Case Report / Olgu Sunumu

Respiratory epithelial adenomatoid hamartoma: a rare nasopharyngeal mass lesion

Respiratuvar epitelyal adenomatoid hamartom: Nadir görülen bir nazofarengeal kitle lezyonu

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Respiratory epithelial adenomatoid hamartoma (REAH) is a rare and nonneoplastic lesion of upper respiratory tract characterized by an abnormal mixture of tissues which are peculiar to the involved anatomic region. The most common site reported is nasal cavity and its nasopharyngeal origin is extremely rare. The lesion can be confused with a variety of benign and malignant entities. In this article, we report a 22-year-old female case of REAH of posterior nasopharyngeal wall. The clinical and radiological features of the lesion are discussed in the light of literature data.

Key Words: Adenomatoid hamartoma; magnetic resonance imaging; nasopharynx.

Respiratuvar epitelyal adenomatoid hamartom (REAH), tutulan anatomik bölgedeki dokuların anormal karışımı ile karakterize, üst solunum yolunun nadir görülen neoplastik olmayan bir lezyonudur. En sık burun boşluğunda görüldüğü bildirilmekle birlikte, nazofarengeal kökeni oldukça nadirdir. Lezyon birçok benign ve malign yapılarla karıştırılabilir. Bu yazıda arka nazofarengeal duvarda REAH görülen 22 yaşında kadın bir olgu sunuldu. Lezyonun klinik ve radyolojik özellikleri literatür verileri eşliğinde tartışıldı.

Anahtar Sözcükler: Adenomatoid hamartom; manyetik rezonans görüntüleme; nazofarenks.

The term 'hamartoma' was first used in 1904. It is derived from the Greek word hamartia, which means 'error'.[1] Hamartomas are benign, non-neoplastic lesions that occur secondary to tissue-development anomalies and may comprise structures such as surface epithelium, seromucous glands, fibrous stroma, and vascular tissue.[2,3] They are most commonly seen in the lungs, kidneys, and gastrointestinal tract. Hamartomas in the head and neck region are extremely rare.[3,4]

Respiratory epithelial adenomatoid hamartoma (REAH) is a rare subtype of hamartoma of the upper respiratory tract. It

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was first described by Wenig and Heffner in 1995.[1,5] Respiratory epithelial adenomatoid hamartoma is characterized by a proliferation of glandular tissue lined by ciliated surface epithelium and glandular invagination into the stromal tissue. The epithelium shows continuity with the surface epithelium and no mitotic activity, atypia, or absence of the basement membrane. The lumen of the gland is usually filled with mucinous or amorphous debris.[1] Respiratory epithelial adenomatoid hamartomas are often localized in the nasal cavity, usually unilaterally in the posterior aspect of the nasal septum.[6] Nasopharyngeal localization has been uncommonly reported; no more than 10 cases have been reported in the English literature.

CASE REPORT

A 22-year-old female presented with long-standing snoring, open-mouth sleeping, and a foreign body sensation. She also complained of frequent nasal itching and sneezing. Her medical history included environmental allergies and chronic sinusitis. Physical examination revealed a polypoid mass covered with mucosa on the nasopharynx. Examination findings of all other systems were normal, and the laboratory work-up was unremarkable. At this stage, nasopharyngeal magnetic resonance imaging (MRI) was performed. T_1- and T_2-weighted MRI revealed a homogeneous, polypoid nasopharyngeal soft-tissue mass that was isointense to lymphoid tissue (Figure 1a, b). The lesion enhanced homogeneously after intravenous gadolinium administration (Figure 2a, b). No invasion into the adjacent neck spaces was observed. The nasal cavity and paranasal sinuses were normal at all levels. Imaging and physical examination findings were related to the primary neoplasm of the nasopharynx. Punch biopsies were taken from the nasopharyngeal lesion, and histopathological evaluation of the biopsy specimens resulted in a diagnosis of REAH. The patient subsequently underwent resection of the lesion.

DISCUSSION

Hamartomas are non-malignant cell proliferations caused by abnormal tissue development and have the features of the involved anatomic site.[2,3] They comprise mature elements and are self-limiting.[7] Hamartomas may originate from various sites, such as the lung, kidney, and gastrointestinal systems. Hamartomas of the head and neck region are extremely rare, and a limited number of cases have been reported in the literature.[2]

Respiratory epithelial adenomatoid hamartoma, a diverse and rare type of hamartoma,

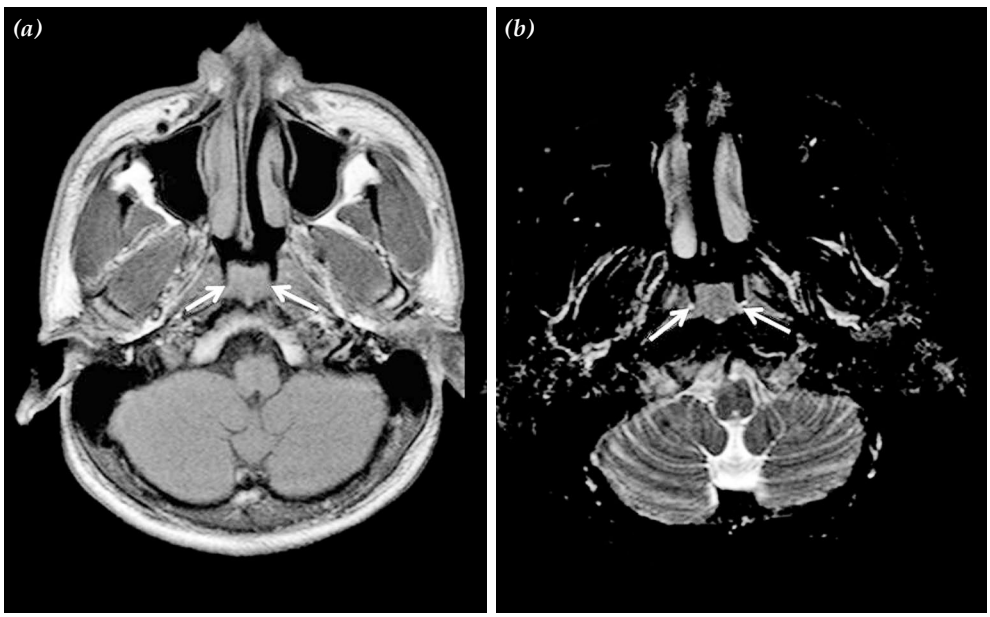

Figure 1. ***(a)*** *T_1-weighted and* ***(b)*** *T_2-weighted fat saturated magnetic resonance images in the axial plane through the nasopharynx and maxillary sinuses. A homogeneous soft-tissue mass was located on the posterior wall of the nasopharynx and was isointense to adjacent lymphoid tissue (white arrows).*

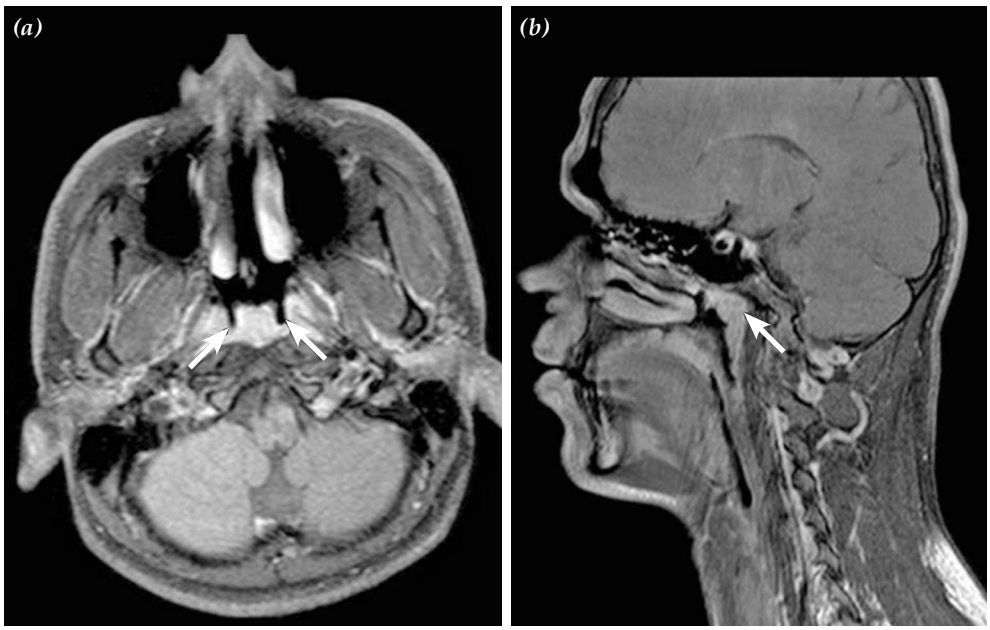

Figure 2. *Post-contrast magnetic resonance images in the* ***(a)*** *axial and* ***(b)*** *coronal planes through the nasopharynx. The mass originated from the posterior wall of the nasopharynx, homogeneously enhanced, and protruded into the nasopharyngeal space (white arrows).*

develops from the sinonasal tract.[2] Wenig and Heffner[1] first described REAH in a case series of 31 patients. They reported a male/female ratio of $^{5}/_{1}$ and showed that REAH most commonly appears in the third to ninth decades of life.[3] Although the mechanisms inducing hamartomas are unknown, cigarette smoking, asthma, nasal polyposis, chronic sinusitis, and inflammation are thought to be possible etiologic factors.[2] Our patient had an allergic background and tendency for rhinosinusitis.

Approximately 70% of reported REAH cases occurred in various parts of the nasal tract.[7] A predilection for the posterior wall of the nasal septum has been noted.[8] Nasopharyngeal localization is extremely rare, and no more than 10 cases have been reported in the English literature. In our case, the REAH originated from the posterior wall of the nasopharynx. Some previously reported REAHs have originated from ethmoid cells, frontal sinuses, olfactory recesses, and, rarely, from the maxillary sinus.[3] Clinical signs of REAH located in the sinonasal region are nasal obstruction, deviation of the nasal septum, repetitive rhinosinusitis, and epistaxis.[4]

Imaging modalities frequently reveal an exophytic soft tissue mass connected to the nasal septum, sometimes with opacification of adjacent sinuses.[1,4,9,10] Calcification may be visualized within the lesion on computed tomographic (CT) images, but bone erosion and invasion into the deep tissues are uncommon. Magnetic resonance imaging shows no characteristic sign because signs vary according to the main element of the hamartoma.[4,10,11]

The final diagnosis of REAH is made histopathologically. In the present case, the surface of the mass lesion was lined by pseudostratified and ciliated columnar epithelium and was continuous with the surface epithelium on histologic images. Atypical or metaplastic cells were not present in the specimen (Figure 3a, b).

The differential diagnosis of nasopharyngeal REAH includes benign pathologies, such as adenoid hypertrophy; antrochoanal polyp; vascular pathologies, such as hemangioma and lymphangioma; lymphoma; and the most common malignant disease of the nasopharynx, squamous cell carcinoma.[12,13] Computed tomographic and MRI findings usually do not aid the differential diagnosis of polyps and adenoid hypertrophy. Nasopharyngeal lymphoma can be diagnosed readily based on a typical radiologic appearance on CT and MRI, and vascular pathologies can be differentiated based on the presence of phleboliths and signal void areas on MRI. Furthermore,

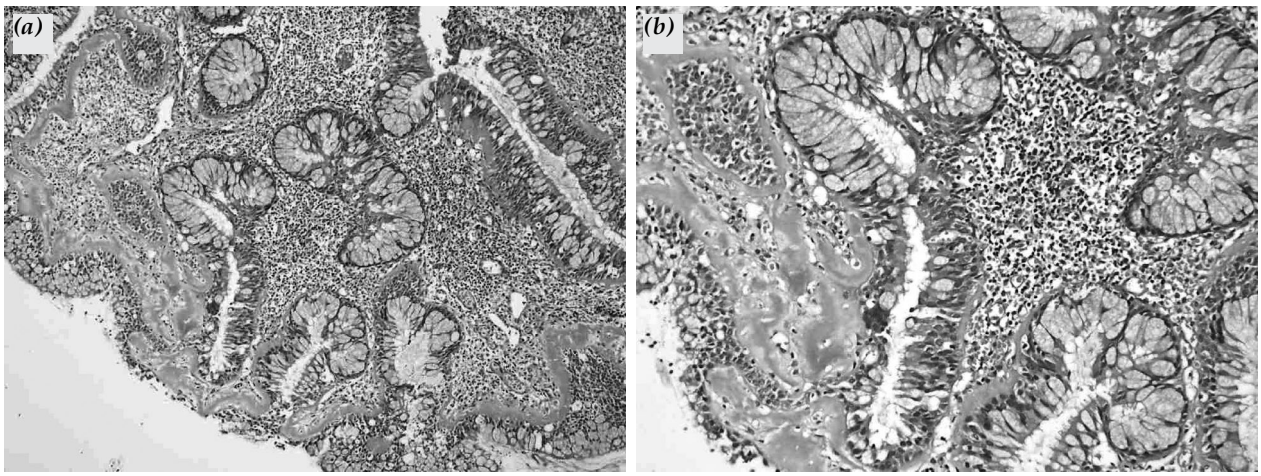

Figure 3. ***(a)*** *The lesion is dominated by glandular proliferation composed of variously sized glands separated by hyalinized stroma. Some glands are in direct continuity with the surface epithelium and invaginate downward into the submucosa. The stroma is infiltrated by lymphocytes and plasma cells (H-E x 100).* ***(b)*** *The glands are lined with multi-layered ciliated respiratory epithelium admixed with mucocytes (H-E x 200).*

nasopharyngeal carcinoma can be distinguished because of its characteristic invasion of deeper tissues. Respiratory epithelial adenomatoid hamartoma typically does not invade deeper tissues or cause bony destruction.[2] The lesion tends to grow slowly, and thus may tend to expand rather than cause destructive bony changes. One report emphasized that REAH shows olfactory cleft enlargement in comparison with normal and sinonasal polyps on CT images.[14]

The treatment for REAH is conservative surgical resection. To date, no metastatic disease or recurrence has been reported when complete resection is performed.[15] Despite the rarity of REAH, it should be considered by radiologists and surgeons in the differential diagnosis of mass lesions of the nasopharynx, especially in the absence of deeper tissue invasion, to avoid extensive surgery and treatment protocols.

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