CASE REPORT

Temporal Bone Carcinoma: A Case Report

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SUMMARY

Temporal bone carcinoma may masquerade as an infective process causing late diagnosis. A delay in treatment as a result of missed diagnosis would carry a poor prognosis as the disease progresses to an advanced stage. We present a lady with history of chronic otorrhea, who developed left sided otalgia associated with hearing loss in her sixth decade. She underwent surgery which revealed left mastoiditis and cholesteatoma. After a year, she had a mass in her left ear and pus discharge which was initially treated as an infection. The biopsy of the mass was proven to be squamous cell carcinoma. High index of suspicion is necessary when encountering patients presenting with a mass in the ear canal with prior history of chronic otorrhea or cholesteatoma. Proper tissue biopsy is crucial. Early referral to tertiary centre is required for further management of the patient.

KEY WORDS:

temporal bone carcinoma, temporal bone resection, palliative radiotherapy, cancer of external auditory canal, mastoiditis

INTRODUCTION

Temporal bone carcinoma is rare, occurring about 0.2% of all cancer of the head and neck[1]. Histologically it can arise from epithelial, mesenchymal or salivary gland. Eighty-six percent of these tumours are squamous cell carcinoma(SCC)[2]. Less frequent types are adenocarcinoma, adenoid cystic carcinoma, mucoepidermoid carcinoma, basal cell carcinoma, ceruminous carcinoma, rhabdomyosarcoma. Chronic otorrhea and inflammation or cholesteatoma within external auditory canal(EAC) and middle ear are associated risk factors for the development of SCC[1].

CASE REPORT

A 62-year-old Indonesian lady of no medical illness presented with a growth in her left ear canal. History went back 20 years ago when she had intermittent left ear otorrhea. Other than itchiness secondary to seafood consumption, she had no other associated ear or nasal symptoms. Her ear discharge would normally resolve without medications. However the problem started when she developed left otalgia about 2 years ago, associated with scanty otorrhea and hearing loss. She underwent left modified radical mastoidectomy at a private centre where intraoperative findings and histopathology revealed left mastoiditis and cholesteatoma. Postoperatively the otalgia subsided but the hearing loss persisted. The left otalgia and otorrhea recurred about a year later. On her subsequent visit to the private centre, a mass was noted in the ear canal. Biopsy of the mass which occupied the posterior part of EAC showed granulation with no malignancy. The patient declined the advice for left mastoid exploration. Her symptoms of otalgia and hearing loss worsened and there was pus discharge coming out from the postauricular scar for 2 months. She also had intermittent fever for a week. No tinnitus, vertigo or symptoms suggestive of raised intracranial pressure were elicited. High resolution computed tomography(HRCT) of temporal bone showed left mastoid mass with extension into the middle and posterior fossa. A referral was made to our centre for further management.

On examination, the medium sized lady was alert, comfortable, afebrile and non toxic looking. A small fistula was seen at the left postauricular area. The overlying skin was not inflamed. The pinna was normal and non tender. The external ear meatus was adequately enlarged. A whitish mass was seen in the bony part of EAC extending into the mastoid cavity and completely occluding the tympanic membrane. It was a solid mass, unpeelable and mildly tender. There was scanty pus, not foul smelling. The right ear was normal. No cervical lymph nodes were palpable. Tuning fork test was suggestive of left conductive hearing loss. The facial nerve and other cranial nerves were intact. Biopsy was taken from the EAC mass at the clinic. The histopathology showed hyperkeratosis and parakeratosis, but no malignancy.

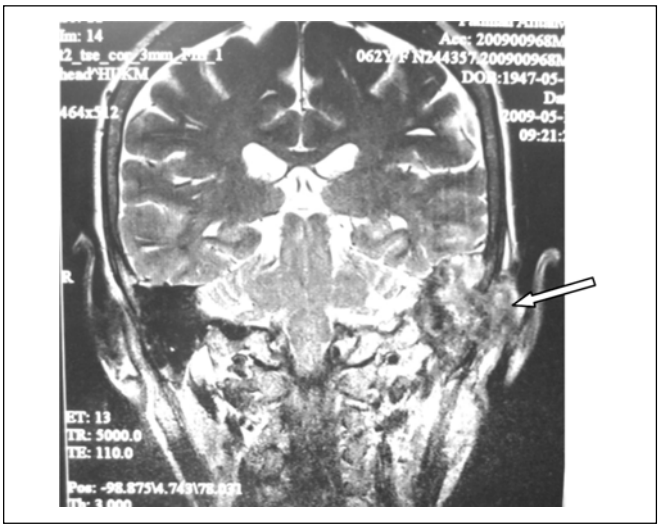

Fig. 1: MRI coronal view of the temporal bone showing mass from the left mastoid area extending out to involve external ear canal and skin.

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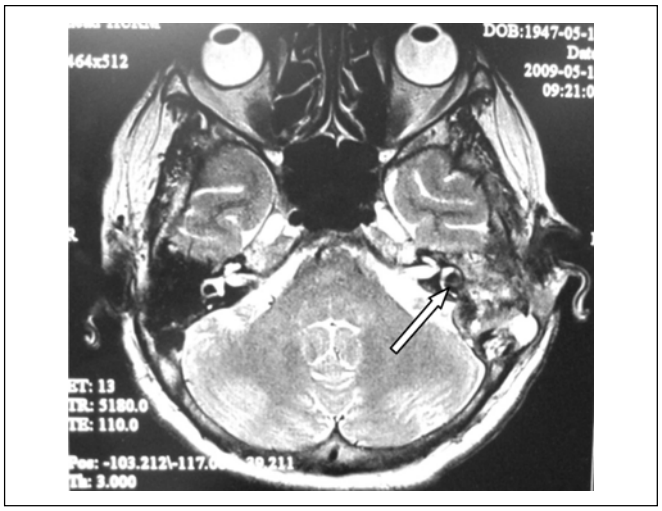

Fig. 2a: MRI axial view of the temporal bone showing tumour eroding left lateral semicircular canal.

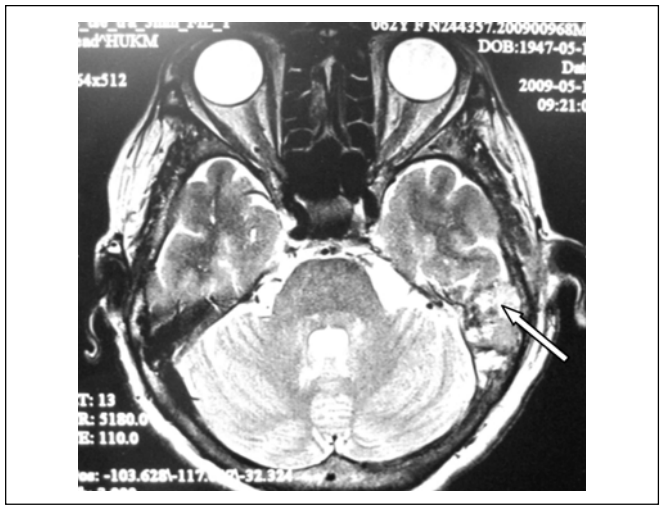

Fig. 2b: MRI axial view showing the left temporal lobe involvement and cerebellar compression.

Blood investigations showed normal total white count and fasting sugar. The erythrocyte sedimentation rate was raised. Pure tone audiogram confirmed left conductive hearing loss (moderate to severe), right moderate hearing loss. Pus from the left EAC cultured Streptococcus milleri. She was treated conservatively with IV Rocephine 1gm daily and analgesics. In the ward, no fever was documented, her conscious level remained the same. The fistula became bigger and exposed the mass within the mastoid. After reviewing again the HRCT and with a clinical suspicion of malignancy, we proceeded with a second biopsy which was two weeks after the first biopsy. It was taken from floor of the EAC and mastoid cavity. The histopathology report revealed well differentiated squamous cell carcinoma. Magnetic resonance imaging showed the heterogenous lobulated mass occupying the left EAC and mastoid, eroded the lateral semicircular canal and the posterior mastoid bone(Figure 1 and 2a). It compressed the cerebellum and extended into the left sigmoid sinus and parts of the left transverse sinus. No clear delineation between the left inferior temporal lobe and the lesion seen (Figure 2b). There was no abscess formation. A final diagnosis of well differentiated SCC of temporal bone $T_4N_0M_0$ was made. The patient was not keen for surgery, she was planned for radiotherapy. Unfortunately she defaulted radiotherapy and lived only for another 4 months.

DISCUSSION

The most common presentations of temporal bone carcinoma are otalgia, bloody otorrhea, ear canal mass and hearing loss. As the mass extends into the inner ear and petrous bone, patient will develop vestibulopathy, cranial neuropathies commonly facial nerve, headache or haemorrhage. The malignancy may continue to spread into the middle and posterior fossa. Extension into petroclival region or cavernous sinus carries a grim prognosis.

Diagnosis may not be readily evident as otologic inflammation due to tumour can masquerade as infection. Hence missed diagnosis can happen causing treatment delay as what possibly occurred to our patient. It should be noted that conditions such as chronic otorrhea or inflammation and cholesteatoma may lead to squamous metaplasia. These as well as human papilloma virus have been implicated as etiologic factors for temporal bone cancers[1]. Tissue biopsy is important in confirming the diagnosis, which sometimes may not be representative as demonstrated in this case. Therefore it is advisable to repeat a biopsy if clinically the mass is highly suspicious of malignancy. The delay in biopsy may only result in progression of disease.

High resolution CT provides detail of invasion into bony structures but cannot distinguish mucosal inflammation from tumour. This is best seen in MRI which could also show the evidence of tumour without bony erosion, tumour with dural and brain involvement. Therefore both CT and MRI have importance in staging the disease and establishing the best surgical plan. PET CT would be useful in distinguishing scar from neoplasm in previously operated area to identify tumour recurrence.

Staging follows the Pittsburgh classification for SCC of the external auditory canal (Table I). Involvement of lymph nodes automatically put patient in advanced stage 3 (T1,N1) and stage 4 (T2,3,4,N1). It has been identified as the strongest indicator for aggressive disease[3]. Distant metastasis similarly places patient in stage 4 disease[2]. Poorly differentiated SCC, brain involvement and salvage surgery are associated with poor outcome.[4]

Depending on the extent of disease, the surgical management would be lateral temporal bone resection(LTBR), subtotal temporal bone resection and total temporal bone resection. Some centres perform modified LTBR(resection lateral to tympanic membrane)[5]. T1 lesions successfully treated with surgery alone have a 95% 5-year survival. T2 and T3 lesions treated with complete surgical removal followed by radiation have 85% 5-year survival. More advanced lesions have 5-year survival of less than 50%[2]. Total resection for T3 and T4 tumour is rarely performed as it carries high morbidity level and lacking in survival benefit. Postoperative radiation is advocated for T2 and T3 lesions, as well as T1 with positive margins. Moffat feels that sleeve resections of EAC is inadequate, so is radical mastoidectomy with postoperative radiotherapy for primary surgery of middle ear or mastoid SCC[3]. Extended modified

Table I: University of Pittsburgh staging system for cancer of the external ear canal[2]

T1	Tumour limited to the EAC without bone erosion; no soft tissue extension
T2	Tumour with limited EAC canal bone erosion (not full thickness) or limited (<0.5cm) soft tissue involvement
T3	Full-thickness erosion of the bony canal with limited (<0.5cm) soft tissue involvement, or tumour involving middle ear/mastoid or facial paralysis
T4	Erosion of the medial wall of the middle ear, cochlea, petrous apex, carotid canal, jugular foramen, or dura or extensive (>0.5cm) soft tissue involvement
Stage 1	T1N0
Stage 2	T2N0
Stage 3	T3N0, T1N1
Stage 4	T4N0, T2–4N1, any M

radical mastoidectomy was performed as palliative procedures for T4N0 disease due to patients unfit for an extended resection[4]. Palliative radiotherapy is given for more advanced cases such as in our patient where the tumour involving the dura and temporal lobe has poor prognosis.

In conclusion, temporal bone carcinoma is rare. Tumour may masquerade as infection. High index of suspicion is required especially when patient has history of chronic otorrhea or cholesteatoma. Missed diagnosis will delay treatment and allow progression of disease. Early diagnosis carries better prognosis for patients.

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