Case study

Spontaneous regression of recurred adenoid cystic carcinoma in the nasal cavity

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Abstract
Adenoid cystic carcinoma (ACC) is the most frequent malignant tumour of the submandibular and minor salivary glands, constituting 10% of salivary neoplasms, but it is an uncommon malignancy in the sinonasal tract. ACC in the sinonasal tract tends to be extensively involved in surrounding structures, including the brain, orbit or carotid artery, and it has a moderate to high-risk of local recurrences. In addition, there is a propensity for discrete regions of tumour infiltration along the cranial nerves. These findings make the clinical course of ACC potentially morbid. Because ACC in the sinonasal tract is uncommon, it is difficult to define the characteristics, but there are a few reported cases of SR of sinonasal tract ACC where the patient did not receive any type of therapy. In this study, we present a case of recurred ACC that spontaneously regressed after a biopsy was performed to confirm the pathologic diagnosis without treatment. We also present a brief literature review.

Introduction
Spontaneous regression (SR) of a malignant tumour is defined as partial or complete disappearance of a tumour in the absence of any treatment or during therapy that does not exert a significant effect on neoplastic disease. SR is known to be very rare, with an estimated incidence of >1 in 60,000–100,000 cases. The mechanisms underlying SR of cancer have not yet been fully determined; however, immunologic action, elimination of carcinogens, hormones, trauma, diet and medication have been reported as possible causes. Among the reported cases of SR of cancer, more than half are related to renal cell cancer, neuroblastoma, malignant melanoma and choriocarcinoma.

Case presentation
In October 2011, a 74-year-old woman was admitted to our department due to complaints of frequent nasal bleeding over a three-month period. The patient had been diagnosed with ACC (pT4aN0M0) of the hard palate in 2006 and had undergone total maxillectomy. She also received adjuvant radiotherapy (7000 cGy) to prevent recurrence and she received regular follow-up for five years without any signs of recurrence. Upon physical examination, there was a 1 × 2 cm-sized haemorrhagic mass on the medial portion of the right middle turbinate, which extended from the sphenoid sinus antrum and tended to bleed easily (Figure 1a). Enhanced perineural spread magnetic resonance imaging (PNS MRI) was performed, which revealed a well-defined heterogeneous enhancing mass between the right middle turbinate and nasal septum (Figure 1b). Therefore, we suspected the recurrence of ACC in the nasal cavity and recommended an intranasal biopsy for pathologic diagnosis and further treatment.

Intranasal biopsy using straight cup forceps was performed (Figure 2) under local anaesthesia, which established the histological diagnosis of recurred ACC (rT1N0M0) of the right nasal cavity (Figures 3a and 3b). Considering the patient’s general weakness, booster radiotherapy was planned and it was scheduled to start after one month.

The following month, physical examination of the patient’s nasal cavity was remarkable. The size of the mass was clearly reduced without the administration of any kind of treatment and nasal bleeding was not noted. Thus, radiotherapy was delayed and we decided to closely observe the changes in tumour size at our outpatient clinic. Follow-up MRI was performed at two months after biopsy, which indicated that the mass had completely regressed (Figure 4a). At six months after biopsy, an intranasal endoscopy showed a barely visible mass lesion in the patient’s nasal cavity (Figure 4b). Since then, the patient has remained in good health and currently shows no signs of recurrence.

Discussion and conclusion
ACC usually originates in the major or the minor salivary gland of the head and neck. Although uncommon,
Figure 4: (a) Perineural spread magnetic resonance imaging (PNS MRI) findings after an intranasal biopsy. PNS MRI was performed at two months after the intranasal biopsy. The enhanced T2-weighted axial view showed that the previous mass lesion on the medial portion of the middle turbinate had disappeared and no suspected malignant lesion was observed. (b) Endoscopic finding after an intranasal biopsy. Endoscopic examination was performed at six months after the intranasal biopsy and healthy nasal mucosa was observed around the middle turbinate and the nasal septum. No tumour-like lesion was detected.

*, middle turbinate; #, nasal septum.

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ACC is known to be a highly invasive malignancy with a typically fatal prognosis. It is also difficult to achieve adequate locoregional control with ACC. In this report, we presented a case of ACC with SR in the sinonasal tract and suggested that surgical trauma may have been the cause of improvement in the clinical prognosis of ACC in our patient.

Abbreviations list
ACC, adenoid cystic carcinoma; MRI, magnetic resonance imaging; PNS, perineural spread; PSN, paraneoplastic sensory neuronopathy; RCC, renal cell carcinoma; SR, spontaneous regression.

References

Tumours may sometimes grow more rapidly than their blood supply, which may cause direct necrosis and regression of the tumours. However, the definite causes for such a rapid growth of tumour cells have not yet been verified.

Paraneoplastic sensory neuronopathy (PSN), which is a rare syndrome causing sensory ataxia in the distal extremities, has been suggested as a cause of SR of small-cell lung cancer in some patients. It has been reported that specific autoantibodies such as anti-Hu, anti-Yo and anti-Ri antibodies react with tumour tissues, resulting in the activation of the T lymphocyte immune response. Although neurologic toxicity is a concern, patients with these autoantibodies tend to have a better cancer prognosis, with the tumour being smaller, less metastatic and more slow growing.

Recent reports have focused on alteration of the immune system such as T-cell dysfunction and immune deficiency syndromes, through the use of biological modulators, or during infection or hormonal changes. In particular, surgical trauma may also induce immune dysfunction of tumour cells, thereby resulting in direct necrosis and SR in patients.

In the case of thoracic malignancies, surgical trauma had also been reported to result in SR. Interestingly, 43% of thoracic malignancies that regressed spontaneously were observed in patients who had received surgical trauma. In those cases, surgical trauma included procedures such as bronchoscopy and incomplete tumour removal. As for the patient in our case, an intranasal biopsy was performed for a pathologic diagnosis and the recurrent ACC regressed without additional treatment one month after the biopsy. We did not find any clinical or physical signs of neurologic toxicity to confirm PSN but estimated that surgical trauma to recurrent ACC may have potentially acted as a trigger for an immune response, resulting in SR.
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