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CASE REPORT

2 Case reports of sinonasal adenoid cystic carcinoma: Review of the literature on surgical approaches

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KEYWORDS

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Abstract Adenoid cystic carcinoma (ACC) of sinonasal is an uncommon tumour that progresses slowly and generally manifested in advanced stage. Surgical management of sinonasal ACC can be challenging to the attending surgeon as they need to outweigh the decision between tumour clearance and morbidity of the patient. Multimodality treatment is the mainstay of treatment for sinonasal ACC and prognosis depends on the histological subtype of tumour, patient factor and treatment modalities opted. We are presenting 2 cases of sinonasal ACC manifested in different age groups in our centre with different types of surgical approaches opted and their outcomes.

1. Introduction

Sinonasal malignancy accounts for 1–2% of all malignancies. Adenoid cystic carcinoma (ACC) is the 3rd commonest sinonasal malignancy. It is common in females compared to males with a ratio of 2:1 and the peak incidence at age of 5th to 6th decade of life. The most common site affected is in the maxillary sinus followed by nasal cavity.[1] Tumors of the sinonasal tract commonly present with symptoms that are identical to those caused by inflammatory sinus disease, such as nasal obstruction, nasal discharge, epistaxis, headache, facial pain and cheek swelling.

ACC exhibits extensive local tissue infiltration and perineural spread, which results in a high rate of recurrence despite aggressive surgical resection. Classically, ACC has a distinct natural history of slow and indolent growth with late development of distant metastasis but tends to have local recurrence.[2] A few studies suggested that subtype of histopathology of

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ACC such as solid, tubular, cribriform and mixed types may determine the prognosis of the survival of a patient.

In general, tumours with perineural invasion, cervical lymph node metastasis, solid histological features and distant metastasis are associated with increased treatment failures and recurrences.[3] Tumour stage according to TNM staging has a significant correlation to the type of modality treatment opted.[4] In view of these tumours being rare and often presenting at an advanced stage, controversies exist as to the most appropriate treatment suitable. Until today, most sources agree that aggressive treatment of these tumours is necessary. Radiation therapy (RT), surgical resection, and combined modality treatments of these tumours have been reported and suggested.

2. Case report

2.1. Case 1

A 23 year-old Malay lady presented with left epistaxis for 8 months. There was no haematemesis and anaemic symptoms. It was associated with persistent left unilateral nasal obstruction associated gradual painless worsening left cheek disfigurement for 2 months. There were no allergic rhinitis symptoms and smell disorder. There was no cheek numbness, loosening of teeth, blurring of vision, or limited mouth opening. There were also no headache, throat or ear symptoms and no associated constitutional symptoms.

Generally patient was stable. There was blunted left nasolabial fold with vague mass overlying it. All cranial nerves examinations were unremarkable. The nasoendoscope examinations revealed a mass in the left nasal cavity which were dry, friable and bleed easily. The mass was sandwiched in between the septum and lateral nasal wall. Otherwise, the right nasal cavity was normal and both the Fossa of Rosenmueller (FOR) was clear.

Computed tomography (CT) scan revealed a soft tissue lesion at anterior left nasal cavity causing mild nasal septum deviation to the right. There was heterogenous enhancement seen post contrast and no soft tissue mucosal thickening seen at bilateral maxillary, ethmoidal, frontal and sphenoid sinuses without bony destruction. There was no turbinate hypertrophy, bilateral osteomeatal complexes are patent (Fig. 1). There were no neck nodes or distant metastases.

Magnetic resonance (MRI) scan showed a well-defined enhancing mass occupying the anterior aspect of left nasal cavity with a tiny hypointense lesion with no enhancement on post-gadolinium T2 weighted image. There was evidence of deep aspect of the overlying nasal ala subcutaneous tissues involvement but preservation superficial subcutaneous fat and skin of the nasal ala. No enhancing flow void was seen within the mass to suggest for an enlarged vessels within. The mass is in contact with the nasal septum. There was minimal focal deviation of the nasal septum to the right. There was no extension to the paranasal sinuses or focal intracranial lesion. There was no perineural invasion.

Biopsy of the mass revealed multiple fragment of tumour tissues, a few of the fragments are lined by stratified squamous epithelium. The tumour cells are predominantly in solid nest (60%) and cribriform pattern (40%). It is composed of small basaloid cells with scanty cytoplasm. Mitosis is frequently

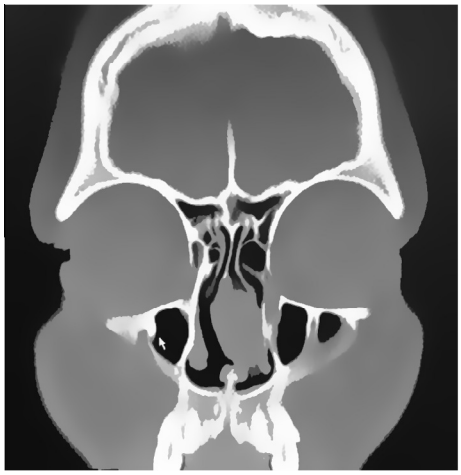

Figure 1 Soft tissue heterogenous enhancement lesion at anterior left nasal cavity causing mild nasal septum deviation to the right. There was heterogenous enhancement seen post contrast and no soft tissue mucosal thickening seen at bilateral maxillary, ethmoidal, frontal and sphenoid sinuses. There was no turbinate hypertrophy, bilateral osteomeatal complexes are patent, orbit and visualized brain is normal. No bony destruction seen.

seen. The lumina contain PAS positive materials that are suggestive of adenoid cystic carcinoma (Fig. 2). All blood investigations were in normal range.

Prior surgery, disease was at stage T3N0M0. Patient underwent tumour resection via combined endoscopic and midfacial degloving, with sublabial incision approach and transfixes incision at interseptal cartilage under general anaesthesia. Intraoperative revealed a mass arised from the anterosuperior septum which was about 1 cm anterior to middle meatus. Both inferior, middle turbinate and OMC were normal. The mucosa of septal cartilage and nasolabial fold which are infiltrated by the mass are resected. Histopathological (HPE) specimen revealed a positive margin of tumour septal root, septal mucosa and nasolabial subcutaneous tissue. The patient was finally diagnosed as having ACC of nasal septum. She then underwent radiotherapy 33 fractions for total 66 Gy. 6 months after surgery follow up showed no recurrence of local and regional of disease.

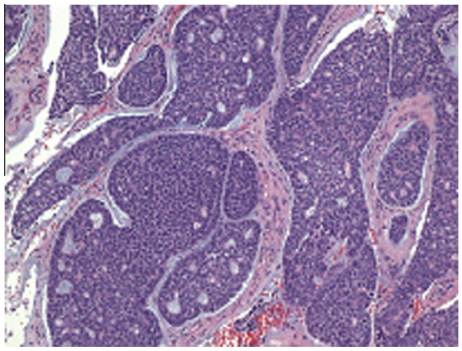

Figure 2 Tumour cells are predominantly in solid nest (60%) and cribriform pattern (40%).

2.2. Case 2

66 year old Malay gentleman with underlying hypertension initially presented with persistent unilateral left nasal obstruction associated with blood-stained foul smelling nasal discharge for 4 months duration. There were no anosmia, facial pain, loosening of tooth and no constitutional symptoms. There was no gross facial asymmetry. Nasoendoscope examination revealed a fleshy mass occupying the left nostril with contact bleeding. The mass seems to be arising from lateral nasal wall. The right nostril showed normal inferior and middle turbinate with clear osteomeatal complex and both Fossa of Rosenmueller were normal.

CT scan showed mass of homogenous soft tissue attenuation within the left nasal cavity extending into left maxillary as well as left ethmoidal sinus with impression of sinonasal polyposis and differential of inverted papilloma or carcinoma (Fig. 3). CT scan staging showed no sign of locoregional and distant metastases. Biopsy of the mass came back as ACC with TNM stage of T2N0M0.

Patient then underwent endoscopic resection of tumour, near total inferior turbinectomy and middle meatal antrostomy under general anesthesia. Intraoperative findings showed a fungating mass occupying the left nasal cavity, its pedicle originating from inferior turbinate which easily bleed. There was also suspicious mass with polypoidal mucosa occupying left maxillary sinus. The anterior and posterior ethmoid cells were normal.

Histopathological examination post-operative showed the tumour tissue fragment consists of malignant cells arranged in cribriform, tubular, cords and solid pattern. The malignant cells are composed of two populations of basaloid and pale cells exhibiting oval to round shaped cells with large hyperchromatic nuclei, scanty cytoplasm and some shows prominent nucleoli (Fig. 4). They are surrounded by desmoplastic stroma, the tubular lumen contains basophilic material. Lymphovascular and perineural invasion was not seen. The oncological tumour margin was clear. The mass in maxillary sinus was reported as benign inflammatory polyps.

Patient underwent 35 fractions of radiotherapy for total 60 Gy. During follow up 3 months post radiotherapy, there was distorted small stump of the left inferior turbinate with no local and regional recurrence.

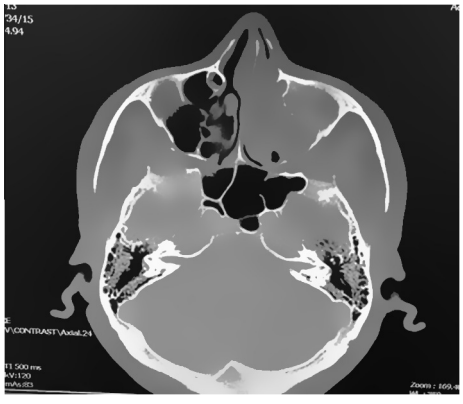

Figure 3 Homogenous soft tissue attenuation within the left nasal cavity extending into left maxillary as well as left ethmoidal sinus.

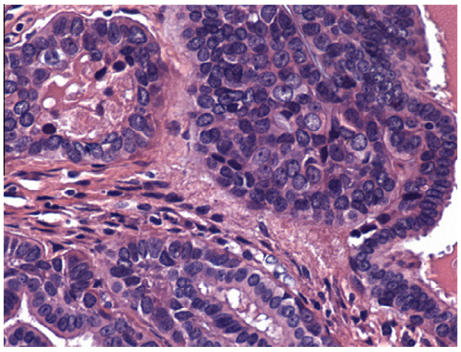

Figure 4 Tumour tissue fragment consists of malignant cells arranged in cribriform, tubular, cords and solid pattern.

3. Discussion

Adenoid cystic carcinoma is the third commonest sinonasal malignancy. It posed challenges in term of its management because of difficulty to access the tumour. Furthermore most patients at presentation are already at advanced stages of the disease.[5] The earlier the diagnosis and treatment received patients will have better outcomes in terms of morbidity, risk of residual or recurrence of disease. A few important factors that need to be considered in the treatment of sinonasal malignancy are oncological clearance of surgery, functional status of the patient and cosmetic acceptability.

Clinical manifestation of adenocystic carcinoma varies but the patient mainly presents with epistaxis and nasal blockage. As in both cases, its clinical presentation is classical and at an early stage, there were no symptoms of local spread. Sinonasal malignancy commonly presented with nasal obstruction, followed by epistaxis and local spread symptoms such as auditory, nerve and visual symptoms.[6]

Surgery is the mainstay of the treatment for sinonasal ACC. It can be either open, endoscopic or a combined approach. The current paradigm of treatment of sinonasal malignancy is towards multimodality and multidisciplinary approaches.[7] Ideally, adjuvant chemoradiation is given 4–6 weeks after surgery.

In our first case, the patient is a young female unfortunately diagnosed at quite moderately advanced stage of the disease; T3N0M0. Although it was postulated that adenoid cystic carcinoma may have distant metastases, but one study showed that a large tumour of adenoid cystic carcinoma often does not have distant metastases.[8] Therefore, oncological clearance of surgery is mandatory with preservation of function and cosmetic acceptability in this patient. She underwent a combined open and endoscopic midfacial degloving and transfix incision at interseptal cartilaginous approach. Post operatively, her HPE showed clear oncological margin of tumour. She then received post-operative radiotherapy. This is in concordant with one of the cancer centre experiences in multimodality treatment for adenoid cystic carcinoma. This centre showed better outcomes in terms of prognosis in comparison with single modality treatments.[9]

For our second case, patient presented to us at stage T2N0M0 and underwent endoscopic approach tumour

resection. HPE result showed oncologic free tumour margin. Although the study showed perineural and perivascular invasion are high in adenoid cystic carcinoma[10], in both our cases there was no invasion seen from imaging and HPE.

Generally, adenoid cystic carcinoma is associated with good prognosis in stage I–II of the tumour when combined with multimodality treatment. There are few factors that give favourable prognosis of ACC such as histopathological findings of solid type of tumour, absent of perineural or perivascular invasion, absent of distant metastases, site of tumour as well as patient and treatment factors such as age and multimodality treatment.[11] In both of our cases upon follow up after radiotherapy, there was no local and distant tumour recurrence.

4. Conclusion

Sinonasal ACC may manifest in various presentations with peak incidence at 5th to 6th decade of life. However, classical symptoms of sinonasal ACC are epistaxis, nasal blockage and cheek swelling. Therefore, any patient with suspicious malignancy needs to be investigated thoroughly by histological diagnosis. The mainstay treatment of sinonasal ACC is surgery followed by radiotherapy. However, option of surgical approach may tailor to the case basis. Tumour factors such as tumour stage, locoregional metastasis significantly affect treatment modalities. Besides that, patient and surgeon factors also influence the outcome of treatment.

References

1. Gandhi AK, Roy S, Biswas A, et al. Adenoid cystic carcinoma of head and neck: A single institutional analysis of 66 patients treated with multi-modality approach. *Indian J Med Paediatr Oncol*. 2015;36(3):166–171.
2. Sung M-W, Kim KH, Kim J-W, et al. Clinicopathologic predictors and impact of distant metastasis from adenoid cystic carcinoma of the head and neck. *Arch Otolaryngol Head Neck Surg*. 2003;129 (11):1193–1197.
3. Lloyd S, James BY, 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.
4. Blanch JL, Ruiz AM, Alos L, Traserra-Coderch J, Bernal-Sprekelsen M. Treatment of 125 sinonasal tumors: prognostic factors, outcome, and follow-up. Otolaryngology–head and neck surgery. *Otolaryngol Head Neck Surg*. 2004;131(6):973–976.
5. Poursadegh M, Poursadegh F, Esmaeili M, Bakhshaee M. Epidemiological survey of sinonasal malignancy in North-East Iran. *Iran J Otorhinolaryngol*. 2015;27(80):225–229.
6. Thompson LDR, Penner C, Ho NJ, et al. Sinonasal tract and nasopharyngeal adenoid cystic carcinoma: a clinicopathologic and immunophenotypic study of 86 cases. *Head Neck Pathol*. 2014;8 (1):88–109.
7. Nicolai P, Castelnuovo P, Villaret AB. Endoscopic resection of sinonasal malignancies. *Curr Oncol Rep*. 2011;13(2):138–144.
8. Touati MM, Ammar H. Giant adenoid cystic carcinoma of the sinonasal cavity. *Pan Afr Med J*. 2015;21:82.
9. Lupinetti AD, Roberts DB, Williams MD, et al. Sinonasal adenoid cystic carcinoma: the M.D. Anderson Cancer Center experience. *Cancer*. 2007;110(12):2726–2731.
10. Hanna E, Vural E, Prokopakis E, Carrau R, Snyderman C, Weissman J. The sensitivity and specificity of high-resolution imaging in evaluating perineural spread of adenoid cystic carcinoma to the skull base. *Arch Otolaryngol Head Neck Surg*. 2007;133(6):541–545.
11. Ko YH, Lee MA, Hong YS, et al. Prognostic factors affecting the clinical outcome of adenoid cystic carcinoma of the head and neck. *Jpn J Clin Oncol*. 2007;37(11):805–811.