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Case Report

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Temporal bone carcinoma in a 25 year old patient

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ABSTRACT

A 25 year old female patient presented to us earache and ear discharge. She was diagnosed with moderately differentiated carcinoma of the temporal bone. The patient was treated with radical mastoidectomy and postoperative radiotherapy. A review of literature has been done.

Keywords: Squamous cell, Temporal bone, Carcinoma, Radiotherapy, Radical mastoidectomy

INTRODUCTION

Temporal bone malignancies are extremely rare, aggressive tumor of the head and neck accounting for less than 0.3% of all head and neck cancers.[1] Middle ear cancers are even rare and form a small fraction of the temporal bone malignancies. It mimics several common ear conditions like otitis externa, unsafe type of COM and malignant otitis externa. Late detection of the tumour and its proximity to the skull base is responsible for poor prognosis. Hence patient should be examined under microscope and tissue must be sent for biopsy if the ear does not respond to medical line of management. Surgery with postoperative radiotherapy remains the treatment of choice. The present case report highlights the need to be vigilante even in a young patient presenting with the given symptoms.

CASE REPORT

A 25 year old female patient presented to our OPD with intense otalgia since and purulent, blood tinged ear discharge since 4 months. Patient noticed decreased hearing on the right side since 2 months. She had no giddiness. She had history of habitually cleaning her ear with matchstick. On examination there was a pale polypoidal mass occupying the medial aspect of the external auditory meatus with foul smelling (fishy odour) purulent discharge obscuring the view of the tympanic membrane. The preauricular and postauricular areas were unremarkable. There was a healed central perforation in the opposite ear. CT scan revealed soft tissue opacity in the middle ear, mastoid antrum and medial aspect of external auditory meatus. There was no bony erosion of the sinodural plate, the labyrinth and cochlea. The bony external auditory canal and the ossicles appeared intact with no evidence of erosion of the temporomandibular joint. Facial canal appeared intact. Pure tone audiometry revealed bilateral moderate sensorineural hearing loss. Since the patient was young, active squamosal type of COM was our first provisional diagnosis. The patient was taken up for a canal-wall-down Mastoidectomy under general anesthesia. The extensive pale polypoidal granulation, which mimicked tubercular otitis media, aroused suspicion and was sent for frozen section. It surprisingly revealed squamous cell carcinoma. As the tumour was limited to middle ear cleft and medial EAC without any soft tissue involvement, facial palsy or lymph node enlargement, it was decided that radical mastoidectomy with a wide meatoplasty would be appropriate surgical modality of treatment. The CT

findings of absence of bony erosion were confirmed during surgery. The patient had immediate pain relief postoperatively. The histopathology report confirmed moderately differentiated squamous cell carcinoma. (Figure 1-3) Cholesteatoma sac was also seen on H and E staining. After 3 weeks the patient was sent for postoperative radiotherapy of 66 Gy in 33 fractions over 6 weeks (Figure 4).

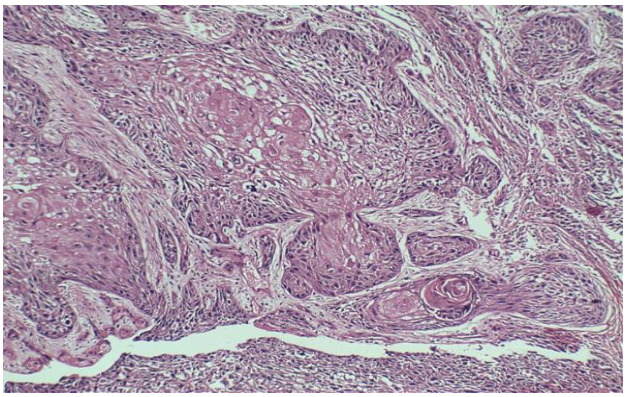

Figure 1: squamous cell carcinoma tumor cell invading the stroma (10 X, H & E).

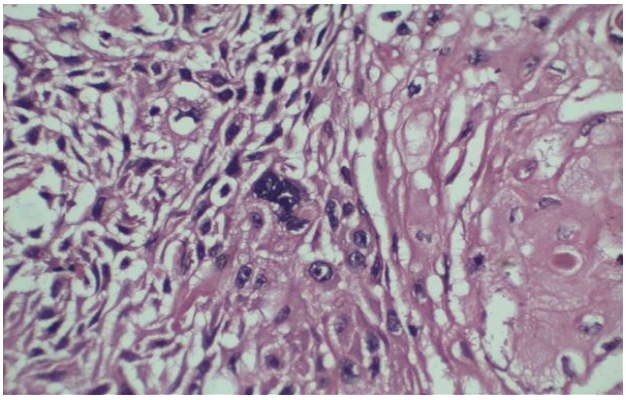

Figure 2: squamous cell carcinoma. Tumor cells showing pleomorphic nuclei and cytoplasmic keratinization (40 X, H & E).

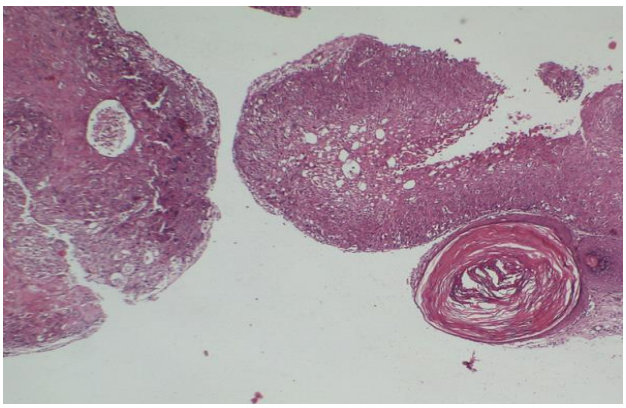

Figure 3: Tiny cholesteatoma and well differentiated squamous epithelium surrounded by tumor tissue (4 X, H & E).

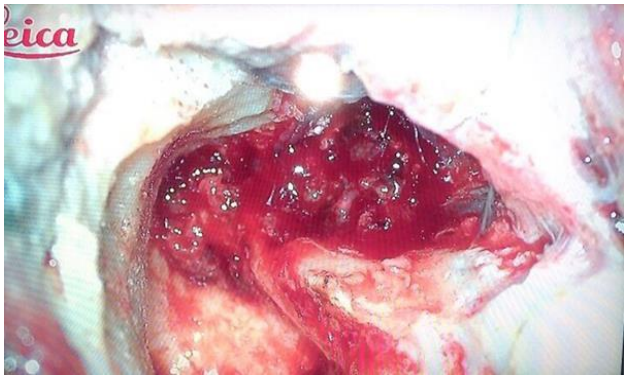

Figure 4: Granulations within middle ear cleft.

DISCUSSION

The incidence of temporal bone malignancies is 6 cases per million population.[2] Carcinoma of the middle ear constitutes 5-10% of ear neoplasms.[3] In most of the cases it is difficult to ascertain the site of origin, whether it is middle or external ear as the tumour is almost always extensive on presentation. Middle ear malignancies usually have a long standing history of ear discharge. It usually presents in the 5th to 6th decade of life with otorrhoea, otalgia, tinnitus and hearing loss. Middle ear carcinoma in a patient as young as 18 years has been reported by Deka RC whereas O.A Lasisi has reported a similar case in a 20 year old female.[4,5] Chronic otorrhoea, as seen in COM may be a major etiological factor. Presence of long standing COM with a recent change in the pattern of symptoms like obstinate otalgia is a pointer towards the unlikely disease. History of radiation exposure should be elicited. Malignancies arising from lateral aspect of auditory canal and pinna could be due to excessive exposure to sun. HPV 16 and 18 infection is known to cause changes at molecular level.[6] Infiltration into temporomandibular joint can lead to trismus. HRCT temporal bone and MRI are useful in staging the tumour and planning surgery. HRCT helps identify bone erosion and soft tissue extension whereas MRI helps identify vascular involvement (Table 1).

Table 1: Prognosticators of temporal bone malignancies.

Poor prognosticators of temporal bone malignancies
Dural invasion
Regional/distant metastasis
Recurrence following surgery or radiotherapy
Extensive soft tissue involvement (T4 tumor)
Histologic type- squamous cell carcinoma has poorer prognosis compared to adenocarcinoma
No surgical or radiotherapy intervention
Age at presentation (>70 years)
Positive margins
Petrous bone invasion
Large size

The modified Pittsburgh's classification is most popular and a comprehensive means for staging temporal bone malignancies. Our patient had a T3 tumour according to modified Pittsburgh classification. There is lack of standardized treatment protocol for squamous cell carcinoma of temporal bone. Most of the studies advocate primary surgery with adjuvant radiotherapy for advanced stage.[3,7] Surgery can range from mastoidectomy, lateral temporal bone resection, extended lateral temporal bone resection, subtotal/ total petrosectomy.[8] En bloc subtotal temporal bone resection is recommended if there is middle ear involvement. Surgery should be done to achieve negative margins all around the tumour. Nodal metastasis may occur in 10-20% of the advanced cases.[9] For stage 3 disease Kollert et al has recommended lateral or subtotal temporal bone resection with parotidectomy and neck dissection.[10] All patients who are fit for surgery should undergo surgery as it improves the 5- year survival rate drastically. Radical surgery causes significant morbidity and reduces the quality of life in terms of hearing and facial nerve function. Hearing rehabilitation with BAHA can be considered when there is enough cochlear reserve. Indications for postoperative radiotherapy are recurrence, positive margins and inadequate excision, regional and nodal spread, perineural and perivascular spread, T2-4 tumor. Adjuvant radiotherapy improves the 5- year survival rates.[11] Palliative radiotherapy is reserved for patients with poor general condition. According to Yeung et al, the 5 year case specific survival for a series of 51 patients was 54% with stage 1, 2, 3 and 4 disease being 90, 45, 40 and 19% respectively.[12] Zhang et al have reported a 5 year survival rate of 69% for T3 and 20% for T4tumors treated with piecemeal technique.[13] The overall five- year survival rate is about 61% in patients who undergo surgery and radiotherapy.[14] Concomitant chemoradiotherapy with TPF regimen (docetaxel, cisplatin, 5-fluorouracil) is also one of the modalities of treatment carrying a significant risk of bone marrow suppression.[15]

CONCLUSION

The anatomic complexities make surgical excision difficult and it is even more difficult to be sure of negative tumour margins all around the tumor. Perineural invasion of facial nerve, proximity to the carotid and skull base worsens the scenario. High index of suspicion in a non-resolving middle ear infection will help in early diagnosis and appropriate treatment of the patient though the prognosis remains poor.

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