

Intraosseous adenoid cystic carcinoma of maxilla: A rare case report

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Abstract

Adenoid cystic carcinoma (ACC) accounts for approximately 6-10% of all salivary gland tumors. Palatal minor salivary glands, parotid, and sub-mandibular glands are usually affected. Rarely, these lesions arising intraosseously have been reported. Mandible is commonly involved than maxilla. The present case is a giant ACC involving the right maxilla. A thorough clinical and radiographic evaluation was performed to assess the involvement of surrounding vital structures along with a meticulous metastatic work-up. Computed tomography showed a giant lesion in maxilla encroaching the left nasal fossa, antrum, buccal space, and oral cavity. No metastasis was noted. Histological evaluation from multiple sites showed both cribriform and solid patterns. Radiotherapy was given as patient did not comply for surgery. Though central ACC is extremely rare, especially in maxilla, it should be included in the differentials for lesions in maxilla. A prompt diagnosis with treatment and long-term follow-up is advised in such cases.

Keywords: Adenoid cystic carcinoma, central salivary neoplasm, cylindroma, intraosseous

Introduction

Adenoid cystic carcinoma (ACC) is one of the most common and best recognized salivary gland tumors. This was first described by Theodor Billroth as “Cylindroma” in 1859.^[1] It represents fifth most common epithelial tumor of the salivary glands and accounts for approximately 6-10% of all salivary gland tumors.^[2] Palatal minor salivary glands are the most commonly affected locations followed by parotid and sub-mandibular glands.^[1,2] This tumor frequently occurs in the fifth decade of life usually affecting women.^[2-4] Rarely, it may arise centrally (intraosseous) causing bony destruction and affects mandible more commonly than maxilla.^[2-5]

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One such rare case, which presented with a large central lesion in maxilla involving the nasal fossa, oral cavity and maxillary sinus causing gross destruction of the structures involved is reported.

Case Report

A 48-year-old female reported to the Department of Oral Medicine and Radiology with swelling over the left side of face since 4 years. There was history of swelling in the same region 5 years ago for which she was operated. The swelling recurred within a year, which gradually increased to attain the present size. Swelling was associated with dull and mild occasional pain on wide opening of the mouth. There was no history of pus/blood discharge, but had caused mild functional difficulty. There was no history of paresthesia, diplopia or change in resonance of the voice. Her past medical and personal histories were non-contributory.

On extra oral examination left side revealed gross facial asymmetry, with a diffuse swelling approximately 5 cm × 4 cm extending superoinferiorly from about 1 cm above the ala-tragus line up to the left angle of mouth and mediolaterally from philtrum up to the level of lateral canthus of left eye. Obliteration of left nasolabial fold and deviation of the left ala of the nose was noted. The overlying surface of the swelling was stretched and shiny but was normal in color. On palpation, no raise of local temperature was noted. Swelling was non-tender and firm in consistency with no pulsations evident. Examination of regional lymph nodes showed no evidence of lymphadenopathy [Figure 1].

On intraoral examination an irregular, but well-defined swelling was noted involving the left vestibule and hard palate approximately measuring 7 cm × 6 cm extending antero posteriorly about 2 cm from the maxillary anterior

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labial vestibule i.r.t 11.21 until the mesial aspect of 26 and medially crossing the mid palatine raphe on the right side approximately 1 cm away from the marginal gingiva of 13 and extending laterally into the left labial vestibule causing obliteration from 11 to 24. Overlying surface had areas of erythema interspersed with a bluish tint. Swelling was non-tender with firm consistency in the palate and cystic consistency in the left labial vestibule on palpation. Hard tissues in the region of complaint showed spacing in-between 21 and 22 and grade I mobility noted with 22, 23, and 24 [Figure 2].

On correlating the history and clinical findings, a provisional diagnosis of low-grade minor salivary gland malignancy was rendered with mucoepidermoid carcinoma on top of the list followed by the ACC and others. Left maxillary antral malignancies and benign odontogenic tumors namely adenomatoid odontogenic tumor, squamous odontogenic tumor and others were considered in the differential diagnosis list.

Panoramic image showed an irregular osteolytic lesion in maxillary anterior left region approximately 6 cm × 5 cm extending from mesial aspect of 12 up to the distal aspect of 26 antero-posteriorly and superiorly had caused erosion and discontinuity of floors of the nasal fossa and maxillary sinus with soft-tissue density seen infiltrating into the respective structures and inferiorly up to interdental alveolus of 22, 23, 24, and 25. The lesion had an irregular moth-eaten internal architecture and had caused diversion of roots of 11, 21 and migration of 22. Intraoral periapical radiographs confirmed radicular findings along with multiplanar root resorptions seen with 21 and 23.

To assess the exact boundaries and invasion into the adjacent areas, computed tomography with contrast of mid-face was performed, which revealed a heterogeneous enhancing mass in the left maxilla. The lesion had caused bony destruction and erosions with the epicenter in the maxilla and the soft-tissue density was seen extending into the surrounding structures namely oral cavity, maxillary sinus, nasal cavity, buccal space, and upper lip equidistantly suggestive of intrabony origin [Figure 3].

Routine hematological investigations were normal except erythropenia with microcytic, hypochromic anemia seen on peripheral smear.

Incisional biopsy was performed from two separate locations namely, palate, and labial vestibule. On histopathological examination, hyperchromatic basaloid tumor cells were arranged in islands with interspersed multiple cystic areas showing eosinophilic material classically resembling cystic “Swiss-cheese pattern” was seen along with a solid pattern. Hence, diagnosis of ACC was rendered [Figure 4].

As a part of metastatic work-up, clinical and imaging investigations were carried out including clinical examination of the oral cavity and neck, chest radiograms, and abdominal ultrasound were carried over, which showed no evidence of secondaries.

The patient was referred to higher regional oncology institute where surgery followed by the radiation treatment was planned. However, as the patient was reluctant to undergo surgery, lone radiation therapy with a total dose of 60 Gy was executed in the fractionation of 2 Gy/day for 5 days/week for a period of 6 weeks. As the treatment was incomplete and palliative, the prognosis was considered poor, nevertheless, follow-up of the case for 3 months after treatment showed no evidence of recurrence [Figure 5].

Discussion

ACC is a malignant salivary gland tumor, which was earlier known as “Basiloma,” coined by Krompecher in 1908.^[6,7] Later in 1954, Ewing termed it as ACC.^[6,7] It is the second most common malignant salivary neoplasm affecting 50-60% of minor salivary glands followed by the major salivary glands.^[1] Intraosseous occurrence of malignant salivary gland tumors is rare with mucoepidermoid carcinoma being most common followed by ACC.^[1,4,5]

Numerous theories have been proposed to explain the origin of central malignant salivary gland tumors, which include: (1) Entrapment of ectopic salivary gland tissues in jaws (2) neoplastic transformation of odontogenic cyst epithelium and (3) neoplastic transformation of sinus epithelium.^[5]

ACCs occur more commonly in middle aged females however, central lesions may occur at any age (24-82 years) with no gender predilection.^[7,8] They commonly arise in posterior mandible, but seldom in the maxilla with typical primary presentations being pain, swelling and rarely paresthesia and numbness.^[5,8] The characteristic features include late onset, slow growth, insidious destruction of surrounding tissues, perineural invasion, and distant metastasis.^[9] In the present case study, it occurred as swelling in middle aged female with involvement of left maxilla and palate.

In 1979, Batsakis proposed diagnostic criteria for primary intraosseous salivary gland neoplasms^[4], which are:

- Radiographic evidence of osteolysis
- Presence of intact cortical plates
- Presence of intact mucous membrane overlying the lesion
- Absence of any primary tumor within major or minor salivary gland
- Histological confirmation of the typical architecture and morphological features of a salivary gland tumor.

All the diagnostic criteria were satisfied in the present



Figure 1: Extraoral swelling

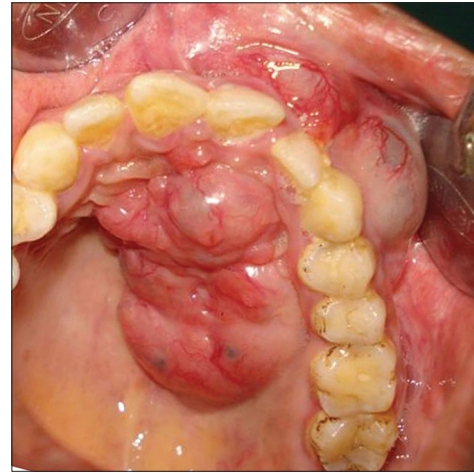


Figure 2: Lobulated, non-ulcerated mass noted over palate and buccal vestibule

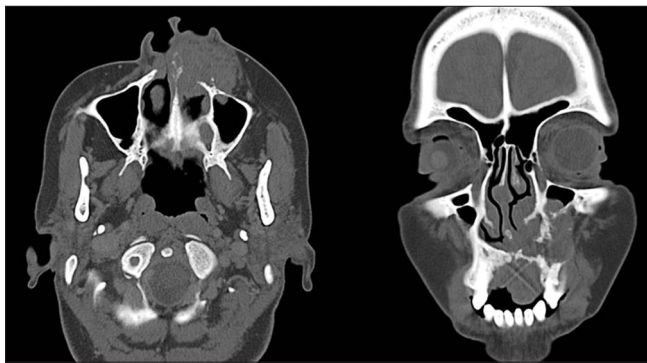


Figure 3: Axial and coronal sections showing irregular destruction of left maxilla with soft tissue mass extending into left antrum, nasal fossa, oral cavity and buccal space

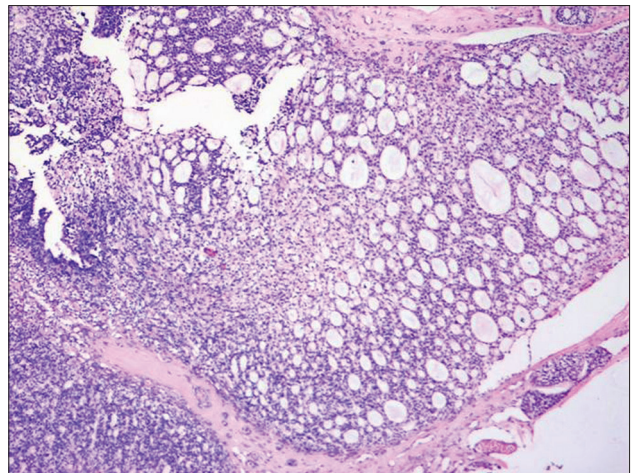


Figure 4: Histopathological section showing "Swiss-cheese" pattern

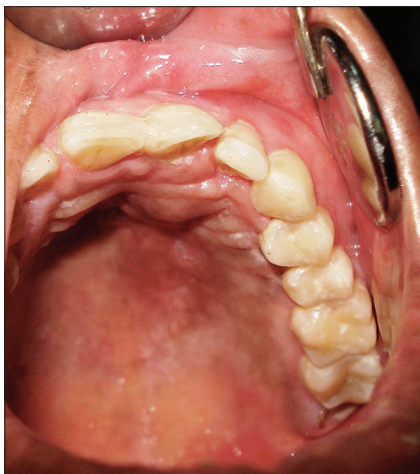


Figure 5: Photograph after 4 weeks of radiation therapy

case except for second and this can be contributed to the chronicity of the lesion, which allowed the lesion to grow to an extensive size. Based on the destruction of bone and cortex, Brookstone and Huvos established a staging system for central salivary gland malignancies^[5] – Stage 1: Lesions with intact cortical plates with no evident bony expansion; Stage 2: Tumors with intact plates, but

intrabony expansion; Stage 3: Lesions associated with cortical perforation or nodal disease. The present case is pertinent to stage three.

Spread to regional lymph nodes is rare, but distant metastasis particularly to lungs and bone are more common and often unpredictable.^[2,10] In the present case, owing to the massive size and a history of 4 years, a thorough metastatic work-up was carried out, which revealed neither regional lymph node nor distant metastasis.

Three histological patterns of growth have been described: Cribriform, tubular, and solid variants. Solid variants of ACC are a high grade lesions with reported recurrence rates of 100% compared to 50-80% for tubular and cribriform variants.^[11] In our case, basilooid cells arranged in both solid and cystic patterns, and the cystic pattern is in the form of "Swiss-cheese" with pseudocysts was observed.

Treatment includes excision with wide margins well beyond

the radiographic margins followed by radiotherapy, especially, when resected tumor borders are not microscopically free of malignancy and when sinonasal areas are involved. Tumors involving nose, paranasal sinuses and maxilla usually have a poor prognosis.^[11]

Conclusion

A rare giant central ACC occurring in the maxilla with all the clinical, radiographic, and histopathological features closely studied have been reported. Central salivary gland tumors though rare should be considered in the differentials for lesions in maxilla. Comprehensive investigations should be carried out for prompt diagnosis and appropriate treatment.

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