

An Outlook on Variants of Squamous Cell Carcinoma Amidst the Dura and Pleura with a Case Report on Ackerman's Tumour of Hard Palate

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Abstract

Squamous cell carcinoma (SCC) is the most frequently encountered malignancy by head and neck surgeons. There are certain variants of SCC that are classified as individual tumours by the World Health Organization (WHO). The histopathologic picture of these malignant tumours guides the surgeon in evaluating the prognosis. These variants require special attention owing to fact that there's gross difference in their presentation and management. Ackerman's tumour is the most commonly encountered variant. A case of Ackerman's tumour involving the hard palate and lower labial mucosa is discussed.

Keywords: Ackerman's tumour, Carcinoma Cuniculatum, Variants of Squamous cell carcinoma.

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INTRODUCTION

Squamous cell carcinoma (SCC) is the most frequently encountered malignant mucosal neoplasm of head and neck, accounting for over 90% of all such neoplasms[1]. It is well established by now that oral SCC has multiple variants which manifests it in various forms and that each variant has a distinct clinicopathological appearance and are associated with risk factors which are unique to individual variants. Treatment options usually include surgery, chemotherapy, radiotherapy, phototherapy and cryosurgery.

CASE HISTORY

A male patient, aged 42 years came to our department complaining of growth in the left back tooth region and difficulty in opening mouth since two months. Patient had a history of pan chewing thrice daily for 25 years. On local examination, a well-circumscribed yellowish white fungating proliferative growth of approximately size 4x3 cm was seen on hard palate extending anteriorly from mesial surface of 26 not crossing the midline; posteromedially to soft palate and anterior faucial pillar and posterolaterally to the

maxillary tuberosity and left buccal mucosa (Figure 1). Another well-circumscribed lesion of approximate size 1x0.5 cm was seen on the lower labial mucosa as well (Figure 2). Blanching was seen bilaterally on the buccal mucosa. Uvula was not involved. On palpation, the lesion was indurated and tender. Vertical fibrous bands were palpable in the buccal mucosa. Left Submandibular lymph nodes were palpable, hard in consistency, non-tender and were not attached to the underlying structures. An incisional biopsy was performed which got reported as Ackerman's tumour (Figure 3). Wide local excision and obturation of the defect by an acrylic plate was the proposed treatment. Patient was not willing for treatment and got discharged against medical advice.



Fig-1: A well circumscribed yellowish white proliferative lesion of hard palate



Fig-2: A lesion of similar presentation as in Fig-1, involving the lower labial mucosa

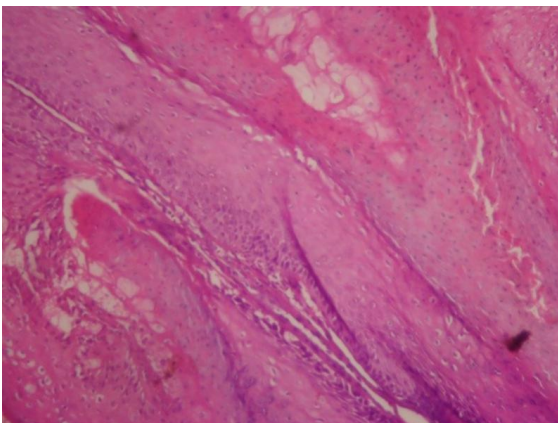


Fig-3: Incisional biopsy revealing Ackerman's Tumour, showing characteristic verrucous surface with hyperparakeratinized stratified squamous epithelium with elongated broad rete ridges and areas of parakeratin plugging

DISCUSSION

Our aim is to discuss the aetiology, clinical features, macroscopic and microscopic features and the management techniques for multiple variants of SCC.

We come across these tumours as ulcerative, exophytic, flat, polypoidal or pebbly lesions. Ackerman's tumour, customarily known as verrucous carcinoma (VC), is a low grade well-differentiated variant of SCC which is typically seen to affect elderly men. Koch BB and associates in their study of 2350 patients demonstrated that tumours originated most frequently in oral cavity (55.9%) and larynx (35.2%), when head and neck sites were considered [2]. They also demonstrated that oral cavity tumours were common among older females. When oral cavity sites were considered, buccal mucosa is the most common site, unlike our case; which involved the hard palate, is an uncommon site for VC. VC is typically considered as an unrushed whitish growth, characterized by finger-like projections, and is usually well-demarcated lesions. They seldom metastasize to regional lymph nodes, but can grow up to 10 cm in size and can cause local destruction of the soft tissues. Hence, bony resection and neck dissections are not performed usually [3, 4]. The typical histologic features of verrucous carcinoma include a verrucous surface with parakeratinized clefts, exo-endophytic growth pattern, bulbous rete ridges, pushing border with no infiltration, subepithelial inflammatory infiltrate with cytologically benign characteristics. Vertical church-spire like keratinization and parakeratin plugging is among the main histological findings of this tumour. Surgical management is the mainstay treatment. Mucosal excision has to be performed taking care to involve the depth of the lesion. The depth of excision in the non-keratinized mucosa would be till the muscle fascia; the buccinator fascia in the buccal mucosa. In the areas of keratinization, resection should be taken till the supraperiosteal plane; VC involving gingiva requires a similar resection. Usually, a 1 cm clinically uninvolved mucosal margin is taken to avoid recurrence. Marx advises a 2-cm margin for the posterior and floor-of-the-mouth resection since Ackerman's tumour is known to spread in this direction and can pursue the salivary flow channels [5]. The role of Radiotherapy (RT) in the management of VC is controversial. Jyothirmayi R and colleagues demonstrated that out of 53 patients who had undergone radiotherapy, 76% had Complete response and 24% had partial response [6]. They demonstrated a five year actuarial disease-free survival of 66% and overall survival of 86%. Mohan and associates noted that compared to Ackerman's tumour treated with surgery alone and SCC treated with surgery and adjuvant RT; VC treated with adjuvant RT had a worse overall survival rate and disease-specific survival. They concluded that, when indicated surgical resection should be performed rather than adjuvant RT if there are positive margins or local recurrence [7-9]. Chen and associates reported treating a case of VC involving the commissure of lip with topical 5-aminolevulinic acid-mediated Photodynamic therapy (ALA-PDT) protocol composed of multiple 3-min fractionated irradiations. It was noted that the lesion showed complete remission in the extraoral aspect of the lip after 6 treatments and

after 22 treatments to the intraoral tumor. No recurrence was identified after 6 months of follow-up [10]. Carcinoma cuniculatum of the oral cavity (OCC) is an extremely rare variant of SCC and was first described by Fliegar S and Owanski T in 1977 [11]. The lesion was named so because it represents crypt-like spaces on histology that resemble burrows created by rabbits (Figure 4). Etiology is linked to chronic irritation; trauma and Human Papilloma Virus (HPV) infection. The World Health Organization has categorized OCC as a separate entity. It presents over wide age groups with a mean of 50 years and a male preponderance [12]. It is encountered as a pebbly slow growing lesion, with deep bony invasion. Unlike VC, it shows well differentiated neoplastic cells with minimal cellular atypia bland cytology, with an endophytic growth pattern. It also shows keratin-filled crypts demonstrating a complex branching pattern. Surgical management is the most advised mode of treatment. Pons and colleagues state that even though it has a locally aggressive behavior, regional metastasis is rare. They conclude that surgical excision of the lesion with free margins provides excellent prognosis [13]. If bony involvement is confirmed, resection of the affected area should be carried out. If clinically involved nodes are present then neck dissection should be considered. Papillary squamous cell carcinoma is a variant of oral squamous cell carcinoma which is not encountered frequently. It exhibits a predilection for the larynx, oropharynx, and sinonasal tract. It is seen commonly in elder men [14]. The tumour is soft to touch, friable and exophytic in nature and is reported from 0.2-4.0 cm in sizes by Suarez *et al.* [15]. The patients usually present with symptoms which are related to the involved site. Local invasion and metastasis into regional nodes are a common finding. Infrequently it is documented to develop from noninvasive papillomas. HPV association is documented recently in a subset of papillary squamous cell carcinomas [16]. It is noted that these HPV related tumours affect younger patients, display a nonkeratinizing morphology, and occur most often in the oropharynx. Even though it is a rare tumour, preliminary studies suggest improved disease-free survival [17]. Typical histological features include exophytic papillary proliferation with fibrovascular cores, atypical squamous epithelial cells (Figure 5). The tumor cells also show nuclear atypia, hyperchromasia, mitotic figures, cancer pearls, apoptotic bodies and individual keratinization. Mild stromal invasion may also be present [18]. Thompson and associates noted that 20.7% of patients exhibiting exophytic pattern had recurrences and succumbed to the disease caused by locally aggressive disease or metastatic tumor. They suggested that these tumors be treated as potentially invasive until proven otherwise [19]. The management technique for papillary carcinoma is similar to that of conventional squamous cell carcinoma, includes wide local excision with nodal dissection for occult nodes in the clinically negative neck [20]. Spindle cell carcinoma also known as sarcomatoid carcinoma of the

head and neck is a biphasic neoplasm, elucidated initially by Virchow in 1865 [21]. Though being a rare form of SCC, it exhibits an aggressive behavioral pattern with an occurrence of 3%. It exhibits a higher male predilection particularly in the 6th and 7th decades of life [22, 23]. Etiology is linked commonly to poor oral hygiene, local irritation by tobacco use and/or alcohol abuse, previous ionizing radiation of the area [24]. The larynx and hypopharynx are the most common sites with oral cavity being an uncommon site. It is usually seen as an exophytic polypoidal growth on lateral surfaces of tongue with high local recurrence rate, and it may involve the regional nodes. Typical histopathological features include atypical spindle cells, pleomorphism, hypercellularity, increased mitotic figures and opacified cytoplasm (Figure 6). Surgical resection with neck dissection is the preferred mode of treatment with or without adjuvant radiotherapy [25, 26]. Adenosquamous carcinoma (ASC) is an extremely rare, aggressive subtype of SCC, exhibiting a peculiar combination of the histological features of both SCC and adenocarcinoma [27]. It was first described as a variant of malignant salivary gland tumor by Gerughty *et al.* in 1968 [28]. It exhibits a higher predilection in smoking males with a peak incidence in the 5th decade of life. HPV association is also documented [29]. It develops in various extraoral sites including lungs, uterine cervix and pancreas and is a rare finding in upper aerodigestive tract. In the head and neck sites, ASC is predominantly seen in the larynx, paranasal sinuses and oral cavity [30]. Tongue is a common site for ASC, though it affects the other areas as well. Clinically, the patient complaints of pain in the initial days due to its tendency to spread by perineural invasion. Other symptoms include odynophagia, otalgia and oozing from the lesion. It is usually seen as keratotic ulcers to nodular, exophytic and indurated masses [31]. Histopathology reveals dysplastic stratified squamous epithelium proliferating deep into connective tissue as nests, islands and ducts. In the connective tissue, the ducts are lined by tall columnar epithelium central area of eosinophilic degenerated cells (Figure 6). ASC has a higher grade of nodal invasion, local recurrences and distant metastases and a dismal prognosis [32]. Adenosquamous carcinoma is aggressive than the conventional SCC and mucoepidermoid carcinoma and treatment modality depends on the site and stage of disease with surgery chemotherapy and radiotherapy. Keelawat *et al.* demonstrated that 65% of patients had metastasis into regional nodes, 47% had local recurrence and 23% had distant metastases. They noted a 5-year survival rate of 13% [33]. Basaloid SCC, first described in the head and neck by Wain *et al.* in 1986, is a rare, distinct variant of SCC [34]. Common sites are hypopharynx, base of tongue and supraglottic larynx. It is seen frequently in elder men. It is aggressive, histologically distinct and the occurrence rate is less than 1%. HPV and HSV are linked to BSCC [35]. It represents 2% of head and neck cancers [36]. Tobacco, alcohol abuse and a previous

history of radiation to the head and neck region are considered as strong risk factors [37]. There are no clinical features of BSCC that can distinguish it from conventional SCC. Microscopically both epithelial-like and basaloid tumor cells are seen (Figure 8). Typical peripheral palisading and hyperchromatic nuclei along with a high nucleus-cytoplasm ratio can be observed in BSCC [38]. Wedenberg *et al.* demonstrated that the distant metastasis rate of BSCC was 52% [39]. Surgical excision and neck dissection followed by radiotherapy/ adjuvant chemotherapy is the preferred treatment [40]. Chemotherapy is suggested, because of the high incidence of distant metastasis and the relatively poor prognosis [41]. Yu *et al.* noted that recurrence rate of the primary tumor post-treatment was 33%. They also noted that overall three year survival rate was 53%, and the 5- year survival rate was 32% [42].

CONCLUSION

It is lucid that the surgeon must have imperative knowledge regarding the variants of SCC and their clinicopathological presentation for treatment planning and prognosis determination.

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