

Carcinoma ex Pleomorphic Adenoma of the Maxillary Sinus

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상악동에 발생한 다형선종유래암종 1예

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다형선종유래암종(Carcinoma ex pleomorphic adenoma)은 타액선 종양 중 드문 종양으로 대부분 이 하선에 발생하며 비강 또는 부비동 내에 발생하는 경우는 드물다. 저자들은 좌측 상악동 내 종물이 있는 81세 여자환자에서 악성 종양 의심 하에 좌측 상악전적출술 및 견갑설골상부 경부절제술을 시행하였다. 수술 후 시행한 병리조직 검사 상 다형선종의 양성 병변이 악성종양으로 변화된 양상을 보여 다형선종유래암종으로 확진하였다. 수술 후 방사선 치료는 시행하지 않았으며 12개월간 재발 소견 없이 외래 추적 관찰 중이다. 상악동 내에 발생하는 다형선종유래암종은 매우 드문 경우로 문헌고찰과 함께 보고하는 바이다.

중심 단어 : 다형선종유래암종 · 상악동.

Introduction

Malignant mixed tumors account for 5% to 12% of all salivary gland malignancies¹⁾ and most commonly involve the parotid gland (73%). They are not usually found in the nose, paranasal sinuses or nasopharynx²⁾. Malignant mixed tumors include three categories and carcinoma ex pleomorphic adenoma accounts for the vast majority of these tumors. Recently we treated a patient with a car-

cinoma ex pleomorphic adenoma that originated in the maxillary sinus. Here we report this unusual case and review the relevant literature.

Case Report

An 81 year-old female patient presented to the hospital complaining of a foul odor in the mouth for 1 month. Physical examination revealed about a 1×1cm perforation of the hard palate and purulent drainage from the perforation site (Fig. 1). Enlarged cervical lymph nodes were not noted and the remaining physical examination was unremarkable. The patient had a history of an excisional biopsy of a hard palate mass, which confirmed a pleomorphic adenoma 10 years previously. The patient

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reported that the perforation of the hard palate developed after the surgery. A punch biopsy at the margin of the hard palate perforation was performed at the outpatient clinic. The histopathological examination revealed only a few inflammatory cells. An inhomogenous expansile mass in the left maxillary sinus was identified by computed tomography scanning (Fig. 2A, B). The mass destroyed the medial, inferior wall of the maxillary sinus and hard palate. Well enhancing lymph nodes were noted at level II of the left neck suggesting metastasis of the tumor (Fig. 2C).

Under general anesthesia, a frozen biopsy was performed around the perforation margin with punch forceps.

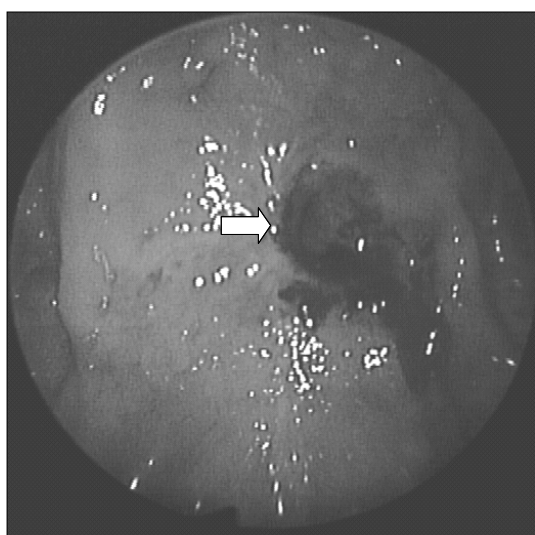


Fig. 1. Image of the palate. Approximately 1 × 1 cm sized perforation is noted of the hard palate.

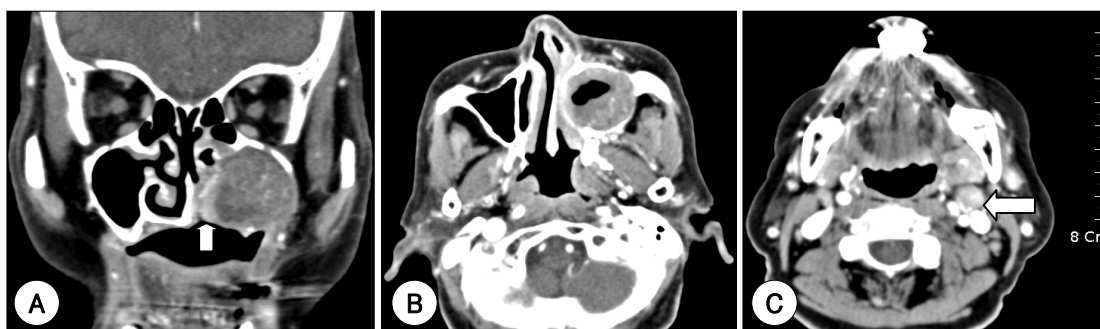


Fig. 2. Radiology findings. A : Neck CT (coronal view). B : Neck CT (axial view) shows expansile mass in the left maxillary sinus. The mass has destroyed the hard palate partially (arrow), however, the posterior wall of the maxillary sinus is relatively intact. C : Neck CT (axial view) shows about a 1.3cm well enhancing lymph node on the left at level II.

The results of the frozen biopsy were reported as a squamous cell carcinoma. A total maxillectomy and supraomohyoid neck dissection of the left side were performed. The tumor was confined within the maxillary sinus. A partial bone defect was noted on the superior wall (infra-orbital wall). However, the tumor did not extend through the defect, because the tumor was well encapsulated. The other wall of the maxillary sinus was relatively intact (Fig. 3A-C). On microscopic examination, features of a pleomorphic adenoma were focally seen in the tumor, while a malignant area showed features of epithelial or myoepithelial carcinoma with squamous differentiation (Fig. 4A, B); the findings were consistent with the diagnosis of a carcinoma ex pleomorphic adenoma. There was no tumor metastasis into the adjacent cervical lymph nodes. We diagnosed the patient as stage II (T2N0M0). After surgery, no postoperative radiotherapy or chemotherapy was performed. At twelve months of follow up the patient has no sign or symptoms of recurrence or metastasis.

Discussion

Carcinoma of the nose and paranasal sinuses accounts for only 3% of head and neck neoplasms. Squamous cell carcinomas constitute the majority of malignant tumors of the nasal and paranasal sinuses, while minor salivary gland tumors and other glandular neoplasms account for 4% to 8% of malignant neoplasm at this site¹⁾. The case presented here, with a carcinoma ex pleomorphic adenoma developing in the maxillary sinus is extremely rare.

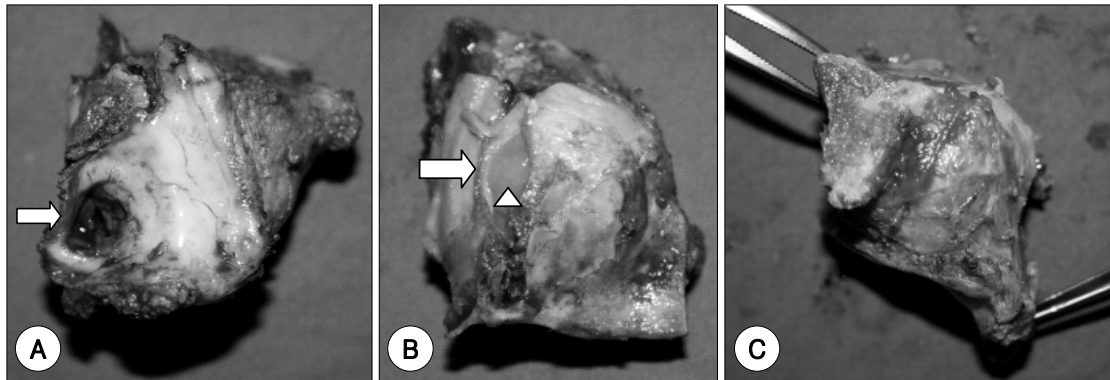


Fig. 3. Gross findings. A : Inferior view. Perforation of the hard palate is noted (arrow). B : Superior view. Infraorbital wall is relatively intact. However partial bone loss is noted (arrow). Yellowish expansile cystic mass is located in the maxillary sinus (arrow head). C : Posterior view. Posterior wall of the maxillary sinus is intact.

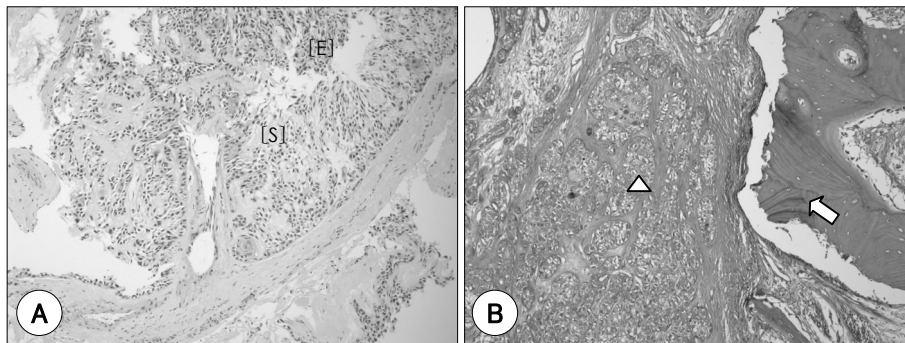


Fig. 4. Pathology findings. A : The features of the pleomorphic adenoma are noted. They show a biphasic appearance composed of an epithelial component [E] and a myxoid stromal component [S] (H&E stain, $\times 100$). B : Tumor nests (arrow head) invading an adjacent bony structure (arrow) is noted (H&E stain, $\times 100$).

Only four cases of carcinoma ex pleomorphic adenoma of the nasal cavity have been reported ; two of these cases developed from the nasal septum and one from the lateral nasal wall²⁾⁴⁾⁵⁾. There has been only one Korean case reported that developed in the maxillary sinus⁶⁾.

Malignant mixed tumors are grouped into three categories, carcinoma ex pleomorphic adenoma, carcinosarcoma, and metastasizing mixed tumors. Carcinoma ex pleomorphic adenoma accounts for the vast majority of these tumors. The diagnosis of a carcinoma ex pleomorphic adenoma requires that (1) at least a focus of benign mixed tumor be identified ; or (2) a prior surgically removed benign mixed tumor was excised from the site where the recurrent tumor is found to be carcinomatous⁷⁾. On gross examination, the tumors appear to be well capsulated, but in some areas the capsule infiltration or disruption of the lesion is usually found. In our case, the

tumor was well capsulated and confined within the maxillary sinus. A partial bone defect was noted on the superior and inferior wall. However, the tumor did not extend through the defect, and the other walls of the maxillary sinus were relatively intact.

Microscopically, foci of benign mixed tumors are associated with characteristic carcinomatous elements that are usually undifferentiated carcinoma or adenocarcinoma, not otherwise specified. Occasionally, other carcinoma types, including mucoepidermoid carcinoma, adenoid cystic carcinoma, polymorphous low grade, epidermoid, clear cell, and acinic cell, tumors occur. In some malignant mixed tumors, most of the tissue lesion has features of a benign mixed tumor and only in a limited number of foci with the carcinoma. In contrast to the presentation of the case reported here, there are some malignant mixed tumors where almost the entire tumor bulk is composed

of a carcinoma and only very limited region shows the presence of a benign mixed tumor. Such tumors are often classified as an adenocarcinoma or mucoepidermoid carcinoma without proper attention to the benign mixed tumor component³). In our case, although the initial frozen biopsy of the tumor suggested a squamous cell carcinoma, it was eventually diagnosed as a carcinoma ex pleomorphic adenoma ; components of a pleomorphic adenoma were observed on the final pathology. Therefore, fine needle aspiration biopsy or punch biopsy would not be sufficient to identify a carcinoma ex pleomorphic adenoma. For such cases the entire mass must be evaluated for the correct diagnosis.

The prognostic parameters include : recurrence, capsular invasion, and metastasis. Malignant mixed tumors metastasize regionally and to distant areas, such as the lungs, hilar and cervical lymph nodes, bone, and central nervous system⁸). The presence of cervical lymph node involvement at the time of diagnosis is an important prognostic predictor of outcome⁹).

Treatment of a carcinoma ex pleomorphic adenoma must be individualized on the basis of the tumor location, involvement of adjacent structures, and histological subtype. Radical tumor resection combined with adjuvant radiotherapy is the treatment of choice. Olsen *et al.* reported that postoperative radiation significantly reduced the frequency of local recurrence¹⁰). In our case, we performed only surgery, due to advanced age, the tumor was limited to the maxillary sinus, and no metastases were detected. No sign of local recurrence has been detected during the twelve months of follow up after surgery. The patient will continue to be monitored closely.

Conclusion

This case of a carcinoma ex pleomorphic adenoma developing in the maxillary sinus is extremely rare. When recurrence and distant metastases occur, the prognosis is poor and survival is limited. Therefore, early and ade-

quate removal of tumor is extremely important. Further study of the natural history, prognosis and treatment of malignant mixed tumors of the nose and paranasal sinus is needed.

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