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Case Report

Surgery Section

Uncommon Presentation of Adenoid Cystic Carcinoma in the Parotid Gland: A Case Report

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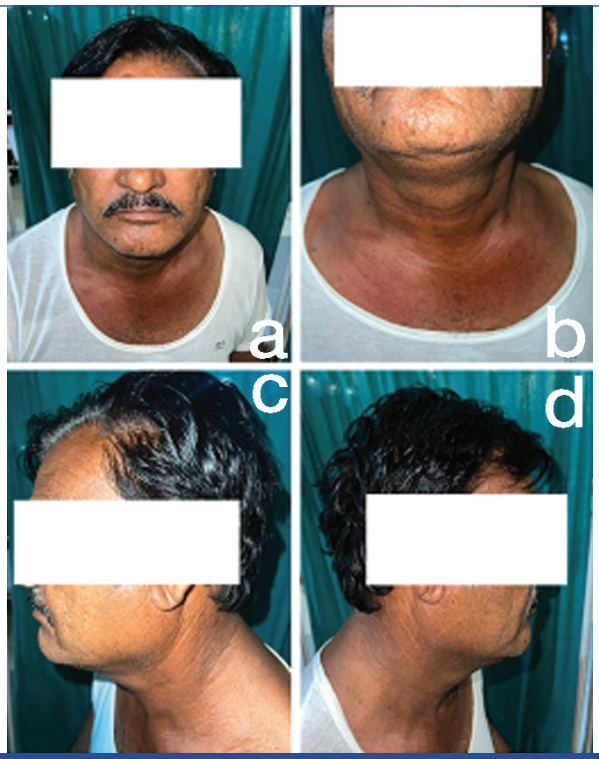
ABSTRACT

Adenoid cystic carcinoma is a rare form of cancer that primarily affects the salivary glands, although it can also occur in various other tissues, including the breast, skin, cervix and prostate. This malignancy is characterised by its slow growth and distinctive histological patterns, often presenting as a painless mass in the affected area. It is most commonly diagnosed in adults aged 40 to 60 years, with a slight predominance in females. The tumour is known for its potential to recur locally and metastasise, primarily to the lungs, although it rarely spreads to regional lymph nodes. Diagnosis typically involves a combination of medical history review, physical examination, imaging studies such as ultrasound, Computed Tomography (CT), Magnetic Resonance Imaging (MRI) and biopsy of the tumour tissue. The biopsy reveals the tumour's histological characteristics, which are crucial for diagnosis. This is a case of a 65-year-old male who presented with a primary complaint of preauricular swelling for two months. The swelling was diagnosed as a low-grade malignant tumour of adenoid cystic carcinoma based on Fine Needle Aspiration Cytology (FNAC). The patient was managed with surgical excision followed by adjuvant chemotherapy and radiotherapy.

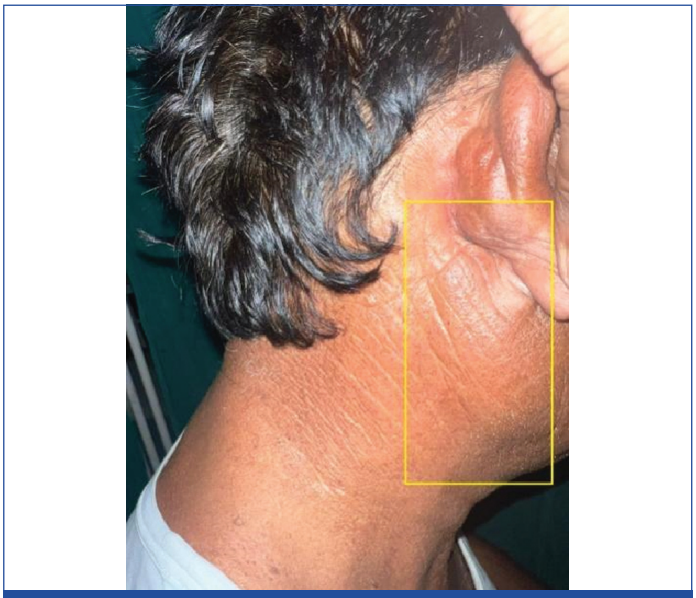
Keywords: Fine needle aspiration cytology, Neck mass, Neck swelling, Preauricular swelling, Salivary gland tumours

CASE REPORT

A 65-year-old male patient presented with swelling in the preauricular region for the past two months. The patient reported being healthy two months prior when he first noticed the swelling in the right preauricular region. The swelling was insidious in onset and progressive in nature, with no aggravating or relieving factors. There was no significant medical or family history. Physical examination revealed a non tender and mobile swelling measuring 4.0×4.0 centimeters, located in the right preauricular region, with no previous scar marks, skin discoloration, active discharge, raised temperature, or bleeding [Table/Fig-1,2].



[Table/Fig-1]: Physical presentation of the patient.

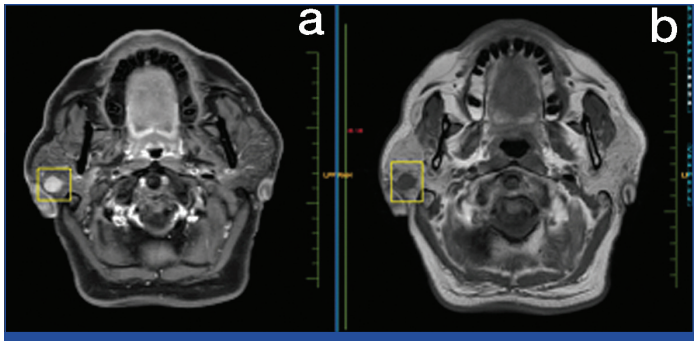


[Table/Fig-2]: Protruded swelling behind the ear in the postauricular region.

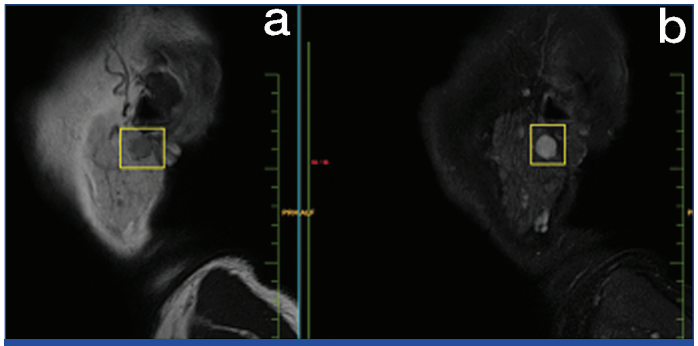
The patient did not have any significant associated medical history, except for being diagnosed with diabetes one year ago, which was managed with oral medications (glimepiride, metformin, tenegliptin and pioglitazone).

The patient underwent ultrasound-guided FNAC of the right preauricular region. The smear showed a few papillaroid structures and sheets of basaloid cells. The cells exhibited enlarged hyperchromatic nuclei with inconspicuous nucleoli, with some cells showing mild polymorphism and rare nuclear raphe. This cytomorphology was suggestive of a low-grade malignant tumour of adenoid cystic carcinoma.

MRI of the parotid region revealed that the superficial lobe of the right parotid gland appeared hypointense on T1FS and hyperintense on T2FS, showing diffusion restriction with dimensions of 13.2×11.5×13.5 millimeters (TR×AP×CC). There was no evidence of any invasion into the deep lobe of the parotid gland [Table/Fig-3,4].



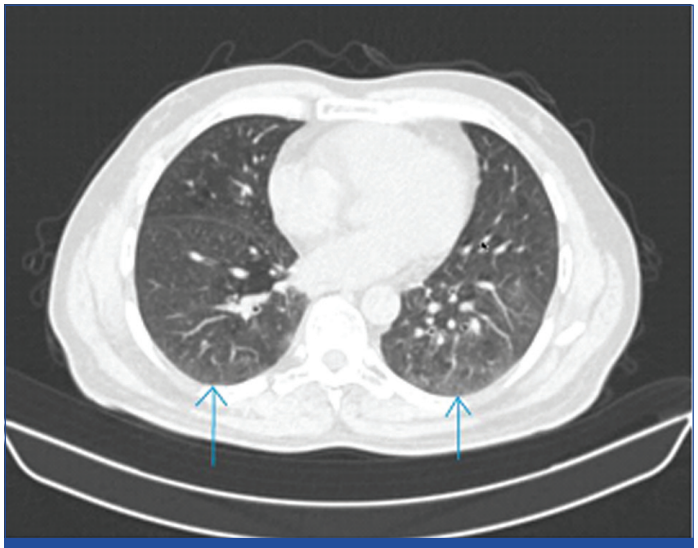
[Table/Fig-3]: Magnetic Resonance Imaging (MRI) (axial plane) evidence of tumour.



[Table/Fig-4]: Magnetic Resonance Imaging (MRI) (Sagittal plane) evidence of tumour.

The patient was further advised to undergo High-Resolution Computed Tomography (HRCT) of the thoracic region to rule out metastasis, which showed mosaic attenuation noted in the bilateral lung fields, with the rest of the area appearing normal [Table/Fig-5]. There was no sign of mediastinal lymphadenopathy. The case was discussed in the tumour board meeting and surgical excision of the parotid gland was recommended [Table/Fig-6], with a follow-up planned after receiving the histopathological report of the specimen.

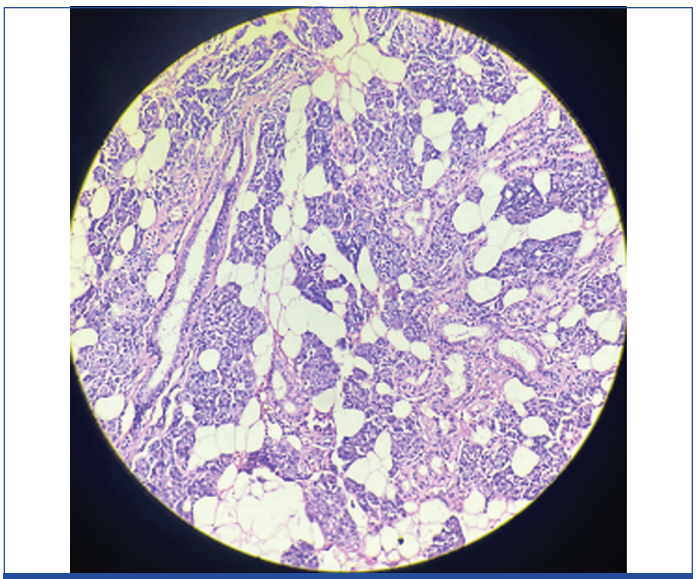
Based on the HRCT findings, the patient was scheduled for surgical management of the tumour through right parotidectomy with ipsilateral neck dissection. Level II and Level III lymph nodes were excised and sent for histopathological examination, along with the excised submandibular gland. The sutures were removed and histopathological analysis predominantly displayed cribriform and tubular patterns. The tumour consisted of well-defined nests of cells with a myxoid or hyaline stroma, often showing a biphasic structure with both luminal epithelial and myoepithelial cells. These histopathological features from the excised sample were noted as a low-grade malignant tumour of adenoid cystic carcinoma [Table/Fig-7].



[Table/Fig-5]: HRCT image showing mosaic attenuation.

Therapy was initiated due to the presence of perineural invasion, which was noted as positive in the histopathological report. Postoperatively,

[Table/Fig-6]: Intraoperative image showing branches of facial nerves preserved during the surgery.



[Table/Fig-7]: Haematoxylin and eosin staining of the excised specimen at 40x magnification.

as per the tumour board discussion, the patient was advised to undergo adjuvant chemoradiotherapy. The dose of radiotherapy (face and neck) is 60 Gy in 30 fractions. The patient is doing well with chemotherapy and radiotherapy. On follow-up at three months, the patient was found to be healthy, with the facial nerve intact and no signs of facial nerve palsy.

DISCUSSION

Adenoid cystic carcinoma is a type of salivary gland tumour characterised by slow growth, primarily affecting the minor salivary glands [1]. Aside from the minor salivary glands, it is commonly observed in the mucinous and submandibular glands. It is a rare occurrence in the parotid gland [2]. Its incidence is approximately 3-5% of all head and neck malignant tumours, with a 10-year survival rate of around 60% [2,3]. Adenoid cystic carcinoma is a rare finding in the parotid gland, accounting for only 2-3% of all tumours in this area [1,4]. There is a reported female predominance in the fifth and sixth decades of life. This carcinoma is commonly characterised by perineural spread, wide local infiltration and a tendency to recur [4].

Adenoid cystic carcinoma has been positively associated with smoking, with the exception of the mucoepidermoid carcinoma subtype; however, there is also evidence suggesting no significant association [2]. It is classified into three types based on histopathology: solid pattern, tubular and cribriform, with solid patterns associated with a poorer prognosis and lower survival rates [1,2,4,5]. Although long-term survival rates are low, the 10-year survival rate has been reported to be about 20% [4].

A MYB-NFIB fusion gene mutation, resulting from a translocation, has emerged as an important marker of adenoid cystic carcinoma. Other genetic alterations in the MYB/MYC pathway have also been reported in relation to the pathogenesis of adenoid cystic carcinoma, as this pathway plays a critical role in its pathophysiology [2,6].

Different clinical presentations of adenoid cystic carcinoma of the salivary glands can include facial asymmetry, external canal stenosis, facial palsy and swellings in the parotid gland and mandible, all of which are progressive in nature [7,8]. However, involvement of the external auditory canal—such as in the earlobes—has been reported as rare [9]. In some cases, these tumours can present as mucoceles in the oral cavity, affecting areas such as the cheeks, lips, or the floor of the mouth [10]. Patients may also report symptoms such as nasal congestion, nasal obstruction and facial pain [11]. In contrast to these findings, this patient only presented with swelling in the preauricular region for a duration of two months. The typical duration of symptoms has been reported to range from two months to two years [7,8,11].

The great auricular nerve can be a suitable graft for the reconstruction of facial nerves, which has been managed effectively in other cases [12]; however, this patient did not require reinnervation and the nerve was preserved. These tumours are very rare in the parotid gland and may be misdiagnosed due to their rarity [4,7].

A comprehensive treatment approach—including surgery, chemotherapy and radiotherapy—is recommended for the management of adenoid cystic carcinoma to achieve better outcomes [9]. This case involved a major complaint of preauricular swelling for two months, which was later diagnosed as adenoid cystic carcinoma based on FNAC and subjected to MRI to rule out any underlying metastasis. This case highlights the importance of a comprehensive approach to the management of adenoid cystic carcinoma, a conclusion supported by findings from a research study that utilised a combined treatment approach (surgical resection and radiotherapy), which led to better outcomes and improved control over the disease [13].

CONCLUSION(S)

Adenoid cystic carcinoma can present challenges due to its slow growth and rarity. Early diagnosis, along with a comprehensive approach to management, is crucial for addressing the recurrence rates and risk of metastasis associated with this malignancy. In this case, the preauricular swelling led to the diagnosis of a low-grade malignant tumour of adenoid cystic carcinoma, which was managed through surgical excision and adjuvant therapy.

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