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Case Report

Squamous cell carcinoma of tympanomastoid region – A rare and often misdiagnosed entity

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ABSTRACT

Squamous cell carcinoma of the temporal bone and external auditory canal is a rare entity. We present a case of 62 year old male, with stage IV squamous cell carcinoma of tympanomastoid region. As the tumour was deemed unresectable, mass debulking was done and patient was given post-operative radiotherapy. The objective to report this case was to emphasise the importance of early diagnosis and prompt treatment for a better survival. A high index of suspicion is necessary as the initial presentation may mimic more common otologic conditions.

Keywords: Carcinoma, Temporal bone, Unresectability

INTRODUCTION

Neoplasms of the tympanomastoid region are a rare entity. Incidence of temporal bone malignancy is 1-6 / 1000,000 cases. Peak age of presentation is 60 years. The most common type of temporal bone malignancies is squamous cell carcinoma.[1] The other histologic types of neoplasms arising in the region are adenocarcinoma, adenoid cystic carcinoma, basal cell carcinoma, mucoepidermoid carcinoma, ceruminous carcinoma, and rhabdomyosarcoma.

Temporal bone neoplasm is often a misdiagnosed disease and symptomatically mimics chronic suppurative otitis media (CSOM), which makes it a more difficult to diagnose and treat entity. CSOM is associated with squamous cell carcinoma (SCC) of temporal bone in 60-70% of cases. Tumours spread through deficiencies in cartilaginous portions of external auditory canal (EAC) namely fissures of Santorini and through foramen of Huschke. Improperly done mastoidectomy can break down the bony barriers and leads to tumour dissemination.

The most common presenting symptom is chronic ear discharge seen in 80% of the patients, 72% patients present with hearing loss and 32% with facial nerve palsy. Others present with tinnitus, headache, vertigo and bleeding mass in external auditory canal.[2]

Important investigations include high-resolution computed tomography (HRCT) of the temporal bone and magnetic resonance imaging (MRI) of the head and neck. Angiography is required in selected cases, to delineate the vessels supplying the tumour.

The temporal bone malignancies are notorious for easy intracranial spread due to their close proximity with skull base and their aggressive behaviour which rapidly turns the tumour unresectable, thus emphasising the importance of early diagnosis and prompt treatment.

CASE REPORT

A 62 year old male presented to the Department of Otorhinolaryngology with chief complaints of discharge

from left ear for past 3-4 months. The discharge was serosanguinous initially and was associated with decreased hearing. The patient also complained of swelling in postaural region for 1 month. There was no history of fever, facial weakness, tinnitus, vertigo, headache or ear pain. On examination, the external auditory canal was filled with a pinkish, firm, fragile mass, which bled on touch (Figure 1). The post aural swelling was firm with some fluctuant areas, with hyperaemia, warmth and tenderness. The patient was clinically diagnosed as a case of complicated CSOM with post aural abscess, and incision and drainage was done as indicated. The pus was sent for microbiological culture, which did not reveal any growth. There were no palpable cervical lymph nodes.

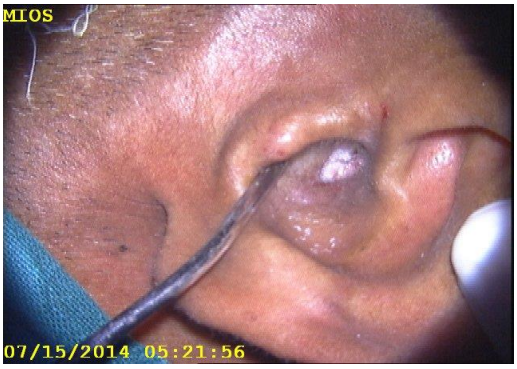

Figure 1: Showing mass filling external auditory canal.

HRCT scan temporal bone of the patient showed a soft tissue mass involving the external auditory canal, middle ear, mastoid with erosion of tegmen tympani and destruction of petrous temporal bone (Figure 2). Pure tone audiometry showed moderate conductive hearing loss in left ear.

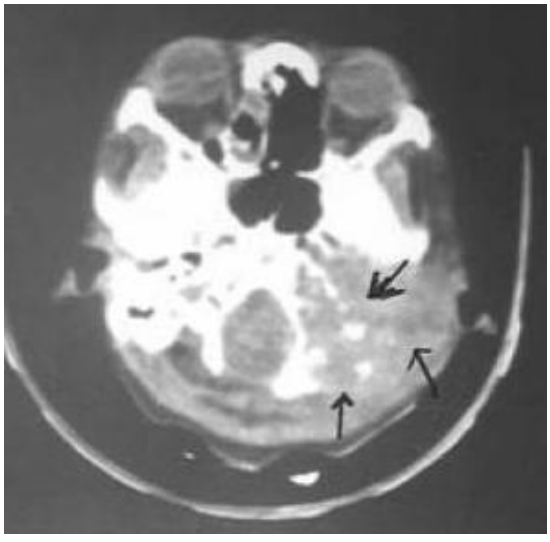

Figure 2: HRCT temporal bone showing extension of disease into middle ear, mastoid with erosion of tegmen tympani and destruction of petrous temporal bone.

The patient was posted for mastoid exploration under general anaesthesia. Intraoperatively, a pink, firm, warty growth was seen filling the external auditory canal, middle ear, destroying the mastoid bowl (Figure 3). Keeping the possibility of a neoplasm, the mass was debulked and sent for histopathological examination.

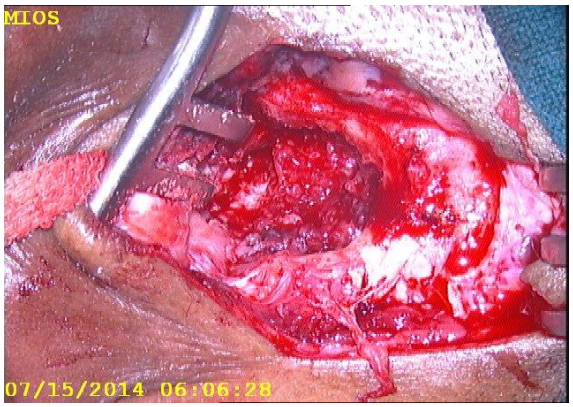

Figure 3: showing intraoperative extensions of SCC temporal bone.

The histopathology showed the mass to be well differentiated (grade I) squamous cell carcinoma, with nests of squamous cells showing dysplasia, along with keratin pearls and individual cell keratinisation.

On the basis of clinical, HRCT, intraoperative and histopathological findings, the tumour was staged as stage IV (T4N0) and the patient was sent for postoperative radiotherapy.

DISCUSSION

Tumours of tympanomastoid region are uncommon, of which the most common is squamous cell carcinoma. The other tumours of this area being adenocarcinoma, basal cell carcinoma, melanoma, rhabdomyosarcoma and metastatic disease.

The predisposing risk factors are chronic otitis media and prior exposure to irradiation for intracranial and nasopharyngeal carcinomas and also exposure to ultraviolet radiation.[1] The incidence rate is 1 in 10,00,000 in women /year as compared to 0.8 / 10,00,000 men/year.

Besides otorrhoea (often blood stained) and mass in external auditory canal, other presenting complaints may be otalgia, periauricular swelling and facial palsy.[3-5] It is aggressive locally as well as shows perineural invasion. In 10 % cases, lymph nodes are involved, and parotid nodes are first to get involved. Distant metastasis occurs to lungs, bones and liver. Involvement of lymph nodes and distant metastasis are poor prognostic indicators. HRCT of temporal bone and magnetic resonance imaging

(MRI) of head and neck are the preferred modes of investigation to look for extensions and proper staging.

Lodge *et al.* suggested that the chronic otitis might promote the development of a carcinoma in the middle ear in a manner analogous to the skin adjacent to draining sinuses resulting from chronic oseomyelitis.[6]

According to a recent study, amongst all the cases of cancer of middle ear cleft, the relative frequency of CSOM was 85%. The first case in Africa was reported in 2005 which highlights the dilemma of diagnosis because of background of CSOM in which it arises.[7] The patient presented with discharge and heaviness in left ear accompanied by discharge and swelling in postaural area. Initially, the diagnosis of CSOM atticoantral type with polyp, complicated by mastoid (zygomatic) abscess was made and incision and drainage was done. The diagnosis of this entity is mainly based on HRCT scan and histopathological examination of the tissue involved.

The rare incidence of the tumour makes it difficult to have an appropriate staging system .Till date, the University of Pittsburg staging system (modified) is the most widely accepted.[8-10]

- T1 – Tumour limited to the EAC without bony erosion or evidence of soft tissue involvement
- T2 – Tumour with limited EAC bone erosion (not full thickness) with limited (<0.5 cm) soft tissue involvement
- T3 – Tumour eroding the osseous EAC (full thickness) with limited (<0.5 cm) soft tissue involvement or tumour involving the middle ear, mastoid, or both
- T4 – Tumour eroding the cochlea, petrous apex, medial wall of the middle ear, carotid canal, or jugular foramen of dura; or with extensive soft tissue involvement (>0.5 cm), such as involvement of the temporomandibular joint or stylomastoid foramen; or with evidence of facial paresis.

Nodal and metastatic disease is staged according to the American Joint Committee System (as it is for other cancers of the head and neck).

Nodal metastasis is extremely important in temporal bone carcinoma as N1 disease in a T2 or higher lesion is considered as stage IV.

Cancer is staged as follows:

- Stage 0 – Tis N0 M0
- Stage I – T1 N0 M0
- Stage II – T2 N0 M0
- Stage III – T3 N0 M0, T1 N1 M0
- Stage IV – T4 N0 M0, T2-4 N1 M0, any T N2 M0, any T N3 M0, any T any N M1

After staging, the treatment still remains a controversy. Many authors have recommended combined therapy (surgery and radiotherapy) for advanced disease.[11-13] 5 year survival whether radiotherapy or surgery is decided as the mode of treatment is 35%, but primary surgery is faced with more morbidity than RT. Primary radiation is also reported to be ineffective as curative treatment. Surgery or radiotherapy alone is not sufficient for most of the cases of carcinoma of the middle ear because of late presentation.[14] At this juncture, it is recommended that radiotherapy has an adjuvant role to surgical clearance of disease. En bloc versus radical mastoidectomy have similar outcomes on lesions confined to the external auditory canal, but en bloc surgery offers better outcomes on middle ear extensions. Dura and temporal lobe invasion are poor prognostic indicators. The main predictor of poor outcome is positive margins.

Radiation associated tumours have got a higher propensity to recur. In such cases, the patient should be counselled in a proper way, the risks and realistic outcomes of each treatment protocol.

This case is reported owing to rare incidence of this condition found mostly in association with CSOM and therefore needs a high index of suspicion and emphasises the importance of early identification of malignancy. Prompt diagnosis and treatment may prolong survival. Recalcitrant cases of CSOM presenting with blood stained discharge and granulations within the canal for more than 3 months not relieved by usual methods should raise a suspicion of malignancy and all the tissues removed during mastoid exploration should be submitted for histopathological examination lest diagnosis of carcinoma in resistant cases of CSOM be missed.

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