Temporal bone malignancies are rare with the reported incidence of 1-6 in 1000000 [1-3]. Auricle is the most common site of origin of ear malignancy (60%), followed by external auditory canal (28%) and middle ear or mastoid (12%) [1]. Average age of presentation of temporal bone malignancies ranges from 48 to 64 years, although it can occur in patients of all ages [6-8]. It does not have gender predilection [1]. Squamous cell carcinoma is the most common histologic type in this region, occurring in 60% to 80% of cases [2]. Basal cell carcinoma is second followed by papilloma of middle ear [8]. Early detection becomes problematic due to simultaneous presence of chronic suppurative otitis media with or without cholesteatoma along with malignancy. Due to non specific symptoms, diagnosis of squamous cell carcinoma of mastoid antrum and middle ear is often delayed. Several patients present with nonspecific sign of chronic inflammation cause difficulty in proper diagnosis. Alterations in character or volume of ear discharge in patients with chronic suppurative otitis media should be evaluated for malignancy, especially when the discharge becomes bloody and serosanguinous [17]. The patients with chronic suppurative otitis media those who do not respond to antibiotics should be evaluated for malignancy [17]. These malignancies retain poor prognosis despite the development of radical surgery and advancement in radiotherapy [17]. Early detection can have a great impact on outcome [17]. Here we are presenting a rare and advanced case of squamous cell carcinoma of mastoid antrum and attic extending to middle cranial fossa and cavernous sinus.

CASE REPORT
A 55 years old male patient presented in ENT OPD, School of Medical Sciences and Research, Sharda Hospital, Greater Noida with history of right ear discharge on and off since childhood, headache on & off since 6 months. There was associated vertigo, hearing loss and facial paralysis of 15 days duration. The general health of the patient was preserved. On examination, there was a subtotal perforation of tympanic membrane with discharge in right external auditory canal without any mass, granulation tissue or foreign body. The patient was afebrile and there was no history of fever. The patient was afebrile and there was no history of fever. The patient was well nourished and looked healthy. The right external auditory canal was discharging and there was a mass on the posterior wall of the auditory canal. The mass was fixed to the bone. The tympanic membrane was bright red in color with a reddish grey mass on it. The mass was fixed to the upper part of the tympanic membrane. The mass was not mobile. The mass was not discharging. The mass was not friable. The mass was not painful. The mass was not tender. The mass was not tender to palpation. The mass was not tender to percussion. The mass was not tender to auscultation. The mass was not tender to palpation. The mass was not tender to percussion. The mass was not tender to auscultation. The mass was not tender to palpation. The mass was not tender to percussion. The mass was not tender to auscultation. The mass was not tender to palpation. The mass was not tender to percussion. The mass was not tender to auscultation. The mass was not tender to palpation. The mass was not tender to percussion. The mass was not tender to auscultation. The mass was not tender to palpation. The mass was not tender to percussion. The mass was not tender to auscultation. The mass was not tender to palpation. The mass was not tender to percussion. The mass was not tender to auscultation. The mass was not tender to palpation. 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Post aural examination was normal. Right sided facial palsy was present (Fig.1). Pure tone audiogram was suggestive of profound hearing loss in right ear. There were no associated nasal, visual and throat symptoms. Left ear, nose and throat appeared normal. There were no palpable lymph nodes. There were no signs of meningeal irritation. No neurological deficit was found except right facial palsy.

Computerized tomography of brain and temporal bone showed soft tissue density lesion involving mastoid air cells extending to middle cranial fossa, sphenoid sinus and cavernous sinus along with destruction of dural plate and sinus plate (Fig. 2). HRCT temporal bone, MRI and angiography were refused by the patient and his attendant.

Patient was posted for mastoid exploration under general anaesthesia. Mastoid exploration revealed a mass involving attic and antrum extending towards the middle cranial fossa with erosion of dural plate and sinus plate (Fig. 3).

Biopsy was taken from the mass and the specimen sent for histopathological examination. Histopathological examination of the specimen revealed moderately differentiated squamous cell carcinoma (Fig. 4). Patient was treated with palliative radiotherapy.

Fig. 1: Picture showing right facial paralysis

Fig. 2: Computerised tomography of temporal bone showing soft tissue density lesion in right mastoid antrum and attic extending to middle cranial fossa, sphenoid sinus and cavernous sinus
The diagnosis of squamous cell carcinoma of the temporal bone is frequently delayed because of the nonspecific nature of presenting symptoms. Squamous cell carcinoma of mastoid and middle ear is often advanced before a correct diagnosis is made. Rarity of temporal bone malignancies limits the large risk assessment studies as a result of which the aetiology of these tumours is not well defined [17]. It is frequently argued that chronic suppurative otitis media is a risk factor for temporal bone malignancy, especially squamous cell carcinoma [18-19]. Carcinoma may arise, in a manner similar to Marjolin’s ulcers, from epithelium damaged by chronic otorhoea or from bacterial toxins that can alter the normal mitotic activity of the epithelial cells [17]. Some authors have found cholesteatoma associated with temporal bone malignancy, but a causal link has not been established [3, 20].

Common signs and symptoms associated with temporal bone malignancy are ear discharge, earache, hearing loss, facial palsy, tinnitus, pruritus, headache, vertigo and aural bleeding [5, 6, 8, 21]. The early symptoms of temporal bone carcinoma are similar to chronic suppurative otitis media that include purulent foul-smelling aural discharge, severe earache, bleeding, and pruritus. Cranial nerve palsy is a threatening sign of tumour spread. Involvement of cranial nerves may result in facial paralysis, facial anaesthesia, hoarseness, visual disturbances, and dysphagia. Spread into the glenoid fossa can result in trismus, and severe pain and headache may occur due to dural involvement. Assessment for cranial nerve dysfunction, balance disturbance, trismus, and periauricular swelling can help in predicting the areas of tumour extension [17]. Lymphadenopathy is uncommon with temporal bone malignancy [5].

High-resolution computed tomographic (CT) scans show the bony anatomy of the temporal bone and demonstrate the extent of bony erosion. CT scans of the neck may also show adenopathy that is clinically undetected. Magnetic resonance imaging (MRI) complements CT scanning that provides a clearer image of tumour, soft tissue structures, dural involvement, invasion of brain parenchyma, neural structure and surrounding soft tissue planes [17].

There is no universally accepted temporal bone malignancy staging system. This lack of a uniform system makes comparison of data among studies difficult [17]. The Pittsburgh system is based on radiographic findings, has been successfully correlated with both clinical outcome and histopathologic examination [22]. The University of Cincinnati system incorporates radiographic and intraoperative findings and has been successfully used as a guide for determining the extent of temporal bone resection required [17].

Politzer in 1883 reported temporal bone carcinoma histologically for the first reported. In early 20th century, radical mastoidectomy followed by radiation was the standard treatment. Campbell and colleagues raised the possibility of temporal bone resection, but Parsons and Lewis in 1954 reported the first successful single-stage temporal bone resection. Classic operations include sleeve resection, lateral temporalbone resection, subtotal temporal bone resection, and total temporal bone resection [17].

The use of radiation for curative treatment of temporal bone malignancy has met with limited success. Zhang and colleagues reported a 5-year cure rate with radiation alone of 28.7% compared with a rate of 59.6% for patients receiving radiation and surgery [21]. Higher doses are limited by toxicity to the brainstem.

Radiation is most often used in concert with surgical resection or for palliation [17]. As a palliative therapy, radiation may have a short-lived efficacy [23]. However, as an adjuvant treatment, radiation has been
found by some authors to improve survival and reduce the incidence of local recurrence [19]. For carcinoma of the temporal bone, a wedged-pair photon radiation field is most often employed. The total dosage given is limited to 7,000 rads, with brain exposure held to 6,000 rads [17].

Fortunately, the rate of regional and distant metastases from temporal bone malignancy is low. The reported rates for the presence of nodal disease at the time of presentation range from 9 to 18% [21, 22, 24]. When nodal disease is present, the upper jugulodigastric and parotid nodes are most commonly involved. Routine radical neck dissection has not been shown to improve survival [17].

Malignancies involving the temporal bone are rare. But they are associated with considerable morbidity and mortality [17]. Early stage of diagnosis makes the prognosis better. Cure rates reduces to 40-60% when the disease extends into the middle ear and mastoid cavity and further fall to 18-25% when tumour extends beyond the temporal bone. Even with the therapeutic and diagnostic advancement, these malignancies bear poor prognosis. The prognosis usually remains poor due to late presentation which is even worse in cases with facial nerve paralysis, positive tumour margins, dural involvement, and regional lymph node involvement [17].

CONCLUSION

Chronic suppurrative otitis media is a very common disease. This case report highlights a rare advanced malignant tumour which is associated with chronic suppurrative otitis media. Early diagnosis and management are necessary to increase chances of patient’s survival in these cases. Here we are trying to highlight how closely the disease mimics chronic suppurrative otitis media. This case report also emphasizes the submission of all soft tissues removed during mastoidectomy for histopathological examination and confirmation of diagnosis. Prompt workup and evaluation of all cases of chronic suppurrative otitis media not responding to treatment are necessary. Conservative surgical approaches, combined with adjuvant therapies, may allow preservation of vital organ and cranial nerve function. The final solution for temporal bone malignancies are still to be achieved.

REFERENCES


