Clinical Case Report

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Sinonasal undifferentiated carcinoma originating from inverted papilloma

A case report

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Abstract

Rationale: Sinonasal inverted papilloma (IP) is a locally aggressive tumor found in the schneiderian membrane, lining the nasal cavity and paranasal sinuses.

Patient concerns: We report a case of a 63-year-old woman with undifferentiated carcinoma (UC) arising in an IP in the left maxillary sinus.

Diagnoses: The preoperative diagnosis was IP of the maxillary sinus.

Interventions: We performed endoscopic medial maxillectomy to remove the remnant mass in the left maxillary sinus.

Outcomes: Histological examination of the specimen revealed typical area of IP and sinonasal UC.

Lessons: Clinicians should consider sinonasal UC in the differential diagnosis of malignancy arising in the sinonasal IP.

Abbreviations: IP = inverted papilloma, UC = undifferentiated carcinoma.

Keywords: inverted, maxillary sinus neoplasms, papilloma, paranasal sinus cancers, sinonasal undifferentiated carcinoma

1. Introduction

Sinonasal inverted papilloma (IP) is a locally aggressive tumor found in the schneiderian membrane, lining the nasal cavity and paranasal sinuses. IP is clinically important due to its high rate of recurrence, locally aggressive nature, and association with malignancy.[1] Synchronous or metachronous malignancy of IP usually occurs in the form of squamous cell carcinoma (SCC).[1] Sinonasal undifferentiated carcinoma (SNUC) is nonsquamous cell epithelial and nonepithelial malignant tumor of varying histogenesis.[2] SNUC is a high-grade malignant epithelial neoplasm with or without neuroendocrine differentiation, but without evidence of squamous or glandular differentiation.[3] The World Health Organization classified SNUC as a highly aggressive and locally extensive tumor.[2] SNUC arising in an IP has never been reported, until now. Here, we report a case of a 63-year-old woman with undifferentiated carcinoma (UC) arising in an IP in the left maxillary sinus.t

2. Case report

A 63-year-old woman was transferred to our hospital due to an incomplete removal of IP. Suffering from a left nasal obstruction and rhinorrhea, she had visited a local clinic and had had endoscopic sinus surgery. A physical examination showed swelling and tenderness in the left cheek, and a rhinoscopy revealed an irregularly shaped mass in the left nasal cavity (Fig. 1). Her medical history was unremarkable, and she displayed no ocular symptoms. A computed tomography (CT) revealed that the remnant soft tissue mass was opacified in the floor of the left maxillary sinus (Fig. 2A). Magnetic resonance imaging (MRI) revealed a heterogenous signal intensity in the T1 and T2 weighted image (Fig. 2B, C). Positron emission tomography (PET) revealed that the mass was hypermetabolic (maximum standardized uptake value [SUV]= 5.96) (Fig. 2D).

We planned an endoscopic medial maxillectomy to remove the remnant mass in the left maxillary sinus; the patient was placed under general anesthesia. A frozen biopsy was performed on the mucosa of the medial maxillary wall, revealing the IP. After a resection of the medial maxillary wall by osteotome and diamond drill, a whitish, irregularly shaped mass was noted, originating on the prominent bony wall in the floor of the maxillary sinus. After the mass was removed, we proceeded to drill the bony wall, electrocauterized the floor of the maxillary sinus, and resected the left inferior turbinate. A permanent biopsy then showed SNUC with IP (Fig. 3A, B).

The patient was treated with a radiotherapy dose of 70 Gy. An 18-months follow-up was uneventful, and no pathologic

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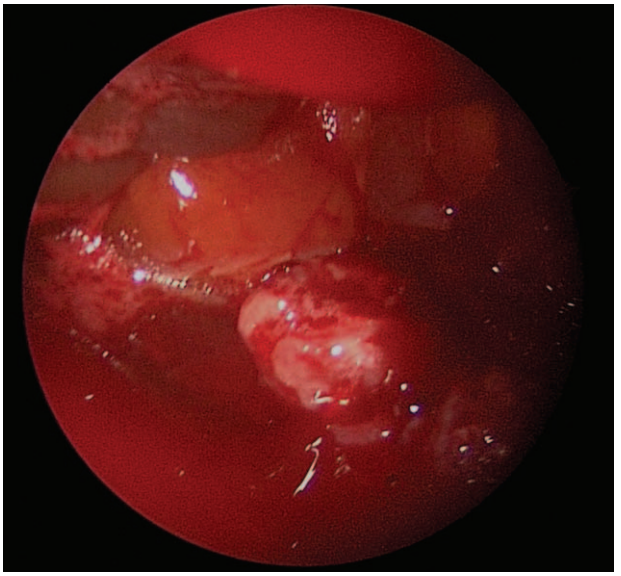

Figure 1. Endoscopic view of the nasal cavity. A huge, whitish irregularly shaped mass was detected from the left maxillary sinus.

lesions of the nasal cavity or maxillary sinus were detected (Fig. 4).

This study was approved by the institutional review board of the Chonbuk National University Hospital. Informed consent was given by the patients.

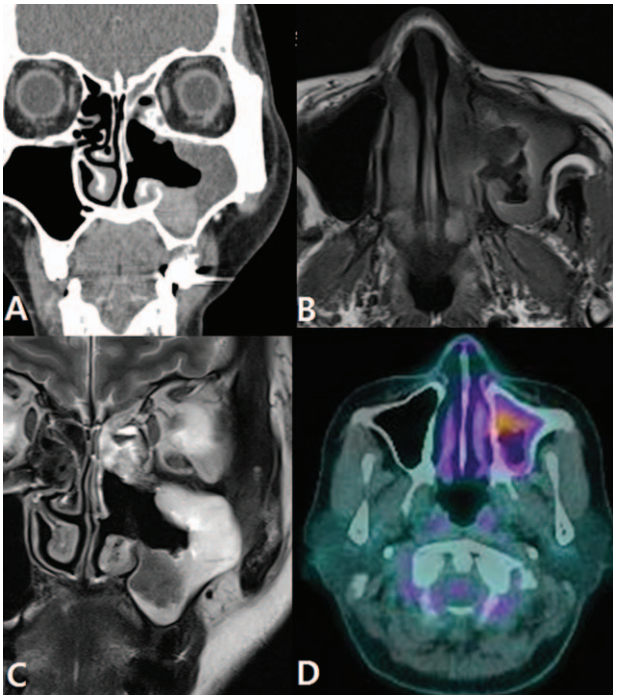

Figure 2. Coronal computed tomogram showing a hyperattenuated remnant lesion is in the left maxillary sinus (A). MRI showing a heterogenous signal intensity in the T1 (B) and T2 (C) weighted image. PET CT revealing a hypermetabolic mass in the left maxillary sinus (maximum SUV = 5.96), but showing no invasion to adjacent structures (D). CT = computed tomography, MRI = magnetic resonance imaging, PET = positron emission tomography, SUV = standardized uptake value.

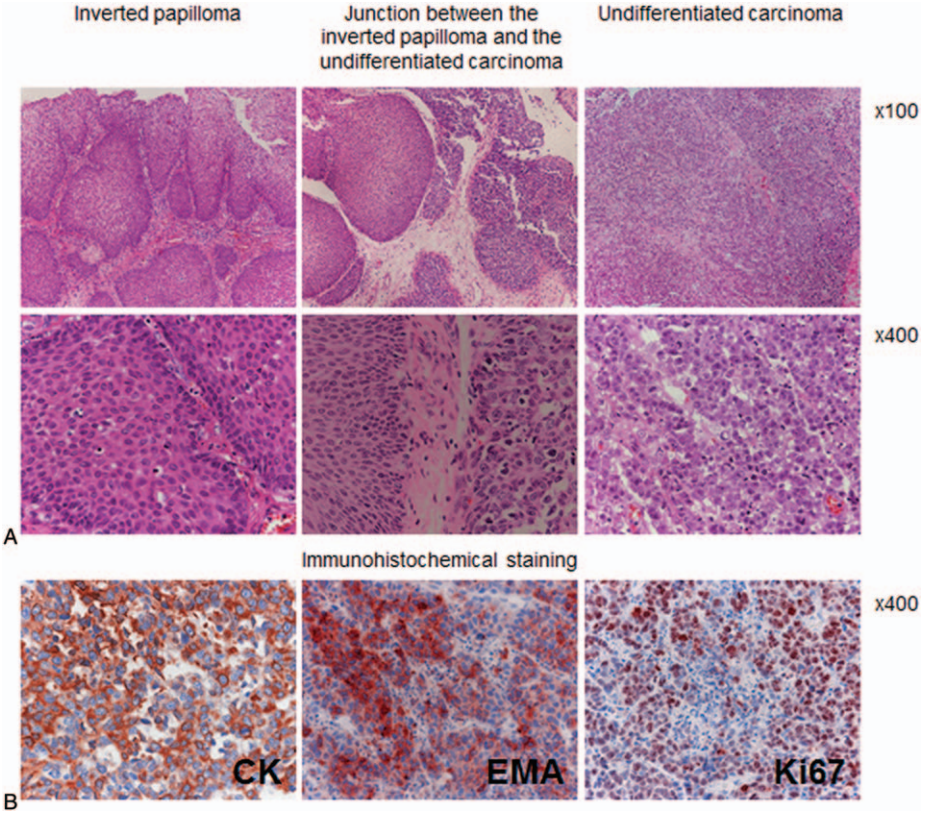

Figure 3. Histologic findings of coexisting inverted papilloma and sinonasal undifferentiated carcinoma. (A) Typical histologic features of inverted papilloma presented in left column and left side of middle column, and there was no cytologic atypia. Right side of the middle column and right column show histologic features of undifferentiated carcinoma with significant cytologic atypia, frequent mitosis, and tumor necrosis. (B) The tumor cells of undifferentiated carcinoma are positive for CK and EMA and have high proliferation index as represented by immunohistochemical staining for Ki67. CK = cytokerain, EMA = epithelial membrane antigen.

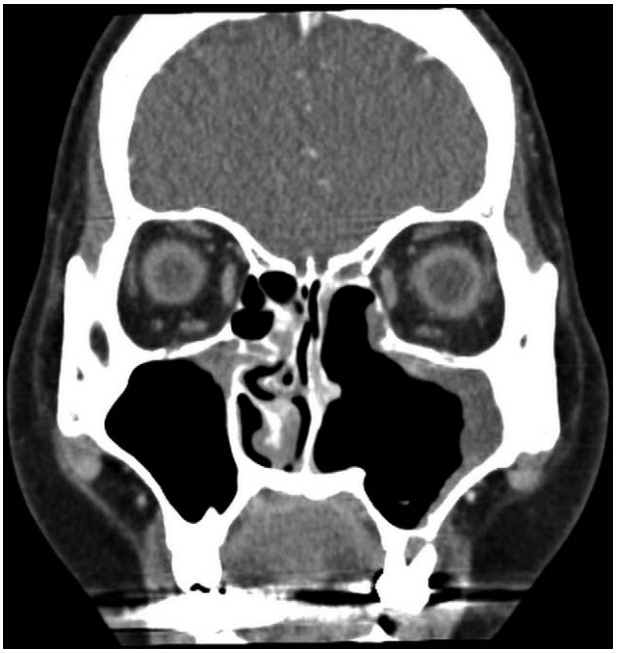

Figure 4. Coronal computed tomogram showed no evidence of tumor recurrence at 18-month follow-up.

3. Discussion

SNUC is an exceedingly rare and aggressive tumor found in the schneiderian epithelium lining the sinonasal tract. Most often, these tumors are diagnosed at an advanced stage and have a poor prognosis.[4] Surgery is the best treatment option for SNUC, although many patients require postoperative radiotherapy for advanced stage and close-tumor resection margins.[5] The diagnosis is difficult because there is no pathognomonic feature associated with SNUCs.[2]

IP is a rare benign tumor found in the schneiderian epithelium and is characterized by local destruction, recurrence, and possible malignant change. IP can be changed to SCC; however, SNUC associated with IP has never been reported until now.

To discriminate the IP from the malignancy, imaging study is necessary. Of all the imaging options, 18F-FDG PET/CT has proven most effective in detecting malignancy. In one study, which reviewed 8 patients with sinonasal IP (with or without SCC), using PET and MRI, the maximum SUVs measured 8.2 and 7.8 for benign IP, and 13.3 to 31.9 (mean $\pm$ SD = 20.2 $\pm$ 6.6) for IP with SCC.[6] In 6 patients who were defined as IP with SCC, the convoluted cerebriform pattern (CCP) in the MRI was investigated, which revealed a diffuse CCP in 2, a partial CCP in 3, and no CCP in 1. There was a wide discrepancy between the MRIs and PET/CTs regarding the area distribution of CCP and SUV. Shojaku et al[7] reported the maximum SUV in IP with SCC varied from 8.9 to 20.9. However, Zhang et al[8] reported that a primary sinonasal IP with SCC showed a maximum SUV of 4.02. From these results, we see that a wide variability of SUV exists in IP with SCC, effectively providing no diagnostic value in predicting malignancy. Felix-Ravelo et al[9] recently reported that SNUC had a mean maximum SUV of 14.2, which is higher than that of other sinonasal malignancies (sinonasal adenocarcinoma [9.9], sinonasal mucosal melanoma [7.0], olfactory neuroblastoma [7.0], adenoid cystic carcinoma [7.0], and sinonasal neuroendocrine carcinoma [4.7]). The maximum SUV of our SNUC with IP was 5.96, which is lower than might be expected. It is unclear whether the relatively lower SUV is due to the synchronous IP, or whether it is related to some other cause. However, we can deduce from this report that the SUV of SNUC is not always high.

To confirm the SNUC arising from IP, histopathology is essential. The tumor was composed of 2 components: typical area of IP and SNUC. The cells of IP did not show cytologic atypia or mitosis. However, several fragments of tumor components composed of anaplastic cells with severe cytologic atypia, frequent mitosis, and necrosis were identified. In addition, there was an area that the cell nests composed of anaplastic cells are adjacent to the IP (Fig. 3A). Immunohistochemically, the tumor cells of undifferentiated carcinoma are positive for cytokerain and epithelial membrane antigen and showed a high proliferation index in Ki67 immunostaining (Fig. 3B). The tumor cells were negative for CD3, CD20, and CD56 immunostaining and in situ hybridization for EBV-encoding RNA.

Since the synchronous IP and SNUC was first reported, our treatment regimen has been focused on the SNUC, due to its aggressiveness and poor prognosis. As previously reported, radical surgery is the best treatment option for SNUC.[5] Although some report found that surgery with chemoradiotherapy has a better outcome than surgery alone, or in conjunction with radiotherapy, our patient refused chemotherapy due to a fear of complications.[10] The origin of the SNUC in our case was the mucosa of the maxillary floor, which led to an inadequate resection in the private clinic. A 2nd surgery was performed 1 week later, requiring radical procedures, including medial maxillectomy with an inferior turbinate. High speed drilling (30000 rpm) and high frequency electrocautery (40 W) were used for trimming the bare bone in the floor of the maxillary sinus. Postoperative radiotherapy was performed with a dose of 70 Gy for 6 weeks.

4. Conclusion

To our knowledge, this is the first reported case of SNUC IP. Due to its rarity and the likelihood of a nonspecific finding at the time of presentation, the diagnosis runs the risk of being delayed. Based on our experience, and in light of its aggressiveness, we propose a radical surgery followed by radiotherapy for the treatment of synchronous IP and SNUC.

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