Case Report

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Respiratory Epithelial Adenomatoid Hamartoma at an Unusual Location: A Case Report and Literature Review

흔치 않은 위치에서 발생한 호흡상피 선종양 과오종: 증례 보고와 문헌고찰

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Respiratory epithelial adenomatoid hamartoma (REAH) in the head and neck is a rare benign lesion containing glandular tissue covered with ciliated respiratory epithelium. In the head and neck, REAH of the nasal cavity, paranasal sinuses, and nasopharynx have been reported in literature. Due to rareness of REAH and insufficient knowledge of its imaging features, the diagnosis can be challenging when we encounter a non-specific cystic mass at an uncommon site in the head or neck. Here, we report the case of a pathologically confirmed REAH showing a cystic mass centered at the buccal space (retromaxillary fat pad) with CT and MRI findings.

Index terms Hamartoma; Head and Neck Neoplasm; Benign Neoplasm; Infratemporal Fossa; Magnetic Resonance Imaging

INTRODUCTION

Respiratory epithelial adenomatoid hamartoma (REAH), a rare glandular proliferative be-

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nign neoplasm, was described for the first time in 1995 (1). It commonly arises in the nasal cavity. It has also been reported in paranasal sinuses (2), nasopharynx (3), and external auditory canal (4). REAH should be distinguished from more aggressive neoplasms because it is a benign entity that can be cured with conservative surgical resection (5). However, only a few reports of imaging findings of REAH have been published (3, 5, 6).

Here, we report a case of REAH centered at the left buccal space (retromaxillary fat pad) with CT and MRI findings.

CASE REPORT

A 22-year-old male presented with nasal obstruction and rhinorrhea with a 2-week duration. Sinuscopy demonstrated nasal septal deviation and scanty purulent discharge. The patient's symptoms were judged to be because of a deviated septum and sinusitis. Septoplasty and endoscopic sinus surgery were planned, and paranasal CT was performed for preoperative evaluation. On CT, a 3.5-cm lobulating cystic lesion was found incidentally in the left buccal space (retromaxillary fat pad) and the masticator space extending to the pterygopalatine fossa and inferior orbital fissure. The lesion was homogeneous hypoattenuation with a smooth margin. Pressure remodeling of the adjacent bony wall was seen with widening of the pterygopalatine fossa. MRI showed a well-marginated lobulating mass with a high signal intensity in the T2-weighted image and a low signal intensity in the T1-weighted image. The mass showed peripheral thin-walled enhancement in the fat-suppressed contrast-enhanced T1-weighted image. The lesion showed no diffusion restriction or internal solid component. Intracranial extension of the lesion was not evident on CT or MRI (Fig. 1).

Based on imaging findings, we considered benign tumors, such as a schwannoma or minor salivary gland tumor with cystic degeneration, as the differential diagnosis.

The lesion was completely resected endoscopically with the Caldwell-Luc (transmaxillary) approach. The soft tissue mass measured 2.8 cm × 1.6 cm × 1.5 cm in size. The mass contained hemorrhagic and focal cystic portions. The cystic mass was microscopically composed of respiratory epithelium with small to medium glandular proliferation and mature smooth muscle bundles (Fig. 1), consistent with REAH.

The patient made a full recovery from the operation without any evidence of recurrence on serial MRI or CT follow-up scans (Fig. 1) until 1.5 years postoperatively.

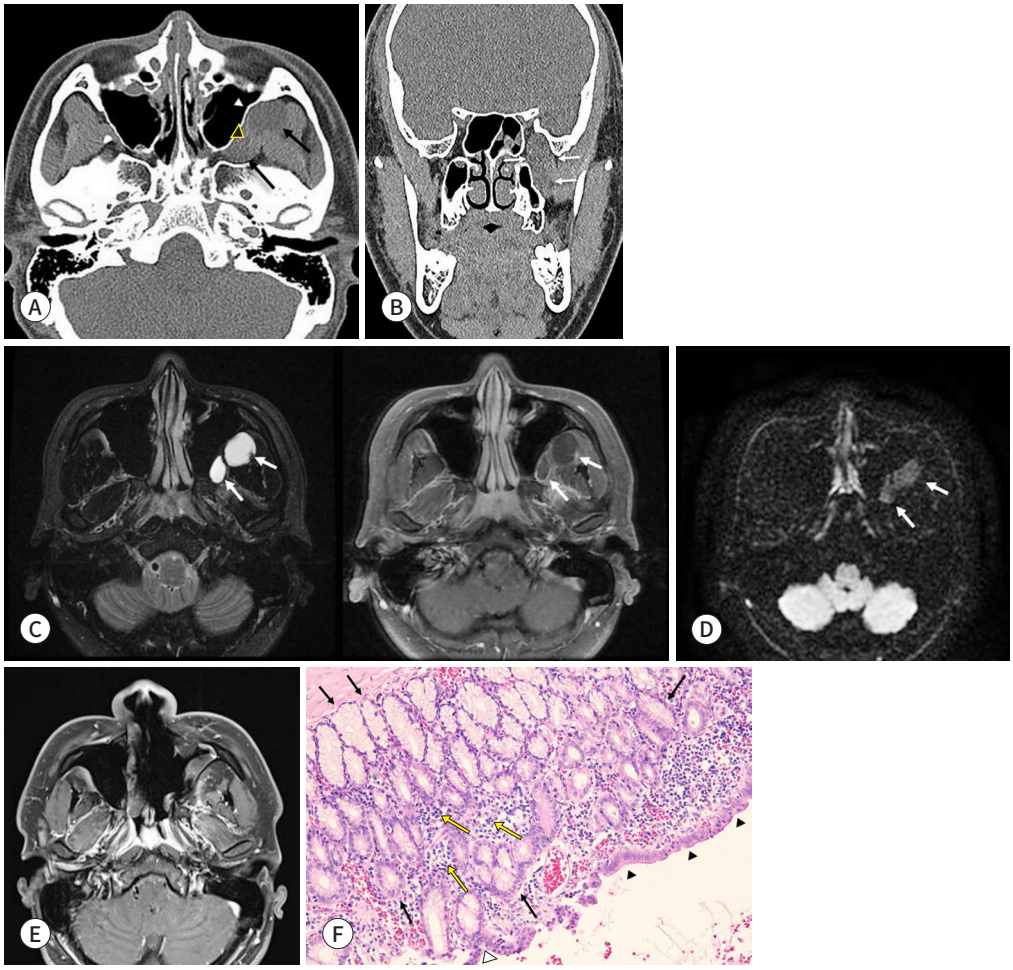
Written informed consent was obtained from the patient.

DISCUSSION

REAH, a rare lesion occurring in the upper aerodigestive tract, was first described by Wenig and Heffner in 1995. The lesion originates from the respiratory surface epithelium that forms prominent glandular proliferations lined by ciliated respiratory epithelium (1, 5). Clinically, REAH is a benign lesion that predominantly affects men. Its occurrence is associated with tobacco use, asthma, and nasal polyposis (2).

Approximately 70% of reported REAHs are located in the nasal cavity (particularly in the nasal septum). REAHs can also be located in the nasopharynx and paranasal sinuses, includ-

Fig. 1. A pathologically confirmed respiratory epithelial adenomatoid hamartoma in a 22-year-old male.
A, B. Preoperative axial (A) and coronal reformatted (B) CT scans show a well-defined lobulating cystic lesion in the left retromaxillary fat pad and pterygopalatine fossa (arrows in A, B). The adjacent bone shows remodeling without any erosive change (arrowhead in A). The left pterygopalatine fossa is widened by the cystic lesion (arrows in B).
C, D. Preoperative axial MRI scans show a well-demarcated lobulating mass in the left buccal space (retromaxillary fat pad). The mass demonstrates homogeneously high signal intensity in the fat-suppressed T2-weighted image (arrows in C, left) and subtle enhancement in the peripheral portion of the mass in the fat-suppressed contrast-enhanced T1-weighted image (arrows in C, right). The mass shows no diffusion restriction in the diffusion-weighted image (arrows in D).
E. Postoperative axial fat-suppressed contrast-enhanced T1-weighted MRI scan obtained 1.5 years postoperatively shows no evidence of tumor recurrence.
F. Photomicrograph of a REAH shows small to medium glandular proliferation (arrows) arising in direct continuity with the surface epithelium (black arrowheads) and invagination downward into the submucosa (white arrowhead). Scattered inflammatory cells (yellow arrows) are seen in the stroma (hematoxylin and eosin stain, × 200).
REAH = respiratory epithelial adenomatoid hamartoma



ing the ethmoid, frontal, and maxillary sinuses (2, 3). A case of REAH in the external auditory canal other than the sinonasal cavity has been reported (4).

The reported typical radiologic feature of REAH on CT is a hypoattenuating homogeneous polypoid soft tissue mass with possible smooth remodeling of the adjacent bones without erosive changes (3, 4). In MRI scans of REAH in previous studies (5-7), REAHs demonstrated non-

specific findings, with a soft tissue mass showing homogeneous or partial gadolinium enhancement, hypo/isointensity with the brain parenchyma in the T1-weighted image, and heterogeneous hyperintensity in the T2-weighted image. REAH rarely extends intracranially (3).

REAH that demonstrates radiologic appearance reported so far can be difficult to differentiate from sinonasal polyposis, inverted papilloma, minor salivary gland cancer, and squamous cell carcinoma, as all appear as soft tissue masses typically arising from the nasal cavity. The only described feature of REAH distinguishing it from nasal polyps is olfactory cleft widening and the predilection of REAH to form in this anatomical location (3).

A preoperative diagnosis of REAH is challenging, particularly when the lesion is in an uncommon location, as seen in the present case.

In a previous study, clinical presentations of tumors within the infratemporal fossa, including the retromaxillary buccal fat pad, were mainly facial hypoesthesia, preauricular/auricular pain, headaches, and jaw deviation. Due to its concealed location, clinical signs and symptoms might present late and progress gradually (8). Furthermore, the buccal space is often underrepresented in the literature because of its small size and prevalence of adipose tissues (9). However, due to its proximity to several important structures, such as the parotid duct, facial artery and vein, lymphatic channels, minor salivary glands, and branches of facial and mandibular nerves, the pathology varies (9). The most common type of pathology seen in the buccal space is minor salivary gland tumors, including benign pleomorphic adenoma and malignant salivary gland tumors, such as adenoid cystic carcinoma and mucoepidermoid carcinoma (9). The differential diagnosis for masses in this region also includes schwannoma, sarcoma, squamous cell carcinoma, lymphoma, lymphangioma/hemangioma (lymphatic or venous malformation), ameloblastoma, and abscess from dental infection (9, 10).

In the present case, the primary location of the lesion in the buccal space, along with its incidental discovery led us to initially consider the possibility of a minor salivary gland tumor or schwannoma with cystic degeneration.

REAH can be treated with complete local resection with little or no risk of recurrence, while malignant minor salivary gland tumors, even the localized type, require surgery with a wide safety margin (5). Therefore, having an idea regarding REAH and considering it as a differential diagnosis for lesions in head and neck spaces before surgical planning could be beneficial to prevent unnecessary aggressive surgical procedures that might require more time for recovery and possibly cause complications.

In conclusion, we presented a case of REAH with an uncommon location and radiologic features compared to previously reported cases. This case emphasizes the significance of considering REAH as a potential diagnosis in cases of a lobulating cystic mass centered at buccal space and masticator space, which can mimic other tumorous conditions with cystic degeneration.

Author Contributions

Conceptualization, K.D.E., K.D.M.; supervision, K.D.M., S.C.J., L.I.H., K.Y.M.; writing—original draft, K.D.E.; and writing—review & editing, K.D.E., K.D.M.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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흔치 않은 위치에서 발생한 호흡상피 선종양 과오종: 증례 보고와 문헌고찰

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두경부의 호흡상피 선종양 과오종은 섬모호흡상피로 둘러싸인 선조직으로 구성된 드문 양성종양이다. 두경부의 비강, 부비동 또는 비인강의 호흡상피 선종양 과오종이 현재까지 보고되었다. 호흡상피 선종양 과오종은 드물고 특징적인 영상 소견이 잘 알려져 있지 않기 때문에 흔하지 않은 위치에 비특이적인 낭성종물로 발생한 경우 영상의학적 진단을 내리기가 쉽지 않다. 저자들은 볼쪽공간(상악후방 지방층)에서 낭성종물의 형태로 나타난 병리적으로 진단된 호흡상피 선종양 과오종의 CT 및 MRI 영상 소견을 증례 보고하고자 한다.

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