Case Report

Adenoid Cystic Carcinoma – A Case of an Aggressive Ulcerated Lesion of the Upper Lip

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Abstract

Adenoid cystic carcinoma (ACC) is an unusual slow growing salivary gland malignancy with higher chances of recurrence. It is characterized by the proliferation of ductal (luminal) and myoepithelial cells in cribriform, tubular, solid, and cystic forms. Standard treatment, including surgery with postoperative radiation therapy, has attained reasonable local control rates, but distant metastases do not allow any improvement in the survival rate. We present a case of a 50-year-old female diagnosed with ACC involving almost the whole of the upper lip, with an aim to highlight its histologic evolution from it being clinically seen as severely ulcerated and necrotised upper lip mimicking a case of squamous cell carcinoma and its prognosis.

Keywords: Adenoid cystic carcinoma (ACC), salivary gland malignancy, World Health Organization (WHO)

Introduction

The World Health Organization in 2005 defined "A basaloid tumour consisting of epithelial and myoepithelial cells in various morphological configurations, including tubular, cribriform and solid patterns. It has a relentless clinical course and usually a fatal outcome" and called it as adenoid cystic carcinoma (ACC)[1,2] ACC, although rare, is a deadly malignant tumor and it can be associated with adverse clinical course and poor prognosis which leaves the clinician and histopathologist clueless to understand its biological behavior.[3] It is mainly observed in minor salivary glands.[1,2,4] It usually manifests as a painless lump, but when it affects the palate, it is observed to be painful and ulcerated.[1,4] Very few cases have been reported having the involvement of minor salivary glands of the upper lip. Common features noted in ACC are its high chances of recurrence, perineural spread and its insidious and distant metastasis.[1,4] Although the histopathological features are histopathognomic, its course and behavior is unpredictable. This case report highlights the histologic evolution of the entity which was clinically seen as severely ulcerated and necrotized upper lip mimicking a case of squamous cell carcinoma, which, after histopathological examination, was diagnosed to be a case of ACC.

Case Report

A 50-year-old female patient presented to the Oral and Maxillofacial Pathology Department of the college for the evaluation of the ulceration on her upper lip, involving the philtrum, which was of 2-year duration. The patient gave the history of chewing tobacco habit, multiple times a day for about 10 years. Clinical examination revealed a large destructive ulceration with induration measuring about 7 cm × 3 cm × 5 cm, extending beyond the middle third of the upper lip area, involving the outer skin and exposing the upper anterior mucosa [Figure 1a and b]. There was no regional lymphadenopathy. She was advised for a computed tomography scan which revealed an ulcero-infiltrative soft-tissue lesion which measures approximately 4.1 cm × 1.8 cm × 1.3 cm involving the upper lip extending across the midline, causing erosion of the outer cortex of the superior

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alveolar arch in the region of the incisor and canine teeth. It was provisionally diagnosed to be squamous cell carcinoma.

Incisional biopsy of the ulceration was performed and was sent to the Oral and Maxillofacial Pathology Department for histopathological examination.

Microscopic examination of the hematoxylin- and eosin-stained tissue revealed cribriform islands with cystic spaces lined by basaloid epithelial cells with hyperchromatic nuclei and scanty eosinophilic cytoplasm. The cells showed mild-to-moderate atypia. The cystic spaces are separated by hyalinized stroma and contained scanty mucoid material. Areas of ductal, tubular, and pseudo-glandular structures lined by epithelial cells and islands of basaloid hyperchromatic cells are evident. The connective tissue was also observed to be hyalinized and interspersed with adipose tissue and extravasated blood elements. Overlined epithelium was seen to be parakeratotic with blunt rete ridges without any dysplastic features [Figures 2 and 3].

The final diagnosis was given to be ACC.

Clinical course

The patient was advised the need of surgery. Her physical status was poor and was also a rheumatoid arthritis patient. Some abnormal auscultatory findings were observed, and hence, it was required for her to consult a pulmonary doctor. It was also predicted to have a high chance of perineural invasion involving neck region and poor prognosis. However, due of financial constraints, she decided to not go ahead with the surgery, instead start with ayurvedic treatment.

Discussion

The very first case of ACC involving the upper lip was reported by Pizer and Dubois in 1985, but initially, it was described by Robin and Laboulbene in 1853.[2,4,5] Primary site of occurrence is the palate comprising all malignant palatal tumors,[4,5] but this can show a rare presence involving minor salivary glands of lower lip, retromolar tonsillar pillar region, sublingual gland, buccal mucosa, and floor of the mouth.[5] Gorlin *et al*. pointed out that the incidence of ACC in the upper lip was ~3%.[6] Very few cases have been reported involving the minor salivary glands of the upper lip.[5] ACC can be circumscribed or infiltrative in nature, comprise multiple cell types (ductal and myoepithelial), and may exhibit a variety of architectural patterns (solid, cribriform, and tubular).[7] In our present study, we have observed a mixed type of pattern, involving all the three architectural patterns. Theodore Billroth described the long amorphous sections as cylinders and named it as "cylindroma."[1,2]

ACC can be graded as:

- Well-differentiated ACC:
- The tumor:
  - Shows a tubular pattern
  - Is composed of tubules or nests enclosed by hyaline material.
- Moderately differentiated ACC

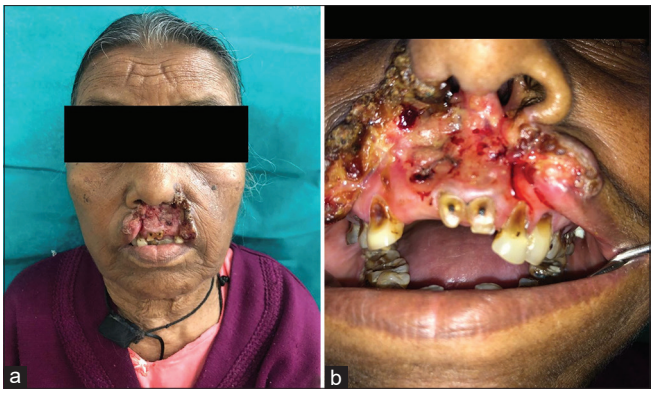

Figure 1: (a and b) a large destructive ulceration with induration measuring about 7 cm × 3 cm × 5 cm, extending beyond the middle third of the upper lip area, involving the outer skin and exposing the upper anterior mucosa

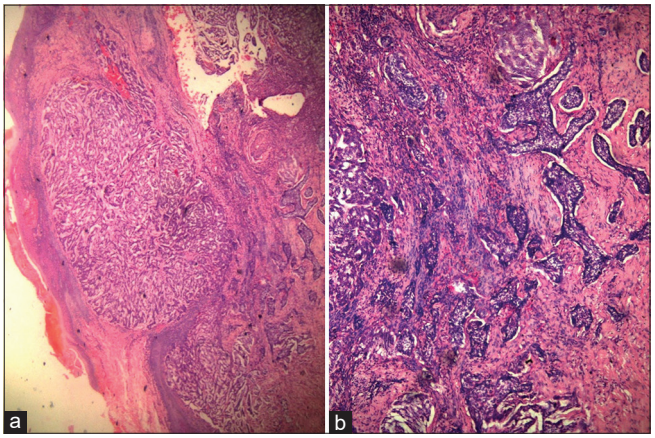

Figure 2: (a) H and E-stained soft-tissue section showing cribriform islands with cystic spaces (×4). (b) H and E-stained soft-tissue section showing cribriform islands with cystic spaces (×10)

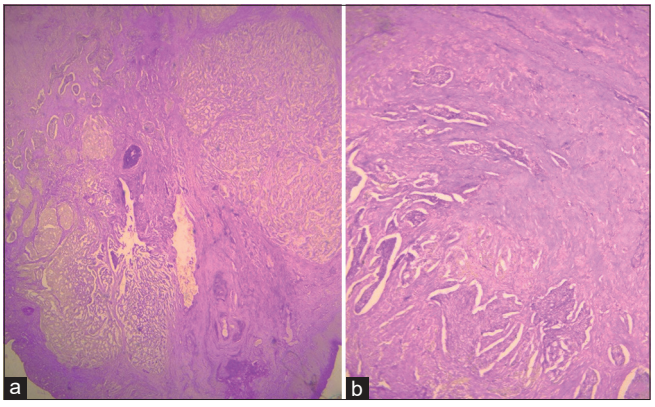

Figure 3: (a) Cribriform epithelial islands surrounded by hyalinization (×4). (b) Cribriform epithelial islands surrounded by hyalinization (×10)

- The tumor:
  - Grows in a cribriform pattern
  - Consists of nests and cords of tumor cells, with multiple cyst-like spaces showing Swiss-cheese pattern.

- Poorly differentiated ACC
- The tumor:
  - Exhibits a solid pattern of growth
  - Composed of anaplastic tumor cells forming solid nests or sheets.[8]

Inside the solid nests, areas of comedo-like necrosis may be evident.

In 1971, Stuart W. Leafsted *et al*. stated that the structural arrangement of the tumor appeared to be dependent on the type of tissue invaded by it.[9] When the tumor invaded open spaces and cavity, glandular or solid patterns were predominant, whereas, when it invaded solid tissue, cord-like arrangement was predominant. The tubular pattern represented the best differentiated form of ACC, where the nests exhibit minimal proliferation of neoplastic cells. As cell proliferation progresses, the units become larger, but the lumens are still maintained, producing a cribriform pattern. In some units, cell proliferation becomes more aggressive and lumens are overgrown, thus forming the solid pattern.[2] Literature suggests that the solid form of ACC is associated with worst prognosis compared with other histological types.[5,10,11] Histologically, our case presents mixed pattern of tumor cell proliferation with areas of hyalinization.

Conclusion

ACC of the upper lip is very rare, and this case report presents on the aggressive ulcerated mass of the upper lip and highlights the need to consider ACC as differential diagnosis to other ulcerated lesions of the maxillofacial region.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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