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Multimodal Approach For Recurrent Adenoid Cystic Carcinoma Of Sublingual Gland - A Rare Clinical Entity

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Abstract

Sublingual salivary gland tumours are rare and most of them are malignant. We report a case of recurrence of adenoid cystic carcinoma of the sublingual salivary gland in a 52 years old male who presented with swelling in the floor of the mouth with a history of excision of the tumour from the same site 18 months back. Magnetic resonance imaging was reported as a heterogeneous mass in the floor of the mouth in the anterior midline. Surgery was performed followed by reconstruction with free flap and regional flap. Histopathology was reported as Adenoid cystic carcinoma. Postoperative adjuvant radiotherapy was recommended and given. Immunohistochemistry was CD117 positive and was started on tablet Imatinib mesylate as adjuvant therapy. An aggressive approach with radical resection is advisable in recurrent tumours as it provides improved control of disease with adjuvant therapy followed by lifelong follow-up for a better prognosis. The patient remains free of tumour after 18 months follow-up.

Keywords: Adenoid cystic carcinoma, Imatinib mesylate, Recurrence, Sublingual salivary gland

Introduction:

Adenoid cystic carcinoma (ACC) accounts for 1-2% of head and neck cancers and 10% of all salivary gland neoplasm [1, 2]. ACC is characterized by its slow and relentless growth having its peak incidence in 6th decade [3, 4]. Incidence in submandibular gland is 15-30%, 30% in minor salivary glands and parotid gland accounts for 5-15% [2, 5]. Sublingual salivary glands accounts for 0.3%-1% of epithelial salivary glands tumours, out of which ACC accounts for 36% [4, 6].

Case Report:

A 52 years old male patient reported with a chief complaint of swelling in floor of mouth from past six months which was insidious in onset, small in size and had gradually progressed to present size. History of excision of a lesion was done 18 months back at the same site which was reported as ACC of right sublingual gland; without any adjuvant treatment. Patient was asymptomatic for one year after which he redeveloped symptoms. On extraoral examination no cervical lymphadenopathy was noted. Intraoral examination, on inspection anterior floor of mouth was raised bilaterally with well-defined solitary swelling of approximately 3cms in size from lower left first molar to right first premolar region. On palpation the swelling was abutting lingual surface of anterior mandible, soft to firm in consistency, with mild tenderness. On investigation, MRI and PET-CT scans was done and MRI was reported as well-defined

heterogeneous mass in the floor of mouth in the anterior midline, extending along the sublingual spaces bilaterally of 30×24×27mm (mediolateral×anteroposterior×craniocaudal) in close relation to the posterior surface of symphysis, involving portions of genioglossus, geniohyoid, insertions of submandibular duct and mylohyoid, no evidence of neural spread (Figure 1). PET-CT ruled out distant metastases. Surgery was opted with radical approach considering recurrence. Bilateral Selective neck dissection (level I-IV), compartment resection of the lesion was done including- mandibulectomy (browns class III), floor of mouth, anterior 2/3 glossectomy, bilateral lingual nerve, bilateral hypoglossal nerve were excised and included in the specimen (Figure 2). Reconstruction was done using fibula flap and pectoralis major myocutaneous flap. Histopathology was reported as ACC, rpT2N0 with solid variant with no perineural and lymphovascular invasion (Figure 3). In Immunohistochemistry it was CD117 positive. Adjuvant radiotherapy of 6600cGy was given. After a month of post radiotherapy tablet Imatinib mesylate 400mg 1OD was started as adjuvant therapy. The patient remains free of tumour after 18 months follow-up.

Discussion:

ACC is uncommon malignancy with persistent recurrence pattern and propensity for distant metastasis [6]. The genomic hallmark of ACC is a recurrent t(6;9)(q23; p23) translocation that results in a fusion between the MYB and NFIB genes [7]. Clinically it is usually asymptomatic with painless swelling, having behavior more like a benign mixed tumour with no features to assist its diagnosis, however with neural involvement is highly suggestive of ACC. Pain, discomfort, ulcerations, paraesthesia is usually noted in advanced and recurrent tumours [6, 8]. Bony involvement in ACC is different as it spreads through marrow spaces with little cortical and trabecular destruction either through direct extension or via embolic metastases [6, 9]. Any submucosal lesion which is non-ulcerating should be considered as ACC until otherwise proved by biopsy. In our case patient presented with submucosal well-defined swelling which was soft to firm in consistency and mild tenderness indicating of the recurrence of ACC. In our case tumour was in sublingual space, intraductal spread of tumour could be a possibility of recurrence [4].

Histologically ACC is biphasic composed of ducts and basal /myoepithelial cells [4,10]. Broadly it can be divided into three types, solid, tubular and cribriform [4]. Perineural invasion is reported in upto 76% of ACC, commonly in advanced and recurrent tumours due to local extension of tumour [5, 6, 11]. In our case there was no perineural invasion which was in favour of good prognosis.

Management is a therapeutic challenge. Aggressiveness of the disease should be considered when it is solid type, size of tumour >3cms, positive surgical margin, positive lymph nodes [12]. For small lesions wide surgical excision including the involved gland and nerve with adequate margin would suffice [13]. Whereas in tumour larger than 2cms, recurrent tumours aggressive approach of radical enbloc resection is advisable [4,13]. Other treatment option could be radiotherapy, chemotherapy, brachytherapy, targeted therapy or combined modalities [4, 12, 14]. In local recurrence salvage surgery is a good option to improve the survival and quality of life whenever feasible [15]. Aggressive salvage surgery was opted in our case considering recurrence and with the aim of long term survival, to get R0 resection with adequate margin. Management of neck is controversial in ACC. Regional lymph node involvement is seen in 4-13% with incidence of occult lymph node metastases is 14-33%, so it is advisable to perform elective neck dissection in T3, T4 cases [6, 14]. In our case neck dissection was performed to facilitate en-bloc resection of the regional nerves, for planning of adjuvant therapy. Surgery with adjuvant radiotherapy is considered to have better control than a single modality alone and was done [2, 13, 14, 15, 16]. Adjuvant radiotherapy of 6600cGy was given in our case considering recurrence.

Targeted therapies have been given, utilizing the C-kit overexpression in the pathogenesis of ACC [17]. Imatinib mesylate acts

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by inhibiting proliferation and causes apoptosis or both in KIT expressing cells [18]. Imatinib mesylate as an adjuvant therapy was started considering CD117 positive immunohistochemistry, local recurrence in a very short span and solid histological variant to prevent recurrence.

Conclusion:

Painless submucosal intraoral swelling should be treated with high index of suspicion for salivary gland tumours, with pre-operative fine needle aspiration cytology confirmation, followed by wide local excision of lesion along with inclusion of nerve with adequate margins. Life-long follow-up must be adopted for all patients with ACC.

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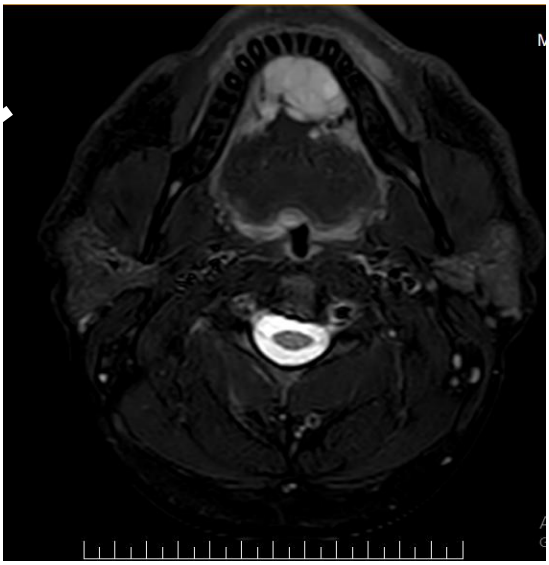

Figure 1: T2w Mri Scan: Arrow Showing Well Defined Heterogeneous Mass In The Floor Of Mouth In The Anterior Midline, Extending Along The Sublingual Spaces.

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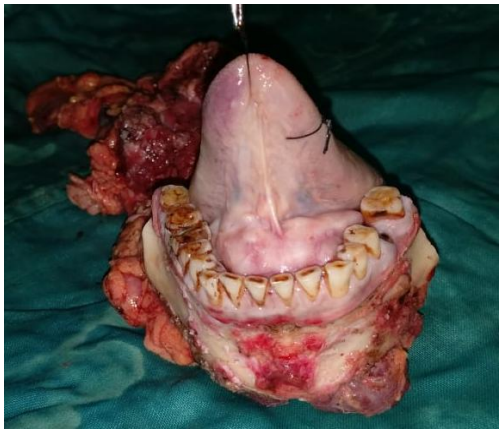

Figure 2: Resected Specimen

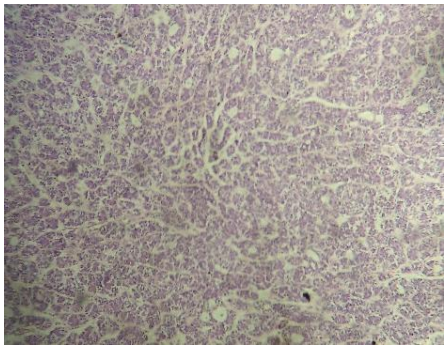

Figure 3: Histopathology: Adenoid Cystic Carcinoma- Solid Variant

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