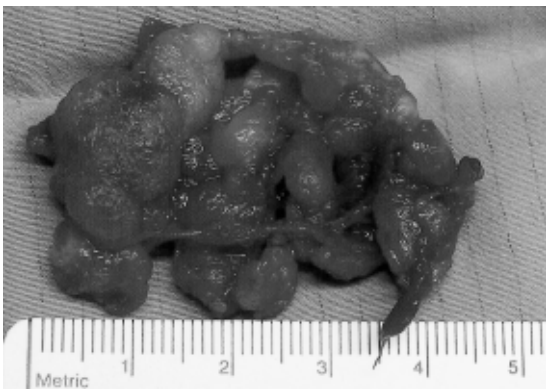


Fig. 2. Gross findings: lesion was circumscribed and polypoid, with smooth surface, rubbery consistency, red-brown appearance, measuring up to 4 cm in maximum dimension. A short stalk was also present.

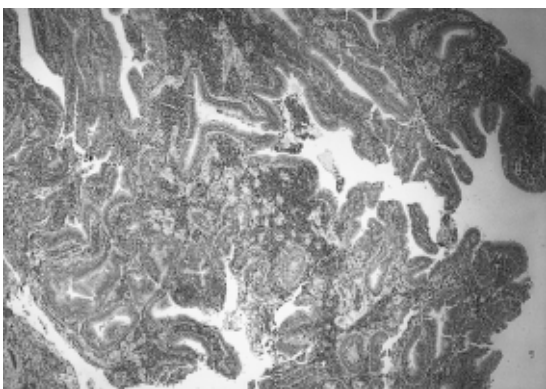


Fig. 3. Respiratory epithelial adenomatoid hamartoma (REAH): glandular proliferation is found in direct continuity with surface epithelium with invagination downward into submucosa (H&E, 2x).

sizes covered with pseudo-stratified ciliated respiratory epithelial cells separated by stromal tissue. The surface

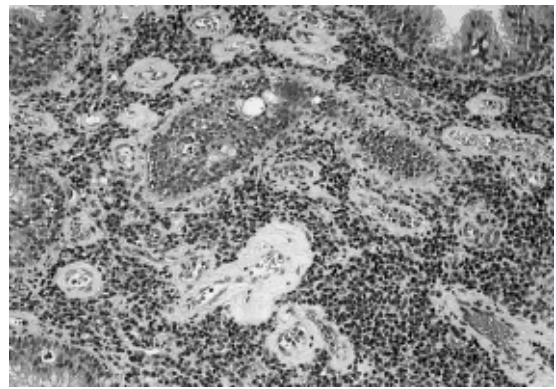


Fig. 4. Stroma contains mixed chronic inflammatory cell infiltrate. NB prominent thickened stromal hyalinization enveloping glands and blood vessels (H&E, 2x).

of the lesion was lined with ciliated respiratory epithelium in a direct continuity with some of the glands, creating a tubular appearance with elongated invaginations into the underlying loose myxoid lamina propria (Fig. 3). Cystic glands containing eosinophilic material or distended with mucus were found. A characteristic finding was stromal hyalinization with a thick eosinophilic basement membrane covering the glands. Other abnormal features included stromal oedema, nodular stromal hyperplasia, seromucinous glands proliferation, increased vascularity and a mixed acute and chronic inflammatory infiltrate (Fig. 4).

The patient is well and without recurrence of symptoms at 10-month follow-up.

Discussion

Hamartomas may be viewed as malformations composed of an excessive proliferation of one or more cellular components specific to a given tissue. They are not clearly neoplastic and are definitely not inflammatory. In contrast to neoplasms, hamartomas do not have the capability of a continuous unimpeded growth⁴. They are, therefore, self-limiting. In general, hamartomas have no malignant potential and no tendency to regress spontaneously.

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 decade²⁻¹³.

In our patient, there was no relationship to any aspecific aetiological agent and no correlation with tobacco use or alcohol abuse could be established.

Presenting symptoms are vague and non-specific, often including nasal obstruction, nasal stuffiness, epistaxis, rhinorrhea and allergy-like symptoms.

CT and magnetic resonance imaging (MRI) show no characteristic signs as they vary according to the main element of the hamartoma^{2,5-7}.

Although the mechanisms inducing a hamartoma are still unknown, it has been speculated that they arise as a result of an underlying inflammatory process⁴. To our knowledge, 39 cases of REAH have been reported in the literature^{2,4-13}. Of these cases, 25 (64%) were identified in the nasal cavity^{2,4,5,7,10} the most common site being the nasal septum, particularly the posterior aspect⁴. In the nasal cavity, the lesions were not limited to the septum, but were seen to arise along the lateral wall, middle meatus, or inferior turbinate. In addition to the nasal cavity, the other cases more often involved the nasopharynx (n = 8)^{4,5,9,13}, the ethmoid sinus (n = 2)^{4,11}, both the ethmoid sinus and frontal sinus (n = 1)⁴, both the maxillary sinus and ethmoid sinus (n = 1)¹².

The involvement of the maxillary sinus alone is quite rare with only two other cases having been reported. In one of these, the lesion was discovered during the course of a dental X-ray, as a peri-apical radiolucency mimicking a peri-apical granuloma and apical periodontal cyst with no evidence of the typical sino-nasal symptoms being apparent⁸. In the other case, only aspecific symptoms were present, like unilateral nasal obstruction, rhinorrhea and headache⁶.

If the adenomatoid proliferation is in the maxillary sinus, the REAH could most likely be confused with an inflammatory disorder and there should be no difference in the treatment between these two lesions.

The treatment for hamartoma is complete local excision. There are no reports of recurrent, persis-

tent, or progressive disease, in the literature⁶. The differential diagnosis of REAH in para-nasal sinus includes inverted papilloma, and adenocarcinoma. The inverted papilloma derives from the schneiderian membrane and is composed of papillary fronds with a delicate fibrovascular core covered by multiple layers of epithelial cells, most often of squamous or transitional cells, in contrast to the REAH, which shows adenomatoid structures lined by a single layer of ciliated columnar epithelium. Sino-nasal low grade adenocarcinoma may produce an architectural pattern mimicking that seen in REAH. However, adenocarcinoma can be distinguished from REAH by the presence of numerous uniform small glands or acini arranged in a back-to-back pattern, cellular pleomorphism, atypical mitoses, keratin pearls, loss of basement membranes and stromal invasion associated with an inflammatory-desmoplastic response¹⁴. The prevalence of occult neoplasia in routine endoscopic sinus surgery is very low¹⁵ but, in our opinion specimens should be sent for histopathologic examination when unilateral nasal polyposis, with unilateral sinus opacification, is present together with intra-operative findings.

Head and Neck surgeons should be aware of this pathological entity as a differential diagnosis for inverted papilloma and adenocarcinoma, in order to avoid unnecessary aggressive surgery. On the other hand, misinterpretation of REAH as a chronic sinus inflammation may result in inadequate treatment.

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