Cancer Association of South Africa (CANSA)

Research • Educate • Support

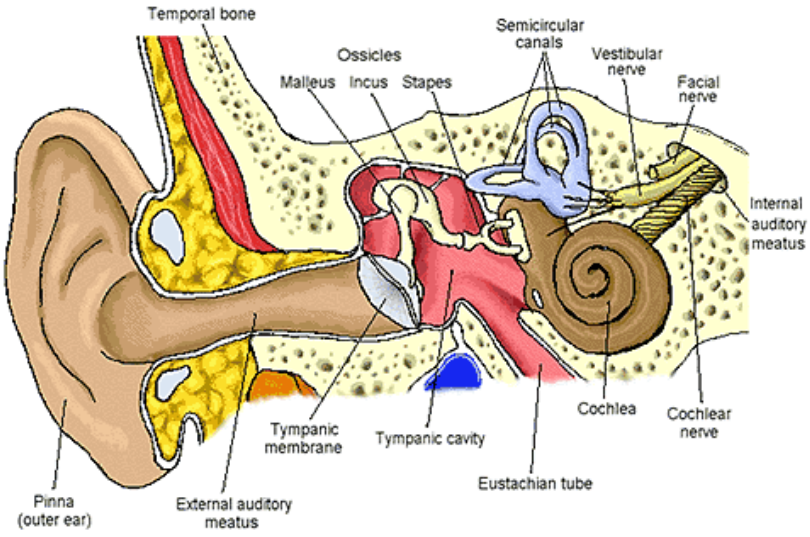
Fact Sheet
on
Cancer of the Ear

Introduction

The ear is part of the auditory system. The ear is the organ that detects sound. It not only receives sound, but also aids in balance and body position. The entire organ is often considered to be the ear, though just the visible portion of the ear may be considered to be the ear. In most mammals, the visible ear is a flap of tissue that is also called the pinna (or auricle in humans) and is plays a role in the first of many steps in hearing. Vertebrates have a pair of ears placed somewhat symmetrically on opposite sides of the head. This arrangement aids in the ability to localise sound sources.

The ear consists of three parts, namely the outer ear, the middle ear and the inner ear.

[Picture Credit: The Ear]



The outer ear - the outer ear is the most external portion of the ear. The outer ear includes the fleshy visible outer ear, called the auricle, the ear canal, and the outer layer of the tympanic membrane, also referred to as the ear drum.

The middle ear - The middle ear is an air-filled cavity behind the tympanic membrane and includes three tiny bones (ossicles): the malleus (or hammer), incus (or anvil), and stapes (or stirrup). The middle ear also connects to the upper throat via the Eustachian tube.

The inner ear - The inner ear is split anatomically into bony and membranous labyrinths. This contains the sensory organs for balance and motion, namely the vestibules of the ear (utricle and saccule), and the semicircular canals. It also contains the sensory organ for hearing, the cochlea.

Researched and Authored by Prof Michael C Herbst
[D Litt et Phil (Health Studies); D N Ed; M Art et Scien; B A Cur; Dip Occupational Health; Dip Genetic Counselling; Dip Audiometry and Noise Measurement; Diagnostic Radiographer; Medical Ethicist]
Approved by Ms Elize Joubert, Chief Executive Officer [BA Social Work (cum laude); MA Social Work]

Brant, J.A., Eliades, S.J., Chen,J., Newman, J.G. & Ruckenstein, M.J. 2017.

INTRODUCTION: Malignancy of the middle ear is a rare condition with limited data available for clinical guidance.

STUDY DESIGN: Retrospective evaluation of a large national database.

SETTING: Deidentified national cancer database.

PATIENTS: Subjects with diagnosis of malignancy of the middle ear in the National Cancer Database between 2004 and 2012.

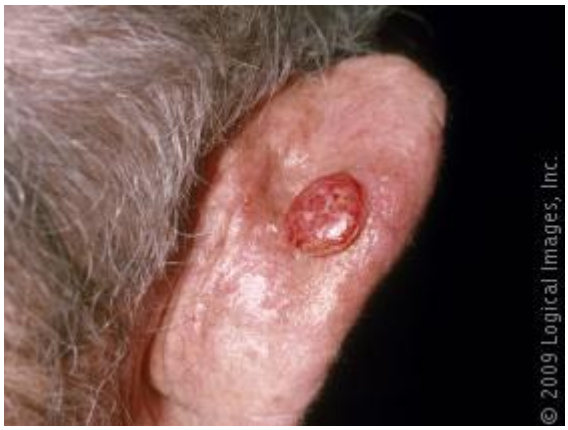
MAIN OUTCOME MEASURES: Demographic information and tumor characteristics were evaluated. The primary endpoint of interest is overall survival.

RESULTS: The most common histology was squamous cell carcinoma (SCC) (50%). Multivariable Cox proportional hazard analysis found the following variables had a significant negative impact on overall survival: age (HR 1.04 95% CI [1.02-1.05]), squamous cell carcinoma, not otherwise specified (NOS) (HR 2.08 95% CI [1.30-3.32]), squamous cell carcinoma, keratinizing, NOS (HR 4.20 95% CI [2.14-8.24]), embroynal rhabdomyosarcoma, NOS (HR 4.96 95% CI [1.17-21.11]), and unknown extension (HR 2.87 95% CI [1.22-6.74]). For patients of SCC who underwent surgery, 30 had positive margins and 29 underwent adjuvant radiation. For these, no survival advantage was found with the addition of chemotherapy, regardless of node status.

CONCLUSION: Malignancy of the middle ear is a rare condition with prognosis that depends on histology. The most common histology, SCC, is associated with the poorest overall survival. Evaluation of large national datasets can add significantly to the understanding of such uncommon tumors.

Cancer of the Ear

Cancer of the ear is a rare cancer. Most of these cancers start in the skin of the outer ear. About 5 out of 100 skin cancers develop on the ear. Those that develop inside the ear are very rare. On average, less than 1 in every million people will develop cancer in the middle ear.



[Picture Credit: Squamous Cell Carcinoma on Ear]

Most cancers of the ear are squamous cell carcinomas. Other types of cancer include:

- Basal cell cancer
- Melanoma
- Adenoid cystic carcinoma
- Adenocarcinoma
- Merkel-cell carcinoma

Sanniec, K., Harirah, M. & Thronton, J.F. 2019.

Background: The ear serves many functional and aesthetic purposes, and its complex structure presents a notable challenge for reconstruction. A paucity of objective data and analysis on reconstruction of acquired ear defects remains. The goal of this study was to evaluate all ear reconstructions and the lessons learned over the past decades in treating these complicated defects in a large clinical Mohs reconstruction practice.

Methods: A retrospective analysis of consecutive patients who underwent ear reconstruction after Mohs cancer excision from 2004 to 2018 performed by the senior author (J.F.T) was conducted. Data

regarding patient demographics, oncologic type, treatment, defect characteristics, reconstructive modalities, number of stages, and complications were collected and analyzed.
Results: Three hundred twenty-seven patients underwent ear reconstruction. Defects most commonly involved the superior one-third of the helix and the antihelix. Approximately half of the patients' defects were reconstructed with full-thickness skin grafts, and approximately one-third of the patients' defects required flap reconstruction. There were 30 complications (9 percent), ranging from partial flap loss to cancer recurrence. There was no difference in complication rates in elderly patients compared with the younger cohort.
Conclusions: Optimizing results when reconstructing ear defects is challenging, and there are multiple preoperative variables to consider. Ear reconstruction is safe in an outpatient setting, and age should not preclude patients from undergoing reconstruction of ear defects. The lessons learned from the past decade of ear reconstructions are demonstrated, and an algorithmic approach to treating these defects allows for a safe and reproducible method for reconstructing acquired ear defects.

Oliver, J.D., Boczar, D., Sisti, A., Huayllani, M.T., Restrepo, D.J., Spaulding, A.C., Gabriel, E., Bagaria, S., Rinker, B.D. & Forte, A.J. 2019.
Background: External ear melanoma (EEM) is a rare condition with controversies in the literature. We analyzed patients with EEM in the United States compared to other head and neck melanomas (OHNMs).
Methods: The National Cancer Database (NCDB) was used to select patients with head and neck melanoma from January 1, 2004 to December 31, 2015. Mann-Whitney and χ tests were used to estimate statistical significance, and multivariate logistic regression to identify independent associations adjusted for confounders.
Results: A total of 137,233 patients met the study criteria. Among them, 16,991 (12.4%) had EEM and 120,242 (87.6%) had OHNM. For patients with EEM, the mean (standard deviation) age was 66.26 (15.798) years. Most of the patients with EEM were men (85.5%), insured by Medicare (52.4%), and treated in Academic/Research Programs (47.7%) or Comprehensive Community Cancer Programs (32.3%). Most of the EEM tumors had invasive behavior (68.0%) were Stages 0 (30.3%) or I (40.3%), and were without ulceration (76.9%). Mean time to receive any treatment was 14.1 days for EEM compared with 14.6 days for OHNM ($P < 0.001$). We noticed a greater proportion of EEM in men (14.8%; adjusted odds ratio [aOR] 2.72 [2.605-2.852]; $P < 0.001$) compared to women (6.22%; reference). EEM was an independent factor for tumor Stage I (14.47%; aOR 1.61 [1.101-1.224], $P < 0.001$) and invasive behavior (13.86%; aOR 1.268 [1.15-1.389]; $P < 0.001$) compared to OHNM.
Conclusion: EEM was associated with higher odds of invasive behavior compared to OHNM. Furthermore, men were found to have a higher likelihood to develop EEM compared to women.

Incidence of Cancer of the Ear in South Africa
The outdated National Cancer Registry (2017), known for under reporting, does not provide any information regarding the incidence of cancer of the ear in South Africa.

Cause and Symptoms of Cancer of the Ear
The cause of cancers in the middle ear is unknown. People with a history of chronic ear infections have a higher risk of developing cancer in the ear. Chronic, in this instance, means for 10 years or more.

The symptoms of cancer of the ear depend on where the tumour is within the ear. Some people may also have swollen lymph nodes in their neck.

Middle ear – the most common symptom is a discharge from the ear which may be blood stained. Other symptoms include hearing loss and earache. Occasionally people cannot move the face muscles on the side of the affected ear.

Inner ear – pain including a headache, hearing loss, tinnitus (ringing or buzzing in the ear) and dizziness.

Diagnosis of Cancer of the Ear

The only way to confirm a diagnosis of cancer is to take a small amount of tissue from the abnormal area of the ear and examine it under a microscope. Doctors call this a biopsy. Before the doctor takes the biopsy he/she will give the patient a local anaesthetic to numb the area so that he/she will not have any pain. Biopsies of the middle ear can be difficult to take and the patient may need to have a general anaesthetic.

If the biopsy shows a cancer, the patient may also have an MRI scan or a CT scan to help the doctor decide which treatment is needed.

Doctors do not take biopsies of the inner ear. This is because it is very difficult to reach the inner ear without causing problems to other structures around it. The doctor will make a diagnosis using MRI scans and CT scans.

Treatment of Cancer of the Ear

The treatment one has for cancer of the ear depends on:

- Where in the ear the cancer is
- The type of cancer one has
- The size of the tumour
- Whether it has spread beyond the area it started in (the stage)
- One's general health

The main treatments for cancers that start in the ear canal or middle ear may include surgery and radiotherapy. Depending on the stage of the cancer the patient may also have chemotherapy. The treatment the patient has depends on:

- Where in the ear the cancer is
- The type of cancer
- The size of the tumour
- Whether it has spread to
- The general health of the patient

People who have cancers that start in the head and neck usually see a team of specialist doctors and other health professionals. They include:

- Head and neck surgeons – including ear, nose and throat surgeons, mouth and facial bone surgeons, and plastic surgeons
- Specialists in cancer drugs and radiotherapy – oncologists
- Dentists
- Specialist nurses, physiotherapists and dieticians

Surgery - the type and amount of surgery a patient needs depends on where the cancer is in the ear and whether it has spread into any of the surrounding tissues, or into nearby structures, such as the bone.

The surgeon will remove the tumour together with an area of tissue surrounding it that is completely free of cancer cells. This is called a clear margin of tissue. It needs to be at least 5mm all-round the cancer. Doing this helps to lower the risk of the cancer coming back.

Surgery may involve having some or all of the following removed:

- The ear canal
- Part or all of the temporal bone
- The middle ear
- The inner ear

[Picture Credit: Temporal Bone]

The temporal bone is the bone at the side of the skull, by the ear. The operation to remove the temporal bone is called a mastoidectomy or temporal bone resection.

Rarely, the surgeon may need to remove the facial nerve. This runs down the side of the face and through the salivary gland. They may also need to remove the lymph nodes nearby in the neck and the salivary gland on that side of the head.

Puyana, C., Ham, P. & Tsoukas, M.M. 2020.
Background: The external ear is composed of thin skin overlying cartilage making melanoma on the external ear difficult to resect while preserving the intricate anatomy. Although surgeons have achieved robust clinical outcomes for nonmelanoma and most recently melanoma skin cancers with Mohs micrographic surgery (MMS), there is still not enough evidence on the MMS application for external ear melanoma treatment.
Objective: The authors examined survival outcomes in patients treated with MMS, narrow margin excision (NME), and wide margin excision (WME) for melanoma on the external ear.
Methods: Data from the NCI SEER program was retrospectively analyzed. Patients who received surgical treatment on the external ear and had microscopically confirmed diagnosis of cutaneous melanoma were included in the study. The effect of different surgery types: MMS, NME, and WME, on melanoma survival was evaluated.
Results: A total of 8,212 melanoma cases of the external ear performed during the years 2000 to 2015 were considered for analysis. There were no significant differences in survival comparing NME and WME with MMS.

Conclusion: Mohs micrographic surgery is at least equivalent to WME for the treatment of melanoma of the external ear.

Radiotherapy - radiotherapy uses high energy rays to treat cancer. The patient may have radiotherapy as the main treatment or may have it if the surgeon has not been able to remove a clear margin of tissue from around the tumour. Then radiotherapy can lower the risk of the cancer coming back.

The patient usually has radiotherapy every day (from Monday to Friday) for between 4 and 7 weeks. At the first appointment the radiotherapy doctor (radiation oncologist) plans the treatment. This planning appointment takes a couple of hours but after that each treatment only takes a few minutes.

Chemotherapy - chemotherapy uses anti-cancer (cytotoxic) drugs to destroy cancer cells. Chemotherapy on its own will not cure cancer of the ear but doctors may use it to relieve symptoms if the cancer comes back or when the patient cannot have other treatments.
To help cure ear cancer, researchers have been looking into giving chemotherapy with radiotherapy before or after surgery. More research is needed to find out how well this works and when it is best to have chemotherapy.

The chemotherapy drugs one may have include fluorouracil and cisplatin.

Shinomiya, H., Uehara, N., Teshima, M., Kakigi, A., Otsuki, N. & Nibu, K.I. 2019.
OBJECTIVE: The purpose of this study was to clarify the impact of superficial parotidectomy and postoperative radiotherapy (PORT) for the surgical treatment of early stage squamous cell carcinoma (SCC) in external auditory canal (EAC).
MATERIALS AND METHODS: Thirty-seven patients with T1 (n = 14) or T2 (n = 19) SCC in EAC treated between 2000 and 2016 at Kobe University Hospital were enrolled in this study. Thirty-three patients were operated with sleeve resection or lateral temporal bone resection.
RESULTS: The 5-year overall survival and disease-specific survival rates were 95% and 100%, respectively. Surgical margin was positive in 4 patients, who were treated by PORT and have been alive without disease. Prophylactic superficial parotidectomy was simultaneously performed at the time of initial surgery in 15 patients, in whom no lymph node (LN) metastasis was observed. Among the other 22 patients, regional recurrence in parotid LN was observed in one patient, who was successfully salvaged by total parotidectomy. Potential parotid lymph node metastasis rates of T1 and T2 SCC in EAC was 0% (0/14) and 5% (1/19) respectively.
CONCLUSIONS: Complete resection without positive surgical margins is essential for the treatment of the patients with T1 and T2 earcancers. Prophylactic superficial parotidectomy or neck dissection is not mandatory for T1 and T2 diseases, as long as precisely extent of disease is assessed preoperatively. PORT should be performed for the patients with positive surgical margins.

Follow-up and Coping with Cancer of the Ear
The patient will have regular check-ups once the treatment has finished. The doctor will examine the affected ear and ask about one's general health. This is the chance to ask any questions one may have and to tell the doctor if anything is causing concern. How often one has check-ups will vary, depending on the situation. They usually start off every 2 or 3 months and become less often as time goes on.

Coping with a diagnosis of cancer can be difficult, both practically and emotionally. It can be especially difficult if one has a rare cancer. Being well informed about the cancer and its treatment can make it easier to make decisions and cope with what happens.

It can also help to talk to other people who have the same condition. But it can be hard to find people who have had a similar rare type of cancer.

About Clinical Trials

Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.

Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers
- Tests to find new ways of screening for cancer

The **South African National Clinical Trials Register** provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details.

For additional information, please visit: www.sanctr.gov.za/

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