

Paparella: Volume II: Otology and Neuro-Otology

Section 3: Diseases of the Ear

Part 3: Middle Ear and Mastoid

Chapter 36: Surgery for Advanced Cancer of the Ear

John S. Lewis

Cancer of the external ear is a disease of primarily elderly white men, usually in their seventh decade, who have been exposed to some external irritant such as frostbite or the actinic rays of the sun being superimposed, as in keratosis. About two-thirds of these lesions are of the basal cell variety and may form large rodent ulcerations of the pinna, mastoid process, and temporal region. The remaining third are most often squamous carcinoma or, rarely, melanoma.

Neoplasms of the auditory canal are most prevalent among women. Those tumors arising in the middle ear and mastoid process have an even sex distribution. The median age of onset for these cancers is about 55 years.

Cancer of the auditory canal and middle ear is associated with a chronic otorrhea in about one-third of cases (Table 2). The remainder have been an associated infection dating back 6 to 12 months. Cholesteatomas have been associated with about one-fourth of middle ear mastoid carcinomas. Two cases of squamous carcinoma of the ear canal have occurred in patients with chronic lymphatic leukemia. A report from the University of Chicago indicated that eight cases of cancer of the mastoid process occurred in radium dial painters (Table 1).

Fortunately, cancer of the ear is extremely rare; the incidence is estimated at between 1 in 5000 and 1 in 15,000 of all otologic pathologic conditions. It may be classified as follows:

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| <p>I. Primary malignant tumors of the ear</p> <p>A. Epithelial</p> <ol style="list-style-type: none">1. Squamous cell carcinoma2. Adenocarcinoma3. Malignant melanoma <p>B. Mesenchymal</p> <ol style="list-style-type: none">1. Sarcoma2. Malignant xanthoma3. Malignant glomus tumor | <p>II. Secondary tumors of the ear</p> <p>A. Direct extension from:</p> <ol style="list-style-type: none">1. Parotid2. Nasopharynx3. Temporomandibular joint <p>B. Distant metastases from:</p> <ol style="list-style-type: none">1. Kidney2. Lung3. Breast4. Prostate. |
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By far the most common type of ear cancer is squamous carcinoma, comprising 82 per cent of all reported cases Table 2. Basal cell carcinoma affects the external ear and only rarely arises from the auditory canal. Adenocarcinoma, usually of the adenoid cystic type, arises from ceruminous glands in the auditory canal. More commonly, adenocarcinoma

involving the deep lobe of the parotid gland extends directly into the temporal bone. Rarely, malignant melanoma, sarcoma, malignant xanthoma, and malignant glomus tumors arise within the confines of the temporal bone and require radical extirpation. Embryonal rhabdomyosarcoma arises from the middle ear in young children and is best treated by a combination of radiotherapy and chemotherapy.

Table 1. Anatomic Site of Origin of Ear Cancer in 100 Patients

Site	Median Age (years)	Number of Patients	
		Male	Female
External ear	70	21	4
Auditory canal	55	28	51
Middle ear and mastoid	56	14	14

Symptomatology and Diagnosis

The erosion of the external ear by neoplasm is usually obvious, and friable bleeding ulceration may be readily biopsied. The chronically infected auditory canal that fails to respond to antibiotics and local medication may present a more subtle problem. In our experience it is estimated that the period of time from the initial symptoms to the diagnosis averages about 6 months. Hearing loss with otorrhea is the earliest symptom. Vertigo and facial paralysis indicate advanced disease. Some cases have external swelling due to invasion of the parotid gland and sternomastoid muscle. Mastoid films and CT scan will usually show the extent of skull and brain involvement. The use of retrograde jugular venography will demonstrate the possible invasion of the lateral sinus and jugular bulb area. Carotid angiography with the use of subtraction technique in the venous phase is an excellent alternative to retrograde jugular venography.

Table 2. Pathology (Total Experience - 132 Cases)

Squamous carcinoma	108
Adenocarcinoma	7
Basal cell carcinoma	5
Embryonal rhabdomyosarcoma	4
Malignant melanoma	4
Spindle cell sarcoma	2
Angiosarcoma	1
Malignant histiocytoma	1

Surgical Considerations

The operation is a combined intracranial-extracranial approach. Usually one or two malleable spinal puncture needles are inserted in the lumbar spinal canal to withdraw 50 to 100 mL of cerebrospinal fluid. If severe osteoarthritis of the spine precludes their use, Lasix may be used to shrink the brain. The craniotomy flap may be based either above or below, the former being used at the present time. If extensive involvement of the external ear and auditory canal is present and involves sacrifice of the external ear, a posteriorly based scalp

flap or bipedicle scalp flap is utilized. High-speed Stryker air drills facilitate the bony dissection. Bleeding from the lateral sinus is frequent and is controlled with vascular silk and Surgicel. Segments of dura are replaced with fascia. The temporal muscle is mobilized and rotated down into the defect to cover the exposed dura. If extensive dural excision and replacement are required, rotation of a scalp flap to cover the defect is necessary.

Operative mortality has been reduced from an initial 10 per cent to less than 5 per cent.

Technique

The following is a summary of the procedure illustrated in the figures.

1. Malleable spinal puncture needles (preferably two) are inserted into the lumbar spinal canal for withdrawal of 50 to 100 mL of cerebrospinal fluid late in the procedure, allowing for adequate exposure of the petrous pyramid.

2. Hypotensive anesthesia is administered.

3. A thorough temporal craniotomy is done to avoid trauma to the temporal lobe and spinal and cavernous sinuses.

4. The superior and inferior petrosal sinuses are cauterized, as are the mastoid emissary vein and middle meningeal vessels, if necessary.

5. At the skull base, injury to the jugular vein, internal carotid artery, and hypoglossal and vagus nerves should be avoided. The facial nerve is sacrificed.

6. Tears of sigmoid sinus are managed by proximal finger pressure and closure with atraumatic surgical silk or a temporal muscle plug. A large sinus tear at the jugular foramen level may be controlled with 2-inch packing left in place for 5 days.

Figures. A. Skin incision with flap, including pinna, based superiorly with modification for neck dissection. A1. Skin incision based inferiorly with auditory meatus circumscribed. A2. Skin incision to sacrifice diseased pinna with bony specimen. If the external ear is sacrificed or a large segment of dura is removed, a posteriorly based scalp flap should be rotated to cover the defect. B. Outline of extent of bony resection. C. Diagram indicating extent of bony resection of squamosa and petrous pyramid. D. Incision is carried through the auricularis and temporal muscles to expose the squamosa and mastoid with muscular attachments. Incision is made through the parotid gland, sacrificing the facial nerve, to the base of the zygoma and the ascending ramus of the mandible. The posterior facial and jugular veins and superficial temporal artery are ligated and divided. The auditory canal is cored widely to be included with the specimen.

A. The zygoma is transected and the ascending ramus of the mandible is sectioned near the joint. The sternocleidomastoid muscle and posterior belly of the digastric are sectioned, exposing the internal jugular vein in the carotid sheath. The styloid process is transected with the stylohyoid muscle. A temporal craniotomy is performed, mobilizing

underlying dura of the temporal lobe of the brain. The lateral sinus and its sigmoid portion are carefully exposed. B. Cerebrospinal fluid is withdrawn and the temporal lobe and sigmoid sinus are retracted from the petrous pyramid. A Styker air drill saw with an orbital blade sections the anterior portion of the middle cranial fossa into the temporomandibular joint. The orbital blade makes the initial incisions on the three surfaces of the petrous pyramid near the junction of its medial and middle thirds. Chisels are directed transversely to complete the transections. When dura is involved, it is freed from the petrous pyramid with the electrocautery knife, the bony resection is completed and then the dura removed and replaced with temporal fascia. It may be necessary to ligate the lateral sinus. This is accomplished by making an incision on either side of the sinus through the dura and then clamping and cutting through the sinus wall in stages. The opening is then closed with continuous 3-0 or 4-0 vascular silk. C. Soft tissue attachments are transected and the bony specimen is removed at the level of the jugular foramen. The operative defect, including brain and dura, petrous remnant, carotid and jugular vessels, hypoglossal and vagus nerve, is shown. D. The incision is closed in layers. The auditory meatal defect is lined in purse fashion with a split-thickness skin graft.

Complications

Hemorrhage. This is usually confined to venous loss from the external jugular vein and petrosal sinus. The median loss is 1500 mL. The use of hypotensive anesthesia has reduced the amount of hemorrhage considerably.

Infection. The use of radiation therapy has produced a high incidence of postoperative infection to the grafted site. The most common causative organism is *Pseudomonas aeruginosa*; it is adequately controlled with carbenicillin administered parenterally in combination with gentamycin and local acetic acid dressings, which help to clear up the problem. Frequently portions of the skin graft are lost and must be replaced. A permanent temporal decompression is present, so that skin coverage is imperative.

Cerebral Herniation. If skin coverage of the dura is not intact, herniation may occur. It then is necessary to reduce the hernia and cover it by both fascia and scalp flap.

Cerebrospinal Fistula. The dura should always be repaired primarily. If a defect exists, grafting with temporal fascia and skin is mandatory. If a cerebrospinal leak persists for more than 10 days the wound should be reopened and the tear repaired. Meningitis and cerebral abscess are hazardous, especially with gram-negative infections.

Facial Nerve Paralysis. Lateral lid fusions are carried out to prevent corneal ulceration. Fascial sling repairs of the affected side with face lift may be carried out at a later date. Faciohypoglossal anastomoses have been carried out successfully.

Deafness. This is complete on the operated side.

Vertigo. This lasts from 5 to 15 days, and there may be a period of unsteadiness for several months.

Carotid Artery Thrombosis. This may occur from trauma or laceration of the internal carotid artery during the operative procedure. In a recent case of the author's, thrombosis occurred 2 days following surgery and resulted in a hemiplegia.

End Results

A total of 132 cases of advanced cancer of the ear have been treated by the author, of which 105 cases were resectable (Table 3). The total experience yielded a cure rate of 28 per cent. The best management program should entail an en bloc resection of the temporal bone followed by 5000 to 6000 rads radiation therapy 4 to 6 weeks after surgery. Surgery alone, which consisted of a temporal bone resection, yielded a cure rate of 28 per cent; with preoperative radiation therapy and temporal bone resection, the cure rate was 25 per cent; with temporal bone resection and postoperative radiation, the rate was 35.5 per cent. The use of chemotherapy has been reserved for unresectable cases or for recurrent cancer and was of palliative value only.

Table 3. End Results of Treatment (Total Experience - 132 Cases)

5-year cure rate			
Radical surgery only	28 cases	37/132	(28%)
Preoperative radiotherapy plus surgery (includes sandwich therapy)	73 cases	18/73	(25%)
Surgery plus postoperative radiotherapy	31 cases	11/31	(35.5%)