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Adenoid Cystic Carcinoma of External Ear Canal: A Case Report

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Abstract **Case Report**

Adenoid cystic carcinoma is a rare tumor, constituting around 5% of primary malignant neoplasms of the external ear canal. It carries a high risk of local recurrence and distant metastases, primarily to the lungs. This is often attributed to delayed diagnosis and to initial symptomatology resembling benign ear affections. Management typically involves surgical resection followed by radiotherapy. Chemotherapy is reserved for palliative purposes in metastatic stages. We present a case of a 71-year-old patient diagnosed with adenoid cystic carcinoma of the external ear canal. The patient complained of otalgia evolving over three years, along with a gradual hearing loss. The patient underwent surgical tumor resection followed by radiotherapy.

Keywords: Chemotherapy, Adenoid cystic carcinoma, tumor, radiotherapy.

INTRODUCTION

Adenoid cystic carcinoma is a rare disease, accounting for 3 to 10% of salivary gland tumors and 1 to 4% of all head and neck neoplasms [1, 2]. It affects more commonly the accessory salivary glands but can also occur in the lacrimal glands, tracheobronchial tree, breasts, esophagus, and external auditory canal (EAC).

Malignant tumors of the external ear canal (EAC) are rare. Squamous cell carcinoma is the most common. Glandular tumors account for 20% of EAC tumors, and adenoid cystic carcinoma (ACC) is exceptionally rare [4, 5]. Symptoms include pain, hearing loss, and a slow-growing nodule in the ear canal. These nonspecific and heterogeneous clinical symptoms often lead to a delayed diagnosis. Metastasis are frequent in advanced stages of these tumors; Therefore, the therapeutic approach still complicated, and it is not well standardized.

CASE REPORT

We report the case of a 71-year-old patient, previously treated for cholelithiasis, and who presented to our ENT department with persistent left otalgia evolving over the past three years. He also reported progressive hearing loss. However, there was no reported history of chronic otorrhea or other otolaryngological symptoms. On otoscopy examination, a hemorrhagic tissue mass was noted, completely occluding the left external auditory canal without extension to the concha or the remaining pinna, which appeared normal (Figure 1). Cervical examination, of lymph nodes, parotid gland, and facial motricity, revealed no abnormalities.

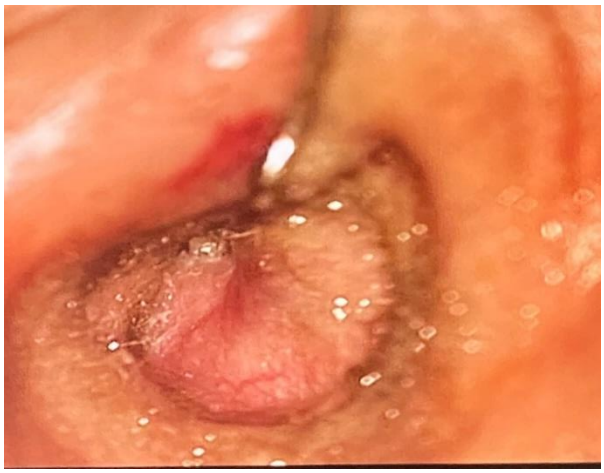

Figure 1: Soft tissue mass in the left external ear canal

A contrast-enhanced facial CT scan, focusing on the petrous bone, demonstrated an isodense mass that entirely occupied the external auditory canal. The mass presents heterogeneous contrast enhancement, with no evidence of bone destruction or extension into the middle ear (Figure 2).

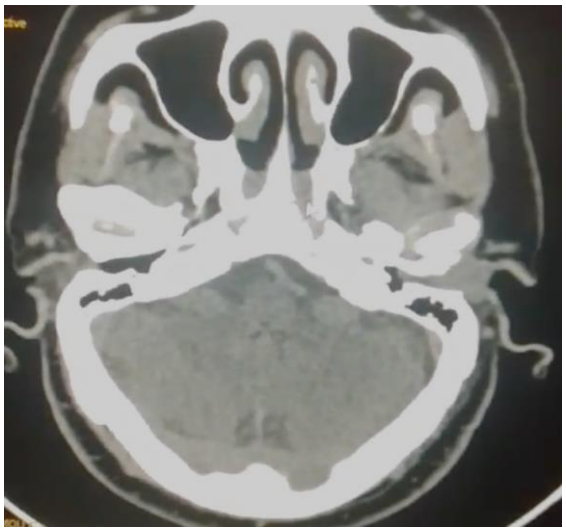

Figure 2: A Tissue process filling the entire external auditory canal, without bone lysis or middle ear extension

A biopsy of the mass confirmed the diagnosis of adenoid cystic carcinoma upon histopathological examination.

An MRI was performed to delineate tumor's extension. The MRI findings indicated a well-defined tissue mass centered on the external auditory canal (EAC), describing regular contours. The mass appeared to have intermediate signal intensity on T1-weighted images, restricted diffusion, and heterogeneous enhancement post-contrast administration. There was no evidence of bone involvement, and the tumor measured 31x17 mm in axial diameter. Furthermore, there were no extensions into the middle ear and mastoid air cells or adjacent structures. No suspicious lymphadenopathy was observed in the cervical or parotid regions examination (Figure 3, 4 & 5).

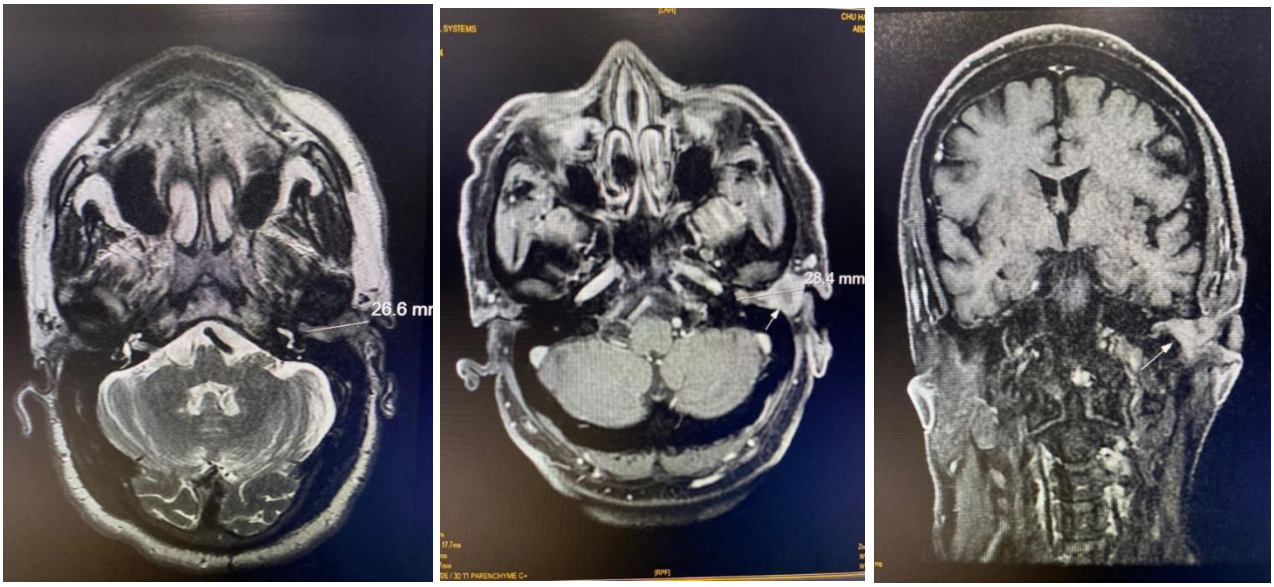

Figure 3, 4 & 5: MRI: EAC Tumor: Intermediate signal on T1, Heterogeneous enhancement after contrast injection. Without extensions to neighboring structures

The patient underwent a complete tumor resection via a retro-auricular approach (Figure 6 & 7), with milling of the canal walls, and reconstruction of the canal lining using a temporal aponeurosis graft (Figure 8 & 9).

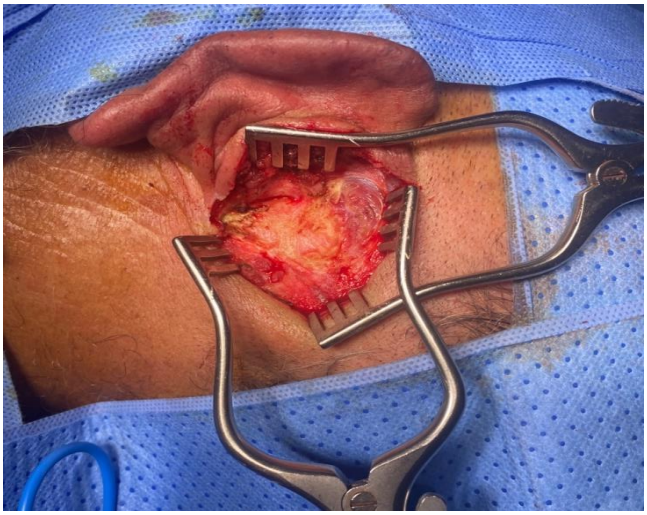

Figure 6: Retro-auricular approach: U-shaped incision of the external auditory canal

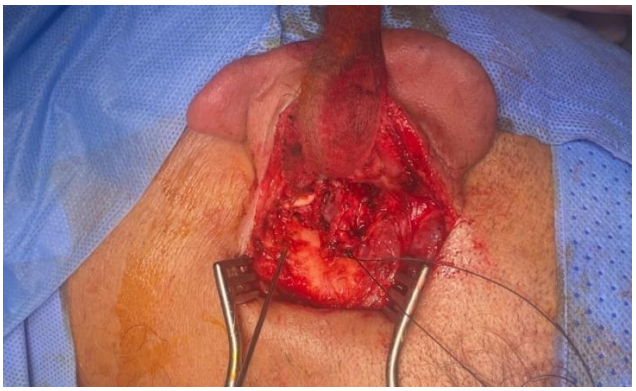

Figure 7: Tumor Marking

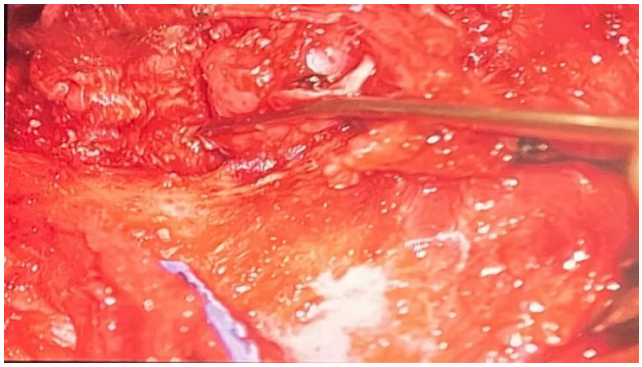

Figure 8: Dissection of EAC skin and removing the tumor

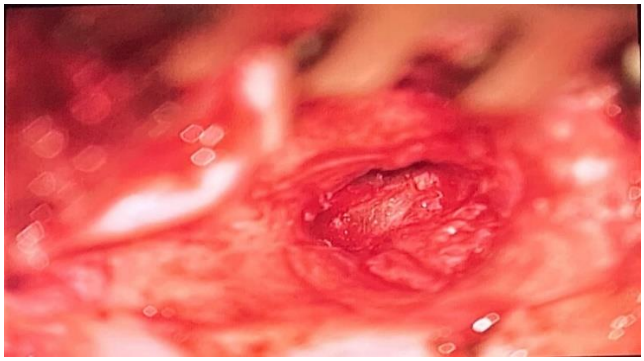

Figure 9: Reconstruction of EAC lining with a temporal aponeurosis.

The histopathological analysis of the specimen revealed a carcinomatous tumor characterized by cribriform, tubular, and solid architectural features. The cribriform structures and tubules appeared cystically dilated, containing an eosinophilic substance. The tumor presented two distinct cell populations: myoepithelial cells with clear cytoplasm and hyperchromatic nuclei, and epithelial cells with variable eosinophilic or clear cytoplasm. These findings are indicative of adenoid cystic carcinoma (Figure 10).

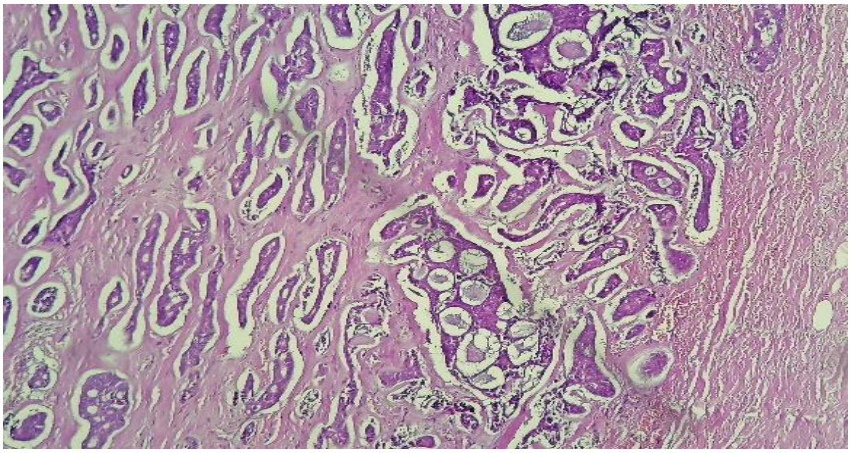

Figure 10: Adenoid cystic carcinoma. Proliferation characterized by cribriform masses composed of basophilic cells (HESx100)

The resection margins were histologically negative except for the internal margin, which had a clearance margin of less than 1mm. Following this, the patient received radiotherapy at a total dose of 70 Gray. A PET scan was conducted to search additional salivary site involvement, and showed no pathological uptake. The patient had a good clinical course with no recurrence observed over a two-year follow-up period.

DISCUSSION

Malignant tumors of the external auditory canal (EAC) are rare, with over 80% of squamous cell carcinomas. Adenoid cystic carcinoma (ACC) is representing approximately 5% of EAC tumors [1]. The head and neck localization typically affects the salivary glands of oral cavity, palate, nasal cavity, and nasopharynx [6]. Adenoid cystic carcinoma (ACC) is an exceptionally rare tumor [4, 5]. Its localization within the external auditory canal is attributed to malignant proliferation of the secretory tissue from the ceruminous glands of the external auditory canal [5-7].

The average age for adenoid cystic carcinoma (ACC) is around 50 years. It is twice as common in women as in men [8]. The majority of patients experience unilateral, intense, chronic otalgia. They also complain of decreased hearing acuity in later stages due to obstruction of the external auditory canal by the mass [9, 10]. The clinical presentation was similar in our case. Adenoid cystic carcinomas clinically present as a polypoid mass or as an epithelial ulceration with granulation tissue [11]. Otalgia may be caused by early perineural invasion by these tumors. Due to their initial symptomatology resembling to other benign ear diseases, they are often diagnosed at a late stage [12]. The progression is marked by symptoms of both local and distant extension. This involves extension to surrounding tissues, mainly intracranial, the parotid gland, and cervical lymph nodes, as well as distant tissues, such as pulmonary metastasis [4-7]. ACC is often associated with early direct extension to the parotid gland [13]. The histological diagnosis of these tumors requires a biopsy that is sufficiently deep and wide to not overlook the characteristics of this tumor [14, 15]. Immunohistochemistry techniques can refine the diagnosis of this tumor [15]. Microscopically, these tumors are made of epithelial cells arranged in various patterns. The typical pattern includes a cribriform architecture combined with areas of tubule formation or solid cellular growth. Perineural invasion characterizes the diagnosis of adenoid cystic carcinoma [11].

Local-regional extension assessment typically involves CT scanning, sometimes in combination with MRI. Evaluation of distant extension requires chest radiography or CT scanning. The diagnosis of primary cutaneous adenoid cystic carcinoma (ACC) can be made only after excluding cutaneous extension from a tumor of other neighboring structure or a distant metastasis from a salivary ACC [25].

Aggressive surgical resection with adjuvant radiotherapy is the standard treatment for local disease control [8-15]. Surgical excision is the primary treatment for ACC. The surgical procedure should be extensive, encompassing healthy tissues and ideally performed in a single bloc. Surgical treatment requires extensive resection, including an expanded mastoidectomy, which may reach neighboring structures, and sometimes can potentially involve subtotal petrectomy [20-22]. The role of elective parotidectomy in improving survival is controversial [16]. Several surgeons prefer to practice a parotidectomy because of the risk of recurrences and

local invasion at this site. This is facilitated by tumor dissemination along the perineural sheath of the facial nerve [17]. Selective lymph node dissection may be considered on a case-by-case basis, depending on tumor extension [18]. Adjuvant radiotherapy is not indicated in cases of wide resection with clear margins. However, given the high risk of local recurrence, intensified radiotherapy using platinum salts may be considered [22]. Adjuvant chemotherapy remains controversial.

The prognosis of ACC of the EAC depends on local resection margins, bone involvement, parotid gland involvement, and nerve invasion [19]. The risk of recurrence due to its metastatic potential is estimated at 30% according to some authors [23]. Secondary locations are mainly pulmonary, but also osseous, renal, and cerebral [24].

CONCLUSION

ACC is a rare tumor of the external auditory canal (EAC). Diagnosis is based on histological examination. Biopsy must be deep and wide. Treatment involves surgical excision, that must be complete, and sometimes radical. A distinctive characteristic of ACC is its neural tropism, underscoring the importance of practicing a parotidectomy into the surgery procedure. The need for cervical lymph node dissection should be discussed. Radiotherapy is recommended by most authors to reduce the risk of local recurrence, which remains the primary prognostic factor.

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