*Galore International Journal of Health Sciences and Research*
*Vol.3; Issue: 2; April-June 2018*
*Website: www.gijhsr.com*
*Original Research Article* *P-ISSN: 2456-9321*

# A Prospective Study of Temporal Bone Malignancy

Dr. Abhinandan Bhattacharjee[1], Dr. Sanchita Kalita[2], Dr. Sumita Dutta Gupta[3], Dr. Devraj Dey[4], Dr. Bhaskar Jyoti Kheria[5], Dr. Bapan Debnath[5], Dr. Nizamuddin Khan[6], Dr. Surjya Sekhar[7]

[1]Assistant Professor (ENT), Silchar Medical College and Hospital (SMCH), Assam, India
[2]Senior Resident, Gauhati Medical College and Hospital, Assam, India
[3]Assistant Professor (Pathology), Silchar Medical College and Hospital, Assam, India
[4]Assistant Professor (ENT), Tezpur Medical College and Hospital, Assam, India
[5]MS ENT (Silchar Medical College and Hospital), Assam, India
[6]Registrar, Jorhat Medical College and Hospital, Assam, India
[7]MD Pathology, Silchar Medical College and Hospital, Assam India

Corresponding Author: Dr. Sanchita Kalita

## ABSTRACT

**Introduction:** Malignancies of the Temporal bone are rare and aggressive type of tumours. Incidence being 1 to 6 per million population years, it accounts to less than 0.2% of all tumors of the head and neck region.
**Materials and Methods:** A prospective observational case series study was carried out in the Department of Otolaryngology & Head and Neck Surgery for a period of three years. All the confirmed cases of Temporal bone malignancy were included in the study.
**Results:** Out of the 6 cases (3 females, 3 males), 4 were Squamous Cell Carcinoma(SCC) and 2 were Mucoepidermoid Carcinoma on the basis of Histopathological Examination. 4 cases were primary in origin, 1 case of Mucoepidermoid Carcinoma developed secondary to ipsilateral Parotid malignancy. Treatment was planned according to the stage of the disease, which included – Surgery, Radiotherapy and Chemotherapy. A delay in diagnosis, masking of the disease by infection, and patient's non-compliance to treatment affected the outcome. The patients have been regularly followed up.
**Discussion:** A review of literature showed that Malignancies of temporomastoid region is a rare and aggressive entity, constituting less than 0.2% of all head and neck tumours. SCC is the most common subtype (70%), as also observed in our study (66.67%). However, Mucoepidermoid Carcinoma of Temporomastoid region is an extremely rare histological subtype.

***Key words:*** Temporal bone, Malignancies, Squamous cell Carcinoma, Mucoepidermoid Carcinoma

## INTRODUCTION

Malignancies of the Temporal bone are rare and aggressive type of tumours. Incidence being 1 to 6 per million population years, it accounts to less than 0.2% of all tumors of the head and neck region. Histologically, it can be classified under epidermal (SCC, BCC, Melanomas), glandular (Adenocarcinoma, Adenoid cystic Carcinoma etc.), sarcoma (Rhabdomyo-sarcoma etc.) and others. About 82% of Temporal bone malignancies are SCCs, followed by BCCs (11%). Less frequent types are Adenocarcinoma (4.3%), Adenoid cystic carcinoma (3.8%), Rhabdomyo-sarcoma (1.4%). Mucoepidermoid carcinoma is classified under the extremely rare types of Temporal bone malignancies under Glandular variety. Secondary malignancies of the Temporal bone are not a common finding. Metastasis, though very rare, can be by lymphatic route, hematogenous route (from breast, lung, kidney, stomach, bronchus, prostate) or by direct spread (from parotid, temporomandibular joint, skin in the pre and post auricular region etc). [3,7,10-12,14]

Apart from UV radiation, genetics, familial association, studies are suggesting that a long standing chronically discharging ear in association with COM might play an

important etiological role in malignancies of the Temporomastoid region, especially those arising from the external auditory canal.

An early diagnosis plays a pivotal role in the outcome of the patient. Till date, there is no universally accepted staging system to classify these tumors. Amongst the ones used, AJCC and Pittsburgh's staging are popular. A Modified Pittsburgh's staging has also been formulated. [6]

## MATERIALS AND METHODS

A prospective observational case series study was carried out in the Department of Otolaryngology-Head and Neck Surgery for a period of three years. All the confirmed cases of Temporal bone malignancy were included in the study. The malignancies involving auricle of ear were excluded from the study. A written consent was obtained from the patients before undergoing any treatment. Staging system used was the Modified Pittsburgh Criteria.

## RESULTS

### Case 1

*Clinical presentation*

A 45 years old female presented with growth in the left external auditory canal(EAC), gradually increasing in size since 7 months, associated with deafness, pain, aural fullness and serosanguinous discharge, with left sided facial deviation. On examination, the growth was irregular, exophytic, bled on touch and seemed to completely fill the external auditory canal. No palpable cervical lymph node was appreciated.

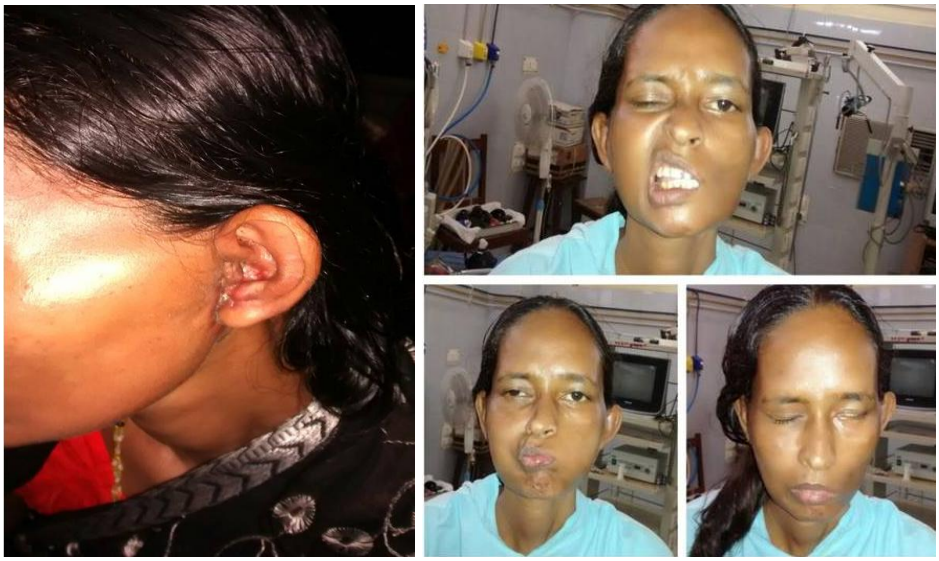

**Figure 1 & 2: Showing a protruding mass from left EAC with left sided Facial Palsy**

*Specific Investigations*

USG and FNAC from the parotid were within normal limits. HRCT scan of temporal bone showed a heterogeneously enhancing mass lesion in the left external auditory canal. There was meatal stenosis because of involvement of the cartilaginous and partial thickness bony canal. Erosion of the mastoid segment of the bony facial canal was also noted. The middle ear and mastoid did not show any mass lesion. No cervical lymphadenopathy was evident. Punch biopsy from the EAC growth showed features of mucoepidermoid carcinoma. According to Pittsburgh's staging, it was a Stage IV disease. Treatment was aimed at surgical resection of the disease from the EAC followed by Radiotherapy and Chemotherapy.

*Intraoperative findings*

Under General Anesthesia (GA), a left post auricular approach was undertaken to achieve complete resection of growth

from the EAC along with Meatoplasty. CWD MRM was done to complete wide margin clearance of the growth. Intraoperatively, the involved part of facial nerve was removed along with the tumour. The resected mass was sent for histopathological examination (HPE). Following postoperative wound healing, the patient received Radiotherapy and Chemotherapy (with Cisplatin and 5 FU).

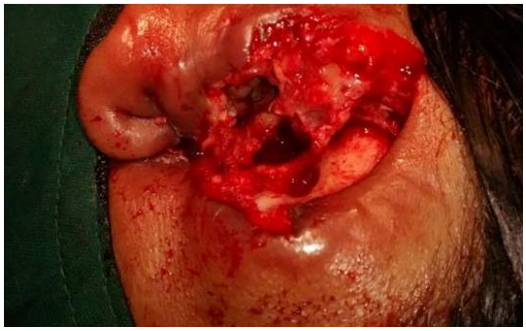

**Figure III: Showing a post auricular approach for resection of the mass from left EAC, mastoid and middle ear**

*Histopathology*

The HPE of the resected specimen showed features of Low Grade (Grade I) Mucoepidermoid Carcinoma.

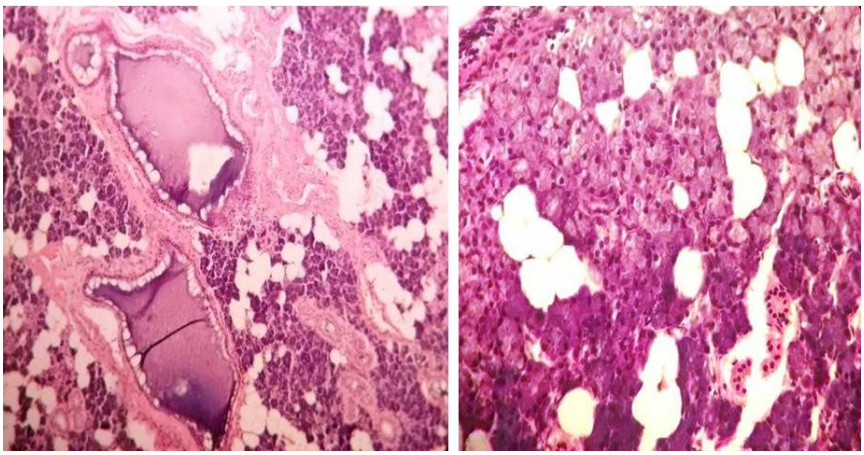

**Figure IV:** HPE showing macrocysts, microcysts, abundant mucin producing cells, lack of pleomorphism and mitosis, mucin pools, with sclerosis, suggestive of low grade (Grade I) Mucoepidermoid Carcinoma

**Case 2**

*Clinical presentation*

A 70 yrs old female, presented with left ear discharge, decreased hearing, blocked sensation and tinnitus and a growth in the left mastoid region. The discharge from the left ear was present since 6 months; it was scanty, purulent, foul smelling and associated with decreased hearing. Also, past history was suggestive of a long term Chronic Otitis Media (COM). Since 2 months, the patient had noticed sanguineous discharge with increasing sensation of ear blockage and tinnitus. On examination, an ulceroproliferative growth, around 2.5X2.5 sq cm, with irregular margins was seen in the left mastoid region; another mass was seen protruding from the left EAC, with overlying purulent discharge. No obvious facial deviation was noted.

**Figure IV: Shows a growth in the left post auricular region and a growth with purulent discharge in the left EAC**

*Specific Investigations*

FNAC from the growth in left mastoid region and left EAC growth showed features suggestive of poorly differentiated SCC. FNAC from the left level II cervical lymph node showed nonspecific reactive

lymphadenitis. HRCT scan of the temporal bones showed a heterogeneously enhancing mass lesion involving left EAC, middle ear cavity encasing the ossicles, mastoid, pre and post auricular soft tissue causing destruction of anteroinferior wall of bony EAC, anterior and lateral mastoid cortex, intrapetrous part of lateral wall of bony carotid canal and bony wall of jugular fossa. Inferiorly, the lesion has extended to the angle of mandible. Laterally, it is infiltrating the skin with loss of subcutaneous fat plane. Anteriorly, it is extending to the left Temporomandibular joint.

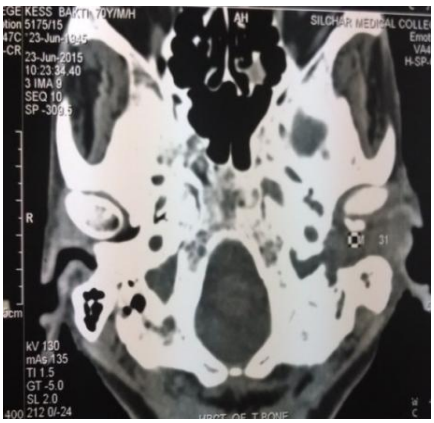


**Figure V: Shows an axial slice of HRCT scan of Temporal bone of the concerned patient with the above mentioned findings.**

Palliative Surgery was planned with the aim of achieving maximum removal of the growth.

*Intraoperative findings*

Excision of the left post auricular growth with wide margins, with Canal Wall Down (CWD) MRM via post auricular approach, Meatoplasty and a left Superficial Parotidectomy was done under GA. The growth was seen to completely fill the left mastoid cavity, extended through the aditus into the middle ear, and continuing into the EAC. Ossicles could not be appreciated because of the growth. Facial ridge was found intact. The three resected specimens of post auricular growth, intratympanic growth and parotid tissue were sent in three separate formalin vials for HPE.

*Histopathology*

Features were suggestive of well differentiated Squamous Cell Carcinoma.

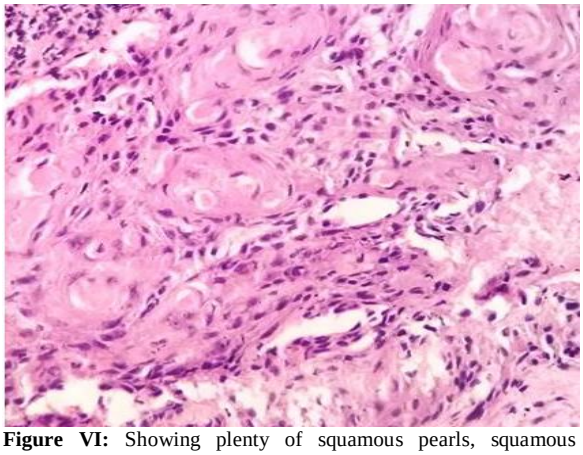

**Figure VI:** Showing plenty of squamous pearls, squamous epithelium showing loss of polarity, absent mucin and intermediate cells, suggestive of well differentiated SCC

Postoperatively, the patient has been started on Radiotherapy and Concurrent Chemotherapy.

**Case 3**

*Clinical presentation*

With a background of Chronic Otorrhoea, A 55 yrs old female, presented with exacerbated right ear discharge since 2 months; which was foul smelling, mucopurulent, yellowish, moderate in amount, severe dull aching pain in right ear radiating to neck since 2 months and left sided facial deviation since 18 months. On examination, right aural polypoidal mass was seen in right external auditory canal, punch was taken and sent for biopsy. Grade 3 right facial nerve palsy was noted.

HPE of the aural mass following punch biopsy showed features of granulomatous disease. HRCT temporal bone revealed bony erosions with soft tissue mass in the right mastoid antrum along with dehiscence of tympanic segment of Facial Canal. Audiometry showed right sided moderate to severe conductive hearing loss.

With the diagnosis of right CSOM with extracranial complication and Granulomatous lesion of right middle ear, Right MRM with facial nerve exploration and Meatoplasty were done under GA. The mastoid antrum and middle ear was filled with soft tissue mass, which was removed

and the tissue sent for HPE and PCR for Tubercular Bacilli.

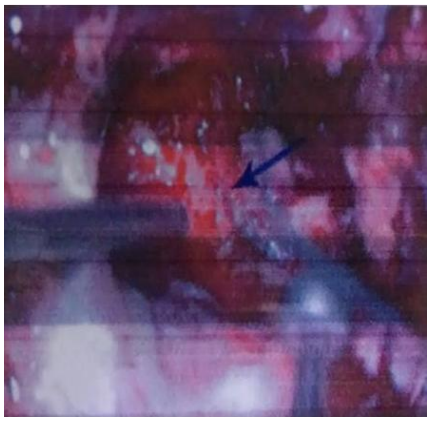

**Figure VII: Intraoperative picture of the granulomatous lesion**

HPE showed a granulomatous lesion and PCR showed detection of Tubercle bacilli. The patient was started on Antitubercular drugs (HRZE).

Post-operative follow up at 3 months showed a proliferative growth, around 5x4 sq cm, over right mastoid, which was friable, irregular and bleeded on touch.

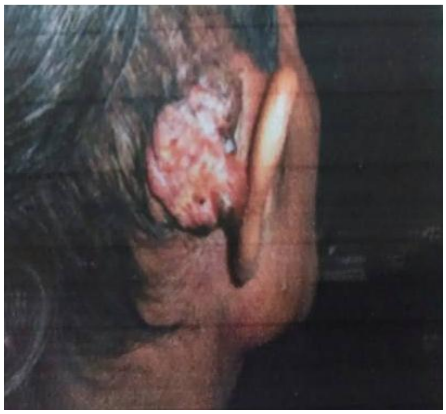

**Figure VIII: Showing a proliferative growth in the right post auricular region**

Punch biopsy from the growth showed features of well differentiated squamous cell carcinoma.

Staging could not be done as the patient party refused further investigations and surgery. The patient had received Radiotherapy and Concurrent Chemotherapy.

**Case 4**

*Clinical presentation*

An 80 yr old male, presented with a fungating growth in the right postauricular region, a protruding mass in the right EAC associated with foul smelling purulent sanguineous discharge, a swelling in the preauricular region, a swelling over the mastoid region, with right sided facial nerve palsy and trismus. All the presenting symptoms were insidious and progressed over a period of 8 months. The patient had a long history of otorrhoea (on and off). On examination, the right post auricular growth was 5x4 sq cm, was friable with irregular margins and bleeds on touch.

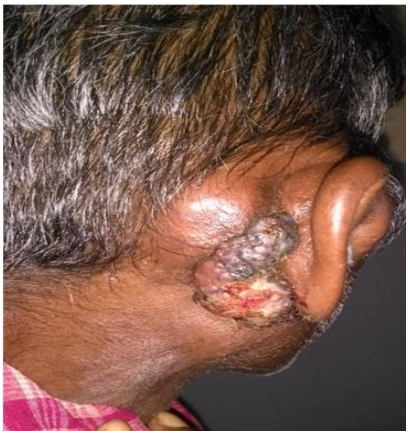

**Figure IX:** Shows two similar proliferative growth, one in the right postauricular area and the other in the right EAC with associated adjacent swellings

The EAC growth completely occluded the canal; dimensions could not be assessed clinically.

A 4x2 sq cm swelling over the right mastoid region was seen, which was non tender, hard, immobile, fixed to the underlying structures and skin. Similar swelling was seen in the preauricular region, 2.5x2 sq cm in dimension. Examination of cervical lymph nodes revealed enlarged palpable right level II, III lymph nodes.

*Specific Investigations*

FNAC from the growth and enlarged lymph node showed features of squamous cell carcinoma. CT temporal bone revealed a heterogeneously enhancing lobulated mass lesion of approximate size 9x8.5x6 cu cm, involving the right mastoid, pre and post auricular area, EAC, masticator space and parapharyngeal space on right side causing

destruction of lateral mastoid cortex, anterosuperiorly to temporal fossa, laterally infiltrating the skin, posteromedially extending to the middle ear cavity encasing the ossicles with erosion of bony lateral semicircular canal, bony facial canal, petrous temporal bone, petrous part of bony carotid canal and foramen lacerum with widening. Inferomedially, the lesion is involving the stylomandibular groove, also infiltrating the medial pterygoid muscle. There is evidence of erosion of right tympanic plate and sclerosis of greater wing of sphenoid bone. Multiple enlarged enhancing lymph nodes are noted along right jugular chain. Right parotid involvement is not noted.

Punch biopsy from the growth showed features of moderate to poorly differentiated Squamous Cell Carcinoma.

It was grouped under Stage IV disease and surgery was planned. However, the patient refused undergoing any surgical procedure. Hence Radiotherapy and Concurrent Chemotherapy was initiated.

**Case 5**

A 60 years old male presented with a gradually enlarging mass in the right external auditory canal since 1 years, extending to the postauricular region, with accompanying right serosanguinous aural discharge, decreased hearing, excruciating headache, vertigo and right facial palsy, with right cervical lymphadenopathy (level II). There was a past history of longstanding right sided otorrhoea. Punch biopsy and HPE had revealed features of Squamous Cell Carcinoma.

CECT scan of temporal bone revealed a heterogeneously enhancing lobulated soft tissue mass involving the right external auditory canal, middle ear and mastoid; with extension into the petrous and squamous temporal bone and into the temporal lobe.

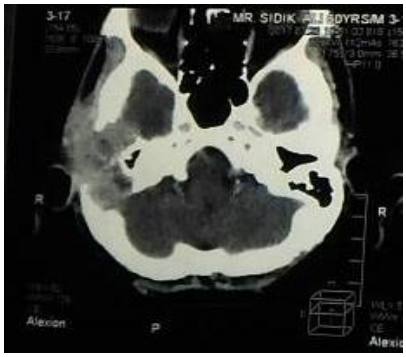

**Figure X: Axial CT scan of temporal bone, with the above mentioned description.**

It was grouped under Stage IV disease and a Lateral temporal bone resection including subtotal petrosectomy with right supraomohyoid neck dissection was planned. However the patient party refused an extensive surgery and opted for concurrent Chemoradiation, which was initiated without delay. The chemotherapy regime of Docetaxel, Cisplatin and 5 Fluorouracil has been completed along with concurrent Radiotherapy. The patient is surviving till now with improved symptoms.

**Case 6**

A 48 year old female, a known case of Mucoepidermoid carcinoma of right parotid, who had undergone Right Total Extended Radical Parotidectomy with cervical cutaneous Augmentation Flap Reconstruction and MRND type 2 nine months ago, and, who had received Radiotherapy and Concurrent Chemotherapy, presented with an ulcerative lesion involving the Right mastoid region, right ear discharge. Right facial nerve palsy was not a new presentation.

**Figure XI: Showing right sided facial palsy**

The ulcerative lesion was about 3x3 sq cm in dimension, covered with foul smelling slough and bleeded on touch. The discharge from right ear was purulent, scanty and foul smelling. EUM showed right EAC occlusion by a protruding mass. HRCT temporal bone revealed a heterogeneously enhancing mass lesion involving the right pre and post auricular soft tissue, parotid space, external ear canal, middle ear cavity encasing the ossicles, mastoid, pre and post styloid space, right parapharyngeal space causing destruction of right mastoid, sinus plate, occipital bone, scutum, tympanic bone, lateral wall of tympanic part of bony facial canal, with extension anteriorly into right TM joint, laterally to right pinna, pre and post auricular skin.

Punch biopsy of the growth from right EAC and post auricular region revealed features of Grade II Mucoepidermoid Carcinoma.

It was a secondary Mucoepidermoid Carcinoma of Temporal bone, primary being the right Parotid gland. As it was in advanced stage, management options were limited. Patient being non compliant to Surgery, Palliative Radiotherapy and Concurrent Chemotherapy was initiated.

## DISCUSSION

In our study, there were 6 cases of HPE confirmed malignancies of the Temporomastoid region. 4 cases were SCC, 2 were Mucoepidermoid Carcinoma. Of the 2 Mucoepidermoid Carcinoma cases, one was primary and the other occurred secondary to ipsilateral Parotid malignancy. 4 of the cases were females, 2 were males. All the cases were above 40 yrs. The 2 Mucoepidermoid variants belonged to relatively lower age groups; one 45 yrs and the other 48 yrs. The 4 SCC cases were above 50 yrs. All our study cases belonged to lower socioeconomic strata. Moreover, the 4 cases of SCCs had past history of long standing COM, pointing towards an etiological significance. Hence a high index of suspicion and a complete investigative work up is warranted in cases presenting with chronic otorrhoea. In our study, Pittsburgh TNM staging was used; all the cases presented in advanced stage. Two of the cases were treated by local resection and CWD MRM with Meatoplasty followed by Radiotherapy and Chemotherapy; in 1 of the cases, additional prophylactic ipsilateral superficial Parotidectomy was done. In the secondary tumour, and in the cases who refused surgery, RT and concurrent CT was given.

A review of available literature showed that malignancies of the Temporomastoid region are indeed rare. Most of the reported cases belonged to SCC variety (60 to 85%). [4,7] But only about 4 cases of Mucoepidermoid Carcinoma of Temporomastoid region have been reported in English literature. [1,8,9,16] One case has been reported in Korea. [19]

Within the limited published literature in this subject, The University of Pittsburgh TNM staging criteria has been widely accepted, followed by AJCC. [1]

No definitive treatment guidelines have been specified. Specific surgeries related to Temporomastoid region malignancy include Partial or Complete Lateral Temporal Bone Resection with/ without Petrosectomy depending on the stage of the disease and choice of the operating surgeon. A conservative approach might be adopted which includes enbloc resection of the lesion with/without Mastoidectomy followed by Radiotherapy. [2,5] Studies have suggested that patients with carcinoma that is limited to the external auditory canal have similar survival, regardless of whether mastoidectomy, lateral or subtotal temporal bone resection is performed, with or without radiotherapy. When disease extends into the middle ear, survival of patients treated with subtotal temporal bone dissection appeared to be improved over those treated with lateral temporal bone dissection or mastoidectomy. The role of adding Radiotherapy, though commonly employed, still remains unclear. [4,13,15,17,18] Lack of awareness, low socioeconomic status,

diagnostic limitations, delay in presentation, masquerading the disease process by infection, patient's uncooperation to plan of surgery, lack of universal staging and treatment guidelines, limited literature are some important factors which affected the outcome in our cases.

A comment on the survival benefit in relation to the type of malignancy, staging and treatment could not be made as the number of cases and the duration of study were limited.

## CONCLUSION

The rarity of malignancies related to the Temporomastoid region limit the literary works and make it difficult to derive a universal guideline for the diagnosis, staging, management; also limiting the study of outcome (morbidity and mortality) in relation to the staging and management. Hereby, we have attempted to contribute to the available literature by studying and following up the cases Temporomastoid region malignancies in our institution.

## ACKNOWLEDGEMENT

Special thanks to Dr. Shams Uddin, Head of the Department of Otolaryngology and Head and Neck Surgery, for his encouragement and guidance.

### *Abbreviations*

BCC=Basal Cell Carcinoma, SCC=Squamous Cell Carcinoma, COM=Chronic Otitis Media, EAC= External Auditory Canal, USG= Ultrasound, FNAC=Final Needle Aspiration Cytology, HRCT=High Resolution Computed Tomograghy, CWD=Canal Wall Down, MRM=Modified Radical Mastoidectomy, 5FU=5 Fluorouracil, PCR=Polymerase Chain Reaction, HRZE=Isoniazid Rifampicin Pyrazinamide Ethambutol, MRND=Modified Radical Neck Dissection, EUM=Examination Under Microscope, TM=Tympanic Membrane, TNM=Tumour Node Metastasis

## REFERENCES

1. Arriaga M, Curtin H, Takahashi H, Hirsch BE, Kamerer DB. Staging proposal for external auditory meatus carcinoma based on preoperative clinical examination and computed tomography findings. Ann Otol Rhinol Laryngol. 1990;99(9 Pt 1):714–72
2. Bared A, Dave SP, Garcia M, Angeli SI. Mucoepidermoid carcinoma of the external auditory canal (EAC) Acta Otolaryngol. 2007;127:280–284
3. Bridges MN, Doval M. Cutaneous squamous cell carcinoma of the external auditory canal. Dermatol Online J 2009;15:13.
4. Gidley PW. Managing malignancies of the external auditory canal. Expert Rev Anticancer Ther 2009;9:1277-82.
5. Jacques E. Gaudet, M.D., Rohan R. Walvekar, M.D.,Moises A. Arriaga, M.D. Michael D. Dileo, M.D., Daniel W. Nuss, M.D.,Anna M. Pou, M.D., Joseph Hagan, M.S.P.H.,2 and James Lin, M.D., Applicability of the Pittsburgh Staging System for Advanced Cutaneous Malignancy of the Temporal Bone, 2010, Skull Base/Volume 20, Number 6.
6. Lasisi O, Ogunleye A, Akang E. Squamous cell carcinoma of mastoid-a report of two cases. Ghana Med J 2005;39:28-32.
7. Lobo D, Llorente JL, Suarez C. Squamous cell carcinoma of the external auditory canal. Skull Base 2008;18:167-72.
8. Magliulo G, Fusconi M, Pulice G. Mucoepidermoid carcinoma of the external auditory canal: case report. Am J Otolaryngol. 2003;24:274–277.
9. Magliulo G, Ciniglio Appiani M. Mucoepidermoid carcinoma of the external auditory canal. Otolaryngol Head Neck Surg. 2010;142:624–625
10. Maneesh SG, Manjula BV, Arun BN, Balasubramanya AM. Adenoid cystic carcinoma of external auditory canal. InternetJOtorhinolaryngol2010;11:2.
11. Mette N, Aksel G. Cancer of the external auditory canal. Arch Otolaryngol Head Neck Surg 2002;128:834-7.
12. Moody SA, Hirsch BE, Myers EN. Squamous cell carcinoma of the external auditory canal: an evaluation of a staging system. Am J Otol 2000;21:582–588
13. Nakagawa T, Kumamoto Y, Natori Y, Shiratsuchi H, Toh S, Kakazu Y, *et al*. Squamous cell carcinoma of the external auditory canal and middle ear: An operation combined with preoperative chemoradiotherapy and a free surgical margin. Otol Neurotol 2006;27:242-8.

14. Permsarp I, Winai W, Songklot A, Phooripan A. Carcinoma of the external auditory canal. J Med Assoc Thai 2005;88:114-7.5.
15. Prasad S, Janecka IP. Efficacy of surgical treatment for squamous cell carcinoma of the temporal bone: a literature review. Otolaryngol Head Neck Surg 1994;110:270–280
16. Rancic D, Nihailovic D, Mijovic Z. Mucoepidermoid carcinoma of the external auditory canal: case report. Arch Oncol. 2003;11:27–29.
17. Schmerber S, Righini CH, Soriano E, Delalande C, Dumas G, Reyt E, *et al*. The outcome of treatments for carcinoma of the external auditory canal. Rev Laryngol Otol Rhinol (Bord) 2005;126:165-70.
18. Wang CC. Radiation therapy in the management of carcinoma of the external auditory canal, middle ear, or mastoid. Radiology 1975;116:713-5.
19. Yoon YH, Park HT, Kim EH, Park YH. A case of mucoepidermoid carcinoma occurring in external auditory canal. Korean J Otorhinolaryngol-Head Neck Surg. 2008;51:822–824.

How to cite this article: Bhattacharjee A, Kalita S, Gupta SD et al. A prospective study of temporal bone malignancy. Galore International Journal of Health Sciences & Research. 2018; 3(2): 60-68.

******