Squamous Cell Carcinoma of the Temporal Bone: Diagnosis, Treatment and Prognosis

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ABSTRACT

Squamous cell carcinoma of the external ear canal is an uncommon condition that is associated with a poor outcome. The development of an accepted staging system has not been forthcoming and this has inhibited the formation of an evidence-based therapeutic protocol.

We report the findings in 14 patients with squamous cell carcinoma of the external ear canal treated in our institutions. The most common presenting symptoms were otorrhoea and otalgia. Four patients had a history of chronic ear discharge and one had previous radiotherapy for nasopharyngeal carcinoma. Five patients had facial palsy which was a poor prognostic sign. Only one patient had clinical neck disease.

Pre-operative imaging with CT or MRI scans was accurate in determining the extent of tumour involvement. The initial T-staging relied heavily on these findings.

With combination treatment involving surgery, radiotherapy and chemotherapy, disease free survival achieved was 69% (9 of 13) over a mean follow-up period of 24.7 months. One patient absconded treatment. Patients with early stage tumours faired better than patients with advanced tumours (100% vs 33%). There was low incidence of involvement of the parotid gland (1 of 7 patients). Patients with facial nerve involvement had a significantly poorer outcome (p=0.035).

Keywords: Squamous Cell Carcinoma, external ear canal, carcinoma staging system

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INTRODUCTION

Carcinoma arising from the temporal bone is uncommon. An age-adjusted incidence of 1/1,000,000 per year in women and 0.8/1,000,000 in men is reported in England, Wales and the USA. Of these, 20 to 30 per cent arise from the external auditory canal. Squamous cell carcinoma accounts for 60 to 90 per cent of all carcinomas of the temporal bone. In Singapore, the incidence of carcinoma of the external ear canal alone is 2.1 per million per year for the past ten years[1]. This is higher than the incidence quoted above which encompasses all carcinomas of the temporal bone.

Because of the relatively few numbers, the development of a reliable staging system has been difficult. Although independent researchers have proposed several staging systems, none of these have been accepted by the Union Internationale Contre le Cancer. Consequently, the absence of a uniformly accepted staging system has prevented the attempt to verify the value of different treatment protocols. All series reported are retrospective and cases are treated empirically.

The aim of this paper is to report the presenting symptoms, clinical, radiological and histological findings in fourteen patients who were treated in our centres for carcinomas of the external ear canal.

MATERIALS AND METHODS

From January 1995 to January 1999, the National University Hospital (NUH) and Tan Tock Seng Hospital (TTSH) in Singapore managed 14 cases of squamous cell carcinoma of the external auditory canal. Information was gathered from the patient clinical notes, radiological, histological and intra-operative reports. The results were recorded under the following headings; presenting symptom and duration, associated symptoms, examination findings (including facial nerve function), initial biopsy result, imaging, modality of treatment, final histology, follow-up period and current clinical status. All statistics used were performed using the Fisher's exact test. A value of $p<0.05$ was considered statistically significant.

RESULTS

Patients

Our series comprised nine males and five females with an age range of 35 to 84 years (median 57 years). The follow-up period ranged from 5 to 44 months with a mean of 24.7 months.

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Table I. Presenting Symptoms and Clinical Findings

Patient	Age/Sex	Main Presenting Symptom	Duration (months)	Associated Symptoms	Hx of Chronic Ear Discharge	Examination	Facial Palsy	Others
1	84/M	Otorrhoea	NR	Otalgia	Y	Multiple aural polyps	N	
2	56/M	Blocked ear	NR	tinnitus	NR	TM perforation, polyp	N	
3	63/F	Otorrhoea	12	Otalgia	NR	Granulations	N	
4	35/M	Otalgia	0.25	Otorrhoea	NR	Granulations	N	
5	38/F	Otorrhoea	NR		NR	Aural Polyp	N	
6	38/F	Deafness	0.5	Blocked ear	NR	EAM Mass	N	Pregnant
7	38/M	Pain upper teeth	2	Otorrhoea, headache	Y	EAM Mass	Y	
8	59/M	Otorrhoea	1	Facial Palsy	Y	TM perforation, polyp	Y	
9	82/M	Otalgia	0.5	Otorrhoea	N	EAM Mass	N	
10	57/M	Facial Palsy	0.25		N	EAM Mass	Y	
11	72/F	Otorrhoea	6	Deafness	NR	EAM Mass	N	
12	57/M	Facial Palsy	0.25	Otorrhoea, headache	Y	Granulations	Y	
13	59/M	Otorrhoea	3	Otalgia, headache Facial weakness, deafness	N	EAM Mass	Y	Previous RT
14	48/F	Blocked ear	1	Otorrhoea, Otalgia	N	Granulations, Preauricular lymph node	N	

NR - Not Recorded, EAM - External Auditory Meatus, RT - Radiotherapy

Presentation

The presenting symptoms described were otorrhoea in 10 patients (71%), otalgia in 5 (36%) and deafness in 3. Facial nerve palsy was the main presenting symptom in two although three other patients had some degree of weakness on examination. Other complaints included pain over left upper teeth, headache and a sensation of blockage in the ear. The duration of symptoms ranged from one week to 12 months although most lasted two weeks to two months. At least five patients gave a history of chronic ear discharge, defined as discharge, intermittent or persistent, of more than one year duration. The main presenting sign was an external canal mass or polyp. A summary of the presenting symptoms and clinical findings are represented in Table I.

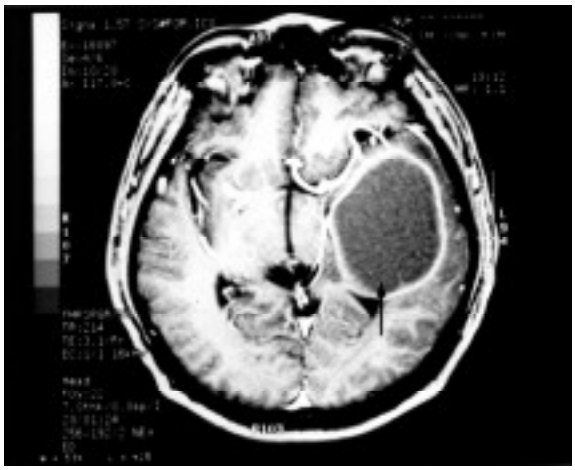

Fig. 1 The arrow indicates a large temporal lobe cyst with an enhancing capsule. There is mild midline shift.

One patient (no. 13) had previous radiotherapy to the nasopharynx for nasopharyngeal carcinoma 11 years prior to presentation for carcinoma of the ear.

One patient (no. 6) was 36 weeks pregnant at the time of presentation. In a one week period, while waiting for the biopsy results to return, the tumour had grown considerably in size. The decision was made to induce the pregnancy and perform MRI with gadolinium contrast and CT scanning soon after delivery. She underwent definitive lateral temporal bone resection four days post-partum. The oestrogen receptor (ER) and progesterone receptor (PR) status of the tumour was negative.

Patient no. 12 presented with a facial palsy and a mass in external canal extending to the middle ear. It was initially thought to be cholesteatoma. However, a CT scan revealed erosion of the anterior wall of the middle ear and a 6 cm temporal lobe cyst (Fig. 1). Biopsy of this mass revealed its true nature.

Investigations

In all patients, the diagnosis was made on biopsy of the external auditory mass under local anaesthesia. In patient 12, the initial biopsy suggested a cholesteatoma. Eight patients had pre-operative CT scan assessment of the extent of disease, four had both CT and magnetic resonance imaging (MRI) performed and two had MRI only.

We found that, in general, high resolution CT scan gave good accuracy in determining pre-operatively, the absence (Fig. 2) or presence (Figs. 3 & 4) of bony

Table II. Findings of Pre-operative Imaging

Patient	Type	External Canal Bone Erosion	Middle Ear Involvement	Parotid Gland	Neck	IJV	ICA	TMJ	Intracranial Extension	Staging
1	CT	-	+	-	na	-	-	-	-	T3
2	CT	-	-	-	na	-	-	-	-	T1
3	CT	-	+	-	-	-	-	na	-	T3
4	CT	-	-	-	na	-	-	-	-	T1
5	CT	-	-	-	-	-	-	-	-	T1
6	CT/MRI	+	-	-	na	-	-	+	-	T3
7	CT/MRI	na	+	na	na	-	+	-	+	T4
8	CT	+	+	na	na	-	-	-	-	T3
9	CT	-	-	na	na	-	-	na	-	T1
10	CT/MRI	+	+	-	-	+	-	-	+	T4
11	MRI	+	+	-	na	-	-	+	-	T3
12	CT/MRI	+	+	-	-	+	-	-	+	T4
13	MRI	+	+	na	na	+	+	+	+	T4
14	CT	+	-	na	na	-	-	-	-	T2

IJV – Internal Jugular Vein, ICA – Internal Carotid Artery, TMJ – Temporomandibular Joint, na – Not Assessed, + is present, - is absent

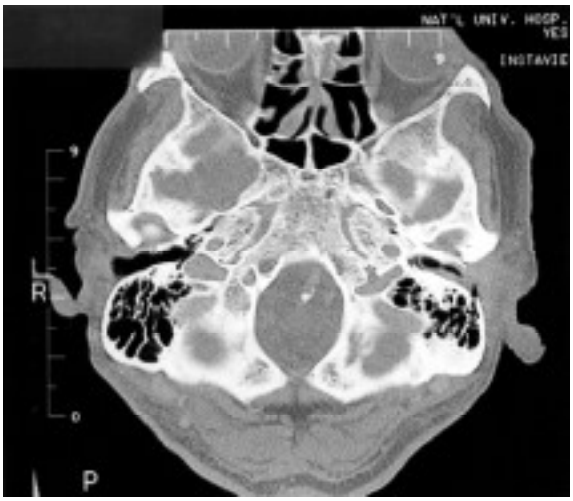

Fig. 2 Bone Windows of a CT Scan showing a soft tissue mass (arrow) confined to the external ear canal with no evidence of bony erosion.

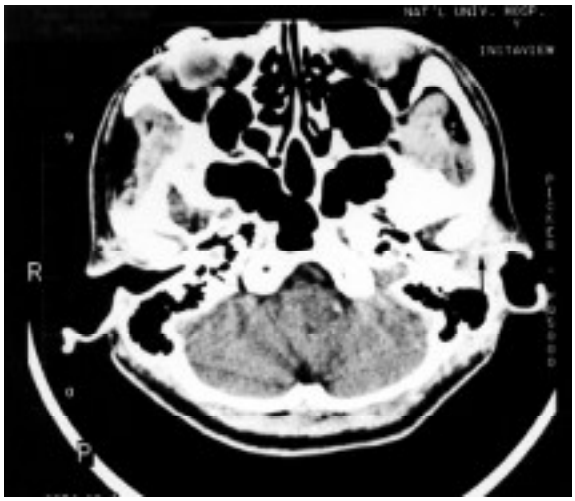

Fig. 3 Soft Tissue settings of a CT Scan revealing a soft tissue mass (arrow) extending into the temporomandibular joint.

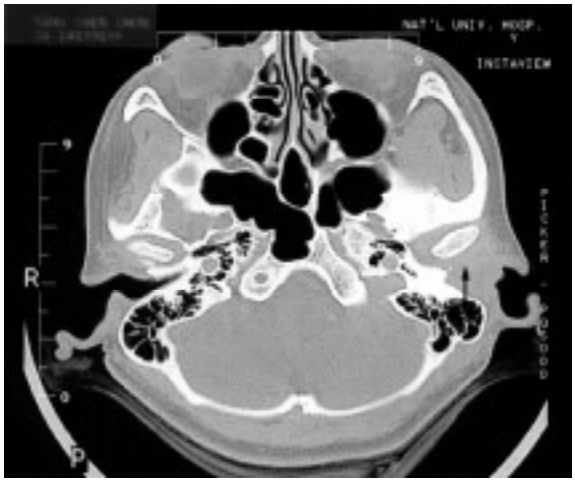

Fig. 4 Bone Windows of the same CT Scan in Fig. 3 depicting the bony erosion of the anterior canal wall (arrow) into the temporomandibular joint but sparing the head of the mandible.

erosion. There was one false positive (patient 14) where bony erosion of the anterior external canal was thought to be present but this was not demonstrated on histological assessment. In addition, there was a false negative finding (patient 3). Although CT scan did not detect any bony erosion, histologically proven bony invasion was seen in the paraffin section.

Distinguishing mucosal thickening from tumour on CT imaging was more difficult. MRI scans were more sensitive in this respect. Assessment of vessel patency, tumour encasement and middle ear involvement was readily demonstrated on MRI scans.

The initial staging of disease relied heavily on the findings of imaging techniques. Based on the T staging proposed by Arriaga et al[2], three were staged as T1, one as T2, five as T3 and four as T4. One patient had clinical neck disease. The final T-stage included the histological findings on the main specimen if surgery was performed. Table II summarises the pre-treatment imaging findings.

Table III. Comparison of Histological Findings, Treatment and Outcome

Patient	Margins Involved	Bone Erosion	Neck Disease	Parotid Gland	TMJ	Middle Ear Involvement	Staging	Treatment	FU (Months)	Status
1	NA	NA	NA	NA	NA	NA	T3	RT	21	NED
2	+	-	NR	NR	NR	NR	T1	SR + RT	22	NED
3	NR	+	NR	-	-	+	T3	LTBR + S Par + RT	44	NED
4	+	-	-	-	-	-	T1	LTBR + T Par + RT	39	NED
5	NA	NA	NA	NA	NA	NA	T1	Nil	NA	Absconded
6	-	+	NR	-	-	+	T3	LTBR + S Par + RT	12	NED
7	NA	NA	NA	NA	NA	NA	T4	Chemotherapy + RT	10	PD
8	-	+	NR	-	-	+	T3	LTBR + S Par + RT	12	NED
9	-	-	-	-	-	-	T1	LTBR + S Par	17	Died(b) NED
10	+	+	-	NR	-	+	T4	STBR + RND + RT	27	NED
11	-	+	NR	-	NR	+	T3	LTBR + S Par + RT	5	NED
12	NR	NR	NR	NR	NR	+	T4	Craniotomy Excision Skull base tumour	36	PD
13	NA	NA	NA	NA	NA	NA	T4	Chemotherapy	34	PD
14	-	-	+(a)	+	NA	-	T1	LTBR + T Par RND + RT	42	NED

NA – Not Applicable, NR – Not Recorded, RT – Radiotherapy, SR – Sleeve Resection, LTBR – Lateral Temporal Bone Resection, STBR – Subtotal Temporal Bone Resection, S Par – Superficial Parotidectomy, T Par – Total Parotidectomy, RND – Radical Neck Dissection, PD – Persistent Disease, NED – No evidence of disease

(a) - preauricular lymph node, level II node, 3 intra-parotid lymph nodes involved

(b) - Died of Tuberculosis, free of disease

Treatment

The decision on treatment was made on a case-by-case basis. Surgery was offered as the first line of treatment in 12 patients. Of these, 10 underwent surgery. In our series, three types of resections were performed and the limits of resection are described by Kuhel et al[3]. The treatment schedules are summarized in Table III. One patient had a sleeve resection, seven had lateral temporal bone resections (LTBR) and one had subtotal temporal bone resection (STBR). The last patient (patient 12) who presented with a temporal lobe cyst, had the cyst drained and excision of tumour via a craniotomy. He was offered post-operative radiotherapy but declined. Eleven months later, a repeat CT scan revealed residual disease extending to the left temporal lobe. This was treated with a course of stereotactic radiotherapy but the disease persisted. In the patient who had a subtotal temporal bone resection (patient 10), gross tumour was left behind in the operative field because the tumour extended deep and no further dissection was possible. He received planned post-operative radiotherapy and is currently free of disease 24 months into the follow-up period.

The decision to add radiotherapy after surgical resection was based on the intra-operative findings and final histological assessment of the tumour. Post-operative radiotherapy was added if:

1. tumour was deemed to have entered the middle ear cleft or
2. it was felt that microscopic tumour was left behind at operation or
3. resection margins were not free of tumour or
4. there was histological evidence of bony invasion.

In two patients (7 and 13), the tumour was considered inoperable because the internal carotid artery was encased by tumour. Both were offered combination chemotherapy and radiotherapy. One patient (no. 7) completed treatment but the tumour had only reduced to 50% of its original size. The other patient (no. 13) developed complications of septicaemic shock and acute myocardial infarction with chemotherapy and a decision to prematurely abort treatment was made. Both continue to have persistent tumour and are expected to succumb to local disease.

Histology

Surgery was performed in 10 patients. The histological extent of the tumour was assessed and the results are summarized in Table III. Of interest was the presence or absence of tumour-free margins. Three of our patients had tumour involvement at the specimen margins and were free of disease 19, 24 and 36 months in the

post-operative period. All received post-operative radiotherapy.

Seven patients had parotidectomy performed as part of the resection. In two of these, a total parotidectomy was performed. In all but one, there was no tumour involvement of the salivary parenchyma or intra-parotid lymph nodes. In patient 14, the preauricular lymph nodes were clinically palpable. The final specimen revealed metastatic disease in that node, along with three intra-parotid lymph nodes in the superficial lobe. The rest of the radical neck dissection specimen and the deep lobe of the parotid were free of metastatic disease.

Outcome

Of the 14 patients, one absconded treatment, was lost to follow up and was excluded from the final analysis. The average follow-up period is 24.7 months (5 to 44 months) with a disease free rate of 69%. The percentage of patients free of disease was higher in patients with early stage disease than those with advanced tumours (Table IV). However, this was not statistically significant (p=0.29). The presence of facial palsy was a poor prognostic sign. In patients who had no facial palsy, all remained free of disease as compared to only 40% when the facial nerve was involved (Table V). This was statistically significant (p=0.035). When treatment modalities were compared, chemotherapy with or without radiotherapy or radiotherapy alone seemed ineffective with only 33% of patients remaining free of disease as compared with patients receiving surgery and post-operative radiotherapy (Table VI). This showed borderline significance (p=0.055) However, this should be interpreted with caution as only patients with unresectable disease was considered for chemotherapy and radiotherapy.

Table IV. Comparison of T Stage and Outcome.

Stage	Number of Patients	% Free of Disease
T1 & T2	4	100
T3 & T4	9	33

p=0.29 (Fisher's Exact Test)

Table V. Facial Palsy vs Outcome.

	Number of Patients	% Free of Disease
Without Facial Palsy	8	100
With Facial Palsy	5	40

p=0.035 (Fisher's Exact Test)

Table VI. Comparison of Treatment Modalities and Outcome.

Treatment Modality	Number of Patients	% Free of Disease
Surgery + Radiotherapy	8	100
Chemotherapy + Radiotherapy	3	33

p=0.055 (Fisher's Exact Test)

DISCUSSION

Squamous cell carcinomas of the external ear canal is rare. The incidence of carcinoma of the temporal bone in England, Wales and USA is 1 per million per year for women and 0.8 per million per year for men[3]. Twenty to thirty percent of these arise from the external ear canal. In Singapore, the incidence of carcinoma of the external ear canal alone is higher, being 2.1 per million per year. Several factors might contribute to this.

1. The greater prevalence of external and middle ear diseases in the East might account in part. As seen in our series, four patients had history of chronic ear discharge.
2. The relative lack of awareness in our population of the need to treat ear disease promptly and the availability of alternative forms of treatment such as traditional Chinese medicine, lead to delay in seeking treatment.
3. Patients from neighbouring countries, where medical treatment for this condition is inadequate, come to our institutions for definitive treatment and add to the incidence.
4. The high prevalence of nasopharyngeal carcinoma is a possible contributory factor since the primary mode of treatment is radiotherapy. Irradiation has been cited as an aetiological factor[4]. Only one patient in our series had such a history.

In our group of patients, the most common presenting symptoms of squamous cell carcinomas were otorrhoea and otalgia. Other series have also reported similar clinical presentations[3,5,6]. Unfortunately, these are also the most common otological complaints with which patients present in routine clinics. This makes it difficult to sieve out patients with carcinoma of the ear from other common otological infections.

On the other hand, patients can present with unusual complaints. Patient 12 presented with ear discharge and facial palsy, which appeared on initial examination to be a cholesteatoma with intracranial and facial nerve complications. Even a biopsy of the granulations in the external ear served to reinforce this notion. It was only after a craniotomy and biopsy to drain the cerebral cyst was the diagnosis of carcinoma made. It appears that only constant suspicion of an

underlying carcinoma of any ear mass is likely to result in early diagnosis and hopefully, better long-term survival. Facial palsy, either at the time of presentation or later, is an ominous sign[7]. Tumours involving the facial nerve were generally extensive and invade other vital structures of the skull base. As seen from our series, when facial palsy was present, only 40% of the patients remained free of disease while all patients without facial palsy remained free of disease.

There are several key issues that remain controversial in the management of patients with carcinoma of the external ear.

1. The development of an accepted staging system. Several authors have proposed different staging systems[2,8] and others have made modifications to these systems[9]. The lack of an internationally accepted staging system makes comparative studies difficult.
2. The pre-operative assessment and its role in staging external ear canal tumours. Arriaga et al[2] based their staging on pre-operative CT imaging. The system is fairly precise with measurements of soft tissue involvement to 0.5cm. However, in a later paper by the same author[10], he recognises the limitations of CT scan in determining the extent of soft tissue involvement in the absence of bony erosion. Other authors[11] claim that pre-operative CT imaging has no place in the assessment of the extent of disease. He proposed that perioperative assessment is most accurate and that the extent of resection should depend solely on it. Our results support the former as we find pre-operative imaging useful in assessing the extent of disease. We further recommend that MRI scan be incorporated in the routine pre-operative imaging protocol when the extent of soft tissue involvement cannot be accurately ascertained.
3. The efficacy of temporal bone resection has yet to be proven. A meta-analysis provided by Prasad and Janecka[12] concluded that carcinoma limited to the external canal had similar survival regardless of the extent of resection (ie, radical mastoidectomy, lateral or subtotal temporal bone resection). It also concluded that patients with middle ear disease faired better if a subtotal TBR was performed compared with lateral TBR. Further prospective controlled trials need to be performed to validate these findings. Furthermore, the addition of radiotherapy as adjunctive treatment to tumours confined to the external canal appeared to confer no additional survival advantage. On the other hand, the role of radiotherapy in patients with middle ear involvement was inconclusive. However, it is generally accepted that radiotherapy as adjunctive treatment has benefit when the primary tumour has extended beyond the confines of the external ear[7].

It is interesting to note that despite the close anatomical relationship of the parotid gland to the external ear canal, only one of the parotid gland specimens in our patients showed pathologic involvement with disease. This is in contrast to other reports[13,14]. Similarly, only one patient in our series had metastatic neck disease, in contrast to other reports[11,15]. But Kinney concedes that routine neck dissection is not recommended. Instead, intra-operative sampling of upper cervical lymph nodes should determine the need for neck dissection[16].

Because of its infrequent occurrence of carcinoma of the ear, it is difficult to perform randomized controlled trials to evaluate the benefit of different modalities of treatment for this condition. At present, there are no randomized trials or even non-randomized controlled studies that can throw light on the best form of treatment[12]. This lack of good scientific evidence has prevented the formulation of an evidence-based protocol of treatment. Spector[13] compared two groups of patients treated before and after 1980. The former group of patients was treated on an individual basis while the treatment plan was formalised for the latter group based on the extent of disease on CT and MRI scanning. He showed improved survival rates with a systematic therapeutic approach based on more extensive and definitive resections and higher doses of radiotherapy.

CONCLUSIONS

1. Patients with early stage tumours faired better than patients with advanced tumours, underlying the importance of early detection and prompt treatment.
2. Facial palsy confers a poor prognosis.
3. Metastasis to neck lymph nodes are uncommon and routine neck dissection is not recommended in patients with absent clinical neck disease.
4. Even though the parotid gland is related closely to the external ear canal, none of the clinically normal parotid glands revealed microscopic invasion by tumour. Superficial parotidectomy, as part of identifying and preserving the facial nerve, may be allowed but routine resection of the deep lobe appears unnecessary.
5. Chemotherapy with or without radiation therapy provided limited palliation in patients with advanced tumours.

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REFERENCES

1. Shanmugaratnam K. Personal communication. 1999.
2. Arriaga M, Curtin, H, Takahashi H, Hirsch BE, Kamerer DB. Staging proposal for external auditory meatus carcinoma based on preoperative clinical examination and computed tomography findings. Ann Oto Rhinol Laryngol 1990; 99:714-21.
3. Kuhel WI, Hume CR, Selesnick SH. Cancer of the external auditory canal and temporal bone. Otolaryngology Clinics of North America 1996; 29:827-52.
4. Shaheen OH. Epithelial tumours of the external auditory meatus. In: Scott-Brown's Otolaryngology Otology 6th Ed. Booth JB. editor. 1997. Butterworth International Editors: 3/22/1-3/22/7.
5. Arriaga M, Hirsch BE, Kamerer DB, Myers EN. Squamous cell carcinomas of the external auditory meatus (canal). Otolaryngology Head & Neck Surgery 1989; 101(3):330-7.
6. Golding-Wood DG, Quiney RE, Cheesman AD. Carcinoma of the ear: Retrospective analysis of 61 patients. Journal of Laryngology and Otology. 1989; 103:653-6.
7. Shih L, Crabtree JA. Carcinoma of the external auditory canal: An update. Laryngoscope 1990; 100:1215-8.
8. Stell PM, McCormick MS. Carcinoma of the external auditory meatus and middle ear: Prognostic factors and suggested staging system. J Laryngol Otol 1985; 99:847-50.
9. Clark LJ, Narula, AA, Morgan DAL, Bradley PJ. Squamous carcinoma of the temporal bone: a revised staging. Journal of Laryngology and Otology 1991; 105:346-8.
10. Arriaga M, Curtin H, Takahashi H, Kamerer DB. The role of preoperative CT scans in staging external auditory meatus carcinoma: Radiologic-pathologic correlation study. Otolaryngol-Head and Neck Surgery 1991; 105(1):6-11.
11. Kinney SE. Squamous Cell Carcinoma of the External Auditory Canal. American Journal of Otology 1989; 10(2):111-6.
12. Prasad S, Janecka IP. Efficacy of surgical treatments for squamous cell carcinoma of the temporal bone: A literature review. Otolaryngology Head & Neck Surgery 1994; 110:270-80.
13. Spector JG. Management of temporal bone carcinomas: A therapeutic analysis of two groups of patients and long-term follow up. Otolaryngology Head & Neck Surgery 1991; 104:58-66.
14. Horowitz SW, Leonetti JP, Azar-Kia B, Fine M, Izquierdo R. CT and MR of temporal bone malignancies primary and secondary of parotid carcinoma. AJNR. 1993; 15:755-62.
15. Testa JRG, Fukuda Y, Luiz PK. Prognostic Factors in carcinoma of the external auditory canal. Archives of Otolaryngology Head & Neck Surgery 1997; 123:720-4.
16. Kinney SE, Wood BG. Malignancies of the external ear canal and temporal bone: Surgical techniques and results. Laryngoscope 1987; 97:158-64.