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Case Report

Sinister growth behind the ear: how can a person be sloppy and shoddy???

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Abstract

Malignancy of ear & temporal bone are uncommon and unusual, but if occurs belongs to destructive variety. It can have both local as well as distant spread from external ear canal to inner ear passages. The spread can involve parotid gland to petrous apex. After a prompt radiological investigation, treatment is scheduled as per staging of the disease process. Early detection with treatment will have good prognosis while delayed recognition will have poor prognosis.

Key words: temporal bone; malignancy; mastoid region; external auditory canal; petrous apex

Introduction

Malignancy of ear & temporal bone are rare, aggressive types of tumours. The incidence has been reported to be 1 in 6 cases per million population years, which is < 0.2% of all tumors of head and neck region. These tumours spread mainly by direct invasion into temporal bone and neighbouring structures (parotid gland, infratemporal fossa, petrous apex, dura and brain). Lymphatic metastasis are uncommon while distant spread is extremely rare. CT and MRI play a vital role in staging and prognosis of the disease process. Treatment is usually limited or en bloc resection with an adjunct radiotherapy modality to surgery or palliation as a curative measure. Good prognosis is achieved for T1 & T2 lesions while less or poor prognosis in later stages of the disease [2,4,6].

Case report

Here, is this adult male aged about 54 years who was encountered in the health camp. He comes with foul smelling discharge from the growth behind the left ear since > 2 months. As per history, the growth was initially size of a pea behind the ear nearly 2 years back which has gradually progressed to have attained the present size. Patient has completely neglected about the growth since the time it has appeared and with no self - hygiene. There is association of pain with this growth occasionally (as per the history from the patient) for which the patient takes analgesics (oral or intramuscular) as and when it pleases him. There was history of otalgia, hearing loss from the left ear. He also gives previous history of recurrent otitis externa taking symptomatic treatment for the same then and there. Patient also gives history of trismus. Patient who is farmer by occupation is a chronic alcoholic, smoker and tobacco/supari /ghutka chewer. There is no h/o any systemic illness.

Patient is normal built and nourished and oriented to time, place and person. His vitals and systemic examination findings are within normal limits. On local ear examination: ulcero-proliferative growth as shown in Figure 1 roughly 4X1 cm seen occupying the left post-auricular region extending vertically from upper edge of pinna upto lower end of lobule of the left ear and horizontally involving the mastoid and temporal bone partly of the left ear. This hard growth has ulcerative margins with areas of necrosis with foul smelling discharge from it. The growth has irregular margins & surface and fixed to the underlying skin. The growth is tender on palpation, sensitive to touch with bleeding from the growth with superficial skin peeling present. The surrounding area of the growth is normal in contour and surface with no induration and with no affect to hair line. Permeatal finding of left ear showed canal erosion and postero-superior retraction of the drum. Biopsy was done under local anaesthesia and later HRCT temporal bone +/- MRI was planned. But the patient did not follow up. The clinical staging in this case was T3. Biopsy report was suggestive of 2 varied diagnosis- Squamous cell carcinoma (as expected most likely) and Haemangioendothelioma (which was just out of the box anticipation).

Figure 1: *ulcero-proliferative growth seen occupying the left post-auricular region.*

Discussion

Tumors of temporal bone include skin cancer of pinna extending to temporal bone, primary tumors of external auditory canal, middle ear, mastoid or petrous apex and metastatic lesions in temporal bone. Primary malignant tumors of temporal bone have an incidence of 0.8-1/10 lakh individuals/year and 60-80% of them are squamous cell carcinoma. Metastatic lesions in temporal bone are rare and originate from breast, lung or kidney tumours [1,3]. Although they can occur at any age, temporal bone tumors are more common in 6^{th}- 7^{th} decade of life and in males. A multifactorial etiology has been suggested for these tumors in whom the risk factors include chronic otitis media, past radiotherapy in the neighbouring regions and exposure to UV radiation (actinic tumours with invasion of EAC) for tumors originating in skin of pinna and EAC, especially in fair-skinned individuals. There can be development of temporal bone carcinoma in patients who have undergone radiotherapy for carcinoma elsewhere in head and neck (i.e nasopharyngeal carcinoma as per Lim et al study). But chronic otitis media has been associated with presence of temporal bone carcinoma with no scientific evidence in etiology till date. Agents such as chlorinated disinfectants or human papillomavirus in cases of carcinomas associated with inverted papillomas have been mentioned as possible carcinogens [2].

Temporal bone tumours manifest with nonspecific symptoms such as otorrhoea, otalgia and/or hearing loss that are often attributed to inflammatory ear diseases. Thus, the ones which have superficial location, diagnosis is often delayed. Tumors of pinna and EAC are known to be more aggressive and have a higher risk of recurrence and lymph node metastasis possibly due to presence of fusion of multiple embryonic planes in this region, which may facilitate tumour dissemination. In addition to clinical examination and histo-pathological analysis, diagnostic imaging assessment of head and neck are essential for accurate tumor diagnosis and staging [4,6]. CT with contrast allows assessing bone erosion and presence of regional adenopathy, whereas MRI with contrast allows a better assessment of its extension to parotid gland, temporomandibular joint, petrous apex and intracranial invasion. In locally advanced tumors, PET allows exclusion of distant metastasis. Currently, there is no universally accepted system for staging of temporal bone carcinoma. The most commonly used is modified Pittsburgh by Moody et al. in 2000 as described below which is based on physical examination, pre-operative CT and presence of facial paralysis [5].

Modified Pittsburgh staging system for temporal bone carcinomas: T1: Tumor limited to EAC without bone erosion, or soft tissue involvement, T2: Tumor with bone erosion limited to EAC (without involving entire thickness) or limited involvement <0.5 cm of soft tissues, T3: Tumor with bone erosion throughout EAC thickness with limited involvement <0.5 cm of soft tissues, or tumor involving the middle ear/mastoid, T4: Tumor with erosion of cochlea, petrous apex, medial wall of middle ear, carotid canal, jugular foramen or dura, or large involvement >0.5 cm of soft tissues involvement of temporomandibular joint, styloid apophysis or evidence of peripheral facial paralysis [1,6].

Treatment of temporal bone tumours is a challenge for otorhinolaryngologists due to presence of significant neurovascular structures in this region. This usually includes extended tumor surgical resection, which according to its length can be a wide local excision, lateral temporal bone resection, subtotal temporal bone resection or total temporal bone resection. This surgical approach may be combined with cervical dissection with superficial or total parotidectomy and/or supplementary radiotherapy and/or chemotherapy, according to disease extent, presence of lymph node metastases, histological subtype, available resources, and surgeon's inclination [2,3,4,5].

Conclusion

Temporal bone malignancies are rare often presenting in the setting of long standing chronic otitis media and often at an advanced stage. The tissue diagnosis is relatively forthright but can be tricky sometimes. However, staging the disease is a intricate task that is best approached with consideration of triad features that is clinical, radiological and

pathological findings. The evidence based management of these uncommon tumors is not well established and as such standardisation of surgical and adjuvant treatment, as well as pathological reporting will contribute to more clear management pathways in the future.

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