Myoepithelial Carcinoma of the Nasopharynx

Nadia Syafereera NASERRUDIN, Timothy WONG, Farah Dayana ZAHEDI, Salina HUSAIN
Department of Otorhinolaryngology - Head & Neck Surgery, Universiti Kebangsaan Malaysia Medical Centre, Malaysia

Background

- Myoepithelial carcinoma (MECA) is a locally aggressive salivary gland tumour.
- This is 4th case reported in English literature of nasopharyngeal MECA with majority reported to have nasal obstruction.
- Majority of MECA are located in the parotid gland but it has previously been reported to occur in breast, lung and orbit.
- The management of MECA of nasopharynx remain a challenge due to its rarity. Treatment involves chemoradiotherapy and surgical excision but with high morbidity due to its close relation to intracranial.

Case Report

A 70 year-old-Indian lady presented with blood stained saliva for past 1 month. She also has left ear blockage with reduced hearing. She has no epistaxis or nasal blockage. She has no tuberculous symptom. Nasoendoscopy revealed a lobulated mass arising from left fossa of Rosenmuller, obliterating the Eustachian tube. Biopsy revealed myoepithelial malignant tumour. CT and MRI of Neck showed 2.0x1.1x2.6cm homogenous enhancing nasopharyngeal mass filling the left fossa of Rosenmuller and obstructing the Eustachian tube. It extended to parapharyngeal space (laterally), anterior wall of left common carotid artery (posterolaterally) and foramen lacerum (superiorly). There were also vertebral metastases. She underwent left extended nasopharyngectomy. Intraoperatively, there was a lobulated mass at the left side of nasopharynx, obliterating the Eustachian tube. The extension were until prevertebral muscle (posteriorly), cartilaginous Eustachian tube (laterally), basioccipitoid (superiorly), internal carotid artery (posterolaterally) and soft palate (inferiorly). The immunohistochemical studies showed the malignant cells are positive for p63, S100, SMA, CKA1/AE3 and CK7, but negative for CD117. She is plan for PET CT scan and post operative chemoradiotherapy.

Discussion

- Myoepithelial carcinoma (MECA) of the nasopharynx is very rare. We believe this is the fourth case reported in English literature; being the oldest age affected at 70 years old, compared to other cases which affect 55 to 60 years old (1).
- The definitive diagnosis of myoepithelial carcinoma by WHO is by reactivity for cytokeratins and at least one other myoepithelial marker (2). The immunohistochemistry in MECA demonstrates positivity in myoepithelial marker such as p63, S100 and SMA.
- MECA also showed positivity towards cytokeratin markers such as CKA1/AE3 and CK7. The case is also EBV negative. Negative expression for EBV and positive expression on myoepithelial marker contradicts the diagnosis of nasopharyngeal carcinoma.
- In view of its rarity, the management and prognosis of this tumour is not well described.
- MECA of nasopharynx have been managed with either chemoradiotherapy alone or surgery and radiotherapy with or without chemotherapy. 2 out of 3 cases did show residual or recurrence of disease.

Conclusion

MECA is very rare and its management is challenging due to its rarity, its close proximity with skull base and being locally aggressive. It should be identified early to achieve a complete tumour excision.

Table 1: Current case and other 3 published cases in English literature on MECA of nasopharynx

<table>
<thead>
<tr>
<th>Cases</th>
<th>Demographic</th>
<th>Presenting Complaint</th>
<th>Tumour Size</th>
<th>Management</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Current case</td>
<td>70, female</td>
<td>Bloodstained saliva, left conductive hearing loss</td>
<td>3cm</td>
<td>Surgery plan for CT/RT</td>
<td>Awaiting PET scan</td>
</tr>
<tr>
<td>Soon et al (2015)</td>
<td>55, male</td>
<td>Nasal blockage, epistaxis</td>
<td>3.2cm</td>
<td>CT, RT</td>
<td>5 months, residual tumour</td>
</tr>
<tr>
<td>Dhusan et al (2011)</td>
<td>60, female</td>
<td>Nasal blockage, hyponasal speech, conductive hearing loss, ear fullness</td>
<td>4.3cm</td>
<td>Surgery, RT</td>
<td>28 months, no recurrence</td>
</tr>
<tr>
<td>Tuncel et al (2004)</td>
<td>60, female</td>
<td>Nasal obstruction, epistaxis, left neck swelling</td>
<td>5cm</td>
<td>Surgery, RT</td>
<td>14 months recurrence with intracranial extension, pulmonary metastasis. Died 1 month later</td>
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References: