Case Report

Primary undifferentiated carcinoma of the temporal bone: A new entity?

WZA Rusydi, BS Goh

Department of Otorhinolaryngology, Universiti Kebangsaan Malaysia Medical Centre Kuala Lumpur, Malaysia.

ABSTRACT

Undifferentiated carcinoma of the temporal bone is an extremely rare disease, and has always been associated with metastatic disease from other sites. We report a case of a 70-year-old gentleman presenting initially with facial nerve palsy and features of chronic otitis media with cholesteatoma. He underwent mastoidectomy and was diagnosed with undifferentiated carcinoma of the temporal bone by the histopathological examination of the biopsies from mastoid and middle ear. A thorough assessment for possible primary lesion including blind biopsy of the nasopharynx was negative. He refused further surgery despite residual tumour in middle ear, and was treated with radical radiotherapy alone and responded well with no recurrence, clinically and radiologically after 5 years of follow-up. Primary undifferentiated carcinoma of the temporal bone is a possible new entity and may carry good prognosis. (Rawal Med J 2013;38:86-89).

Keywords: Temporal bone, undifferentiated carcinoma, facial nerve, surgery, radiotherapy

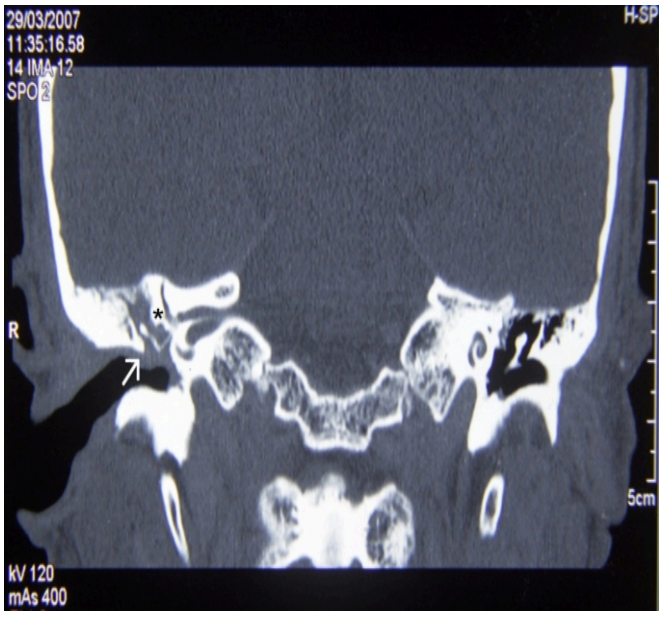
INTRODUCTION

Malignant disease of the temporal bone is rare with estimated incidence of 6 cases in a million populations per year, or about 1 in 5,000 20,000 otologic cases.[1] Its rarity combined with the clinical presentations which are similar to infective ear diseases frequently causes delay in diagnosis. Histologically, squamous cell carcinoma is most common, consisting of 62-69% of all primary malignancy, followed by basal cell carcinoma, adenocarcinoma and other malignancy of salivary gland or mesenchymal origin.[2-3] However, undifferentiated carcinoma of the temporal bone had always been associated with metastatic disease from other sites. Here, we present a case of undifferentiated carcinoma of the temporal bone with no primary found.

CASE PRESENTATION

A 70 year-old Chinese gentleman presented with history of progressive hearing loss in the right ear for six months associated with intermittent ear discharge. He also developed right lower motor neuron facial nerve palsy, House-Brackmann grade V for two months duration. He had history of left inactive chronic otitis media for few years.

Fig. 1 CT scan (coronal view) of the temporal bone showing soft tissue density on the right within the osseous part of the external auditory canal and middle ear cavity, with erosion of the scutum (arrow) and the floor of the horizontal facial canal (inferior to *).

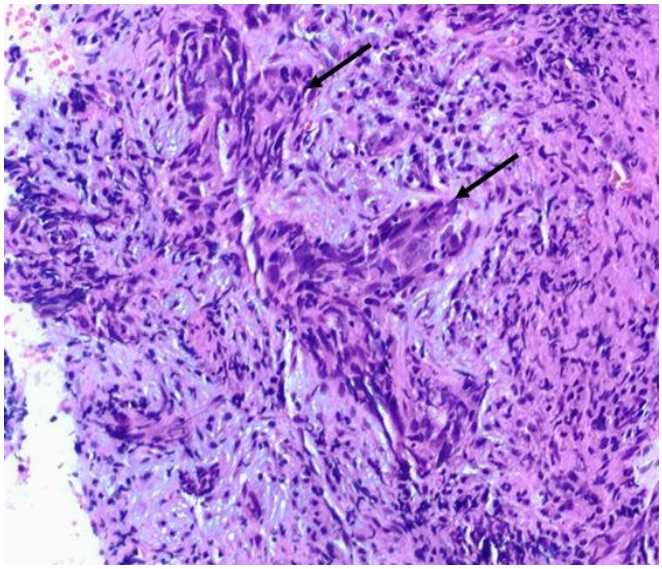


Otoscopic examination revealed sagging of the posterosuperior wall of the right external ear canal (EAC) obscuring the tympanic membrane. Central perforation of the left tympanic membrane was also

noted. No regional lymphadenopathy was present. Pure tone audiometry showed moderate to severe mixed hearing loss with air-bone gap of 35 dB on the right and mild to moderate mixed hearing loss with air-bone gap of 20 dB on the left. Computed tomography (CT) scan of temporal bone revealed soft tissue density mass within the osseous part of the right EAC and middle ear cavity (Fig. 1).

Scutum and horizontal portion of facial nerve canal was also eroded. Diagnosis of right ear cholesteatoma was made and modified radical mastoidectomy was performed in April 2007. Intra operatively, there were soft tissues filling the whole middle ear, mastoid antrum and mastoid cavity and were removed except tissues around the stapes foot plate. Ossicles were all intact and there was no evidence of cholesteatoma. Facial nerve was identified and noted dehiscence at the horizontal segment of the facial canal. Tissues from the right middle ear histopathological examination was reported as undifferentiated carcinoma, and immunohistochemistry showed the malignant cells were positive towards cytokeratin. He was staged as T4N0M0 according to the Pittsburgh system.[4]

Fig. 2 Infiltration of the malignant cells arranged in syncytium (arrow) in a background of inflamed fibrocollagenous tissue by lymphocytes. (H&E x100).



He was investigated for other possible primary malignancy through clinical examination, imaging, tumour markers and targeted biopsy of the nasopharynx but the findings were all negative. He was advised for temporal bone resection for tumour clearance but refused further surgery. He was planned for radiotherapy but it was delayed due to some logistic factors. He finally underwent radical radiation therapy to the right mastoid region in October 2007, and received 60 Gy in 30 daily fractions within 6 weeks duration.

On follow up six months after completion of radiotherapy, surveillance CT scan and MRI did not show any evidence of tumour recurrence. There was no local recurrence or signs of other malignancy seen at 60 months after completing the radiotherapy. However, right facial nerve palsy persists.

DISCUSSION

Undifferentiated carcinoma of the temporal bone had never been reported as a primary disease in English literature searched on Pubmed. Streitmann and Sismanis described five cases of undifferentiated carcinomas out of 141 metastatic temporal bone malignancies by reviewing literatures from 1902 to 1994: 1 each from lung, prostate and bladder, and 2 of unknown primary.[5] Gloria-Cruz et al did a review of autopsy record of patients with primary nondisseminated malignant neoplasm and found that 47 out of 212 patients had metastases to the temporal bone.[6] Out of these, 4 had undifferentiated carcinoma: 2 from lung, 1 from prostate, and 1 unknown primary. These studies also showed the common primary sites for temporal bone metastasis in general are breast, lung, prostate, kidney, stomach and bronchus. Undifferentiated carcinoma in the middle ear has also been reported in 2 patients with undifferentiated nasopharyngeal carcinoma.[7]

Clinical presentation of temporal bone carcinoma may not be distinguishable from infective otologic condition such as chronic otitis media with or without cholesteatoma. Otorrhoea, pain and reduced hearing are common, followed by occlusion, tinnitus, facial nerve palsy and vertigo. As in this case, misdiagnosis is common. Examination may reveal ear canal mass, as most of the primary tumour arises from EAC.[8] However, distinguishing between EAC and middle ear

malignancy may not be possible in advanced disease.

In hematogenously spread metastatic tumours, the most commonly affected site is the marrow-containing petrous apex, which is capable to filter out tumour cells from circulating blood.[5] It is reported to be as high as 83%, followed by internal acoustic canal and mastoid. Therefore, up to 36% of the patient may be asymptomatic of otologic or vestibular symptoms.[6] Tissue biopsy from the tumour is needed to confirm the diagnosis. High-resolution CT scan of the temporal bone allows accurate evaluation of the bony erosions. MRI offers better assessment of the soft tissue spread. Combination of both modalities provide best radiographic tumour mapping.

To date, there is no universally accepted staging system due to the rarity of the disease. The Pittsburgh staging system[4] has been shown to estimate prognosis well.[2 9-10] This system is originally meant for squamous cell carcinoma of the temporal bone, and is similar to TNM system, which classifies cases into T1-T4. N and M classification are similar to other head and neck malignancy.

Standard curative treatment for temporal bone carcinoma is *en bloc* excision with clear margins. This includes lateral temporal bone resection (LTBR) for T1 & T2 tumour, or subtotal temporal bone resection (STBR) for tumour extending beyond tympanic membrane. The need for more extensive surgery should be discussed individually with the patient as it carries higher morbidity. Postoperative radiotherapy with or without chemotherapy is generally used as adjuvant in patients with extensive disease. Because of this bias, it is difficult to assess the effectiveness of these modalities on its own.[1] Nakagawa et al suggested the use of preoperative chemoradiotherapy to facilitate a complete resection.[10] Poor prognostic factors include presence of regional lymphadenopathy, histologically squamous cell carcinoma, moderate to severe facial nerve paralysis, and positive surgical margins. Overall 5-year survival for T1 & T2 ranges from 50% to 100%, T3 46% to 80%, and T4 13 to 38%.[1-3,10-11]

Despite of the presence of several poor prognostic factors, the presented case has been disease-free for 5 years after the radical radiotherapy. It is possible that the undifferentiated carcinoma discovered is a primary tumour of the temporal bone and more sensitive to radiotherapy effect compared to squamous cell carcinoma, similar to other head and neck undifferentiated carcinomas. However, further analysis is difficult because of its rarity. Authors would also like to suggest that if clinical examination and imaging study did not show any possibility of primary lesion in nasopharynx, blind biopsy may not be necessary. In conclusion, Primary undifferentiated carcinoma of the temporal bone is extremely rare. It may be a new entity on its own and responds well to radiotherapy as primary treatment modality.

Author Contribution:
Conception and design: Wan Rusydi
Collection and assembly of data: Goh Bee See
Analysis and interpretation of the data: Wan Rusydi
Drafting of the article: Wan Rusydi
Critical revision of the article for important intellectual content: Goh Bee See
Final approval of the article: Goh Bee See
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Corresponding author Email: irenegbs@yahoo.com
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