

CERUMINOUS ADENOCARCINOMA OF THE EXTERNAL AUDITORY CANAL: A RARE ENTITYGeeta Yadav¹, Rajanikant R. Yadav², Riddhi Jaiswal³, Satya Prakash Agarwal⁴, Madhu Mati Goel⁵¹Assistant Professor, Department of Pathology, King George's Medical University, Lucknow.²Assistant Professor, Department of Radio-diagnosis, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow.³Associate Professor, Department of Pathology, King George's Medical University, Lucknow.⁴Professor & HOD, Department of Otorhinolaryngology, King George's Medical University, Lucknow.⁵Professor, Department of Pathology, King George's Medical University, Lucknow.**ABSTRACT**

Ceruminous adenocarcinoma is an extremely rare malignant tumour arising from apocrine ceruminous glands of the external auditory canal. Histologically, ceruminous adenocarcinomas are similar to adenocarcinomas elsewhere, except that the glandular luminal tumour cells show apical snouts or blebs indicating an apocrine origin. Central comedo necrosis and stromal invasion helps differentiate these tumours from benign ceruminous adenomas. It may be difficult to differentiate ceruminous adenocarcinomas from other adenocarcinomas occurring in the external auditory canal and from benign ceruminous adenoma if small samples are submitted for histopathological examination. We report on a case of ceruminous adenocarcinoma in a 70-year-old male who presented with an infiltrating growth involving his left external auditory canal along with longstanding painless ear discharge. Incisional biopsy was suggestive of adenocarcinoma; however, postoperative histopathological examination confirmed the tumour to be ceruminous adenocarcinoma.

KEYWORDS

Ceruminous Adenocarcinoma, External Auditory Canal, Histopathological Examination.

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INTRODUCTION: Ceruminous glands are modified apocrine glands confined to the skin of the cartilaginous part of the external auditory canal (EAC). Tumours of the ceruminous glands are extremely rare. Less than 150 cases have been reported to date.^{1,2} These tumours are classified under benign and malignant categories. The benign category encompasses ceruminous adenoma, ceruminous pleomorphic adenoma and ceruminous syringocystadenoma papilliferum. The malignant category includes ceruminous adenoid cystic carcinoma, ceruminous adenocarcinoma not otherwise specified (NOS) and ceruminous mucoepidermoid carcinoma which occur in decreasing order of frequency. Ceruminous adenocarcinoma of the EAC is extremely rare. Less than 40 cases of ceruminous adenocarcinoma have been reported in published English literature to date.³ We report on a case of ceruminous adenocarcinoma of the EAC in a 70-year-old male patient who presented with an infiltrating growth involving his left EAC along with longstanding painless ear discharge.

CASE REPORT: A 70-year-old male presented with an ulcerated growth involving his left ear associated with painless watery discharge. The ear discharge had been present for 35 years. He noticed a small nodule in his left EAC about 10 years ago. The patient had not taken any treatment for his complaints and presented to us when he

noticed an enlarging ulcerated growth involving his left external ear. The physical examination of the ear showed a mass in the left EAC and infiltrating into the left pinna and skin anterior to the external ear with surface ulcerations (Fig. 1). There was no evidence of left facial nerve palsy. No lymph nodes were palpable in the neck. Examination of the oral cavity, pharynx and larynx was unremarkable.



Fig. 1: Clinical Photograph showing an Ulcerated Growth Involving the left EAC and Infiltrating the Pinna and Skin Anterior to the External Ear

Pure tone audiometry showed conductive hearing loss of 60 decibels. Computerised Tomography (CT) of the head and neck showed a hypodense mildly enhancing soft tissue density mass arising from the cartilaginous part of the left EAC and infiltrating the left pinna (Fig. 2). There was no involvement of the bony EAC, tympanic membrane, middle ear, temporomandibular joint or left parotid region.

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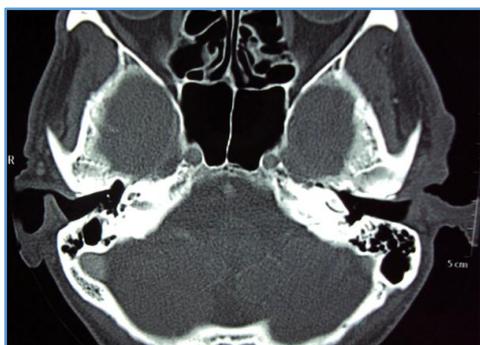


Fig. 2: CT scan Section through the EAC showing a Soft Tissue Density mass in the Cartilaginous Part of the Left EAC with Thickening of the Pinna

An incisional biopsy was taken from the growth. Histopathological examination (HPE) showed a malignant tumour composed of glands, acini and dilated ductular structures lined by multilayered cuboidal epithelium. Individual tumour cells were pleomorphic round to oval to polygonal with abundant eosinophilic cytoplasm, high nuclear cytoplasmic ratio, irregular nucleoli and showed frequent mitosis and stromal invasion. A histopathological diagnosis of adenocarcinoma was rendered.

Local wide excision of the mass including the cartilaginous portion of the left EAC, pinna, a part of the left parotid gland along with level Ib, level II and level III lymph nodes was done. The facial nerve was explored and was found to be uninvolved. Anatomic continuity of facial nerve was maintained. The wound was closed with an occipital rotation flap. On gross examination, the mass was firm, yellowish grey measuring 2.5 cm x 2 cm, filling the left EAC (Fig. 3). The cut surface was solid white with focal areas of necrosis and haemorrhage. On HPE, multiple sections from the growth showed a malignant tumour of apocrine gland of same morphology as seen in incisional biopsy. Additionally, few lobules showed central comedo necrosis and apical snouts or blebs indicating an apocrine origin (Fig. 4). The parotid tissue and all resected lymph nodes were free from tumour invasion. Margins of resection were clear. A final diagnosis of high-grade ceruminous adenocarcinoma (NOS) was made. Postoperative followup of this case after 8 months of surgery was uneventful.

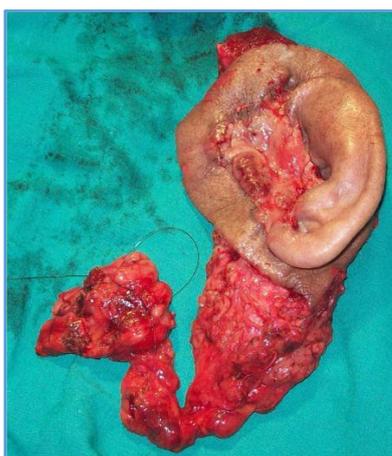


Fig. 3: Gross Specimen Showing the Left External Ear with the Infiltrating growth along with Resected Lymph Nodes

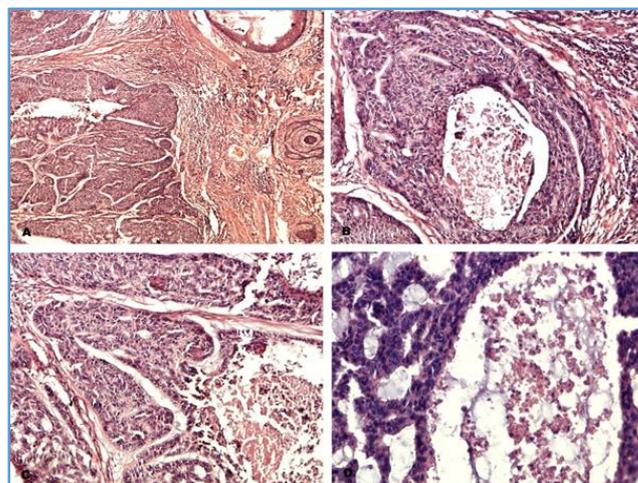


Fig. 4

Fig. 4: Micrographs of tumour tissue in the subepidermal zone shows tumour cells arranged in nests and lobules with stromal desmoplasia (A). Tumour nests show central comedo necrosis and lobular cribriform patterns (B, C). Tumour cells show vesicular nuclei, prominent nucleoli and abundant cytoplasm. Few tumour cells show apical snouts and blebs indicating apocrine origin of the tumour (D).

DISCUSSION: Ceruminous adenocarcinoma presents in the adult age group and usually exhibits a prolonged subclinical phase, often lasting for years before presentation.⁴ The most common clinical symptoms are pain, mass and hearing changes (hearing loss, tinnitus). Other symptoms include discharge, bleeding, cranial nerve palsies and paraesthesias.^{3,4} Otoscopic examination usually demonstrates nodules covered by normal skin, with or without otorrhoea, haemorrhage. Temporal bone CT or MRI is recommended to determine the extent of the lesion and the presence of osseous invasion, and to exclude extrapetrosal disease or metastasis.^{4,5}

Histologically, ceruminous adenocarcinoma shows tumour disposed in back to back glandular and acinar pattern, lined by inner luminal and outer spindled myoepithelial cells with hyperchromatic nuclei. Characteristically, the luminal tumour cells show "decapitation secretions" or prominent "apical caps" supporting a ceruminous origin for these tumours. The characteristic golden-yellow-brown pigment (cerumen granules) within the cytoplasm of normal ceruminous glands and benign ceruminous neoplasms is not seen in ceruminous adenocarcinoma.³

Ceruminous adenocarcinomas can be classified as a low or high-grade depending on the degree of atypia. High-grade adenocarcinomas extensively infiltrate the surrounding tissues as irregular glands and sheets of overtly malignant cells. They show marked nuclear pleomorphism and abundant mitotic figures and may exhibit little or no evidence of apocrine derivation.³⁻⁵ Low-grade adenocarcinoma by virtue of its well-differentiated histology may be indistinguishable from a ceruminous adenoma. The pathognomonic finding for the malignant type is stromal invasion and desmoplasia; a sufficient tissue sample must therefore be obtained to diagnose adenocarcinoma.^{4,5}

The main histological differential diagnosis are direct extension from salivary (parotid) gland neoplasm and metastasis (though rare) to the EAC from breast, lung, prostate, renal, colonic and rectal adenocarcinoma.^{6,7} In this case, CT of the parotid region was unremarkable and evidence of any primary of the above-mentioned sites was ruled out preoperatively.

On immunohistochemistry (IHC), these tumours stain diffusely positive for keratin cocktail, epithelial membrane antigen, CD 117, p 63 and CK 7. IHC may aid in highlighting the individual components i.e. dual cell population of epithelial and myoepithelial cells in ceruminous neoplasms, but it does not aid in the diagnosis of ceruminous malignancies.³ We did not perform IHC as the histological picture for the anatomical site was classical.

Distant metastases from ceruminous adenocarcinoma to the lung, liver, kidney, cervical lymph nodes, and bones have been reported, and metastases tend to maintain their primary histological identity.^{3,4} Surgery and radiotherapy are the mainstay of treatment for ceruminous adenocarcinoma.^{4,8} The outcome of treatment is variable depending upon the extent of disease with frequent and often late recurrences and metastases.^{4,5,9}

CONCLUSION: Ceruminous adenocarcinoma is a rare malignant tumour of the EAC. It should be kept as a differential when an adult patient presents with a prolonged history of painless otorrhoea. Histologically, the tumour cells resemble secretory cells and luminal tumour cells show decapitation secretions or prominent apical caps. Presence of stromal invasion and desmoplasia distinguishes ceruminous adenocarcinoma from benign tumours of the EAC. Differentiating benign from malignant tumours of the EAC may be difficult when samples submitted for HPE are small as tumour cells showing prominent apical caps and stromal invasion may not be detected in small samples.

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