

Early Stage Adenoid Cystic Carcinoma of the Nasal Septum

Oh Jin Kwon, MD¹, Seung Hoon Woo, MD¹, Jin Pyeong Kim, MD¹ and Jung Je Park, MD^{1,2}

¹Department of Otorhinolaryngology, ²Institute of Health Sciences, Gyeongsang National University, Jinju, Korea

– ABSTRACT –

We report an extremely rare case of early stage adenoid cystic carcinoma (ACC) of nasal septum and its recurrence after surgery alone. A case report and a review of world literature concerning early stage ACC of nasal septum and treatment are presented. As shown in the case report, a wide excision with negative surgical margin was performed but the tumor recurred. Thus for early stage ACC in the nasal septum, surgery with radiotherapy, and other combined modality treatments should be considered and long-term follow-up was required. (J Clinical Otolaryngol 2013;24:251-254)

KEY WORDS : Adenoid cystic carcinoma · Nasal septum · Surgery · Radiotherapy.

Introduction

Knowledge of the natural history of adenoid cystic carcinoma (ACC) of the nasal cavity is limited because the disease is uncommon. The nasal cavity symptoms are generally non-specific with nasal obstruction, epistaxis, discharge, facial swelling, and pain. ACC is most commonly described as arising from the seromucinous salivary tissue in the major and minor salivary glands. It is rare for this tumor to arise within the nasal cavity where the lateral nasal wall is the most common site. In the literature to date, only a few cases of early stage ACC arising from the nasal septum has been reported.¹⁻³⁾

Case Report

A 42-year-old man was presented with the unilateral recurrent epistaxis and nasal obstruction. Exami-

nation revealed a polypoid tumor measuring 1.5 × 1 cm arising from the left nasal septum (Fig. 1). The remaining routine examinations were unremarkable. A CT scan demonstrated a polypoid mass based in the nasal septum (Fig. 2A), no cervical lymphadenopathy, and no contact with sinus or turbinate. Biopsy from the lesion was performed under endoscope for pathological diagnosis. The pathology report revealed ACC. Further investigations, including MRI, did not show any cranial nerve or brain involvement (Fig. 2B). There was no evidence of distant metastasis by imaging study. Operation was performed under general anesthesia, the tumor was removed by through and through septum total excision without preserving opposite mucoprechondrium and nasal mucosa. The margins were controlled by frozen sections. Histopathology showed the typical appearances of an ACC with an infiltrating, unencapsulated lesion composed of an island of cytological bland cells with hyperchromatic nuclei arranged around cystic space containing a mucoid substance (Fig. 3). Perineural invasion was not identified. The patient was discharged from the hospital on the sixth postoperative day after an uneventful recovery. During the 4th year follow up observation after surgery, a mass lesion was detected in the left inferior turbinate,

논문접수일 : 2013년 7월 31일
 논문수정일 : 2013년 9월 16일
 심사완료일 : 2013년 10월 28일
 교신저자 : 박정제, 660-702 경남 진주시 칠암동 90
 경상대학교 의과대학 이비인후과학교실
 전화 : (055) 750-8178 · 전송 : (055) 759-0613
 E-mail : capetown@hanmail.net

and thus under local anesthesia, histological test by nasal endoscopy was performed (Fig. 4). The result of histological test showed the recurrence ACC and radi-

cal excision with radiation therapy were recommended. However, the patient refused more surgical treatment and radiotherapy only is planned.

Discussion

ACC of sinonasal cavity is a slowly progressive neoplasm with a high incidence of both local recurrence and distant metastasis, regardless of treatment modality. The lung is the most common site of metastasis and the less common sites include the bone, liver, brain, and kidney. Cervical lymph node metastasis of ACC is rarely seen.

Recurrence and metastasis can occur over a period of decades after treatment of the primary tumor which



Fig. 1. Nasal endoscopic finding. The 1.5 × 1 cm sized polypoid tumor (arrow) arising from the left nasal septum showed. IT : inferior turbinate.

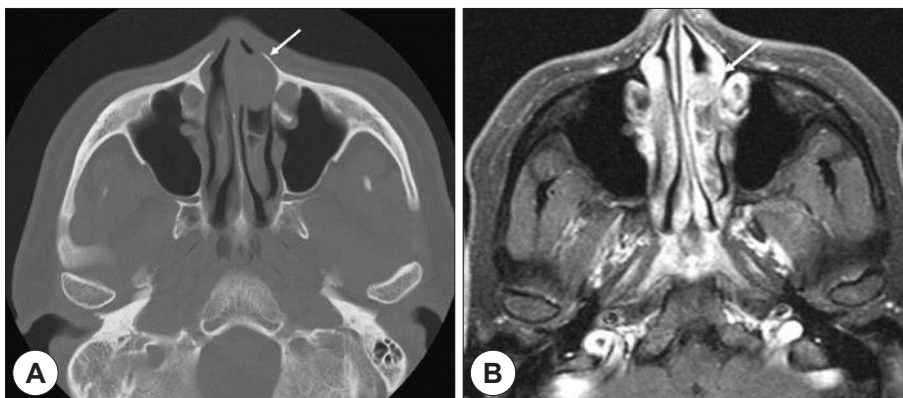


Fig. 2. A : Axial CT image (bone window) shows a well-defined, smooth mass arising from septum (arrow). B : Coronal gadolinium enhanced T1-weight spin echo image. An enhancing soft tissue mass is seen in the left nasal septum (arrow).

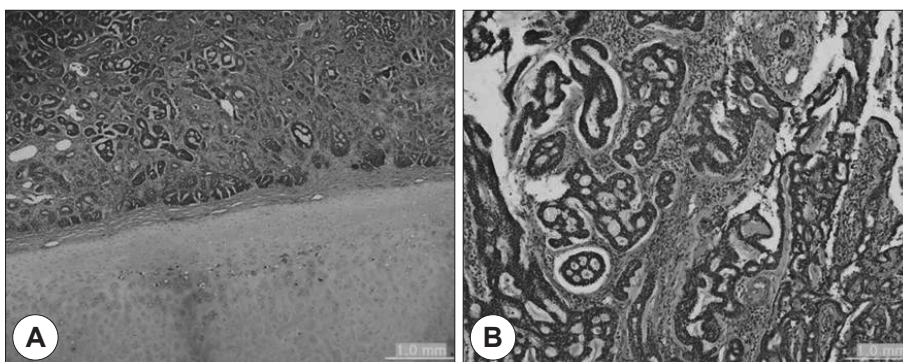


Fig. 3. A : Photomicrograph of the excised specimen shows the tumor located adjacent to the cartilage of the nasal septum (H & E stain × 40). B : Tumor cells are arranged in a cribriform pattern with microcystic space filled with hyaline or basophilic mucoid material (H & E stain × 100).

behavior can be partially explained by its tendency to extend submucosally and perineurally along major and minor nerves. The complexity of the local anatomy can make it difficult to treat. The proximity of the tumors to the skull base and the major nerves, combined with the tumor's tendency to spread perineurally contributes to the difficulty to obtain surgical margins of the disease, and may result in a high morbidity. Therefore, accurate pre-operative imaging is required to select patients for surgery and to determine surgical borders. An MRI is a sensitive method to detect perineural tumor spread. The accuracy of perineural spread on MRI was slightly superior to that on CT. The sensitivity, specificity and accuracy for the detection of perineural spread were 73%, 100% and 77% on MRI.⁴⁾

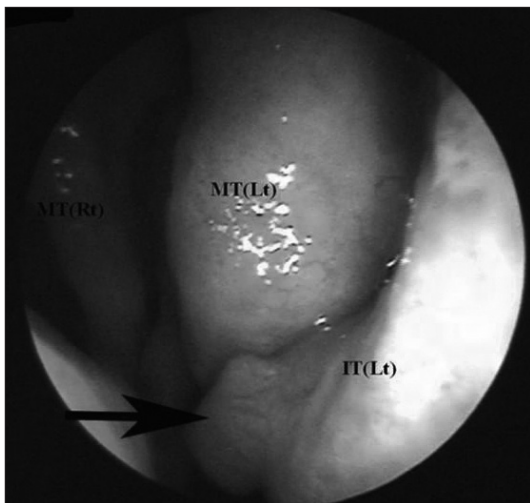


Fig. 4. Left nasal cavity after nasal septum excision. The recurrent tumor (arrow) is located in the inferior turbinate. MT : middle turbinate, IT : inferior turbinate.

Because these tumors are rare and they often present at an advanced stage, some controversy to the most appropriate treatment in early stage ACC exist.

We performed a MEDLINE review of the English language literature of all reported cases of ACC of the nasal septum. Four cases were found to have histologically confirmed ACC originating from the nasal septum. The tumors were staged as T1 in three cases and T3 in one case. One case of T-Stage III was not considered in this report due to the advanced stage.⁵⁾ The details of these three cases together with the details of our case reported here are presented in Table 1. Of the 4 patients, three were male and one case was female with the mean age 57 years (age range, 42–66 years). In all the cases, nasal obstruction was presented as an early symptom, and three cases were accompanied by frequent nasal hemorrhage. Treatment was performed in two cases by wide excision only and the other two cases by wide excision with radiation therapy. The follow-up after the end of treatment was between 9 and 48 months. Disease recurrences were noted in our case, whereas one case never presented a local, regional, or distant recurrence. This case had recurrence of the ACC in the inferior turbinate at the 48 months follow-up.

Contrary to other studies, surgery with radiotherapy did not have an effect on overall survival, rate recurrence, or time to recurrence when compared to patients treated with surgery alone.⁶⁻⁹⁾ It would be make no difference because of advanced stage, positive surgical margins. However, Observations suggest that for cases showing early stage ACC in the nasal septum, surgery only should be avoided and instead more aggres-

Table 1. Reported cases of adenoid cystic carcinoma of the nasal septum

Authors	Year	Sex/Age (years)	Stage	Symptoms	Treatment	Follow-up	Recurrence
Schneiderman TA, et al. ¹⁾	2002	M/66	T1	Nasal obstruction	Excision	9 month	No
Sivaji N, et al. ²⁾	2003	F/64	T1	Nasal obstruction, Frequent epistaxis	Excision and post op RTx	Unknown	Unknown
This study	2004	M/42	T1	Nasal obstruction, Frequent epistaxis	Excision	48 month	Yes
Tai SY, et al. ³⁾	2007	M/56	T1	Nasal obstruction, Epistaxis	Excision and post op RTx	12 month	No

RTx : radiotherapy

sive treatment such as surgery with radiotherapy and long-term follow-up should be required.¹⁰⁾ This would most likely decrease the development of local recurrence such as early stage ACC in nasal septum.

REFERENCE

- 1) Schneiderman TA, Chaudhury SI. *Adenoid cystic carcinoma of the nasal septum. Otolaryngol Head Neck Surg* 2002; 127(3):251-2.
- 2) Sivaji N, Basavaraj S, Stewart W, Dempster J. *Adenoid cystic carcinoma of the nasal septum. Rhinology* 2003;41(4): 253-4.
- 3) Tai SY, Chien CY, Tai CF, Kuo WR, Huang WT, Wang LF. *Nasal septum adenoid cystic carcinoma: a case report. Kaohsiung J Med Sci* 2007;23(8):426-30.
- 4) Shimamoto H, Chindasombatjaroen J, Kakimoto N, Kishino M, Murakami S, Furukawa S. *Perineural spread of adenoid cystic carcinoma in the oral and maxillofacial regions: evaluation with contrast-enhanced CT and MRI. Dento-maxillofac Radiol* 2012;41(2):143-51.
- 5) Freeman SR, Sloan P, de Carpentier J. *Carcinoma ex-pleomorphic adenoma of the nasal septum with adenoid cystic and squamous carcinomatous differentiation. Rhinology* 2003; 41(2):118-21.
- 6) Silverman DA, Carlson TP, Khuntia D, Bergstrom RT, Saxton J, Esclamado RM. *Role for postoperative radiation therapy in adenoid cystic carcinoma of the head and neck. Laryngoscope* 2004;114(7):1194-9.
- 7) Chen AM, Bucci MK, Weinberg V, Garcia J, Quivey JM, Schechter NR, et al. *Adenoid cystic carcinoma of the head and neck treated by surgery with or without postoperative radiation therapy: prognostic features of recurrence. Int J Radiat Oncol Biol Phys* 2006;66(1):152-9.
- 8) Lloyd S, Yu JB, Wilson LD, Decker RH. *Determinants and patterns of survival in adenoid cystic carcinoma of the head and neck, including an analysis of adjuvant radiation therapy. Am J Clin Oncol* 2011;34(1):76-81.
- 9) Kim YW, Cho KS, Kim BH, Goh JY, Roh HJ. *Comparison of treatment outcomes between endoscopic surgery only and endoscopic surgery with external approach in surgical malignancies. J Clinical Otolaryngol* 2011;22(1):59-64.
- 10) Roh HJ, Ahn JH, Lee HS, Cho KS. *Endoscopic management of sinonasal malignancy. J Clinical Otolaryngol* 2005; 16(2):258-63.