Rare Case of Temporal Bone Carcinoma with Intracranial Extension

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Abstract Temporal bone cancer, a relatively rare disease, accounting for less than 0.2% of all tumors of the head and neck and is associated with a poor outcome; often presents in a subtle manner, which may delay diagnosis. It should be suspected in any case of persistent otitis media or otitis externa that fails to improve with adequate treatment. Despite advances in operative technique and postoperative care, long-term survival remains poor). It includes cancers arising from pinna that spreads to the temporal bone, primary tumors of the external auditory canal (EAC), middle ear, mastoid, petrous apex, and metastatic lesions to the temporal bone. Here is a report on a case of temporal bone carcinoma presenting with right otalgia, otorrhea and facial paralysis. The patient was initially diagnosed as mastoiditis and later the clinical impression was revised to temporal bone carcinoma (undifferentiated type), based on the pathologic findings.

Keywords Temporal bone carcinoma · Intracranial · Temporal bone surgery · Facial nerve palsy

Case Report

M.F.B.M is a 24-year-old Malay male, with no known medical illness, presented to ENT clinic early 2008 complaining of right ear persistent discharge for 5 months, the ear discharge was mucopurulent stained with blood, offensive associated with sever otalgia referred to the

temporal area with progressive reduce hearing on the right side. There was no history of tinnitus or vertigo. There was an aural polyp arising from the right middle ear, which was confirmed the tissue growth arising from the middle ear.

Patient underwent right modified radical mastoidectomy on May 2008; HPE at that time was consistent with aural polyp.

He was well till November 2008, presented with sudden onset right lower motor neuron facial nerve palsy associated with right temporal area pain radiate to the right side of the face and right ear there was no ear discharge, no tinnitus, no vertigo no worsening of the reduce hearing .

He have no history of significant nasal symptoms like (blockage, epistaxis) no neck swellings no throat symptoms, no nausea, no vomiting, no diplopia no reduce vision.

No past medical history, no known allergies, no family history of cancer, working as a dispatch boy, non smoker, non alcoholic.

On examination : vital signs were normal, alert, conscious, right facial nerve palsy grade 2 noted, post auricular scar on the right side, otoscopic examination revealed a wide EAC on the right side with a big mastoid cavity filled with a soft tissue mass occupying the mastoid as well as the mesotympanum extending superiorly as well as anteriorly, no pus seen, Rinne test was negative on the right side positive on the left side, Weber was lateralized to the right, no cervical lymph node was palpable, left ear, nose, throat examination were unremarkable.

Imaging (CT and MRI as shown in Fig. 1) showed soft tissue mass filling the right temporal bone and middle ear with bony erosion of the ossicles, anterior wall of the lateral SCC, genu and tympanic portion of the facial canal were dehiscence, the mass extend superiorly eroding the tegmen tympani to the right middle cranial fossae, inferiorly erode the inferior wall of the EAC, posteriorly erode

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Fig. 1 CT and MRI noted a soft tissue occupying the right middle ear extending to the right temporal lobe

the right pyramidal eminence, no mass seen in the nasopharynx and fossae of rosenmueller.

In view of these findings he underwent re-exploration and debulking of the tumor 2/09 HPE revealed undifferentiated carcinoma of the right temporal bone, all blood investigations, ultrasound of the abdomen, chest x-ray was all normal.

Staged as T4N0M0 received 2 cycles of chemotherapy and completed 2 phases of radiotherapy (40 G 35 fraction for the temporal area and 40G 30 fraction for the neck) on 20/7/09. still on regular follow up under ENT.

Discussion

Carcinoma of the temporal bone represents one out of 5000 to 20,000 otologic cases [1, 4], with an incidence between 1 and 6 cases per million population per year [2, 3]. Most carcinomas of the temporal bone occur in the fifth and sixth decades of life. Otorrhea is the primary symptom, and otalgia, hearing loss, and bleeding may be frequent as well.

Diagnosis is usually delayed because symptoms are quite similar to other benign otologic conditions such as chronic suppurative otitis media. These tumors have an aggressive nature and spread along preformed vascular and neural pathways, invading adjacent structures. Aggressive tumors can erode through the tegmen tympani or mastoid into the middle or posterior fossa. The sigmoid sinus may become involved. The dura, although somewhat resistant to invasion, portends a grave prognosis if involved. The facial nerve and the stylomastoid foramen are metastatic routes to the soft tissues of the neck and the parotid. Proximal extension along the facial nerve leads toward the inner ear and posterior fossa. Leonetti et al. [9] offer an excellent review of the invasion patterns of temporal bone cancer.

Nodal metastasis is uncommon in early disease but may occur in 10–20% of cases of advanced disease [6].

Treatment usually combines surgery with free margins and radiotherapy. Surgery alone may be used for very early stages (T1), whereas radiotherapy alone is rarely used. The extent of disease, positive margins, dural involvement, facial nerve paralysis, cranial nerve involvement, or moderate to severe pain on presentation have been associated with poorer outcomes [5]. Early surgical attempts to remove these tumors usually involved high morbidity and mortality. These mortality rates have substantially decreased from Lewis's 8 series to the more recent series of Moffat et al. and Yin et al. [7] as a result of advances in surgery and radiotherapy in the last three decades. The overall 5-year survival rates rank between 40 and 70%, but only reach 20% when advanced stages are considered. The main cause of death is local recurrence rather than regional or metastatic disease [8].

Conclusion

Patients with early stage tumours fared better than patients with advanced tumors, underlying the importance of early detection and prompt treatment. Facial palsy confers a poor prognosis. Metastasis to neck lymph nodes are uncommon and routine neck dissection is not recommended in patients with absent clinical neck disease. Chemotherapy with or without radiation therapy provided limited palliation in patients with advanced tumors.

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