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A massive adenoid cystic carcinoma of nasal septum progressed into the skull base

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ABSTRACT

Background: Adenoid cystic carcinoma (ACC) is a malignant tumor commonly occurring in the major salivary glands. ACC of the nasal septum is exceeding rare.
Methods: The case of a 42-year-old woman with ACC of the nasal septum is presented. Her chief complaint was nasal obstruction and dysosmia for two month. CT and MRI demonstrated a massive mass occupying the nasal septum infiltrating the palate, vomeronasal, anterior skull base, and dura mater. Combined anterior cranial surgery, endoscopic intranasal surgery, and transpalatal surgery were selected due to the size and location of the tumor. A negative surgical margin was achieved without cosmetic deformity or functional disorder.
Results: She had postoperative radiotherapy with no recurrence or distant metastasis during the follow-up period.
Conclusion: The tumor location and the perineural spread pattern should be considered to determine the treatment plan for septum ACC. Post operative radiation is now generally recommended. ACC has a high incidence of local recurrence and distant metastasis rate; therefore, long-term follow-up is necessary.

Cancers of the nasal cavity are uncommon; in particular, those arising in the nasal septum are very rare. Squamous cell carcinoma and adenocarcinoma are the common histologic type, although adenoid cystic carcinoma (ACC) of the nasal septum is exceeding rare. In the English literature to date, only a few cases have been reported. In this report, we describe a case of massive adenoid cystic carcinoma arising from the nasal septum, which progressed into the skull base and palate.

1. Case report

A 42-year-old woman was referred to the Department of Otolaryngology, Faculty of Medicine, Kagawa University in January 2011 because of intractable nasal obstruction and dysosmia lasting for two months. The referring otolaryngologist suggested allergic rhinitis. Her symptoms were not improved after one-month treatment with a nasal steroid spray, anti-thromboxane, and anti-histamine drugs. An intravenous olfaction test (prosultiamine injection test) showed attenuation of smell (latent and duration times were 10 and 25 s, respectively), and recognition and detection thresholds by T&T olfactometry were 4.4 and 5.8, respectively. Anterior rhinoscopy revealed pale swelling of the bilateral septal mucosa and inferior turbinate, and a well-defined smooth mass was observed in the posterior nasal septum. CT scan showed a mass occupying the nasal septum, and bone erosion of the palate, vomeronasal and anterior skull base. MRI showed tumor invasion into the cranial dura mater (Fig. 1). A biopsy was performed of submucosal tissue of the right side of the nasal septum, and the pathological report revealed adenoid cystic carcinoma showing a cribriform growth pattern. In addition, invasion of the surrounding nerve and vessels was observed in the same specimen. Positron emission tomography and systemic CT scan revealed no cervical lymph node or distant metastasis.

A combined otolaryngological and neurosurgical procedure was undertaken to excise the nasal septum tumor under general anesthesia. Bilateral maxillary artery embolization had been performed the previous day. First, anterior cranial surgery via a coronal incision, including the olfactory nerve, dura mater, and anterior cranial fossa, was performed. The dura was reconstructed immediately. Endoscopic nasal surgery was then performed via an upper labial incision. Bilateral anterior and posterior ethmoid sinuses and sphenoid sinuses were opened, the anterior end of the septum was cut, and then the posterior end and vomeronasal bone were drilled. A hard palatal mucosal flap was made through a vertical midline incision, the palatal bone was cut, and the tumor was removed *en bloc* with the cranial fossa and palatal bone by a transoral approach. The septum bone tip, nasal bone, hard palatal mucosa, and upper alveolar bone were spared. The anterior skull

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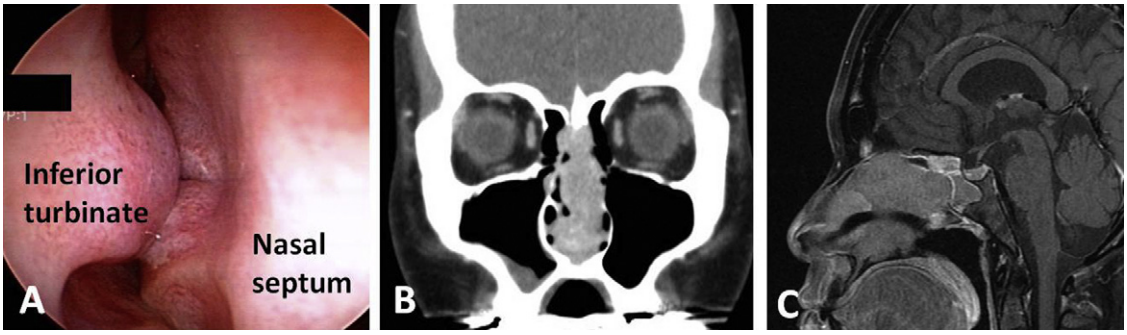

Fig. 1. (A) Right nasal cavity: Well-defined, smooth mass is observed in the posterior nasal septum. (B) Coronal enhanced CT demonstrated a septal mass with septum and skull base bone destruction. (C) Sagittal T1-weighted enhanced MRI demonstrated a septal mass infiltrating the dura matter and extending to the sphenoid sinus.

base defect, which reached to just anterior to the sphenoid sinus, including the bilateral cribriform palate, was covered by a galeal pericranial flap. Abdominal fat tissue was packed into the intracranial and extracranial areas, and then the fibrin combination (TachoComb; CSL Behring Japan, Tokyo) was placed under the fat tissue. The tumor measured 4.5 cm × 4.5 cm × 1.7 cm. Histology showed the typical cribriform pattern of ACC (Fig. 2). The tumor was relatively well circumscribed beneath the respiratory epithelium. Most of the units showed a cribriform pattern; however, several solid nests of tumor cells could also be identified (approximately 10%). A negative surgical margin was confirmed. No infiltrative growth was observed in the skull base bone. Postoperative radiotherapy with a total dose of 50 Gy was administered to enforce locoregional control. Postoperatively, the patient's course was uneventful, and she had no facial cosmetic deformity or no clear functional disorder. Local recurrence did not been observed by follow-up MRI (Fig. 3). There have been no signs of local recurrence or distant metastasis during 9-month follow-up.

2. Discussion

ACC is one of the most common malignant tumors of salivary tissue. The most affected patients are women in their fifth and eighth decades [1], and it commonly occurs in the major salivary glands, palate, and paranasal sinuses [1,2]. ACC arising in the nasal septum is exceeding rare. Young reported 43 malignant tumors of the nasal septum, but only one case of ACC was included [3]. Another clinical report showed similar results that ACC was only one of 85 cases [4], and only a few cases have been reported in the English literature [5–8] (Table 1). ACC typically grows slowly, and the predilection to spread hematogenously and perineurally but not lymphatically has been noted [9].

Its symptoms are usually nonspecific, but nasal bleeding or obstruction is a common symptom [5–8]. Although advanced ACC arising from the paranasal or nasal cavity is believed to be common at initial evaluation [10], early detection of nasal septum ACC may be possible because such symptoms may appear early in the disease. On the other hand, care should be taken in diagnosis because the nasal appearance may lead to an incorrect diagnosis of high septal deviation [8]. Our case presented with dysosmia with nasal obstruction, suggesting tumor invasion of the olfactory fibers or nerve. Dysosmia is one of the possible symptoms of advanced septal ACC depending on its perineural progression pattern.

Complete excision is selected as the primary treatment for the majority of ACC arising in the nasal cavity and paranasal sinuses. Wiseman et al. described that primary treatment patients with negative margins had a lower local recurrence rate than those with a positive margin. On the other hand, they discussed that clear margins might not correlate with better survival due to metastasis [10]. These suggestions indicate that aggressive and extensive surgery should be performed to achieve complete resection, but surgery which markedly reduces the quality of life is undesirable. Lateral rhinotomy is usually selected for anterior septal tumors, and sublabial incision plus the Denker approach is selected for posterior septal tumors. If the tumor is invading the palate or lateral side, further Weber–Ferguson incision may be needed [11]. In our case, the tumor invaded most of the septum, anterior skull base and palate. The patient was a young woman; therefore, both she and we hoped to avoid facial incisions such as lateral rhinotomy or Weber-Ferguson incision. Complete resection and *en bloc* removal were a possible combination with the transoral approach alternative to the transcutaneous approach. In addition, the patient required no oral prostheses, which depended on the preservation of the palatal mucosa and tooth. No cosmetic deformity (e.g. saddle nose) or functional disorder (e.g. abnormal occlusion, dysarthria) occurred; therefore, good patient satisfaction was obtained. We think that our surgical approach for a primary tumor was appropriate.

Histologically, ACC is divided into three groups: solid, cribriform, and tubular growth patterns. The cribriform pattern is the most common histologic subtype and all septal ACC previously reported was the cribriform pattern [5,7,8]. Perzin et al. reported

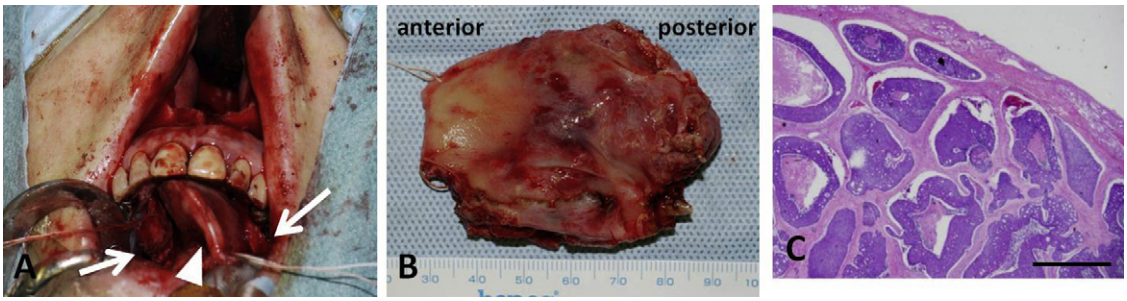

Fig. 2. (A) Intraoperative view: Tumor was excised *en bloc* and removed transorally. Arrows show the palatal mucosal flap. Arrowhead shows the nasal septum within the tumor. (B) The tumor was 4.5 cm × 4.5 cm × 1.7 cm. (C) Histological section: cytologically bland tumor cells showed a typical cribriform growth pattern. Scale bar = 1 mm.