A Case of Rapid Growing Myoepithelioma of the Maxillary Sinus

Jang Won Oh1, Chae Jung Park2, Yoon Jung Choi3, and Jong-Gyun Ha4

1Department of Otorhinolaryngology, National Health Insurance Service Ilsan Hospital, Ilsan; and Departments of 2Radiology and 3Pathology, Yonsei University College of Medicine, Yonin, 4Department of Otorhinolaryngology-Head and Neck Surgery, Chung-Ang University College of Medicine, Gwangmyeong Hospital, Gwangmyeong, Korea

Myoepithelioma is a rare tumor of the salivary glands. Occurrence of myoepithelioma in the nasal cavity is even rarer, and there has been no case wherein the change in the size of the tumor was confirmed by imaging. In this report, a 84-year-old female was found to have a rapidly enlarged tumor on the preoperative image, but she had shown no significant size change in a medical examination six months ago or in the paranasal sinuses view three months ago. The image showed a rapid increase in the size of the cystic component, so the endoscopic medial maxillectomy was performed. There was no recurrence of the tumor at follow-up six months after surgery. Although myoepithelioma is rarely found in the maxillary sinus of the sinonasal cavity, it is important to suspect myoepithelioma and consult a pathologist if it shows a similar pattern to the current case.

Keywords Magnetic resonance imaging; Myoepithelioma; Nasal cavity.
id growth of the cystic component of a tumor. This is the first report of a myoepithelioma in the maxillary sinus of the sinonasal cavity in Korea.

**Case**

An 84-year-old female with hypertension, stenotic coronary artery disease, right cerebral infarction visited the outpatient clinic because of left facial swelling without tenderness and left epiphora that had occurred 1 month ago. Coincidentally, she had brain MRI for health check-up six months ago, and there was an unreported 2.7 cm-sized cystic and solid mass in the left maxillary sinus abutting the medial wall of left maxillary sinus (Fig. 1A and B). This mass probably originated from the medial wall of the left maxillary sinus. After visiting the internal medicine clinic for a postnasal drip 3 months ago, there was a round mass-like haziness of left maxillary sinus on paranasal sinus (PNS) view (Fig. 1C). It grew slowly, but no rapid growth of size compared to the previous Brain MRI 3 months ago. She did not complain of facial swelling or epiphora at the time. She underwent PNS CT and neck MRI, which revealed a well-defined cystic and solid mass that is approximately 3.8 cm in size filling the left maxillary sinus. MRI showed that the mass had solid portions at the medial and lateral aspects, which showed relatively low T2 signal intensity with heterogeneous contrast enhancement (Fig. 2). The cystic portion of the mass showed internal septations with marginal contrast enhancement in a T1-weighted gadolinium-enhanced image. CT showed soft tissue density in the left nasal cavity and maxillary sinus. The center of mass has low density without enhancement by a contrast medium, but the outside is enhanced. It means that the center of mass has a cystic component and the outside has a soft tissue component (Fig. 2). The bony remodeling and erosion of the posterior wall of the left maxillary sinus was demonstrated by CT. In the retrospective view, the cystic portion of the tumor shown by the MRI increased within three months of the interval. The patient underwent septoplasty for correction of septal deviation to left side, and endoscopic medial maxillectomy with a safety margin. Endoscopic medial maxillectomy was performed including nasolacrimal duct, anterior stump of inferior turbinate, medial wall of maxillary sinus.

Histological examination showed plasmacytoid myoepithelial cells with cords and sheets growth patterns in fibromyxoid stroma without ductal differentiation, nuclear atypia or mitosis (Fig. 3A and B). Immunohistochemically, the labeling index of Ki-67 positive cells was about 7%. The cellular area near the cystic portion showed a relatively high Ki-67 labeling index (Fig. 3C). The tumor cells were partly positive for smooth muscle actin (SMA), and glial fibrillary acidic protein (GFAP) and diffusely positive for the p40, p63, and S-100 protein. (Fig. 3D). The histologic findings were consistent with myoepithelioma. The patient was followed up for six months after surgery, and there were no suspicious findings of tumor recurrence (Fig. 4).

The Yongin Severance Hospital IRB approved wavier of consent for this study (IRB No. 9-2022-0002).

**Discussion**

Myoepithelioma is a tumor that originates from myoepithelial cells. These cells are mainly seen in the intercalated ducts.
Fig. 2. Preoperative MRI and CT images. A and B: Coronal (A) and axial (B) planes of CT demonstrating heterogeneous contrast enhancement by the tumor. The bony remodeling and erosion of the posterior wall of the left maxillary sinus was demonstrated at CT. C: Axial T1-weighted image without gadolinium enhancement showing a hypointense lesion occupying the left nasal cavity. D: Axial T2-weighted image showing solid portions at the medial and lateral aspects of the lesion showing relatively low T2-signal intensity and cystic portions in the center of the lesion showing high T2 signal intensity. E and F: Axial (E) and coronal (F) T1-weighted gadolinium-enhanced image showing a low-intensity signal was observed in the center of the lesion, and contrast enhancement was observed in the outer part.
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Fig. 3. Microscopic view. A and B: Histologic findings showing cords or sheets of typical plasmacytoid myoepithelial cells among fibrous-myxoid stroma without mitosis or cytological atypia (hematoxylin and eosin staining; A, ×100; B, ×200). C and D: Tumor cells were positive staining with the cellular area around the cystic portion for Ki-67 labeling index (C), about 7%, and diffusely positive for the S-100 protein (D) (immunohistochemical studies; C, ×100; D, ×200). Arrow, plasmacytoid myoepithelial cell; these cells have bright eosinophilic with eccentric nuclei; arrowhead, hyalinized stroma.

Fig. 4. Nasal endoscopic images. A: Nasal endoscopic view before surgery. Bulging of the medial wall of the left maxillary sinus was observed. B: Intraoperative nasal endoscopic view. After left middle meatal antrostomy, cystic portion of the tumor was observed in the medial wall of maxillary sinus, the mucosal surface of lesion was smooth and the inside was filled with fluid. C: Postoperative nasal endoscopic view six months after surgery. There were no suspicious findings of tumor recurrence. Asterisk, cystic portion of the mass; MT, middle turbinate; IT, inferior turbinate.
clear cell types. Cells are classified as spindle, epithelioid, plasmacytoid, and cells with solid, reticular, or myxoid patterns of growth. The of homogenous, large plasmacytoid cells, prominent eosino-

In this case, a myoepithelioma of maxillary sinus origin was re-

port for the first time in Korea.

The WHO suggests that a diagnosis of myoepithelioma should be made if the neoplasm contains less than 5% ductal components. Myoepithelioma is composed of myoepithelial cells with solid, reticular, or myxoid patterns of growth. The cells are classified as spindle, epithelioid, plasmacytoid, and clear cell types. The growth pattern and cell type do not cor-

relate with the prognosis of the disease. In this case, sheets of homogenous, large plasmacytoid cells, prominent cosmo-

philic cytoplasm and fibromyxoid stroma was seen on hema-

toxylin and eosin (H&E) staining (Fig. 3A), so pleomorphic ade
da and myoepithelioma can be considered. However, it was closer to myoepithelioma because of the absence of ductal differen-
tiation.

As known as myoepithelial cells which exhibit a wide range of differen-
tiation, the myoepithelial tumor cells are positive for a variable epithelial and mesenchymal immunohistochemi-

cal markers. This tumor stains positive for cytokeratins (AE1/ AE3, CK14, and CK5/6), S-100 protein, p63, p40, vimentin, calponin, GFAP, SMA, and carcinoembryonic antigen. Immunohistochemically, the S-100 protein is the main marker for myoepithelioma, and a diagnosis will rarely be made if this stain is negative. In our case, tumor cells were partly positive for SMA and GFAP and diffusely positive for the p40, p63 and S-100 protein.

Ki-67 index means cell proliferative activity. So, the Ki-67 marker is useful for diagnosing malignant myoepithelioma. In this case, the Ki-67 labeling index was low, about 7%. In addition, considering the absence of abnormal mitosis, vascular invasion, and cellular polymorphism on H&E, it was diag-

nosed as benign myoepithelioma. Although it was not ma-
lignant, it showed rapid growth of the cystic component within 3 months. The reason is thought to be because most of the Ki-67 staining, which indicates cellular proliferation, is seen in the cystic component and the myoepithelial cells are dense-

ly located around it (Fig. 3C). Above-average Ki index, 7% also seems to have contributed to the faster growth than in other benign myoepitheliomas ranged from 0.9%–9.1% (mean: 5.4%; standard deviation: 3.1%). Therefore, we think that high cell proliferative activity and the high cellularity of the cystic portion as indicated by the Ki-67 staining were the reason for the rapid growth of the tumor. And it seems that the secre-
tions of these cells have accumulated inside the rapidly prolif-
erating tissue. In the future, the tumor seemed likely to pro-
gress to malignancy.

Macroscopically, myoepitheliomas are usually well-encap-
sulated tumors; therefore, complete excision is important. The treatment choice for myoepithelioma is generally complete surgical excision. In the current case, the endoscopic wide excision of the tumor was performed with clear margins. No recurrence was observed during the six-month follow-up pe-

riod without additional treatment.

Benign myoepithelioma is usually an asymptomatic mass that slowly increases in size over several months or years. It is known that malignant myoepithelioma increases in size, but there has been no confirmation of the growth rate from imaging evidence in papers reporting benign myoepithelioma. In the current case, cystic components and solid compo-

nents were mixed.

The maxillary sinus tumor in MRI and CT six months be-

fore the visit was not growing significantly in PNS view three months later. In the next three months, the cystic component expanded rapidly, showing bone remodeling and facial swell-
cing caused by the mass effect. Therefore, most myoepithelio-

ma is slow growing, but in the case of myoepithelioma, where cystic components are mixed, the possibility of rapid growth should be kept in mind.

In other words, if a clinician or pathologist does not con-

sider the possibility of myoepithelioma, an appropriate imu-

munohistochemical test may not be performed, and diagno-

sis may be difficult. Therefore, although the occurrence of myoepithelioma in the sinonasal cavity is rare, it is impor-
tant to suspect myoepithelioma and consult a pathologist if it shows a similar pattern to the current case.

Acknowledgments

This study was supported by a faculty research grant of Yonsei University College of Medicine for 6-2020-0118.

Author Contribution

Conceptualization: Jong-Gyun Ha. Data curation: Jang Won Oh. Writing—original draft: Jang Won Oh. Writing—review & editing: Chae Jung Park, Yoon Jung Choi, Jong-Gyun Ha.

ORCID

Jong-Gyun Ha https://orcid.org/0000-0002-2712-1297
REFERENCES