Case Report

Intraosseous adenoid cystic carcinoma of the mandible: A rare presentation

Sudip Indu[1], Indranil Deb Roy[2]

[1]Division of Oral Pathology, Army Dental Centre (R&R), [2]Commandant, Armed Forces Dental Clinic, New Delhi, India

Abstract

Central adenoid cystic carcinoma (ACC) of the mandible is a rare entity arising from minor salivary glands. Their clinical and radiographic features may be similar to any odontogenic/nonodontogenic pathology, thus making their precise diagnosis wearisome. ACC is well documented for its protracted clinical course, perineural invasion, multiple recurrence rates and its propensity for distant metastases. The aim of this study is to report a rare case of primary central ACC of the mandible with an unusual presentation in terms of location, however, demonstrating the classical features of ACC in histopathology and positivity in immunohistochemistry to S100, calponin and CD117. A thorough clinical, radiographic, CT evaluation and meticulous metastatic workup along with long-term follow-up is advised in such cases. Although central ACC is extremely rare, especially in the anterior mandible, it should be included in the differentials for lesions of the mandible.

Keywords: Central adenoid cystic carcinoma, immunohistochemistry, mandible

Address for correspondence: Dr. Sudip Indu, Division of Oral Pathology, Army Dental Centre (R&R), New Delhi, India.
E-mail: indusudip30@gmail.com

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INTRODUCTION

Salivary gland carcinomas located centrally within the mandible are rare, comprising <0.4% of all salivary gland carcinomas.[1] Intraosseous adenoid cystic carcinoma (ACC) is an extremely rare neoplasm with only a few cases reported till date causing expansile bony destruction and affecting the mandible more commonly than the maxilla.[2] This tumor frequently occurs in the fifth decade of life usually affecting women and causing pain due to characteristic perineural invasion.[3,4] In terms of site, the palatal minor salivary glands are the most commonly affected locations followed by parotid and submandibular glands. Epidemiologically, the mucoepidermoid carcinomas (MECs) are the most frequently reported types of primary central salivary gland carcinomas of the mandible, followed by ACCs, adenocarcinomas and acinic cell carcinomas.[5]

We hereby report a case of central intraosseous ACC atypically involving the anterior mandible with no distant metastasis in spite of its unusual size.

CASE REPORT

A 71-year-old female patient reported to the outpatient department with the chief complaint of pain in the lower jaw. There was a history of extraction of lower front teeth 31, 32, 33, 41 and 42 5 months back because of continuous pain, which was resistant to analgesics.

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On intraoral clinical examination, an irregular, mild diffuse swelling was noted on the buccal aspect of the anterior mandible. The overlying surface of the swelling was normal in color and showed no associated color changes or gingival inflammation. On palpation, no rise of local temperature was noted. The swelling was nontender and firm in consistency with no pulsations evident. Submental lymph nodes were palpable but nontender [Figure 1].

Panoramic radiograph showed an irregular osteolytic lesion measuring about 6 cm × 3 cm involving the anterior mandible extending from the mesial aspect of 42 regions up to the mesial of 33 regions anteroposteriorly. No diversion or root resorption was noted. All remaining teeth were vital [Figure 2].

To assess the exact boundaries and invasion into the adjacent areas, cone-beam computed tomography (CT) and chest radiograph were advised. Distant metastasis was ruled out with no other lesions of the major or minor salivary glands. Routine hematological investigations were normal [Figure 3].

On correlating the history and clinical findings, a provisional diagnosis of an odontogenic cyst/tumor or an aggressive osteolytic lesion was considered. Differential diagnosis of intraosseous malignancy, minor salivary gland tumor like MEC, ACC were postulated.

On performing incisional biopsy, histopathology revealed moderately collagenous stroma lined by strands of cells forming multiple ductal tubule-like structures with pale eosinophilic mucinous material within the lumen. A focal stromal area shows the presence of numerous ducts such as tubules and pseudocysts mimicking a cribriform pattern in most of the areas. The high-power view revealed cuboidal cells with prominent hyperchromatic nuclei and minimal cytoplasm lining the duct-like tubules. A few cells show clear cell changes as well. The stroma shows the presence of abundant plump fibroblasts, numerous blood vessels and few lymphocytes interspersed among fine moderately fibrillar collagen bundles. No clear perineural invasion was seen. Majority of the areas (>70%) showed a cribriform pattern; hence, the diagnosis of ACC was made [Figures 4 and 5].

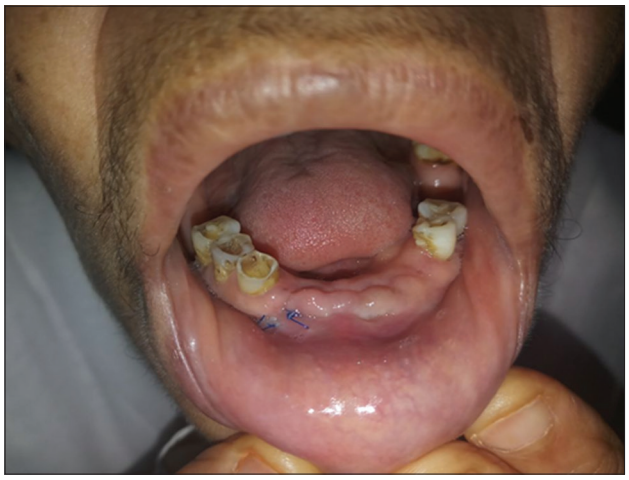

Figure 1: Diffuse swelling on the buccal aspect of the anterior mandible with missing 31, 32, 33, 41 and 42

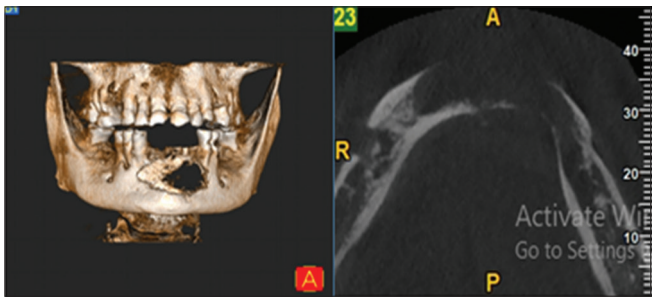

Figure 3: Cone-beam computed tomography images showing a buccal and lingual cortex disrupted

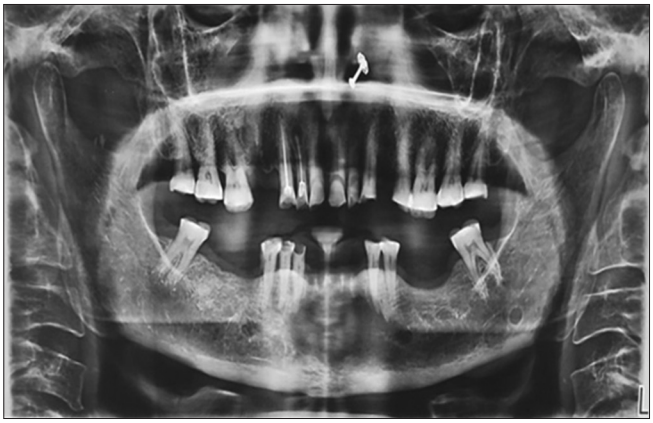

Figure 2: An irregular osteolytic lesion involving the anterior mandible

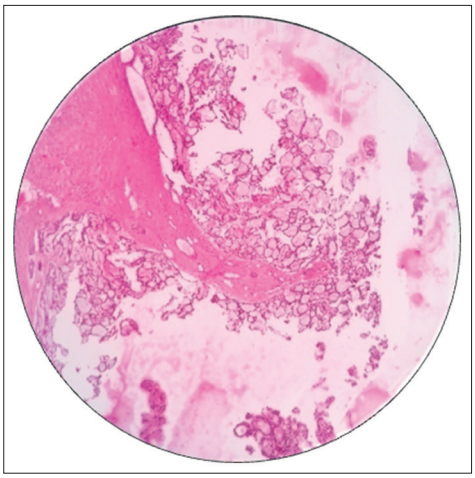

Figure 4: Numerous ducts such as tubules and pseudocysts mimicking cribriform pattern (H&E) (×40)

Focal positivity for periodic acid-Schiff was noted for the eosinophilic material within the lumen of the ductal tubules. Tumor cells showed diffuse immunopositivity with respect to S100, calponin and CD117 [Figures 6-8].

Postoperative radiotherapy was given. Follow-up after 15 months since the initial diagnosis showed no sign of recurrence.

DISCUSSION

ACC is a malignant salivary gland tumor, which was earlier known as "basiloma," given by Krompecher in 1908.[6,7] Later in 1954, Ewing termed it as ACC.[6,7] Intraosseous occurrence of malignant salivary gland tumors is rare with MEC being most common followed by ACC and the most common site being the posterior body or angle of the mandible, contrary to our case reported here which was found in the anterior mandible.[8]

Owing to the exceedingly rare occurrence of primary intraosseous ACC, the diagnosis of such neoplasms is troublesome and, regardless of the histology, should rely on the application of strict diagnostic criteria. In 1979, Batsakis offered certain identification features for a possible diagnosis of central salivary gland neoplasm, namely

- 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.[3,9]

All the identification features were satisfied in our case as it was presumed that the tumor first occurred in the mandible and later expanded widely throughout the medullary spaces of the mandible.

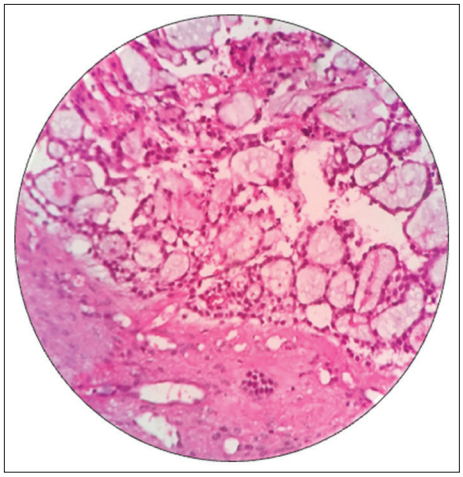

Figure 5: Cuboidal cells with prominent hyperchromatic nuclei and minimal cytoplasm lining the duct-like tubules (H&E) (×100)

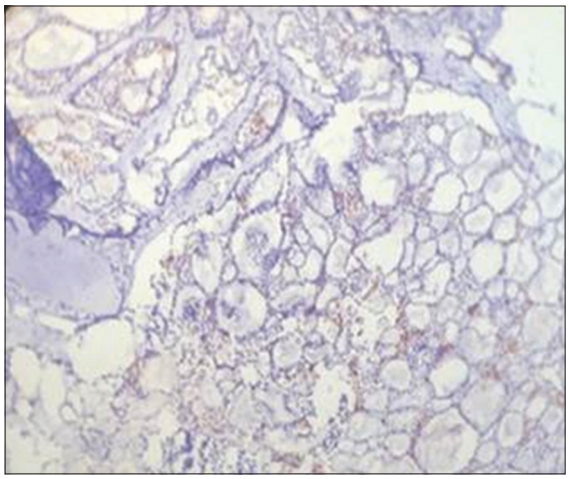

Figure 6: S100 immunopositivity

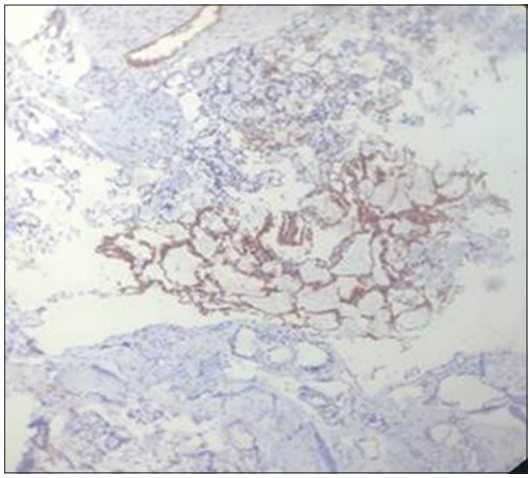

Figure 7: Calponin immunopositivity

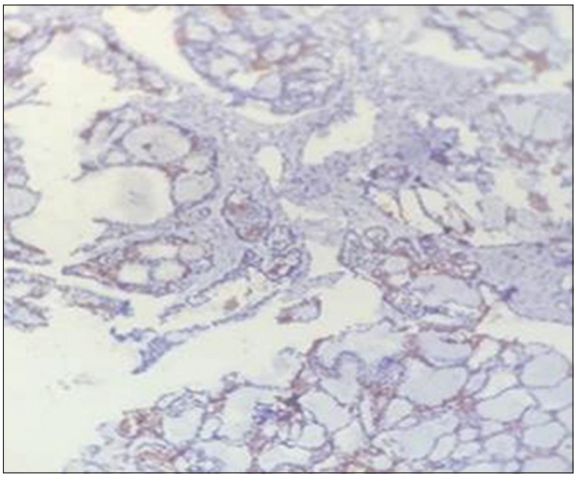

Figure 8: CD117 immunopositivity

The pathogenesis of central salivary gland neoplasms is unknown. Some of the researchers have proposed the following factors which could possibly play a role in the origin and formation of central malignant salivary gland tumors, which include

- Entrapped ectopic salivary gland tissues in jaws during embryological development of the jawbones
- Neoplastic transformation of the mucus-secreting cells commonly found in the epithelial linings of dentigerous cysts or any sinus pathology
- Submandibular and sublingual glands closely apposed in bony defects or cavities in the lingual cortex of the mandible or fragments of these glands that have undergone embryologic evagination.[10]

Histologically, the cribriform or tubular growth patterns of ACC are known to have a healthier prognosis, in contrast to the solid variant of areas more than 40% indicating aggressive clinical course.[8] The present tumor belonged to ACC category with predominance of cribriform and tubular pattern.

According to Brookstone and Huvos, lesions which are lying within the intact cortical bone and undisturbed overlying periosteum with no signs of cortical expansion offer the best prognosis and therefore indicate Stage I disease. Stage II disease is characterized by lesions surrounded by intact cortical bone that has undergone some degree of expansion. Cortical perforation with breakdown of the overlying periosteum or lymphatic spread is categorized as Stage III disease.[2,11]

Radiographically, the lesions, in general, are poorly defined and have infiltrative margins. The center of the lesion, as was the case with our patient [Figure 2], usually shows low density on the CT scan and radiograph. ACC infrequently occupies almost the whole of the bony fragment. The differential diagnosis for ACC should include polymorphous low-grade adenocarcinoma (PLGA), basaloid squamous carcinoma (BSC), MEC, acinic cell carcinoma, malignant mixed tumors, adenocarcinoma and squamous cell carcinoma.[12]

PLGA shows similar histological patterns (solid, cribriform and tubular), growth pattern and perineural spread. Polymorphous architecture, single-file/single-cell infiltration and foci of papillary growth are some of its main characteristic features. Histological features of high grade, i.e., increased mitotic figures, coarse chromatin, apoptosis and necrosis, are commonly associated with ACC (solid) than PLGA. Solid ACC also needs to be differentiated from BSC. Both tumors produce basement membrane-like material, but the latter tends to dissect between tumor cells rather than forming cribriform spaces. Necrosis and basaloid cells with prominent nucleoli and coarse chromatin are common features, although single-cell necrosis, rapid mitotic rate and greater degree of nuclear atypia are more frequent in BSC.[11]

Surgery is the predominant treatment modality for central salivary gland tumors of the mandible. It ranges from enucleation or curettage to *en bloc* or radical excision. Postoperative radiation therapy enhances local and regional control in ACC. Our patient was also treated surgically and was recovering well for this follow-up period of 1 year. There was no evidence of recurrence or distant metastases for 15 months from the original diagnosis. However, long-term follow-up is essential regardless of the site because of the tumor's susceptibility for late recurrence and metastasis.

Spread to regional lymph nodes is rare, but distant metastasis, particularly to lungs and bone, is more common and often unpredictable.[13] In the present case, owing to the massive size and an innocuous history, a thorough metastatic workup was carried out, which revealed neither regional lymph node nor distant metastasis.

In the present case, no metastasis was discovered, which otherwise is a common finding in such cases. The extensive involvement of the mandible could possibly be due to the insidious clinical course that characterizes ACC as it infiltrates nerves, perineural spaces and tissue planes.

CONCLUSION

ACCs have a variable prognosis. The 5-year survival rate is 75%, but the 10-year survival rate is only 20%, and survival at 15 years is about 10%.[14] Although salivary gland tumors arising within the mandible are rare, their unique morphology and clinical behavior make them one of the groups of lesion that should be considered in the differential diagnosis of aggressive lesions of the mandible.

A multidisciplinary diagnostic approach should always be adopted in the evaluation of this group of the lesion. Although in our case, there is no recurrence after 15 months, long-term follow-up is essential for ACC in order to avoid metastasis in addition to recurrence of this lesion. This case underlines the importance of early detection, total surgical resection and long-term follow-up as it is essential in clinical management due to their higher recurrence rate. Comprehensive investigations should be carried out for prompt diagnosis and appropriate treatment.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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