Case Report / Olgu Sunumu

Respiratory epithelial adenomatoid hamartoma associated with inflammatory nasal polyposis: A case report

Enflamatuvar nazal polipozis ile ilişkili bir respiratuvar epitelyal adenomatoid hamartom: Olgu sunumu

Öner Sakallıoğlu, M.D.,[1] Özgen Arslan Solmaz, M.D.,[2] Sertaç Düzer, M.D.,[1]
Zeliha Kapusuz, M.D.,[3] Sultan Pehlivan, M.D.[2]

Departments of [1]Otolaryngology, [2]Pathology, Elazığ Training and Research Hospital, Elazığ, Turkey;
[3]Department of Otolaryngology, Medical Faculty of Bozok University, Yozgat, Turkey

Respiratory epithelial adenomatoid hamartoma (REAH) is an uncommon benign lesion of the nasal cavity and paranasal sinuses. The etiology is unclear, however it is considered to be secondary to chronic sinonasal inflammation. Although it is rare, REAH should be taken into consideration in differential diagnosis of the nasal lesions. Complete surgical excision of the lesion is generally enough for the cure. A detailed pathological examination is necessary to prevent unnecessary surgical interventions. In this article, we present a 60-year-old female patient with REAH in the left nasal cavity associated with inflammatory polyp in the right nasal cavity.

Key Words: Nasal polyposis; respiratory epithelial adenomatiod hamartoma.

Respiratuvar epitelyal adenomatoid hamartom (REAH), nazal kavite ve paranazal sinüslerin nadir rastlanan benign bir lezyondur. Etyolojisi bilinmemekle birlikte, kronik sinonazal enflamasyonlara sekonder olabileceği düşünülmektedir. Her ne kadar nadir görülse de REAH nazal lezyonların ayırıcı tanısında akla getirilmelidir. Lezyonun tümüyle cerrahi olarak çıkarılması genellikle tedavi için yeterli olmaktadır. Gereğinden fazla cerrahi girişimi önlemek için ayrıntılı bir patolojik inceleme yapılmalıdır. Bu yazıda, sağ nazal kavitede enflamatuvar bir polip ile ilişkili, sol nazal kavitede REAH olan 60 yaşında bir kadın hasta sunuldu.

Anahtar Sözcükler: Nazal polipozis; respiratuvar epitelyal adenomatiod hamartom.

Hamartomas are benign, non-neoplastic lesions constituted by a mixture of tissues which are indigenous to the region. They are commonly seen in the lung, kidney, liver, spleen and intestines but are very rare in the upper aerodigestive tract. Hamartomas do not clearly represent either a neoplastic or an inflammatory disorder.[1] Hamartomas of the head and neck region, in particular the nasal cavity and paranasal sinuses, are very rare.[2] Respiratory epithelial adenomatoid hamartomas (REAH) are a distinct and rare type of hamartoma occurring in the sinonasal tract and are characterized by glandular proliferation lined by ciliated epithelium originating from the airway epithelium.[3] The REAH was first described by Wenig and Heffner in 1995 on a retrospective analysis of 31 cases of hamartomas of the nasal cavity. A subgroup of hamartoma named

Received / *Geliş tarihi:* September 18, 2011 Accepted / *Kabul tarihi:* January 18, 2012

Correspondence / İletişim adresi: Öner Sakallıoğlu, M.D. Elazığ Eğitim ve Araştırma Hastanesi Kulak Burun Boğaz Hastalıkları Kliniği, 23100 Elazığ, Turkey.
Tel: +90 424 - 238 10 00 Fax *(Faks):* +90 424 - 212 14 61 e-mail *(e-posta):* onersakallioglu@yahoo.com

'respiratory epithelial adenomatoid hamartoma' is defined as tumors originated from the surface epithelium, with glandular elements arising from this epithelium and not from seromucous glands.[4] In the nasal cavity, REAH mostly involve the posterior nasal septum. The involvement of maxillary sinus, ethmoid sinus, frontal sinus and nasopharynx is extremely rare.[5]

In this article, we described a case of a REAH associated with inflammatory nasal polyposis. To our knowledge, there are only two case reports in the literature concerning an association between REAH and inflammatory nasal polyposis.

CASE REPORT

A 60-year-old woman presented with a history of nasal obstruction, decreased sense of smell, and headache. She had no history of allergic bronchial asthma or previous nasal surgery. Informed consent was obtained from the patient. On endoscopic nasal examination, a yellowish polypoid mass filled the left nasal cavity, occupying the right osteomeatal complex region. The lesion in the right nasal cavity was not seen on direct nasal examination using a nasal speculum. Computed tomography (CT) of the paranasal sinuses and nasal cavity showed a soft-tissue mass in the anterior left nasal cavity, right osteomeatal complex region and right ethmoid sinuses, with opacification of bilateral maxillary sinuses (Figure 1). The patient underwent functional endoscopic sinus surgery under general anesthesia. The lesion was found to have originated from the left lateral nasal wall behind the middle turbinate. The polypoid lesion occupied the ostium of the left maxillary sinus. The polypoid lesion was excised completely. After left uncinectomy, the ostium of the left maxillary sinus was broadened posteriorly, and the left maxillary content was discharged completely. The excised mass was elastic and harder than ordinary nasal polyp with a smooth surface. In the right nasal cavity, the polypoid tissues were excised and then right uncinectomy, bullectomy and anterior ethmoidectomy were performed. The excised tissues grossly resembled ordinary inflammatory nasal polyps. Histopathological examination of the left nasal lesion revealed that a polypoid formation with glandular proliferation was lined by ciliated respiratory epithelium. The round-oval shaped glands were of various sizes and there was a mucoid-amorphous material in the lumens of the glands. No dysplasia or atypia were observed. The mixture type inflammatory cell infiltration with edematous stroma was observed. We diagnosed the lesion in the left nasal cavity as REAH (Figure 2, 3) and diagnosed the lesion in the right nasal cavity as inflammatory nasal polyp (Figure 4). On postoperative clinical course, the symptoms of the patient were resolved.

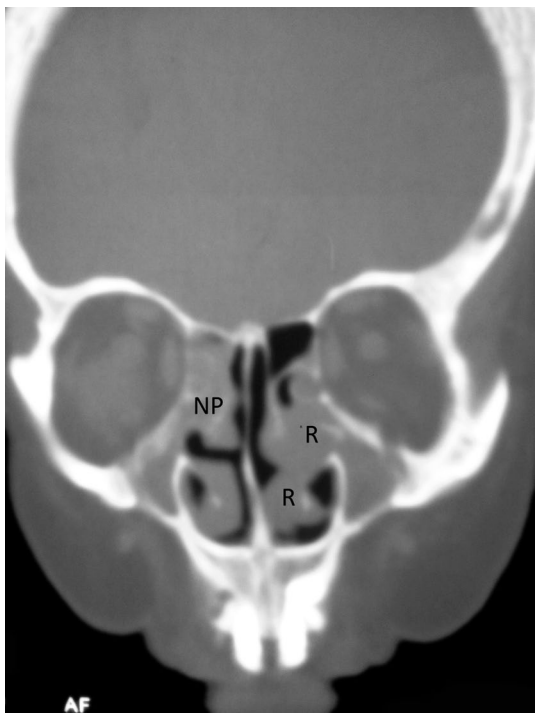

Figure 1. *Coronal view of the computed tomography scan. Respiratory epithelial adenomatoid hamartomas (R) is seen in the left nasal cavity. Inflammatory polyps (NP) is seen in the right nasal cavity. The right ethmoid sinuses and bilateral maxillary sinuses are opacified.*

DISCUSSION

Hamartomas are defined as an aberrant differentiation which may produce a mass of disorganized but mature specialized cells or tissue indigenous to the particular site.[6] They are not clearly neoplastic and definitely not inflammatory.[7] Respiratory epithelial adenomatoid hamartomas is a benign lesion predominantly affecting male adults and the male-to-female ratio is 5-7:1, with age ranging from the sixth to the ninth decade of life.[4,8,9] Hulsmann et al.[10] reported two pediatric patients with hamartoma- one newborn and another

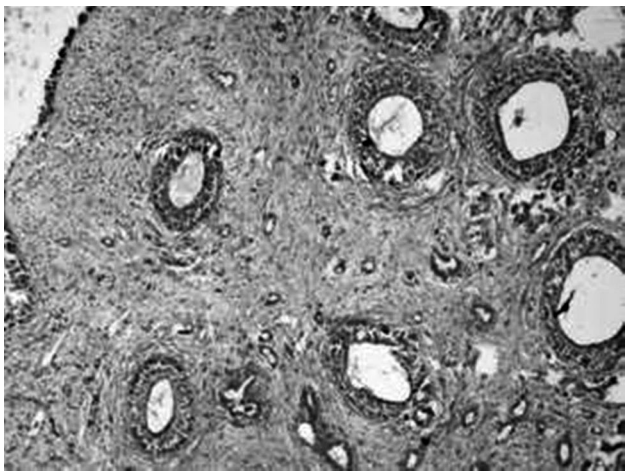

Figure 2. *Microscopic appearance of respiratory epithelial adenomatoid hamartomas (H-E x 100). Glandular proliferation and chronic inflammatory cell infiltration in the stroma.*

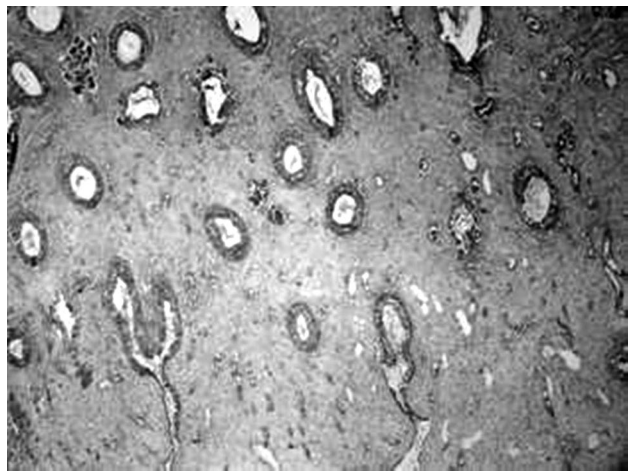

Figure 3. *Microscopic appearance of respiratory epithelial adenomatoid hamartomas (H-E x 40). The dilated glandular structures lined by ciliated respiratory epithelium and edematous stroma.*

three-month-old girl. Seol et al.[11] reported a 60-year-old woman who presented with headache, nasal congestion, an altered sense of smell, and stuffiness in both ears. Our case was a 60-year-old woman and she had headache, decreased of sense smell and nasal obstruction.

The REAH share many features with sinonasal inflammatory polyps, including clinical presentation, histopathologic changes, treatment and behavior. Hamartomas are rare in the nasal cavity, and in contrast to inflammatory sinonasal polyps, REAH mostly involve the posterior aspect of the nasal septum.[12] Besides nasal septal localization of hamartoma, it has been reported that REAH could be rarely localized in the frontal sinus, oro- and nasopharyngeal cavity and olfactory recess.[1,10,11] In our case, the REAH was found to have originated from the left lateral nasal wall behind the middle turbinate. Generally, their growth is self-limiting and most of the resultant sequelae are secondary to mass effect rather than erosion and invasion. As reported by Athre and Ducic[1] extensive progression of the disease to involve the orbit and intracranial area is very rare. Our presented case had no deformity of the surrounding paranasal sinuses.

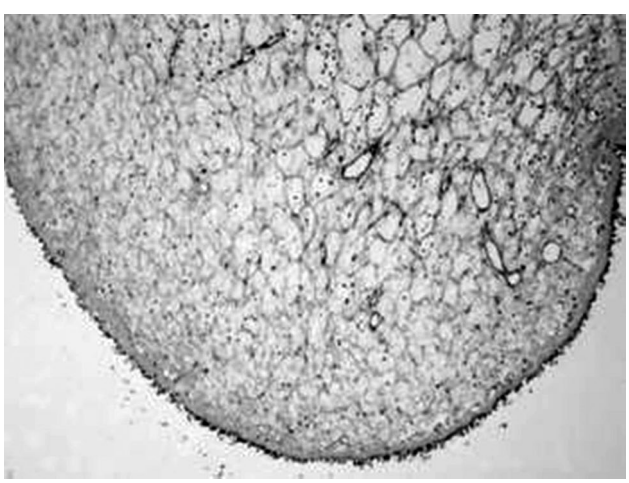

Figure 4. *Microscopic appearance of inflammatory nasal polyp (H-E x 40).*

Grossly, REAH may appear as an edematous polypoid mass resembling an inflammatory polyp. Characteristic histopathologic features include the proliferation of glands that originate from surface epithelium, are lined by ciliated respiratory epithelium, and are surrounded by fibrosis in an edematous stroma.[4] These proliferations are directly connected to the surface epithelium and are not derived from seromucous glands, as seen in the setting of reactive hyperplasia associated with inflammatory polyps. The epithelial cells show no dysplasia or atypia, and the epithelium unrelated to the adenomatoid proliferation is normal.[11] In contrast to inverted papilloma, REAH involves respiratory epithelium that forms glands in a single layer. Inverted papilloma arises from the invagination of surface epithelial cells into underlying supportive tissue and is characterized by a thickened epithelium, local invasion, and bony erosion.[13]

The treatment of REAH is complete surgical resection.[12] Therefore, it is important to distinguish REAH from more aggressive neoplastic diseases

like inverted papilloma. Otherwise, misdiagnosis of this benign lesion may lead to unnecessarily aggressive surgical procedures.[13] We excised the nasal mass completely and postoperative recovery was uneventful. Hamartomas have no capacity for continuous unimpeded growth and therefore their proliferation is self-limiting. They have no malignant potential and do not have a tendency to spontaneously regress.[4,14] No recurrence of nasal hamartoma was mentioned in the literature.[14]

It is important to differentiate REAH from other pathologic diagnosis: inflammatory polyps, inverted papillomas, and adenocarcinoma.[15] Other pathologic lesions entities commonly confused with REAH are inflammatory lesions, including nasal polyps.[12] Long-term chronic inflammation and polyposis of the respiratory epithelium may be an etiologic precursor of REAH.[1,4] Delbrouck et al.[12] and Liang et al.[15] reported REAH cases associated with inflammatory nasal polyposis and our case has features with this association. Our case is the third case of REAH to be associated with inflammatory nasal polyposis.

In conclusion, our case provides strong evidence that chronic inflammatory changes may contribute to the development of REAH. Therefore, biopsy is not only indicated in case of suspicion of malignancy but also differential diagnosis of the nasal lesions. The preoperative biopsy and endoscopic nasal examination must be performed especially in patients with unilateral nasal mass and, although rare, REAH must be taken into consideration in the differential diagnosis of nasal lesions. Either clinician or pathologist must be aware of this benign entity in order to avoid misdiagnosis. This is important to prevent unnecessarily aggressive surgical procedures.

REFERENCES

1. Athre R, Ducic Y. Frontal sinus hamartomas. Am J Otolaryngol 2005;26:419-21.
2. Endo R, Matsuda H, Takahashi M, Hara M, Inaba H, Tsukuda M. Respiratory epithelial adenomatoid hamartoma in the nasal cavity. Acta Otolaryngol 2002;122:398-400.
3. Terris MH, Billman GF, Pransky SM. Nasal hamartoma: case report and review of the literature. Int J Pediatr Otorhinolaryngol 1993;28:83-8.
4. Wenig BM, Heffner DK. Respiratory epithelial adenomatoid hamartomas of the sinonasal tract and nasopharynx: a clinicopathologic study of 31 cases. Ann Otol Rhinol Laryngol 1995;104:639-45.
5. Kessler HP, Unterman B. Respiratory epithelial adenomatoid hamartoma of the maxillary sinus presenting as a periapical radiolucency: a case report and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2004;97:607-12.
6. Kumar V, Abbas AK, Fausto N. Neoplasi. In: Kumar V, Abbas AK, Fausto N, editors. Robbins and Cotran pathologic basis of human disease. 7th ed. Philadelphia, PA: Elsevier-Saunders Publishing Company; 2005. p. 272-98.
7. Di Carlo R, Rinaldi R, Ottaviano G, Pastore A. Respiratory epithelial adenomatoid hamartoma of the maxillary sinus: case report. Acta Otorhinolaryngol Ital 2006;26:225-7.
8. Avilés Jurado FX, Guilemany Toste JM, Alobid I, Alós L, Mullol I Miret J. The importance of the differential diagnosis in rhinology: respiratory epithelial adenomatoid hamartoma of the sinonasal tract. Acta Otorrinolaringol Esp 2012;63:55-61. [Abstract]
9. Metselaar RM, Stel HV, van der Baan S. Respiratory epithelial adenomatoid hamartoma in the nasopharynx. J Laryngol Otol 2005;119:476-8.
10. Hulsmann AR, de Bont N, den Hollander JC, Borgstein JA. Hamartomas of the oro- and nasopharyngeal cavity in infancy: two cases and a short review. Eur J Pediatr 2009;168:999-1001.
11. Seol JG, Livolsi VA, O'Malley BW Jr, Chen JY, Loevner LA. Respiratory epithelial adenomatoid hamartoma of the bilateral olfactory recesses: a neoplastic mimic? AJNR Am J Neuroradiol 2010;31:277-9.
12. Delbrouck C, Fernandez Aguilar S, Choufani G, Hassid S. Respiratory epithelial adenomatoid hamartoma associated with nasal polyposis. Am J Otolaryngol 2004;25:282-4.
13. Melroy CT, Senior BA. Benign sinonasal neoplasms: a focus on inverting papilloma. Otolaryngol Clin North Am 2006;39:601-17.
14. Mortuaire G, Pasquesoone X, Leroy X, Chevalier D. Respiratory epithelial adenomatoid hamartomas of the sinonasal tract. Eur Arch Otorhinolaryngol 2007;264:451-3.
15. Liang J, O'Malley BW Jr, Feldman M, Newman JG. A case of respiratory epithelial adenomatoid hamartoma. Am J Otolaryngol 2007;28:277-9.