Case Report

Unusual presentation of adenoid cystic carcinoma (ACC) on lip mimicking mucocele: A rare case report with review

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Abstract

Adenoid cystic carcinoma (ACC) is a relatively rare, slow-growing low-grade malignant salivary gland tumour with high recurrence. It accounts for 1% of all malignant tumour of the oral and maxillofacial region. A high incidence of cases occurs between the fifth and sixth decades of life. Surgical excision with required radiotherapy, chemotherapy, and combined therapy are possible treatment choices. In this present case, a 36-year-old male patient was diagnosed with ACC on the upper lip. The clinical presentation of this lesion resembled mucocele. There are only a few cases reported of ACC involving the lip. Complete surgical removal was done under local anaesthesia, and post-operative healing was uneventful. After 6 months of follow-up, no sign of recurrence was observed.

Keywords: Adenoid cystic carcinoma, ACC, chemotherapy, radiotherapy, recurrence

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INTRODUCTION

Adenoid cystic carcinoma (ACC) is a rare malignant salivary gland neoplasm. The most commonest site of ACC is the palate.[1] Clinical behaviour of ACC depends on the site involved and the affected nerve, and common clinical features are slow growing, small, painless, and occasionally aggressive in nature.[2] The present reported case is a cribriform type of ACC on the upper lip mimicking mucocele without neural involvement, which was surgically excised under local anaesthesia without radiotherapy and chemotherapy. After 6 months of follow-up, it showed no recurrence.

CASE REPORT

A 36-year-old male patient came to the department of Oral Medicine and Radiology with a chief complaint of diffuse swelling on the right upper lip region for 2 years. The swelling gradually increased in size and reached the present size. Past medical history and dental history were non-significant.

Intraoral examination of right upper lip showed painless, fluid-filled swelling with colour ranging from bluish to black and soft in consistency. It was measuring about 7–10 mm extending supero-inferiorly 4 mm away from the vermillion border of the lip to the labial sulcus area and 11 mm from the upper labial frenum to 4 mm away from the buccal frenum on the right side of the lip antero-posteriorly [Figure 1].

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On correlating the history and clinical features, a provisional diagnosis of 'Mucocele' was given. Routine haematological examinations were performed before surgical excision, which was within normal range. Complete surgical excision was planned and performed under local anaesthesia followed by post-operative medication for 5 days. Single bit of soft tissue specimen was fixed in 10% buffered formalin which was greyish, firm to leathery in consistency, and cheese-like material filled inside the sac, measuring about 21 mm in length × 15 mm in breadth × 11 mm in height [Figure 2].

Paraffin-embedded Haematoxylin and Eosin (H&E) stained soft tissue sections under light microscopic view showed variably sized nests of myoepithelial and duct-like cells discretely separated from the uniform band of the collangenised stroma. These nests consisted of small darkly stained uniform basal and myoepithelial cells arranged in a cribriform pattern [Figures 3 and 4]. The cribriform nests were composed of cyst-like spaces filled with periodic acid schiff-positive mucoid material [Figure 5]. In some areas, the luminal and myoepithelial cells had similar appearance and they appeared slightly larger than usual. The connective tissue stroma was hyalinised and surrounded the tumour cells forming a structural pattern of a cylinder. The connective tissue showed no perineural invasion of the nerve bundles [Figure 6]. Based on histopathological findings, a final diagnosis of 'ACC' was made. The patient was not willing to radiotherapy and chemotherapy. Surgical excision was performed under local anaesthesia. The patient was prescribed orally amoxicillin 500 mg with clavulanic acid 125 mg (TDS), metronidazole 400 mg (TDS), Pantoprazole 40 mg (OD), and diclofenac sodium 50 mg (TDS) for 5 days. Post-operative healing was uneventful, on 6 months follow-up there was no sign of recurrence [Figure 7].

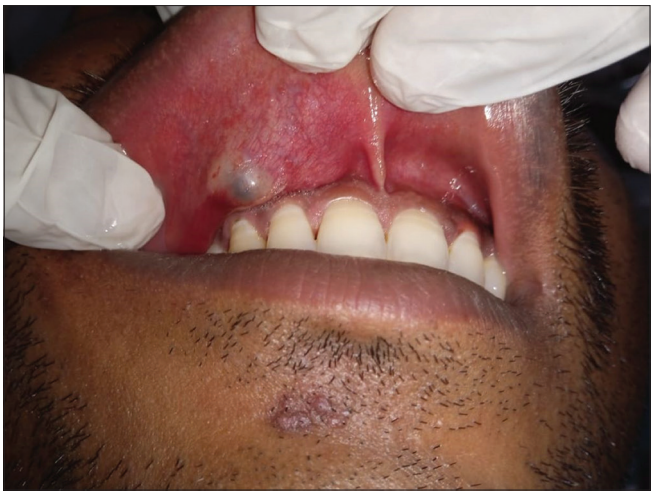

Figure 1: Clinical photograph showing swelling on the right upper lip

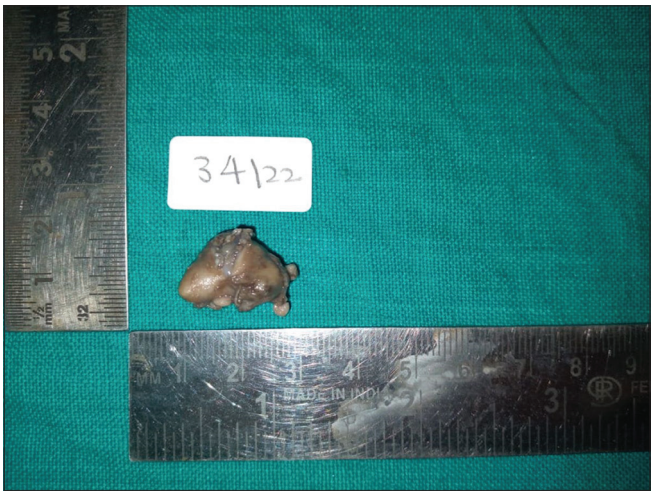

Figure 2: Excised tissue specimen from the upper lip

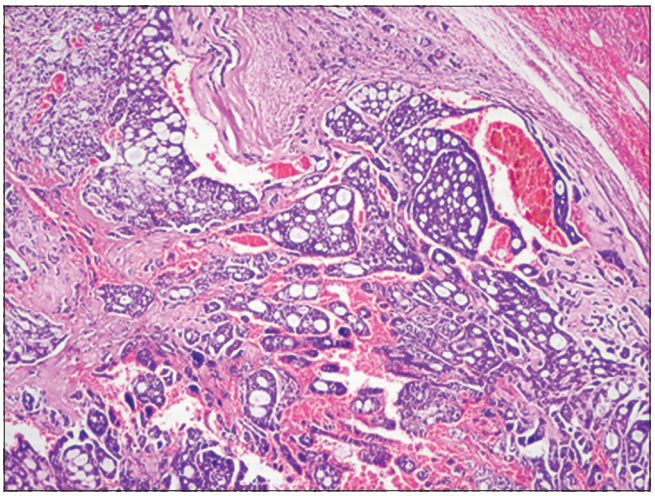

Figure 3: Photomicrograph of the section showed tumour islands circumscribed by fibrous connective tissue capsule (H&E stain × 40)

DISCUSSION

The World Health Organisation in 2005 defined ACC as 'a basaloid tumour containing epithelial and myoepithelial cells in diverse morphological configurations, such as tubular, cribriform, and solid patterns. Its clinical course is relentless and usually has a fatal outcome'.[3] ACC is a rare epithelial tumour with a nonfunctional but persistent growth pattern. ACC occurs predominantly in the fourth to sixth decade of life with a slight female predilection. In our case, a 36-year-old male was diagnosed with ACC. Clinically, ACC is a slow-growing tumour with the property of peripheral nerve invasion. It has a high recurrence rate and metastases to other vital organs. Pain is usually a common and important associated symptom but not always,[2] occasionally pain might be present before the clinical appearance

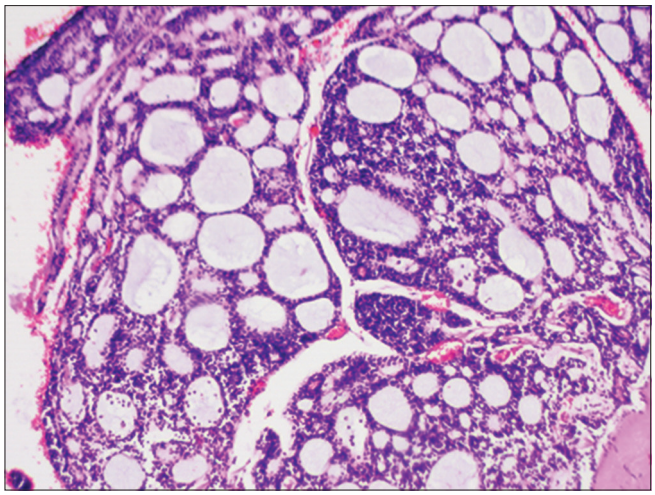

Figure 4: Photomicrograph of the section shows basal and myoepithelial cells arranged in a cribriform pattern (H&E stain ×100)

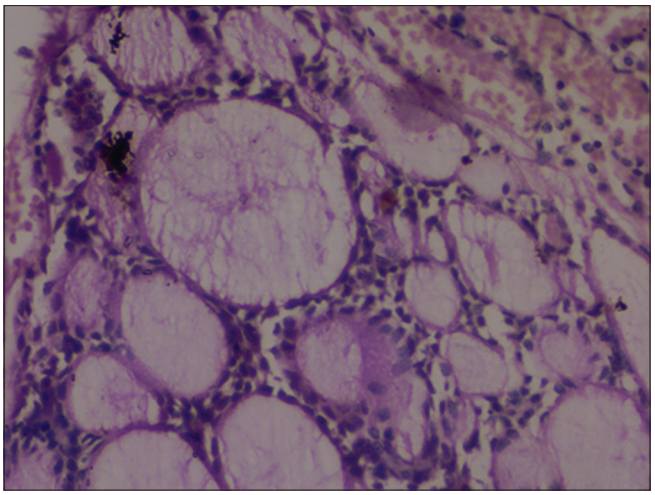

Figure 5: Photomicrograph of the section shows cyst-like spaces filled with periodic acid schiff-positive mucoid material (PAS stain ×400)

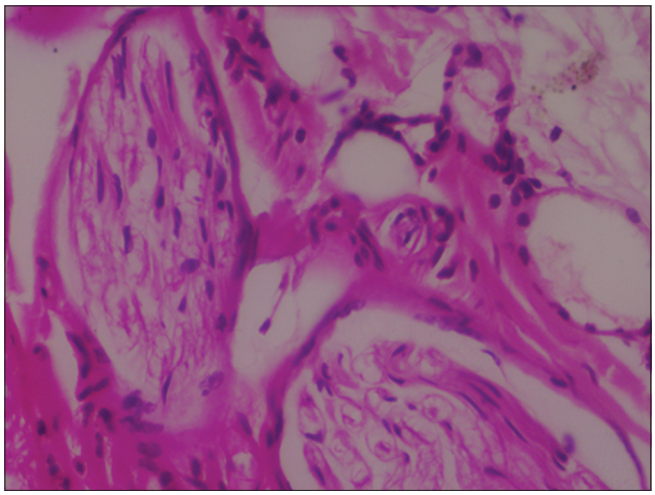

Figure 6: Photomicrograph of the section shows no perineural invasion of the tumour cells (H&E stain ×400)

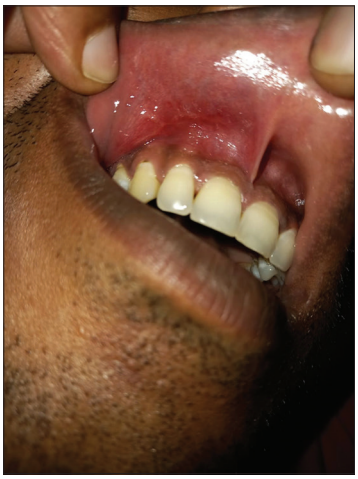

Figure 7: Clinical photograph showing proper healing of the surgical site and no recurrence was seen after 6 months of follow-up

of the disease. Neoplastic cell neurotrophins cause pain, which was not evident in our case.[4] Both major and minor salivary glands could be affected by ACC, among the minor salivary glands, the palate is a more commonly involved site near the greater palatine foramen with a worse prognosis.[4] The most common minor salivary gland neoplasms are mucoepidermoid carcinoma (21.8%) followed by polymorphous low-grade adenocarcinoma (PLGA) (7.1%) and ACC (6.3%).[5] A total of 11 cases of ACC involving the lip have been reported in the literature and only one case was reported in India [Table 1].[2,6,8-14] The first case was reported by Appel, El Attar *et al.* in 1976.[6] The differential diagnosis of ACC includes PLGA, basal cell adenoma, mixed tumour, basaloid squamous cell carcinoma[3], and mucocele (James A and Gunasekaran N *et al.* 2021).[7] This present case had also an appearance similar to a mucocele.

Oral mucocele is defined as mucus-filled pathological cavities in the oral cavity. Mucocele is the 17th most common salivary gland lesions found in the oral cavity. It can involve any part of the oral cavity like lip, cheeks, and floor of the mouth. Clinically, most of the mucoceles are asymptomatic and soft in consistency, pink or bluish, size may vary from 1 mm to several centimetres, and transparent in nature.[15]

Histopathologically, ACC is of three types—cribriform, tubular, and solid. In most ACC cases, all three patterns can usually be observed in a single lesion. In addition, the pattern may vary in different areas of the same mass. The most important and classical feature of it is the 'cribriform' pattern where nests of tumour cells have a 'Swiss cheese' appearance. A second major pattern observed in ACC is the 'tubular' pattern in which elongated tubular structures with a central lumen are

Table 1: Total ACC cases involving lip.[2,6,8-14]

Author's name and year	Place	Age and gender	Clinical presentation	Final Diagnosis	Treatment and follow-up
Appel *et al*. (1976)[6]	United States	62 Years/F	Soft tissue masses in the upper lip.	ACC	Surgical excision/no recurrence
Owens *et al*. (1981)[8]	United States	67 Years/F	A painless, non-tender, slowly growing mass on her left upper lip.	ACC	Wide local excision/Had two time recurrence before surgery, no recurrence after 6 years of a third surgery.
Pizer *et al*. (1985)[9]	United States	NA	A firm mass in the right upper lip.	ACC of the upper lip.	Radiation therapy without surgery/recurrence after 9 months.
Rubio *et al*. (2006)[10]	Spain	65 Years/M	Lesion in the upper lip.	ACC	Radiotherapy/recurrence after 10 years of follow-up.
Mosqueda-Taylor *et al*. (2010)[11]	Mexico	65 Years/M	Growth located in the upper lip.	ACC	Wide excision/no recurrence
Nigri *et al*. (2014)[12]	Brazil	79 Years/F	A 2.0 cm submerse fibrous lobulated nodule covered by normal oral mucosa on the upper lip.	ACC	Surgical excision/no recurrence after 12 months.
Dogra *et al*. (2016)[13]	India	62 Years/M	A solitary, oval-shaped ulcerative growth with induration on the upper lip.	ACC of the upper lip.	Surgical excision/no recurrence after 6 months of follow-up.
Sanchez *et al*. (2017)[2]	Spain (3 cases)	66 Years/M	Painless sub-mucosal nodule in the upper lip.	ACC predominant cribriform pattern.	Surgical excision/NA
		80 Years/F	Nodule on labial commissure.	ACC predominant solid pattern.	Surgical excision/NA
		74 Years/M	Swelling in the right lateral upper lip.	ACC predominant solid growth.	Wide excision/Recurrence after 2 years.
Singer *et al*. (2021)[14]	Turkey	60 Years/M	A 1.5 cm, firm nodule with a slight depression in the inner surface of the right lower lip.	ACC	Wide wedge resection with radiotherapy/no recurrence without 4 years of follow-up.

seen. The third pattern is the 'solid' pattern where the tumour islands are completely filled with basaloid tumour cells without cystic spaces. The cystic spaces may contain basophilic mucinous material or eosinophilic material. Usually, the tumour islands present are surrounded by fibrous connective tissue stroma or hyalinised stroma. Tumour is graded according to microscopic appearance, cribriform or tubular (grade I), <30% solid (grade II), or >30% solid (grade III).[3] Possible treatment choices for ACC are surgical excision, radiotherapy, chemotherapy, and combined therapy. The most common treatment choice is excision with the widest surgical margins, possible as tumour cells extend well beyond the clinical and radiographic margins to prevent a recurrence. Neutron therapy can also use as a primary therapeutic modality for local control. Immunotherapy along with chemo-radiotherapy has recently shown good results.[5] The prognosis of ACC also depends on its infiltrative growth pattern and perineural invasion. This is the reason for the high risk of recurrence of this neoplasm, which could also be due to an incomplete surgical removal.[5]

Immunohistochemistry reveals that the luminal tumour cells are diffusely positive for cytokeratin, epithelial membrane antigen, carcinoembryonic antigen, and CD117 (c-Kit) are positive for ductal lining and those that surround the pseudo cysts show positivity for S-100, smooth muscle actin, calponin, p63, and variable positivity for cytokeratin suggestive of myoepithelial cells. Overexpression of p53 and Ki-67 was found with tumour progression and recurrence.[4]

The long-term survival rate of grade III tumours is particularly low. Distant metastasis occurs in 25–50% of patients, and the lung is the most common site for metastasis followed by bones, liver, brain, and least in the lymph nodes. Five years of survival rate after proper treatment is 75%, but long-term survival rates are low (10 years–20% and 15 years–10%). Tumours limited to lips without neural invasion can show good results as in the present case.[2,3,5] Our case was followed-up for 6 months after surgery and showed no recurrence or complications.

This presented the case in a 36-year young adult diagnosed with ACC, after surgery 6 months of follow-up showed no recurrence and without any complications.

CONCLUSION

The case presented here is an example of a clinical diagnostic dilemma. Clinical findings were suggestive of a benign cystic mass and diagnosis of ACC was not realised until pathologic sections were studied. The present case indicates the need for awareness of the unusual presentation of ACC as a soft and fluctuant cystic mass.

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 initial s 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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