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Case Report

A Rare Case of Sublingual Adenoid Cystic Adenocarcinoma - A Dormant but Aggressive Tumour: Clinical Insights

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Abstract

Adenoid cystic carcinoma (AdCC) is a slow-growing, extremely malignant tumor with significant recurrence rates. Although less common in the parotid gland than mucoepidermoid and acinic cell carcinomas, it is the most common malignant tumor in the minor salivary glands [1]. It is often aggressive and perineural in nature, with no chondromyxoid stroma or squamous metaplasia present. Malignancies of the sublingual glands are extremely rare, accounting for approximately 0.4%-0.6% of all salivary gland cancers. In general, the smaller salivary gland, the more aggressive the tumor and the worse the prognosis. Malignancies originating in the sublingual glands are exceedingly rare, representing only 0.4%-0.6% of all salivary gland cancers. Generally, the smaller the salivary gland involved, the more aggressive the tumor and the poorer the prognosis [2].

AdCC is the most common malignant tumor of the sublingual salivary gland, accounting for about 2% of all AdCC cases in salivary glands [3]. This case report presents a rare instance of ACC arising from the sublingual salivary gland in a 52 year old female, who presented with a slow- growing, painless swelling on the left floor of the mouth, without ulceration, for three months. She underwent wide local excision without any neck dissection or neoadjuvant chemotherapy. Her histopathology turned out to be pT3N0 disease with R0 Resection. She is on regular follow- up for a year without any clinical/radiological evidence of recurrence. We have done a comprehensive review of the literatures on this topic till date, focusing on clinical characteristics, treatment approaches, and prognosis of sublingual AdCC. This report aims to enhance awareness and improve early detection and management of this rare malignancy.

Keywords: Adenoid Cystic Carcinoma (AdCC); Sublingual Salivary Gland; Excision Biopsy; Wide Local Excision; Perineural Invasion; Metaplasia; Epithelial Salivary Gland Tumors; R0 Resection

Introduction

Adenoid cystic carcinoma (AdCC) is an uncommon and aggressive tumor that developsin the secretory glands, specifically the salivary glands. AdCC accounts for around 1% of all headand neck cancers and 10-15% of all salivary gland tumors [3]. The parotid and submandibular glands account for the vast majority of instances, with minor salivary glands accounting for a smaller proportion. AdCC of the sublingual gland is extremely uncommon, with just a few examples reported in the literature, representing just 0.4% to 0.6% of all salivary gland cancers [1].

While Adenoid Cystic Carcinoma (AdCC) are the most common types in the sublingual gland, they account for only 2% of all ACC cases [3]. The palate is the most common location for AdCC, followed by the tongue, buccal mucosa, lips, and mouth floor. AdCC is more prevalent among women in their fifth and sixth decades of life [4].

The rarity of AdCC in the sublingual gland, along with the tumor's aggressive nature and proclivity for perineural invasion, makes it difficult to identify and treat [5]. This report describes acase of sublingual AdCC in a 52yr old female, as well as the diagnostic technique and treatment strategy, and concludes with a review of literature of AdCC.

Case Report

A 52 year old female presented to our outpatient with a slow-growing, painless swelling on the left floor of the mouth, without ulceration, for duration of three months. On examination, a firm to hard swelling was noted on the left side of the floor of the mouth measuring 4 x 3 cms with variable consistency (Figure 1). No obviously clinically palpable cervical lymphnodes. Ultrasound abdomen and pelvis was normal so was the Chest X-Ray. CECT Neck showed few enlarged lymphnodes at level I and II of left cervical group with ill define soft tissue attenuation in pre and para mandibular region with thinning of the underlying mandibular bone with no e/o erosion and with varying degree of enhancement suggestive of neoplastic etiology. FNAC of the lesion revealed poorly cohesive epithelial cells in fibrillarfibromyxoid stroma suggestive of pleomorphic adenoma of sublingual gland.

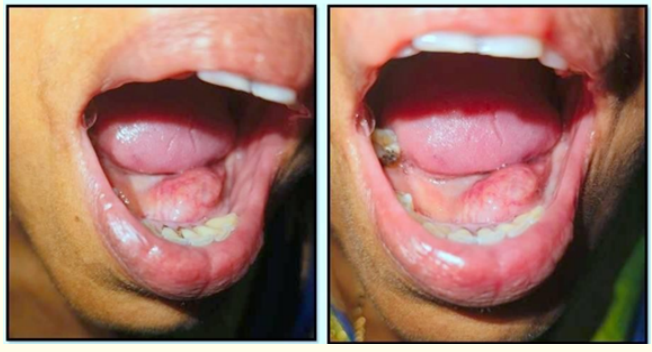

Figure 1: Preoperative presentation of swelling over the floor of the mouth.

After thorough preoperative evaluation, she underwent wide local excision of the sublingual gland and the accompany ducts without any neck dissection or neoadjuvant chemotherapy (Figure 2, Figure 3 and Figure 4). Her histopathology turned out to be unifocal pT3N0 disease of adenoid cystic adenocarcinoma (AdCC) showing 41-50% tubular pattern, 51-60% cribriform pattern and solid component of 1-10% (Figure 5, Figure 6). There was no high grade transformation or lymphovascular invasion with R0 Resection. Oncologist advised for a bilateral modified neck dissection with hemimandibulectomy as a part of completion surgery. After a thorough discussion with the surgical team and reviewing the literatures, it was decided tokeep the patient on follow-up and if any evidence of recurrence, it was planned for adjuvant radiotherapy. She is on regular follow-up now for a year and without any clinical/radiological evidence of recurrence (Figure 7, Figure 8).

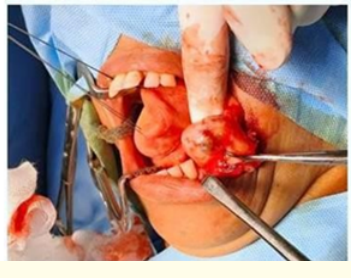

Figure 2: Showing enmasse removal of the tumor along with the accompanying duct.

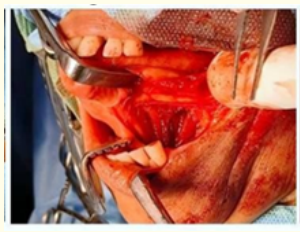

Figure 3: Showing floor of the mouth with underlying muscles after wide local excision.

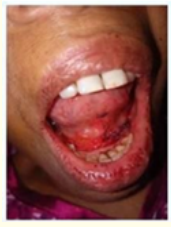

Figure 4: Shows mucosal closure post procedure.

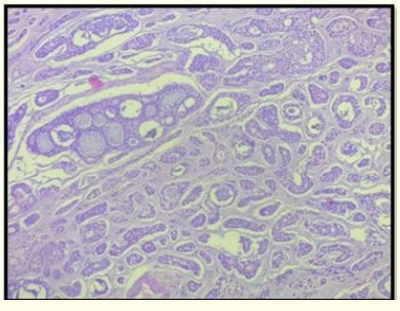

Figure 5: Histopathology showing adenoid cystic adenocarcinoma (AdCC) showing 41-50% tubular pattern, 51-60% cribriform pattern and solid component of 1-10% (with 10X).

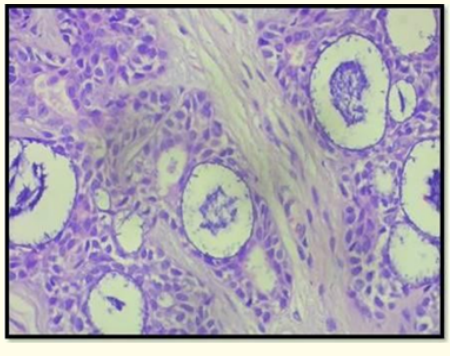

Figure 6: HPE with 40X.

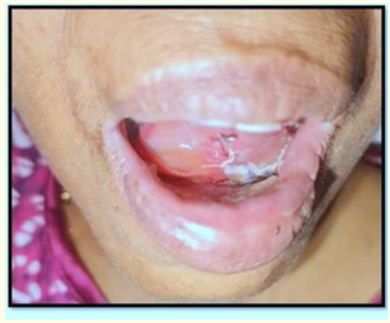

Figure 7: Immediate postoperative image showing sutures over the excised part of the floor of the mouth.

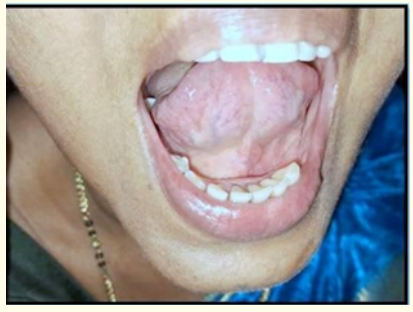

Figure 8: Follow-up image after 1 year post surgery

Discussion

Adenoid cystic carcinoma (AdCC) is a distinct and rare kind of salivary gland malignancy distinguished by its slow but aggressive activity, high rate of perineural invasion, and proclivity for late distant metastases, notably to the lungs. AdCC of the sublingual gland is extremely rare entity, accounting for less than 1% of all salivary gland carcinomas among which AdCC of the minor salivary gland has the worst prognosis [5]. Robin, Lorain, and Laboulbene originally reported AdCC's cylindrical shape in two studies in 1853 and 1854 [6]. In 1856, Billroth used the term "cylindroma" to describe AdCC lesions. The lesion's cribriform appearance was caused by tumor cells with cylindrical pseudolumina or pseudospaces. ACC was utilized to identify the tumor [7,8].

AdCC is histologically classified into three patterns: cribriform, tubular, and solid [9]. The cribriform pattern is the most prevalent and is often linked with a better prognosis than the solid pattern, which has a higher risk of recurrence and metastasis [10]. Perineural invasion, a characteristic of AdCC, is present in the majority of patients and contributes to the high recurrence rate. Microscopic examination of AdCC reveal a malignant tumor consisting of myo-

epithelial and ductal cells organized in tubular patterns in most places. Solid nests and linear cords of single files were seen. Tumor cells exhibited hyperchromatic basaloid nuclei, which were also pale-staining and vesicular in certain locations [11]. Numerous mitotic figures were also observed. The stroma was mucoid and exhibited hyalinization in some areas. The lesion was covered by parakeratinized, atrophic stratified squamous epithelium.

Immunohistochemistry (IHC) analysis shows P63 positivity in peripheral cells of islands and ducts, C-KIT positivity in most tumor cells, and Ki67 positivity in 30% of tumor cells in hotspots [12]. AdCC mostly causes pain due to nerve involvement. Most metastases spread through the lymphatic system rather than the bloodstream. The lungs, bones, liver, and brain are the most common sites for distant metastases. The 5-year survival rate for individuals with head and neck ACC is 90.3% [13,14].

Ultrasonography, CT, and MRI are imaging techniques used to detect AdCC in terms of invasion or abutment in the perineural, vascular, or skull base regions [15]. On ultrasonography, the cystic space of the AdCC is echo-free. However, ultrasonography alone is insufficient to assess deep neck lesions. MRI is the most appropriate method (more accurate than CT and ultrasonography) for diagnosing AdCC. Fat-suppressed T2-weighted MRI is an effective method for evaluating AdCC because it distinguishes lesion intensity based on AdCC histology. Solid ACC has low signal intensity, whereas tubular and cribriform AdCC have higher signal intensities and better outcomes [16].

Tumors arising from the sublingual glands can be diagnosed as mucoepidermoid carcinoma, acinic cell carcinoma, or pleomorphic adenoma. Pleomorphic adenomas are the most common benign tumors in the salivary glands. Pleomorphic adenomas are highly characterized in MRI images, with lobular or spherical shapes and appearing as homogeneous- density masses. Hemorrhage, cystic alterations, and calcification can cause heterogeneous lesions. Acinic cell carcinoma is the second most common cancer in the parotid gland. Mucoepidermoid carcinoma arises in the ductal epithelium. Acinar cell carcinoma and mucoepidermoid carcinoma are comparable to low-grade pleomorphic adenoma on CT and MRI, with neither having any distinguishing characteristics [17]. High-grade tumors are diagnosed by CT and MRI for perineural invasion, which is defined as fatty alterations in the neural foramen and nodular enhancement with a thicker nerve. It is challenging to distinguish benign tumors from malignancies in the sublingual gland based solely on traditional radiographic findings. On the other hand, novel MRI methods like diffusion-weighted MRI, proton magnetic resonance spectroscopy, or dynamic contrast-enhanced MRI might be beneficial.

The most typical treatment for AdCC is radical surgery/wide local excision along with adjuvant radiotherapy, while additional treatments include chemotherapy and concomitant radiotherapy [18]. The impact of adjuvant radiation on the survival rate of AdCC patients is widely discussed [19]. It is now obvious that AdCC is a highly challenging condition to treat. There is no single treatment that can address all of the needs of AdCC management [20].

Conclusion

Lesions from the sublingual glands are rare. In 33% of cases, FNAC leads to incorrect diagnosis such as pleomorphic adenoma, as our patient experienced. Using pre-surgical diagnostic tools like incisional biopsy, MRI, and frozen sectional biopsy during surgery can aid in identifying the pathology and treatment planning. Because of its rarity, there is little information available about the predisposing risk factors and the therapy of patients with severe disease. There is only limited literary work on AdCC of Sublingual gland and no single treatment option is considered as treatment of choice. Also further studies and case reviews have to be carried out to bring in consensus on treatment protocol. Due to the rarity of sublingual gland AdCC, the data on topography of nodal disease is scarce. Hence, there is no consensus as to which nodal levels should be included or the extent of therapeutic neck dissection for such cases.

Author Contributions

Conceptualization, writing original draft – Anand Bhandary Panambur Formal analysis, supervision and writing review - Thoppil Reba Philipose and Pallavi S Compilation of relevant references and editing – Paraashar Rai and Anand Bhandary Panambur

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Conflict of Interest Statement

Authors declare that there is no conflict of interest.

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