The Journal of Laryngology & Otology (2015), ***129*** (Suppl. S2), *S95–S97*.

doi:10.1017/S0022215114002485

Hyalinizing clear cell carcinoma of the nasopharynx operated by trans-oral and trans-palatal approach

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Abstract

Background: Hyalinizing clear cell carcinoma is a rare minor salivary gland neoplasm. The treatment of choice is surgical resection with or without post-operative radiotherapy. This tumour often demonstrates a good prognosis.

Case report: We report a case of hyalinizing clear cell carcinoma arising in the nasopharynx. A 27-year-old female presented with progressive hearing disturbance and tinnitus. On examination, an expansile mass was observed in her nasopharynx. Biopsy was performed and the pathology results returned as clear cell carcinoma.

Results and conclusion: Surgical resection was performed trans-orally accompanied by trans-palatal approach. She has no recurrence during more than two years of follow up.

Key words: Hyalinizing Clear-cell Carcinoma; Salivary Gland; Trans-palatal Approach

Introduction

Hyalinizing clear cell carcinoma is a rare, low-grade neoplasm that accounts for less than 1 per cent of tumours of the salivary gland.[1] Most of the cases are presented as a mass of the pharynx or oral cavity, indicating its origin as a minor salivary gland tumour. The limited number of reported cases indicates that wide local excision is the treatment of choice. Occasional cervical lymph node metastasis is observed and the role of post-operative radiotherapy (RT) or neck dissection is controversial. The clinical behaviour of hyalinizing clear cell carcinoma is often indolent with low frequency of metastasis.[1] However, recent reports following accumulation of sporadic cases, neck metastasis including delayed metastasis is not rare.

We report a rare case of hyalinizing clear cell carcinoma arising from the nasopharynx, whose initial symptom was otitis media with effusion. The clinical, histopathologic characteristic of hyalinizing clear cell carcinoma and the operative procedure of the case are described.

Case report

A 27-year-old female was referred to our hospital with a six-month history of right tinnitus and hearing disability. Pure tone audiometry indicated a conductive hearing loss of 40 dB. On physical examination, her right ear drum revealed otitis media with effusion. On nasal endoscopic examination, there was an expansile tumour in the right side of the nasopharynx obstructing the eustachian tube orifice (Figure 1).

Magnetic resonance imaging examination showed a 30 mm × 25 mm expansile mass in the nasopharyngeal region (Figure 2a) without extension to the parapharynx. Mild 18F-fluorodeoxyglucose uptake to the tumour was observed in a positron emission tomography–chemotherapy (Figure 2b) There was no cervical lymph node metastasis.

Biopsy of the tumour was performed. The tumour revealed proliferation of carcinoma cells with round to polygonal nuclei and clear or eosinophilic cytoplasm. Hyalinised eosinophilic materials were observed within or around the tumour cells (Figure 3). Mitotic figures were rare and the MIB-1 labelling index was about 5 per cent in the hot spot. Further molecular analysis was performed and ESW exon 11–ATF exon 3 fusion gene transcript was detected by Reverse Transcription-polymerase chain reaction. These pathologic and molecular findings led to the diagnosis of hyalinizing clear cell carcinoma.

The patient underwent a wide surgical resection. Preoperative findings indicated that the tumour was localised in the nasopharynx, and a trans-oral, trans-palatal approach was utilized. The soft palate was split in the midline and part of the hard palate was drilled to obtain a good surgical field (Figure 4). The tumour was resected with a 1 cm margin by electric cautery. The soft palate was sutured after resection of the tumour.

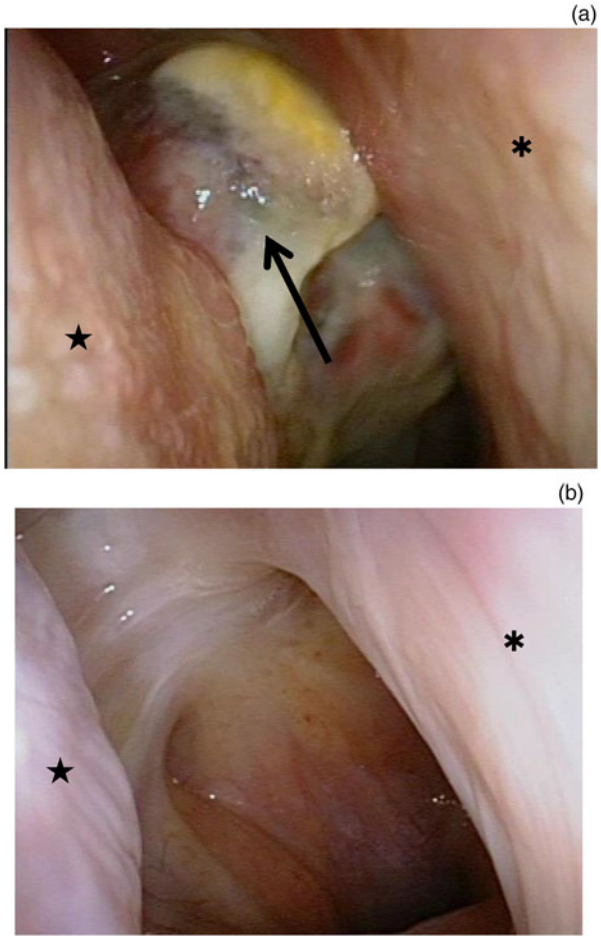

FIG. 1

(a) Nasal endoscopy revealed a partially necrotic, reddish tumour arising from the right lateral wall of the nasopharynx. (Arrow: tumour, ★: inferior turbinate, *: nasal septum.) (b) Endoscopic view of the nasopharynx one year after surgery. (★: inferior turbinate, *: nasal septum.)

Otitis media and hearing disturbance disappeared one month after the surgery. She has no complaint of nasopharyngeal reflux. No adjuvant RT was given. There is no recurrence more than two years after the operation.

Discussion

Hyalinizing clear cell carcinoma was first described by Milchgrub in 1994, as a distinct entity of salivary gland tumours.[2] Since then, hyalinizing clear cell carcinoma of the minor salivary gland has been reported in the oral cavity, larynx and nasal cavity. Hyalinizing clear cell carcinoma originating from the nasopharynx is rare.[3] Definitive diagnosis is based on histological examination following biopsy or resection of the tumour. Hyalinizing clear cell carcinoma cells have clear or eosinophilic cytoplasm arranged in small nests or sheets/cords. The tumour cells are often surrounded by hyalinized eosinophilic material. Mitotic figures are often low. Differential diagnosis should include other salivary gland malignancies such as mucoepidermoid carcinoma, acinic cell carcinoma or clear cell odontogenic carcinoma.[4] In addition, metastatic tumours, particularly metastatic renal cell

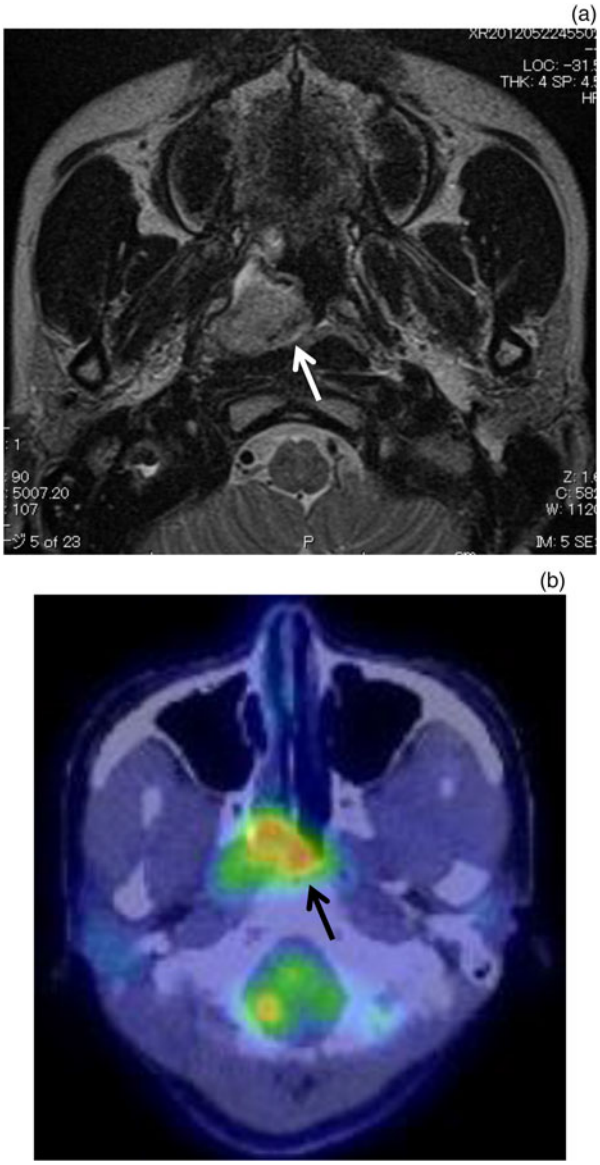

FIG. 2

(a) Magnetic resonance imaging, T_2-weighted image indicates an expansive tumour in the nasopharynx (arrow). Invasion into the muscle layer was not obvious. (b) positron emission tomography–-chemotherapy fusion image shows mild uptake of 18F-fluorodeoxyglucose in the tumour.

carcinoma that commonly appears as a glycogen-rich clear cell, should also be ruled out. Recently, Antonescu *et al.* reported that EWSR1–ATF1 fusion is a consistent finding in hyalinizing clear cell carcinoma[5] suggesting the importance of molecular evaluation for the diagnosis. It may be important for pathologists as well as head and neck surgeons to recognise this unusual salivary gland neoplasm.

We report a case of hyalinizing clear cell carcinoma that was confirmed on molecular basis by detecting the EWSR1–ATF1 gene fusion. The tumour was localised in the nasopharynx and a trans-oral/trans-palatal approach was useful for operation. This surgical approach allows the operation field to the nasopharynx, up to the skull base, and clivus. It is utilized by neurosurgeons combined with a trans-pharyngeal approach to

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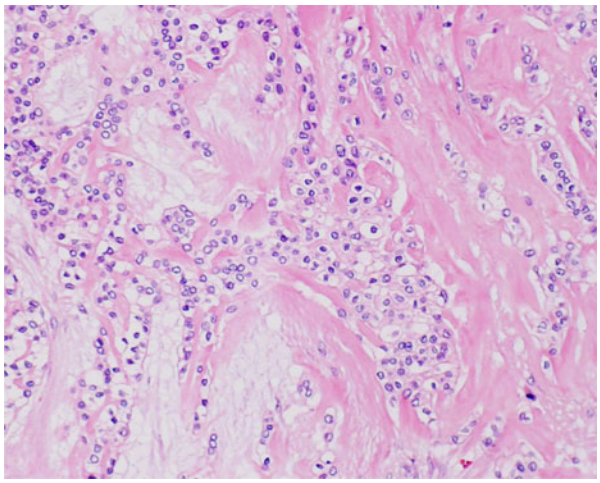

FIG. 3

Haematoxylin–eosin staining of the tumour. The tumour demonstrates proliferation of carcinoma cells with round to polygonal nuclei and clear or eosinophilic cytoplasm. Hyalinized eosinophilic materials were observed within or around the tumour cells. (original maginification ×400)

reach the craniocervical junction.[6] When approaching the nasopharynx, velopalatine incompetence is one of the major complications of this surgical approach, which we did not observe in our patient. The trans-palatal approach is an useful method when operating a localised tumour of the nasopharynx.

Little is known about the role of post-operative RT because of the rarity of hyalinizing clear cell carcinoma. Hijjawi *et al.* reported a recurrent case of hyalinizing clear cell carcinoma with was effectively treated by chemoradiotherapy.[7] Our patient did not receive post-operative RT, since there was neither cervical involvement nor invasive character of the primary tumour. The effect of RT for hyalinizing clear cell carcinoma needs to be further discussed.

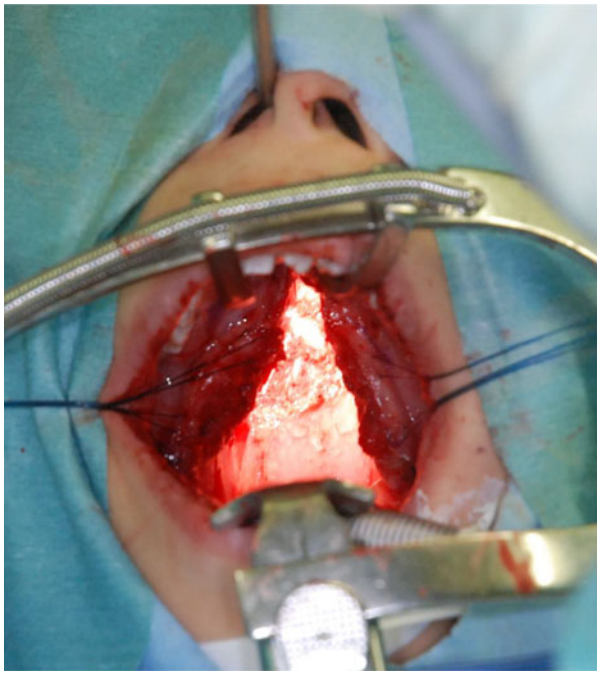

FIG. 4

Intra-operative view of the trans-palatal approach extending the surgical field up to the nasopharynx.

- **This is a case report of a patient with hyalinizing clear cell carcinoma of the nasopharynx**
- **The patient was operated upon utilising a trans-palatal approach**
- **The diagnosis and management of this rare minor salivary gland tumour is discussed**

In general, hyalinizing clear cell carcinoma demonstrates good prognosis with low frequency of metastasis. However in 2009, Solar *et al.* reviewed 52 cases of hyalinizing clear cell carcinoma from previous reports and observed that 11.5 per cent of the patients developed recurrent disease.[8] A case with delayed cervical lymph node metastasis has also been reported recently by Su *et al.*[9] These findings indicate that long-term follow up is desirable. Since the number of reported hyalinizing clear cell carcinoma cases is still small, accumulation of cases is required to discuss the role of multimodality treatment for treating this disease.

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Competing interests: None declared

https://doi.org/10.1017/S0022215114002485 Published online by Cambridge University Press