Neoplasms of the Ear Canal

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KEYWORDS

- External auditory canal • Neoplasm • Bony • Ceruminous • Cutaneous

KEY POINTS

- Osteoma and exostoses are related in histology, often asymptomatic, diagnosed incidentally and might not require intervention.
- Benign and malignant glandular EAC neoplasms are rare, can present similarly and can share histologic characteristics.
- Squamous cell carcinoma is the most common malignant neoplasm of the EAC and should be considered in patients with unresolving otitis externa or non-healing ulcerative or friable lesions.
- Sleeve resection of the EAC skin has a limited role in the surgical treatment of cutaneous malignancies.

INTRODUCTION

Neoplasms of the EAC can be described by their cell of origin and further divided into lesions arising primarily from within the EAC or involving the EAC secondarily from a separate primary location. Given the histologic components of the EAC, primary neoplasms can be broadly classified into bony, glandular and cutaneous (Table 1). Neoplasms that secondarily involve the EAC include metastasis and those arising from within the middle ear, mastoid or jugular foramen. This article will review these classifications, focusing on histopathology, clinical characteristics, prognosis, and management.

Bony

Exostoses are benign generally broad-based rounded bony growths that arise circumferentially as multiple lesions in the medial EAC, often bilateral and symmetric, and consisting of layers of subperiosteal bone containing no bone marrow.[1] Their formation is correlated with cold water exposure and thought to arise due to cold-induced periostitis. Clinical presentation varies with the degree of EAC occlusion and can

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Table 1	
Neoplasms of the ear canal classified by cell of origin	
Bony	
Benign	Osteoma
	Exostoses
Glandular	
Benign	Ceruminous gland adenoma
	Ceruminous pleomorphic adenoma
	Ceruminous syringocystadenoma papilliferum
Malignant	Ceruminous adenoid cystic carcinoma
	Ceruminous adenocarcinoma NOS
	Ceruminous mucoepidermoid carcinoma
Cutaneous	
Benign	Squamous papilloma
	Pilomatrixoma
Malignant	Squamous cell carcinoma
	Basal cell carcinoma
	Melanoma
	Merkel cell carcinoma
Metastatic	Parotid, colorectal, bronchogenic, esophageal adenocarcinomas; small cell, hepatocellular, prostate, renal cell carcinoma
Arising from secondary site	Paraganglioma, sarcoma, schwannoma, hemangioma, Langerhans cell histiocytosis, lymphoma, extramedullary plasmacytoma, leukemia, solitary fibrous tumor

include conductive hearing loss and otitis externa.[2] High resolution computed tomography (CT) of the temporal bone will delineate the depth of bony involvement as well as the presence of any medial cholesteatoma formation. Management includes observation for asymptomatic individuals or exostectomy and canalplasty for symptomatic individuals. A variety of approaches using the drill or osteotome have been described. Advantages of the osteotome include decreased risk of sensorineural hearing loss and postoperative stenosis, while the drill might reduce the risk of tympanic membrane perforation.[2] Preservation of native canal skin is paramount to reducing the risk of stenosis.[3]

Osteomata are benign generally solitary, pedunculated smooth, round lesions arising along the tympanomastoid and tympanosquamous suture line. Histologically they consist of lamellar bone with outer cortical and inner bone marrow spaces, differentiating osteomata from exostoses.[1] Osteomata are often diagnosed incidentally. Most individuals are asymptomatic. For symptomatic individuals, surgical removal can done be via a transcanal or postauricular approach using a drill or osteotome. Preoperative CT imaging can delineate the medial extent of the osteoma and the presence of medial debris (**Fig. 1**). Similar to exostectomy, skin flaps are created with the intention of skin preservation and the bony lesion is drilled to its base. Complete removal of the base is recommended to prevent recurrence.[3]

Glandular

Glandular neoplasms of the EAC are uncommon. Formerly called ceruminomas, they are thought to arise from ceruminous glands, which are concentrated in the lateral membranous portion of the EAC. The World Health Organization (WHO) currently classifies both benign and malignant EAC glandular tumors.[1]
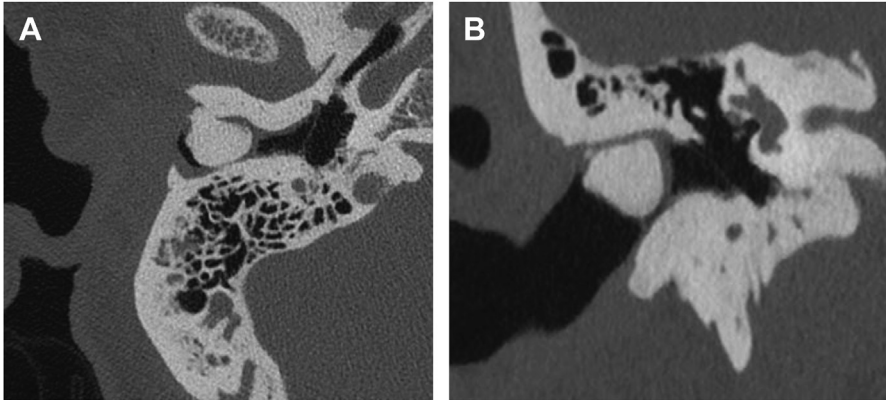
Fig. 1. High resolution CT temporal bone of the right external auditory canal in the axial (*A*) and coronal (*B*) planes demonstrating a large superiorly based osteoma originating from a relatively narrow stalking, causing near complete canal occlusion.

Benign

Benign EAC glandular tumors include ceruminous gland adenoma, ceruminous pleomorphic adenoma and syringocystoma papilliferum. Patients are often asymptomatic but found by otologic exam to have a soft well-circumscribed lesion occupying the membranous EAC, not associated with ulceration or destruction. Rarely are these lesions identified in pediatric patients.[4,5] On imaging, benign lesions will be well-circumscribed with homogeneous enhancement with or without cystic changes and no bony infiltration.[6] Once biopsy confirms the histology, a wide local excision and reconstruction is recommended.

Ceruminous gland adenomas, or more recently termed adenoma not-otherwise-specified (NOS), are the most common benign glandular EAC tumors.[1] They are firm nonencapsulated masses with smooth surfaces and are composed of well-differentiated glandular structures lined by epithelium. The average age of presentation is in the 6th decade.[5,7] There is no sex predilection.

Ceruminous pleomorphic adenoma (CPA) is the second most common EAC benign glandular neoplasm.[7] Tumors are firm, nonencapsulated, and well-circumscribed and are composed of both epithelial and mesenchymal elements, similar to pleomorphic adenomas in the head and neck.[7] Typically, primary neoplasms present as enlarging masses with occlusive and compressive symptoms. There appears to be a slight male predominance, though the average age of presentation is less than that of patients with adenoma NOS.[6]

Ceruminous syringocystadenoma papilliferum (SCAP) is an extremely rare tumor of the EAC with only a few case reports in the literature.[8–12] Tumors can be polypoid, lobulated or ulcerated, containing multiple short, thick papillae.[13] Presenting symptoms are similar to those of other benign EAC neoplasms. Most reported cases were diagnosed in the 7th to 8th decade of life.

Malignant

There is clinical and histologic overlap between benign and malignant EAC glandular tumors, though malignant neoplasms appear to be slightly more common.[13] In descending order of frequency, these include ceruminous adenoid cystic carcinoma, adenocarcinoma NOS, and mucoepidermoid carcinoma.[13] Patients can be asymptomatic or present with canal occlusion, otalgia and facial weakness. Tumors are

more likely to ulcerate and exhibit perineural invasion. They can extend through the fissures of Santorini into the parotid gland, into periauricular soft tissue, through the tympanic membrane or into the bony and cartilaginous EAC. Adequate depth of biopsy is needed to distinguish between benign and malignant neoplasms. CT and MRI should be obtained to delineate the extent of invasion.

Ceruminous adenoid cystic carcinoma

The origin cell-type of ceruminous adenoid cystic carcinoma of the EAC is unknown but it shares features with malignant salivary gland tumors. Tumors are unencapsulated, diffusely infiltrative and invasive into deep tissue and perineurium, and made up of monomorphic basaloid cells arranged in tubular, cribriform or solid patterns. The solid pattern carries the worse prognosis.[14,15] Alterations of the MYB transcription factors seen in salivary gland adenoid cystic carcinoma might also occur in ceruminous adenoid cystic carcinoma,[16,17] however tumors that do not stain for MYB might have a worse prognosis.[18]

Ceruminous adenoid cystic carcinoma is approximately twice as common in females as males, and presents earlier in life than other ceruminous gland neoplasms.[19] Histologic features which correlate with worse prognosis include perineural or bone invasion, solid pattern, involvement of the parotid gland, duration of symptoms for greater than 2 years and positive resection margins.[19] Regional lymph node and distant metastases are not uncommon, but of all distant sites, the lungs are the most common.[20,21] Treatment is a lateral temporal bone resection and superficial parotidectomy. A selective neck dissection is indicated for imaging consistent with regional lymphadenopathy. Adjuvant radiation is recommended for close or positive margins, perineural or lymphovascular invasion, bone invasion, solid pattern histology, or lymph node involvement. In one series of 43 patients, the 5-year survival rates for patients with clear surgical margins was 89% and for patients with positive margins was 54%. The 5-year survival rate for patients who received radiation was 62% and those who did not was 86%.[22] Over-all survival is estimated to be approximately 75%.[23]

Ceruminous gland adenocarcinoma not-otherwise-specified

Ceruminous adenocarcinoma is nearly identical to ceruminous adenoma. Microscopic examination will show irregular clusters, nests and sheets of atypical diffusely invasive epithelial cells.[13] Tumors can be classified as high-grade and low-grade tumors depending on the extent of glandular differentiation, but no specific histologic feature has been shown to correlate with patient outcomes.

Ceruminous adenocarcinoma appears to be more common in males and presents most commonly in the 6th to 7th decades of life. Treatment includes complete surgical resection, but it is unknown whether adjuvant radiation improves overall or disease-free survival.[24] Recurrence and metastasis occur frequently despite adequate surgical resection with negative margins.[25] Over-all survival is estimated to be approximately 50%.[23]

Ceruminous mucoepidermoid carcinoma

Ceruminous mucoepidermoid carcinoma of the EAC is extremely rare with only case reports documented in the literature.[26–32] Histologically, it is identical to its salivary gland counterpart. Tumors can be low, intermediate, or high-grade depending on the growth pattern; infiltrative tumors with lymphovascular or perineural invasion, tumor necrosis, high mitotic rate, and cellular pleomorphism all indicate higher grades.

Complete surgical resection is recommended. Robust quantitative survival data is lacking given the rarity of the disease.[23]

Cutaneous

Benign
Squamous papilloma. Squamous papilloma is a benign lesion rarely found in the EAC thought to be caused by the human papilloma virus type 6 and 11.[33,34] The route of transmission is unknown.[34–36] Tumors are fungiform or polypoid with variably sized bases and comprised of well-differentiated stratified squamous epithelium arranged in stalks with a central fibrous core.[33,34] Complete surgical resection is recommended.[37]

Pilomatrixoma. Pilomatrixoma, previously described as calcifying epithelioma of Malherbe, is a benign cutaneous neoplasm arising from primitive hair matrix cells.[38,39] Occurring in the membranous canal, they are solitary firm, cystic, and well-circumscribed. They consist of basaloid cells intermixed with "ghost cells" that have distinct borders but a central unstained area.[40] The majority of diagnoses occur in pediatric patients. Treatment is complete excision.

Malignant
The most common cause of malignancy in the EAC is from extension from the pinna, followed by primary squamous cell carcinoma, basal cell carcinoma, melanoma and Merkel cell carcinoma. Patients can present with symptoms of chronic otitis externa, leading to delays in diagnosis. A high degree of suspicion is warranted, and biopsy should be considered for patients with unresolving symptoms.

Squamous cell carcinoma

Squamous cell carcinoma (SCC) on the helix arises secondary to actinic exposure. SCC arising from the EAC is thought to be related to chronic inflammatory states.[41] Tumors are friable or ulcerated scaly, irregular, and raised. They are characterized by pleomorphic polygonal cells with eosinophilic cytoplasm and intercellular bridging.[42] Tumors are locally invasive and can spread through the cartilaginous canal into the parotid gland and infratemporal fossa, into the postauricular sulcus, through the tympanic membrane and into the middle ear, mastoid, inner ear, and jugular foramen.

The mean age at presentation of lesions on the pinna is the 6th decade of life and for primary EAC lesions, the 5th decade of life. Chronic bloody otorrhea, deep otalgia, facial palsy or sensorineural hearing loss should raise suspicion for invasive malignancy. A timely biopsy followed by a CT to assess for bony invasion and MRI to evaluate for the extent of soft tissue involvement, depth of invasion and perineural invasion are warranted. Regional metastases are common for advanced tumors so regional or full-body imaging are usually performed.

There is no universally accepted staging system for SCC of the ear and temporal bone, however the modified University of Pittsburgh staging system is the most used.[43] In general, tumors limited to the EAC (T1 and T2) have a better prognosis than tumors with the involvement of the middle ear, mastoid, or facial nerve (T3 and T4). Surgery is the standard of care and should involve a lateral temporal bone resection, subtotal temporal bone resection or total temporal bone dissection depending on disease extent. Complete excision with adequate margins is favored, and there is no literature supporting the use of en bloc versus piecemeal resection for either subtotal or total temporal bone resection. Sleeve resections have

fallen out of favor because of a high associated recurrence rate.[43] Direct tumor involvement of the parotid gland necessitates a parotidectomy but both elective parotidectomy and neck dissection remain controversial.[43] Adjuvant radiation is recommended for positive margins, perineural invasion, bone invasion and lymph node involvement. Chemotherapy and immunotherapy are both emerging as potential alternatives to surgery followed by adjuvant radiation, but neither are widely utilized yet.[43] The reported 5-year disease free survival rates for combined T1 and T2 tumors and combined T3 and T4 tumor range from 67% to 100% and 41% to 59%, respectively.

Basal cell carcinoma

Basal cell carcinoma (BCC) is the second most common EAC malignancy but accounts for less than 30% of tumors in most series.[44] Actinic exposure is thought to be the primary etiology. Lesions are typically well-circumscribed, displaying a nodular irregularity with rolled edges and a central crusting ulcer but can extend subcutaneously, lacking well-defined margins. Tumors display palisading basaloid cells marginally with central necrosis and ulceration. Histologic subtypes include nodular, sclerosing, morpheaform, superficial spreading, and infiltrative, but tumors can display a mix of subtypes. Most of the BCC of the ear are of the nodular and invasive subtype, but up to 25% might be of morpheaform or sclerosing subtypes.[44] Morpheaform lesions have a propensity for deeper infiltration.

Patients often present in the 6th decade of life with symptoms related to EAC occlusion, but up to one-third might be asymptomatic.[44] Males are affected more commonly than females.[44] Biopsy is required to obtain a diagnosis, and both CT and MRI are used to assess for bony invasion and depth and spread of soft tissue invasion (Fig. 2). Though the modified Pittsburgh staging system is commonly used to direct treatment, it appears that BCC overall portends a better prognosis than that of SCC. Surgical resection is the standard of treatment with the choice of limited resection with skin graft, lateral temporal bone resection or composite resection with neck dissection be made based on tumor size and surrounding soft tissue involvement (see Fig. 2).[44] Adjuvant radiation should be considered for perineural invasion. The 5-year
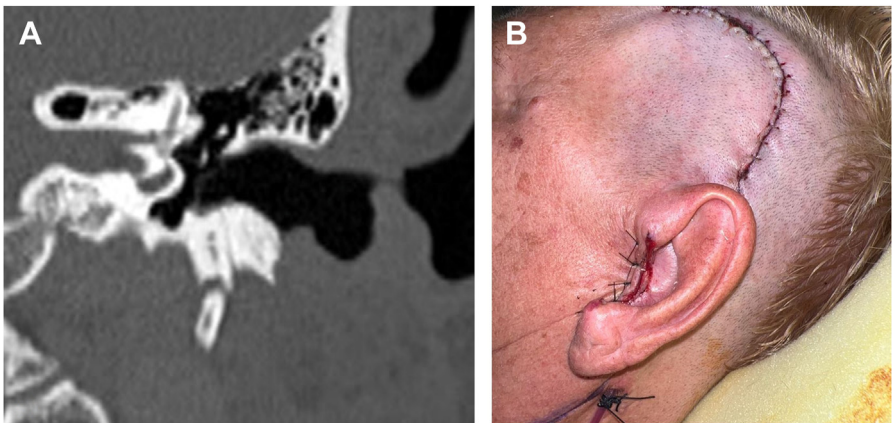

Fig. 2. Preoperative CT temporal bone in the coronal plane (*A*), demonstrating left superior bony external auditory canal soft tissue thickening in a patient diagnosed with primary basal cell carcinoma of the external auditory canal. She underwent a left lateral temporal bone resection, external auditory canal closure and temporalis rotational flap (*B*).

reported disease-free survival is approximately 80%, and the 5-year overall survival is estimated at 78%.[44]

Melanoma

Malignant melanoma arises from melanocytes, derivatives of neural crest cells. Melanomas arising primarily from the EAC make up approximately 5% of primary EAC malignancies.[45] Tumors can be pigmented with changes in color or size or ulceration (Fig. 3). Each of five subtypes - superficial spreading, nodular, lentigo, desmoplastic and mucosal – has variable gross and histologic appearances as well as behavior. Superficial spreading is the most common and nodular is the most aggressive. Lentigo maligna has variable pigmentation and desmoplastic may be amelanotic. Tumors are comprised of atypical melanocytes and stain positive for HMB-45, Melan-A, S-100 protein, and vimentin.[45]

Patients often present in the 5th decade of life.[46] The incidence is higher among males than females. Sun exposure is a known risk factor, but there might also be a genetic predisposition.[47] Excisional biopsy of any suspicious lesion should be undertaken. Complete surgical resection is recommended with appropriate margins. Frozen section pathology cannot reliably detect tumor free margins. Primary melanomas might have a propensity for higher stage at diagnosis than melanomas extending from the auricle. En bloc lateral temporal bone resection is recommended.[46] Additionally, current guidelines recommend consideration of a sentinel lymph node biopsy for lesions that are 0.8 mm thick with adverse features and strongly recommend performance of sentinel lymph node biopsy (SLNB) for all lesions greater than 1.0 mm (T2a).[48] Advanced melanoma of the EAC is often treated with adjuvant radiation with improved locoregional control.[49]
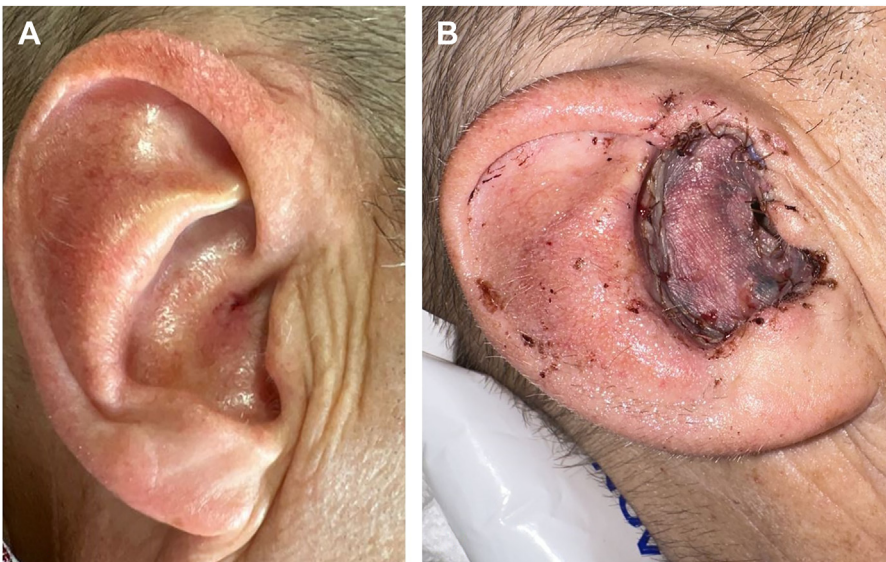

Fig. 3. Clinical photograph of a patient presenting with a discolored, raised lesion centered at the right helical root with extension from the concha cavum toward the meatus (*A*), consistent with malignant melanoma from a dermatologic shave biopsy. He underwent a wide local excision with 1 cm margins involving the meatus and was reconstructed with a split thickness skin graft (*B*).

Several immunotherapeutic agents have been approved for use as well. Distant recurrence is common and disease-free and overall survival outcomes at 1 year are dismal.[46]

Merkel cell carcinoma

Merkel cell carcinoma (MCC) is a rare, aggressive cutaneous neuroendocrine carcinoma.[50] Tumors appear as solitary cutaneous or subcutaneous nodules. Histologically, tumors are comprised of cords, strands or clusters of small, large or intermediate cell sizes, with variably distinct borders, high nucleus to cytoplasmic ratio, salt and pepper appearing nuclei with a small nucleolus. Immunohistochemical staining will differentiate these tumors from other cutaneous malignancies.[50] Risk factors include sun exposure, immunocompromised states or exposure to the polyomavirus.[51,52]

The mean age at presentation is in the 7th decade and males are affected more than females.[53] In addition to developing a rapidly growing, painless nodule of varying hues, approximately 30% of patients will present with regional metastasis.[53] Prompt biopsy and imaging are necessary. Tumor size, infiltrative pattern, thickness and lymphovascular invasion, as well as the presence of regional and distant metastasis are associated with poor prognosis. Recommended treatment includes a wide local excision with 1 to 2 cm margins along with an SLNB or therapeutic neck dissection, followed by adjuvant radiation.[54–57] Local recurrence rates have been reported to be between 40% and 50%.[57,58] Immunotherapy is emerging as a durable option for recurrent and metastatic disease.[57] The estimated 5-year overall survival rates for MCC of all sites is 50% for local disease, 36% for regional metastasis and 14% for distant metastasis.[59]

Metastatic

Metastatic EAC lesions are rare but should be considered in patients with symptoms of chronic non-resolving otitis externa and EAC occlusion with a history of both systemic and nonsystemic malignancies. Metastatic EAC lesions have been reported to arise from parotid, colorectal, bronchogenic, esophageal, rectal, and hepatocellular adenocarcinoma, extrapulmonary small cell carcinoma, prostate carcinoma and renal cell carcinoma.[60–69] Metastatic lesions, most commonly from the breast, lung and prostate, to other regions of the temporal bone might also present as EAC lesions.[70] Up to one-third of patients might be asymptomatic, but of those who are symptomatic, hearing loss, facial paresis and otalgia are the most common symptoms.[70]

Rare neoplasms and those with secondary involvement of the external auditory canal

Primary neoplasms of the middle ear, mastoid, and jugular foramen can present initially as EAC lesions. These include paraganglioma, sarcoma, schwannoma, hemangioma, Langerhans cell histiocytosis, lymphoma, extramedullary plasmacytoma, leukemia, and solitary fibrous tumor. Clinical presentation varies by tumor cell type and extent of disease at the primary site.

SUMMARY

Primary EAC neoplasms include benign and malignant lesions of bony, glandular or cutaneous origin. Benign bony lesions are often asymptomatic, diagnosed incidentally and might not require intervention. Both malignant and benign neoplasms of cutaneous and glandular origin can present with symptoms of chronic otitis externa, leading to delays in diagnosis. Prompt biopsy of soft tissue nodules or lesions associated

with non-resolving otitis externa are warranted. Because of the thin EAC skin, even early-stage malignant neoplasms require aggressive surgical treatment. Metastatic neoplasms and lesions arising from other regions of the temporal bone can present in the EAC as well. Therefore, in addition to prompt biopsy, local and regional imaging is helpful to understand disease extent and origin.

CLINICS CARE POINTS

- Osteoma and exostoses do not warrant intervention unless leading to canal obstruction and conductive hearing loss.
- Wide local excision with canalplasty is acceptable management for symptomatic benign cutaneous and glandular neoplasms.
- Cutaneous malignancy should be considered for patients with unresolving otitis externa or non-healing ulcerative or friable lesions.
- Sleeve resection of the EAC skin has a limited role in the management of cutaneous EAC malignancies.

DISCLOSURE

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