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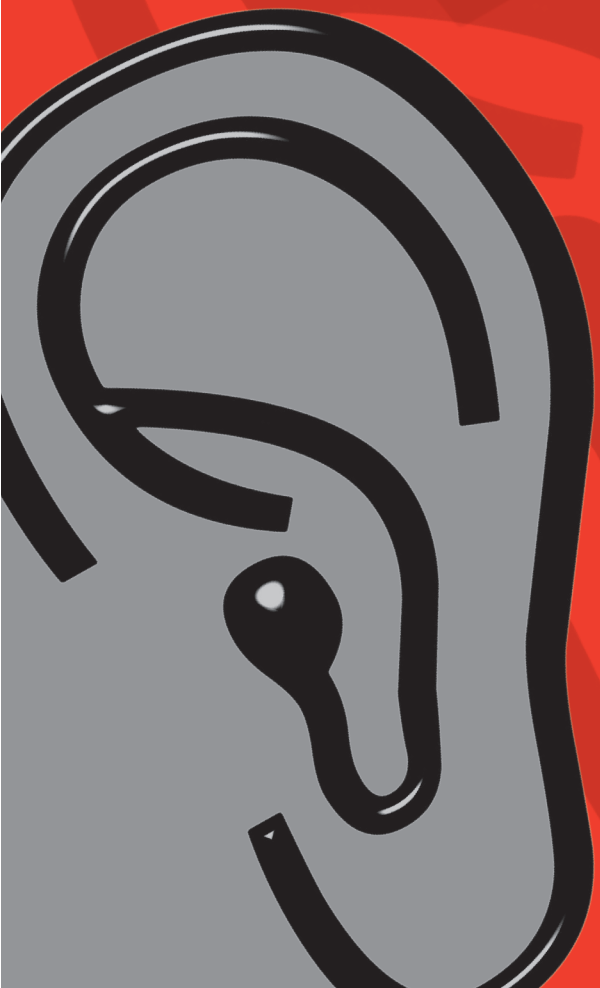

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ORIGINAL ARTICLE

Fatal unresectable malignant neoplasms of temporomastoid region – A case series

Somnath Saha, Sudipta Pal, Saibal Misra[1]

Department of ENT, Head and Neck Surgery, R G Kar Medical College and Hospital, Kolkata, [1]Department of ENT, Head and Neck Surgery, NBMC and Hospital, Siliguri, India

Abstract

Objective: To highlight the importance of early detection of the fatal and highly invasive malignant neoplasm of the temporomastoid region and to identify the criteria that signify their unresectability. We present six cases of unresectable neoplasm of the temporomastoid region, of which three cases are embryonal rhabdomyosarcoma, two cases are squamous cell carcinoma and one is a case of adenoid cystic carcinoma. **Study Design:** Retrospective case series. **Materials and Methods:** Clinical, radiographic and pathologic slides of patients with extensive temporomastoid neoplasm were studied. **Results:** Of the six cases, five patients died and only one patient is alive and is living with the disease. **Conclusion:** The neoplasms of temporomastoid region are predisposed to intracranial spread due to their close proximity to the base of skull as well as their aggressive behavior, factors which rapidly turn the tumor unresectable. Hence, it is very important for the treating surgeons to correctly diagnose these cases at an early stage and identify the unresectable neoplasm so as to avoid unnecessary surgery on these patients which only adds to their morbidity and jeopardizes the quality of life.

Keywords: Temporal bone malignancy, Temporal bone excision, Unresectability, Chemo-Radiation

Introduction

Temporal bone neoplasm is an unusual disease. Histologically, a variety of tumors affect the temporal bone, each of which is a rare entity by itself. The most common neoplasm affecting the temporal bone is acoustic neuroma, followed by glomus tumor. These neoplasms are slow growing in nature, producing few symptoms until they are well advanced. Malignancy involving this area is very unusual, and very often the symptomatology mimics chronic otitis media which delays presentation and diagnosis of the cases, making them unresectable at the time of presentation itself.

Materials and Methods

Medical records, films and pathologic reports of six patients with temporal bone neoplasm at the Department of ENT, Medical College, Kolkata, were studied. The clinical presentation, radiographic features, histopathologic findings, treatment, and outcome of all cases were recorded.

Address for correspondence: Dr. Somnath Saha, Sundaram Apartments, Barat, Lake Town, Kolkata – 700089, India. E-mail: sreekar_saha@hotmail.com

Results

The results are shown in Table 1.

Discussion

Malignancy of the temporomastoid region is a rare entity with an incidence of about 1–6 in 1,000,000.[1-6] External auditory canal (EAC) is the commonest site for these tumors if the auricle is excluded. Patients are usually in the 5th–7th decade; however, no age group is exempted.[3-5] There is usually no gender predilection. In the pediatric age group, rhabdomyosarcoma is the commonest malignant neoplasm of the tympanomastoid region and it is mostly of embryonal variety. In adults, squamous cell carcinomas constitute about 70% of all the malignancies of tympanomastoid region.[3-7] In the present series, the lowest age is 2 years and the highest is 65 years [Table 1].

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Table 1: Summary of presentation, clinical and radiological findings, histopathology, treatment and outcome of the patients included in the series

Age	Sex	Clinical features	Otoscopy	Histopathology	CT findings	T/T treatment	Outcome
27	F	Mass in (L) EAC, swelling in infra-auricular and parotid area with L-sided facial palsy	Pinkish mass with smooth surface occupying (L) EAC	Low- grade adenoid cystic carcinoma	Erosion of petrous apex with involvement of cerebellum	Incision biopsy	Alive
65	M	Foul-smelling Discharge (L) ear, headache, severe SNHL, post-auricular dis. sinus	Bleeding granular mass in (L) EAC	SCC	Involvement of petrous bone and post cranial fossa	Chemotherapy	Died before the 2nd cycle
2	M	Blood-stained Discharge from (R) ear and 7–12th cranial nerve palsy in (R) side	Fungating mass in (R) EAC	Embryonal RMS	Temporomastoid involvement, erosion of first cervical vertebra and clivus	Chemotherapy	Dead
5	M	Discharge (R) ear, 7th Crania; nerve palsy, RICT	Mass in (R) EAC	Embryonal RMS	Erosion of petrous apex with IC invasion	MRM + FND	Dead
5	F	Blood-stained discharge (R) ear and^ 3rd, 4th, 6th, 7th cranial nerve palsy PLPR-abs	Grayish bleeding mass in (R) EAC	Embryonal RMS	Heterogeneous mass in TM region with involvement of *(R) earorbit and maxilla	Supportive	Dead
28	M	Foul-smelling Discharge (L) ear (L) facial palsy fever, RICT	Mass in (L) EAC	SCC	Peripherally enhan cing lesion at (L) CP angle, erosion of petrous bone (cerebellar abscess)	MRM + FND + abscess drainage	Dead

L: Left , R: Right, EAC: External auditory canal; SCC: Squamous cell carcinoma; SNHL: Sensorineural hearing loss; RMS: Rhabdomyosarcoma; RICT: Raised intracranial tension; MRM: Modified radical mastoidectomy; FND: Facial nerve decompression; CP: Cerebellopontine

There is no well-defined risk factor described for the causation owing to the rarity of this neoplasm. However, squamous cell carcinoma has been associated with chronic otitis media in a significant number of cases (40–60%). On this basis, Whitehead argued for chronic infections as the causative factor of squamous cell carcinoma.[3] Unlike the malignancies of auricle, scalp and face where UV rays have been associated as a causative factor, no such association has been seen with malignancies of tympanomastoid region. In between 1940 and 1950, a high incidence of tympanomastoid malignancy had been found in workers of radium watch dial maker industry.

Complexity of the anatomy of the tympanomastoid region is related to the behavior exhibited by the tumors belonging to respective compartments, namely, EAC, middle ear, mastoid cavity and petrous apex. These tumors can easily spread anteriorly through deficiencies in the cartilaginous portion of the EAC, namely, fissures of santorini and through foramen of Huschke in the bony canal to involve the infratemporal fossa and the parotid area. Tumors of middle ear and mastoid cavity can also involve the internal carotid artery as it passes in its vertical course through the anterior part of the petrous bone. As the bone of the otic capsule is resistant to the tumor spread, inner ear involvement occurs mainly through the neurovascular channels.[4,5] Facial nerve can be involved either by invasion of tumor into the mastoid, destruction of the tympanic segment or by extension into the parotid gland. Improperly done mastoidectomy can also break down the bony barrier and may help in tumor dissemination.

The clinical symptoms in most of the patients mimic chronic otitis media. Otorrhea with foul-smelling serosanguinous discharge and otalgia are the commonest complaints that bring the patients to the OPD (80%). 72% patients have hearing loss and 32% present with facial nerve paralysis. Other signs and symptoms include tinnitus, headache, vertigo, bleeding and mass in the EAC.[4]

Evaluation of the exact tumor extension can be made from high-resolution computed tomography (HRCT) and magnetic resonance imaging (MRI) scan of the temporal bone.[1,5,8] CT scan gives an idea of bone erosion, whereas MRI tells about soft tissue involvement. Angiographies can be done to confirm any vessel involvement, particularly the internal carotid artery and the internal jugular vein. CT scan of neck helps to assess regional lymphadenopathy.

Staging of the temporomastoid malignancy is still a matter of controversy. No standard protocol for staging these tumors is present in the world literature though a lot of recommendations have been made.[1,5,6] The staging system proposed by the University of Pittsburgh is the one that is widely accepted and frequently used. American Joint Council for Cancer staging system is also used, but is primarily for carcinoma only. The Pittsburgh staging system describes the lesion as:

- T1 – Tumor limited to the EAC without bony erosion or evidence of soft tissue involvement
- T2 – Tumor with limited EAC bone erosion (not full thickness) with limited (<0.5 cm) soft tissue involvement
- T3 – Tumor eroding the osseous EAC (full thickness) with limited (<0.5 cm) soft tissue involvement or tumor involving the middle ear, mastoid, or both
- T4 – Tumor eroding the cochlea, petrous apex, medial wall of the middle ear, carotid canal, or jugular foramen of dura; or with extensive soft tissue involvement (>0.5 cm), such as involvement of the temporomandibular joint or stylomastoid foramen; or with evidence of facial paresis

The Pittsburgh staging system has been further modified by some authors after review of patients from an extended series. In the modified system, presence of facial nerve weakness categorizes the tumor as a T4 lesion.

Nodal involvement is classified as it is for other cancers of the head and neck:

- N1 – Single ipsilateral lymph node, size less than 3 cm
- N2 – Single ipsilateral node, size 3–6 cm
- N2b – Multiple ipsilateral nodes, all less than 6 cm
- N2c – Bilateral or contralateral nodes, all less than 6 cm
- N3 – Nodes involved greater than 6 cm

Cancer is staged as follows:

- Stage 0 – Tis N0 M0
- Stage I – T1 N0 M0
- Stage II – T2 N0 M0
- Stage III – T3 N0 M0, T1 N1 M0, T2 N1 M0, T3 N1 M0
- Stage IV – T4 N0 M0, T4 N1 M0, any T N2 M0, any T N3 M0, any T any N M1

In clinical practice, temporal bone malignancies with limited extensions, where complete tumor removal by surgery can be attempted, are a rarity. Ideal treatment is total or subtotal temporal bone resection followed by postoperative radiotherapy which has been uniformly accepted.[1,9] Planning of the treatment depends on the histological type of the neoplasm, staging of the disease, and loco-regional spread, along with distant metastasis and general condition of the patient. Subtototal or total temporal bone resection should be attempted when the tumor is limited to the confines of the temporal bone only. It is felt by most of the authors that invasion of the cavernous sinus, internal carotid artery, infratemporal fossa, paraspinous musculature and widespread extension into the brain parenchyma render these tumors surgically incurable. Regional lymph nodes can be dealt with radical neck dissection, but patients with distant metastasis are poor candidates for surgery.

A systematic review of the cases in the present series highlights the causes of inoperability of this type of cases as also the factors that hasten the demise of the patients. In this series, we encountered three children (all within 5 years of age) each of whom was suffering from embryonal rhabdomyosarcoma [Table 1]. All of them presented with mass lesion in the external ear along with otorrhea and facial nerve palsy [Table 1]. A 2-year-old boy had erosion of the petromastoid bone with extension into and erosion of the first cervical vertebra and the clivus region. Consequently, the patient had ipsilateral 12th nerve paralysis along with the 7th nerve palsy. Chemotherapy was given, but the patient succumbed after the first chemotherapy due to the adverse reactions [Table 1]. The second case of rhabdomyosarcoma had multiple cranial nerve paralysis[1,2,6,7] at the time of presentation, along with proptosis and aural symptoms. This 5-year-old girl had no vision in the affected eye. CT scan revealed a huge heterogeneous mass arising from the right petromastoid region with invasion of the right maxilla and orbit [Figure 1]. The poor general condition of the child allowed only supportive care till the girl died from Cerebro Vascular ccident [Table 1]. The third child (5-year-old boy) was initially diagnosed as a squamous type of chronic otitis media and treated with modified radical mastoidectomy along with facial nerve decompression [Table 1]. However, only when the otorrhea did not subside after surgery, a CT scan was done which revealed a mass in the temporal bone that had already eroded the petrous apex with invasion of the internal carotid artery. The first histopathological examination report was inconclusive, but the repeat biopsy proved the case as embryonal rhabdomyosarcoma. The patient had developed signs of raised intracranial tension (RICT) by the time of confirmation of diagnosis and ultimately succumbed from respiratory failure due to raised ICT. The fourth patient was a 65-year-old male patient having a post-auricular discharging sinus along with a granular mass in the left EAC [Table 1]. CT scan revealed mass lesion in the petrous apex, which had eroded the bone to spread inside the cerebellum. This, being a biopsy-proven case of squamous cell carcinoma, the patient was started with chemotherapy, but the patient died after the first chemo. The fifth patient of our series initially presented to some other institute with otorrhea, facial nerve palsy, high fever and signs of RICT [Table 1]. All these symptoms led the treating otologist to diagnose the patient as a case of chronic otitis media having a brain abscess. He underwent emergency drainage of brain abscess with mastoidectomy and facial nerve decompression. This proved to be a futile effort only adding to the misery of the patient. The discharge never subsided, fever did not respond to medications and the patient was referred to our institute, where CT scan was done followed by biopsy of granulation tissue from the postoperative cavity. Tomography showed a neoplastic mass in the left cerebellopontine (CP) angle with surrounding bone destruction and cerebellar abscess [Figure 2]. Histopathology showed it as squamous cell carcinoma. The patient died soon before starting chemotherapy. The sixth and final case of our series was unique because she is the only one alive and, that too, without receiving any definitive treatment [Table 1]. The patient was a 27-year-old female having a low-grade adenoid cystic carcinoma extending from the external ear up to the petrous apex and intracranially into the cerebellum [Figures 3 and 4]. Adenoid cystic carcinoma is a very rare tumor to affect the temporo-mastoid region and is characterized by very slow progression of the disease.[10] The patient did not have any prostrating symptoms and refused any further treatment. The patient is still alive after 15 months of diagnosis without any remarkable deterioration in her condition [Table 1]. These were the cases that were beyond the scope of any rational surgery. Our department has successfully operated on other cases of tympanomastoid carcinoma which were resectable. This review is intended to highlight the disease factors that rendered the patients inoperable. Broadly speaking, tumors extending beyond the confines of the middle ear cavity with bony breach are poor candidates for surgery. Internal carotid

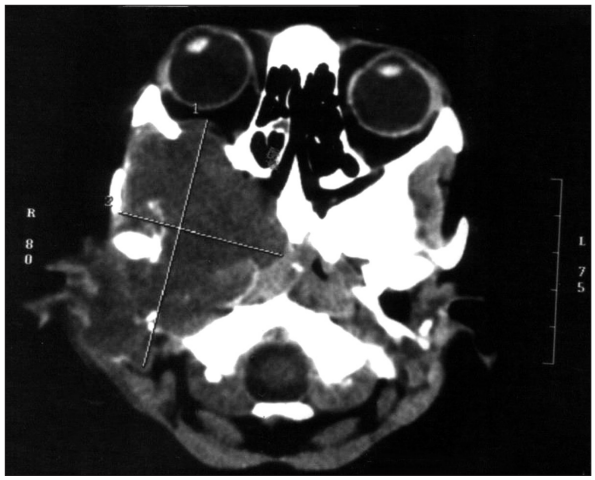

Figure 1: CT scan of a 5-year-old girl having embryonal rhabdomyosarcoma with extension into the orbit

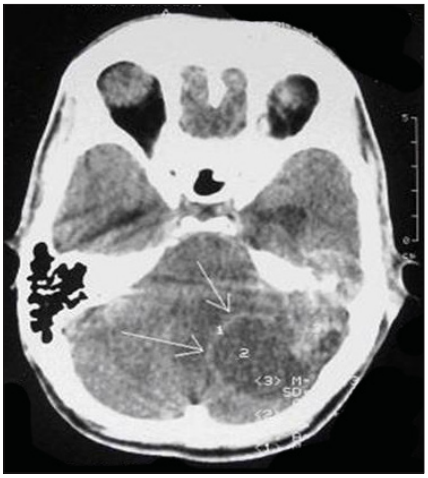

Figure 2: CT scan showing cerebellar abscess in a patient with squamous cell carcinoma of the temporomastoid region

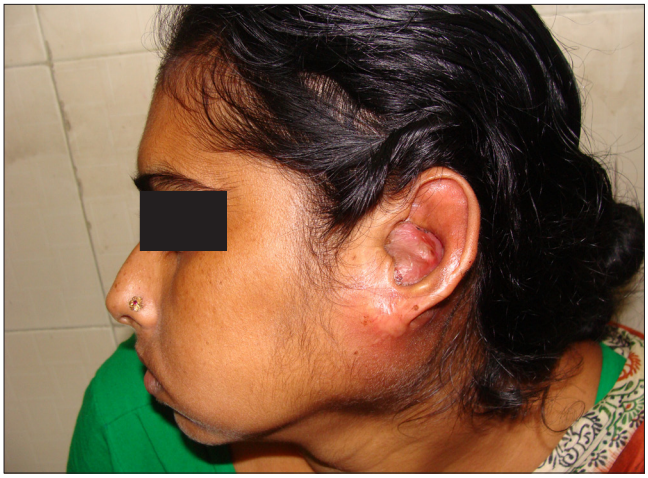

Figure 3: Clinical photograph of adenoid cystic carcinoma of temporal bone in a 27-year-old female, showing the mass in the left external auditory canal – the only surviving patient of the series

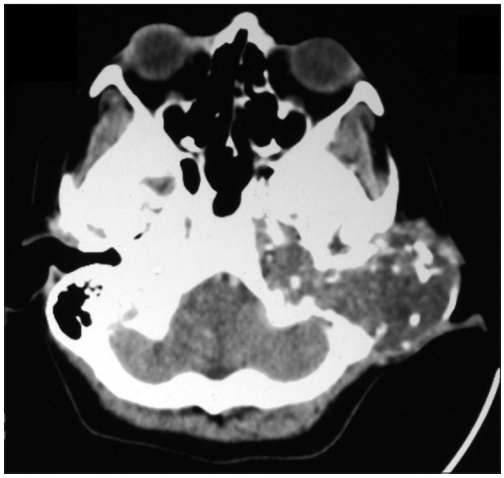

Figure 4: CT scan of the adenoid cystic carcinoma patient showing destruction of the petrous bone with involvement of the external ear

artery involvement, infratemporal spread, orbital invasion, and intracranial involvement render these tumors incurable. The 5-year survival rate for tumors extending beyond the temporal bone is 18–25% when the cure rate is taken into account.[4,11,12] However, dural involvement tends to decrease this value to less than 10%.

Radiation as a curative modality of treatment has met with very limited success. The limitations have been due to poor vascularity of the temporal bone, infected bed of tumor, low oxygen tension and toxicity of brainstem at higher doses.[9] Wedged photon therapy has been the treatment of choice with the dosage of 7000 rad (6000 rad when the brainstem is included). Radiation also has side effects, most severe of which includes osteoradionecrosis, brain necrosis, facial nerve paralysis, etc.

Conclusion

This study provided valuable insight into the progression of tympanomastoid malignancies across the entire spectrum from resectable to unresectable. Overall, the prognosis remains grim for these patients. Radiology plays a very important role in determining the operability of the lesion, with CT scan being the imaging modality of choice. CT pictures showing spread of tumor into the brain, orbit, infratemporal fossa or invasion of the carotid artery, cavernous sinus or the paraspinous musculature contraindicates any surgical treatment. Last, but not the least, any case of non-reducing otorrhea, even after mastoid exploration, should raise the suspicion of indolent tympanomastoid malignancy and proper investigative protocol should be taken up immediately.

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