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Oral variant of Acantholytic Squamous Cell Carcinoma: Report of a case and review of the literature

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Abstract

Adenoid (acantholytic) squamous cell carcinoma (ASCC) is an uncommon histological variant of squamous cell carcinoma. It was first invented by Lever in 1947. It is usually seen among males which mainly occurs in the sixth and seventh decade of life. ASCC has been reported to discover in the sun exposed skin of the head and neck and in other sites. However, ASCC located in the oral cavity is very rare. It is characterized by a combination of classic SCC and pseudo glandular structures, dyskeratotic cells and prominent acantholysis. Here, the patient was a 50-year-old man who presented with an ulcer on the right maxillary alveolar mucosa. The biopsy was diagnosed as ASCC. Histopathologically, acantholytic pattern was seen all over the tumor.

Keywords: Acantholytic squamous cell carcinoma, adenoacanthoma, adenoid squamous cell carcinoma, oral cavity, squamous cell carcinoma.

Introduction

Oral squamous cell carcinoma (OSCC) is the most common malignancy encountered in the oral cavity.[1] It has several histopathological variants, such as spindle squamous cell carcinoma, basaloid squamous cell carcinoma, adenosquamous cell carcinoma and papillary squamous cell carcinoma.[1, 2] These variants have usually been found to have a more malignant course with a poorer prognosis. ASCC differs in histologic features and aggressiveness. The overall incidence of ASCC is only 0.1%. The lesion on the skin appears as elevated nodules that may show crusting, scaling or ulceration. It is commonly seen in the sun exposed areas of adults. Their

occurrence in the oral cavity is uncommon and confers bad prognosis. The peak incidence of the oral ASCC is in the sixth decade. [3] Adenoid squamous cell carcinoma is derived its name from the pseudo glandular appearance resulting from acantholysis and degeneration within the islands of SCC. But there is no evidence of glandular differentiation, secretory activity or products. [1] We report a case in a 50-year-old male and review the pertinent literature.

Case Report

A 50-year-old male presented with a painful bleeding ulcer in the upper right back region of oral cavity which he observed since 2 months. Patient gave history of smoking since 25 years who smoked 1.5 packs of beedies per day. The general condition of the patient was normal and his medical history was noncontributory. On extraoral examination there was facial asymmetry detected. Intraorally on inspection an ulceroproliferative growth was noted on the right maxillary alveolar mucosa measuring 3 × 3 cm. The ulcer had uneven margins and a necrotic base covered with yellowish slough [Figure 1]. On palpation the lesion was firm in consistency, with smooth surface, movable and not fixed. Cervical lymph nodes were normal. To evaluate the bone involvement both paranasal sinus view and orthopantomography was taken which revealed a hazy radiolucency and destruction of floor of the maxillary sinus. Based on the clinical findings and history a provisional diagnosis of carcinoma of maxilla was made. Excisional biopsy was done and histopathological examination showed lesional tissue with proliferation of highly dysplastic epithelium into connective tissue showing nuclear hyperchromatism, altered nuclear cytoplasmic ratio, numerous normal and abnormal mitotic figures. Individual cell keratinisation was also evident. Nests of malignant squamous cells revealing acantholysis in the centre and focal pearl formation giving pseudo glandular/ alveolar appearance [Figure 2 and 3]. Focal areas of typical squamous cell carcinoma were also found. Intervening connective tissue was scanty and was diffusely infiltrated with chronic inflammatory cell infiltrate. Dilated vessel like spaces was observed. These were lined by a single layer of unusual epithelioid cells. The overlying stratified squamous epithelium showing dysplastic features is also evident in the lesional tissue. A final diagnosis of acantholytic SCC was arrived at based on these findings. The patient was referred to a higher specialized maxillofacial surgery for further diagnostic work-up management.

Discussion

ASCC synonyms include, adenoid squamous carcinoma, angiosarcoma-like squamous cell carcinoma, pseudo glandular squamous cell carcinoma, squamous cell carcinoma with gland like (adenoid) features, adenoacanthoma, pseudo vascular adenoid squamous cell carcinoma and pseudoangiosarcomatous carcinoma. [4, 5] It differs from general OSCC in histologic features and its aggressive nature. Histopathologically, cystic degeneration of the neoplastic epithelium, producing a prominent alveolar pattern and pseudo glandular structures with acantholytic cells. [6] This finding is associated with loss of immunohistochemical expression of E-cadherin, causing loss of cell adhesion in the center of the tumor nests. Acantholysis develops as a consequence of the loss of desmosomal adhesion proteins. [6, 7] The acantholytic cells may appear extremely bizarre, large, or multinucleated and mitotic figures are variably present. The loss of the cell-to-cell adhesion leads to morphological changes that mimic other cell types. It is for this reason that acantholytic SCC is mentioned in the literature with numerous different names

(adenoid SCC, pseudo vascular adenoid SCC, pseudoangiosarcomatous SSC, pseudo glandular SCC). [2]

The discohesive tumor cell nests provide resemblance of gland like structures of adenocarcinoma and adenosquamous carcinomas. ASCC is differentiated from adenocarcinomas particularly, adenosquamous carcinomas by lack of true glandular formations and negativity for mucin stains. Histologically, adenosquamous variant of SCC consists of invasive sheets, columns, and strands of dyskeratotic squamous cells which merge with glandular structures with epithelial mucin production. Although ASCC simulates glandular elements, they lack the presence of epithelial mucin production. [7, 8]

ASCC can resemble adenoid cystic carcinomas due to the presence of glandular spaces and fibrin in these spaces may masquerade as mucin. However, in ASCC the glandular spaces frequently have angular appearance and mucin stains do not demonstrate evidence of epithelial mucin. ASCC is always accompanied by foci of conventional SCC, suggesting the correct diagnosis. In addition, adenosquamous carcinomas illustrate only focal glandular formations, whereas ASCC show pseudo glandular formations throughout the lesion. If these lesions are seeming as glandular and a squamous component also is identified, mucoepidermoid carcinoma (MEC) may enter the differential diagnosis. Low and intermediate grades of MEC demonstrate easily definable mucin and rounded glandular spaces. High grade MEC never has profuse glandular formations. ASCC differs from common squamous cell carcinoma not only histologically but also by its aggressive nature. When it occurs in the oral cavity, it is related with a poor prognosis. [8] The review of cases reported in the literature has showed that the prognosis of these lesions is relatively poor when compared to conventional squamous cell carcinoma. Therefore, precise histopathological diagnosis can help the clinician to plan accurate treatment for the patients.

Very few immunohistochemical studies are implemented on oral ASCC. ASCC of the oral cavity as described in a study showed positivity for CK7, CK8, CK19, E - cadherin and p53 but was negative for vimentin, CK 20 and S100 protein.[9] The major role of immunohistochemistry in this scenario is to differentiate it from other confounding entities.

Conclusion

ASCC is a variant of squamous cell carcinoma which occurs within the oral cavity infrequently. ASCC definitely poses diagnostic dilemma. Close examination to the morphological pattern of acantholysis will reveal the true nature of this lesion. The review of case reported in the literature has showed that the prognosis of these lesions is relatively poor when compared to conventional squamous cell carcinoma. Therefore, precise histopathological diagnosis can help the clinician to plan a more aggressive multidisciplinary treatment for these patients.

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Figure 1: Erythematous ulcerative growth involving the maxillary right alveolar ridge.

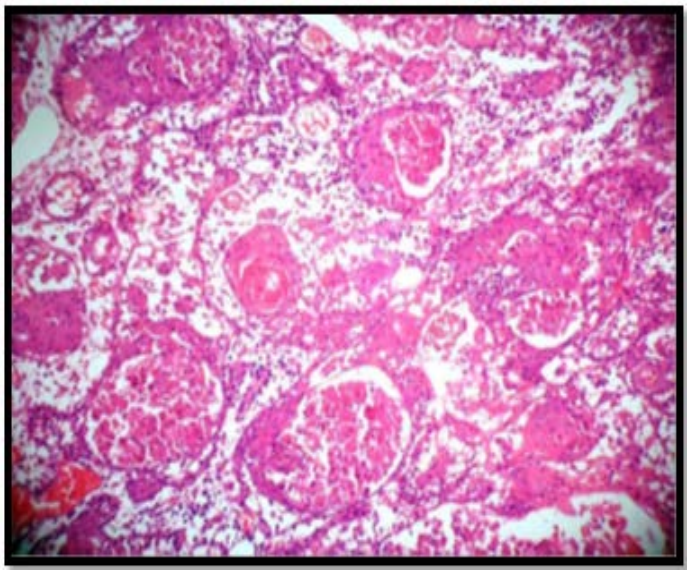

Figure 2: Epithelial tumor islands within connective tissue showing acantholysis and loss of cellular attachment (H&E, 10x).

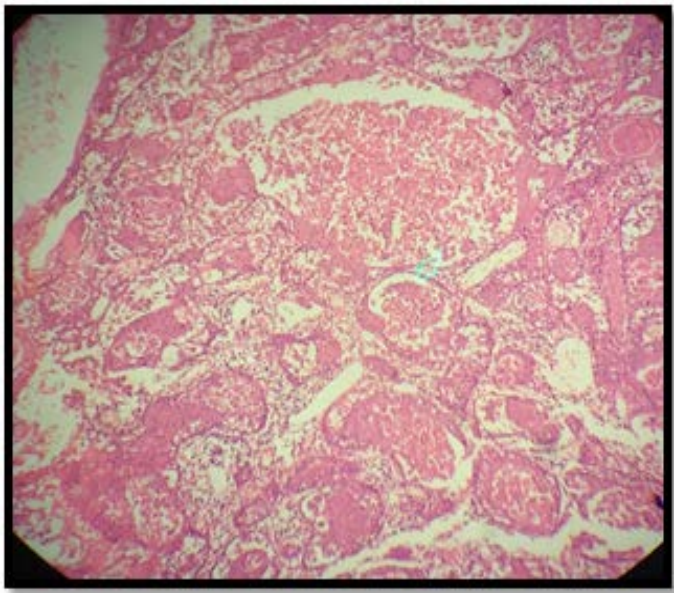

Figure 3: Space giving the pseudo glandular presentation (H&E, 10x)