

## ADENOID CYSTIC CARCINOMA OF EXTERNAL AUDITORY CANAL- A RARE ENTITY

Sushil Raj Shrestha<sup>1</sup>, Bhinyaram Jat<sup>2</sup>, Amit Kumar Tyagi<sup>3</sup>, Sushovit Sharma Luitel<sup>1</sup>, Vetrivel G<sup>1</sup>, Ankita Semwal<sup>1</sup>, Reshma Eugene<sup>1</sup>

<sup>1</sup>Junior Resident, All India Institute of Medical Sciences, Rishikesh, India

<sup>2</sup>Senior Resident, Post Graduate Institute, Chandigarh, India

<sup>3</sup>Associate Professor, All India Institute of Medical Sciences, Rishikesh, India

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Corresponding Author:  
**Dr. Amit Kumar Tyagi,**  
Email: ashuu.06@gmail.com

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### Abstract

Adenoid cystic carcinoma of the external auditory canal is a rare entity. Most patients reported otalgia, otorrhoea, and hearing loss. Usually, patients are diagnosed later which worsens the prognosis. Suspicion is the key to early diagnosis. Surgical intervention and chemoradiation are the standard treatment protocol.

## INTRODUCTION

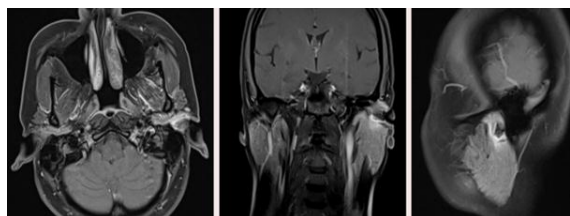
External auditory canal (EAC) carcinomas are a rare type of cancer, with squamous cell carcinomas (SCC) accounting for approximately 80 percent of cases. Conversely, adenoid cystic carcinoma (ACC) is much rarer, occurring in only about five percent of cases.<sup>[1]</sup> Traditionally associated with neoplasms originating from salivary glands, oral cavity, and oropharynx, ACC's occurrence within the realm of otology remains uncommon.<sup>[2]</sup> Nevertheless, sporadic cases of ACC have been documented in atypical anatomical sites such as lacrimal glands, tracheobronchial tree, breasts, and oesophagus.<sup>[3]</sup> Diagnosing EAC ACC can be challenging as its clinical features often mimic chronic otitis media (COM) or benign neoplasms of the EAC. Commonly reported symptoms include hearing loss, the presence of an EAC mass, otorrhea, and otalgia.<sup>[1,4]</sup> The tumour grows slowly and is less likely to spread to lymph nodes.<sup>[4]</sup> Unfortunately, late diagnosis is common, which worsens the prognosis. EAC ACC is notorious for its potential to metastasize distantly, infiltrate nerves and bones, and extend intracranially. Owing to the scarcity of this condition, the number of reported cases remains limited, posing a challenge to the establishment of standardized diagnostic and treatment protocols. Nonetheless, early detection, followed by a combination of surgical intervention and chemoradiation, appears to confer improved survival rates compared to surgery alone.<sup>[1]</sup> It is crucial to screen suspected EAC ACC patients for metastases in the lungs, brain, and liver during

diagnosis and follow-up, as the tumour has a high recurrence rate, even years after treatment.<sup>[4,5]</sup>

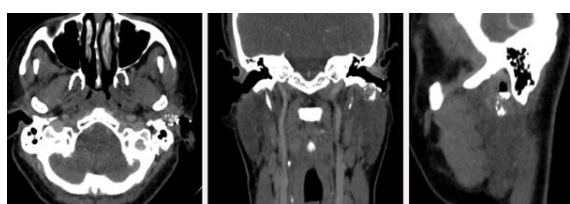
## CASE REPORT

A 31-year-old woman with no known comorbidities presented at our outpatient department with a history of left-sided progressive otalgia persisting for five years. She also experienced intermittent swelling on the left side of her face and occasional scanty discharge but did not report any hearing loss. Upon conducting a routine examination of her ear, we observed purulent discharge in the left ear canal and identified a tender, fleshy growth after cleaning. Pure tone audiometry indicated mild conductive hearing loss in the affected ear. A contrast-enhanced magnetic resonance imaging (CEMRI) of the brain revealed a mass in the left EAC with a smooth interparotid interface, accompanied by level II cervical lymphadenopathy, but no signs of intracranial extension [Figure 1]. High-resolution computed tomography (HRCT) of the temporal bone showed thickening of the soft tissue in the EAC without any associated bone erosion [Figure 2]. Subsequent biopsy results confirmed ACC, precisely the Cribriform type, with positive immunohistochemistry (IHC) markers for CD117, SMA, and P40. Surgical intervention was planned, and the patient underwent a left lateral temporal bone resection (LTBR), a left superficial parotidectomy, and a left selective neck dissection. During the surgery, the tumour was observed occupying the anterior, superior, and inferior walls of the EAC adjacent to the superficial lobe of the left parotid

gland. Post-surgery histopathological examination (HPE) of the specimen confirmed the preoperative diagnosis of Low-grade ACC, and 18 lymph nodes were identified, all free of tumour cells. Following the surgery, the patient experienced marginal mandibular nerve paresis (House-Brackmann Grade III) [Figure 3] and managed conservatively. The patient was discharged in a stable condition.



**Figure 1: CEMRI showing post contrast heterogeneously enhancing lesion around the left EAC.**



**Figure 2: HRCT showing multiple nodular high density focus in the left pre-auricular region suggestive of calcification.**



**Figure 3: Immediate post-op and after 3 months of surgery**

### Follow-up and outcomes

Marginal mandibular paresis subsided following three weeks of follow-up [Figure 3]. The patient was recommended adjuvant radiotherapy (RT), but they refused to undergo RT. A follow-up MRI performed six months later showed post-operative changes, with no signs of tumour recurrence.

## DISCUSSION

EAC tumours are rare, with 20% being glandular in origin. Most are SCC, but ACC is the common glandular type. EAC's ACC shares similarities with the small salivary gland ACC,<sup>[6,7]</sup> but the origin of the tissue has been a source of controversy till date.

ACC can appear in three patterns: solid, cribriform, or tubular.<sup>[8]</sup> The tubular pattern shows the most differentiation of ceruminous glands, while the solid pattern has the least based on IHC staining and

electron microscopy analyses. These patterns are linked to prognosis when originating from salivary glands, with the tubular form having the best prognosis.<sup>[9,10]</sup> It's unclear if the histological state of ACC in the EAC correlates with clinical behavior. In this case, a cribriform pattern was reported.

ACC commonly presents with severe otalgia, otorrhea, hearing loss, and an EAC mass. It can lead to local and distant complications, including intracranial extension, parotid gland and neck node extension, and distant tissue metastasis.<sup>2</sup> ACC may even exhibit early direct parotid extension in stage-1 lesions.<sup>[11]</sup> In this case, the main symptom was also otalgia and otorrhea, but it was not severe, and the tumour was localized to soft tissue without any bony erosion or irregularity but with a smooth inter-parotid interface. Diagnosis may be delayed, as the tumour can grow for years before detection.<sup>[6,7]</sup> Gender predominance in tumour occurrence is a debated topic. Triantafyllidou K et al. found it more common in women, while Lucia A et al. observed an increase in males.<sup>[6,12]</sup>

The management of ACC in the EAC is primarily based on the surgeon's experience due to the lack of evidence-based guidelines. Surgical therapy depends on the extent of the disease and may range from local resection to lateral and sub-total temporal bone resection (LTBR and STBR). In our case, left LTBR, a left superficial parotidectomy, and a left selective neck dissection were done.

RT can be used as adjuvant therapy after surgery for EAC ACC, depending on the tumour's stage, although the outcome is uncertain. Post-operative RT has not shown significant benefits, leading to a 24% decline in 5-year survival.<sup>[13]</sup> Evidence for chemotherapy in EAC ACC is limited.<sup>[14]</sup> The 5-year survival rate is 70%, with staging and margin clearance being important prognostic indicators. Survival rates range from 85% for T1 to 30% for the T4 stage. Good margin clearance can improve 5-year survival rates by 35%.<sup>[13]</sup>

## CONCLUSION

ACC of the EAC presents with otalgia, hearing loss, and a smooth mass. Misdiagnosis is common due to the lack of distinct symptoms. Preoperative HRCT, CEMRI of the temporal bone, and lung CT are crucial for assessing tumour size and extent. For a better prognosis, adequate surgical clearance with a negative margin is required. LTBR is recommended over local resection to reduce recurrence risk. Regular post-operative follow-up is advised for early recurrence detection.

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