Case Report

Primary intraosseous adenoid cystic carcinoma of the mandible: A rare clinical entity

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Abstract Central intraosseous adenoid cystic carcinoma (ACC) of the mandible, formerly known as cylindroma, is a rare neoplasm with only 47 cases reported in the literature. We present a case of central ACC involving the mandible of a 55-year-old male patient.

Keywords: Adenoid cystic carcinoma, intraosseous head-and-neck neoplasm, salivary gland neoplasms

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INTRODUCTION

With a reported yearly incidence of 3–4.5 cases per million, adenoid cystic carcinoma (ACC) is an uncommon tumor, accounting for about 1% of all head-and-neck malignancies and about 10% of all tumors of the salivary glands.[1] In 1856, Billroth suggested the term "cylindroma" for this tumor; the current name of "adenoid cystic carcinoma" was introduced by Spies in 1930.[2] It is a clinically and pathologically well-defined entity which occurs primarily in the oral accessory salivary glands, particularly the palate and only 2% of cases in the parotid gland.[3] ACC which is so rarely seen in the jaw bones usually occurs in the posterior mandible of adults in 4th to 5th decade, causing pain due to perineural invasion.[4] The report of 26 cases of primary ACC of the mandible till April 2009 in the literature suggests its rarity.[5] About 40%–60% of participants develop distant metastasis (lung, bone and soft tissues) despite local control of the tumor.[6] The present paper reports an unusual case of primary intraosseous ACC of the mandible in a 55-year-old male patient with clinical and radiological features masquerading as a malignant lesion of odontogenic infection, indicating that a proper diagnosis of such a lesion is dependent not only on thorough clinical and radiographical examinations but also on the accurate interpretation of biopsied material. The difficulties of accurate diagnosis and the potential for such a condition should constitute an alert to clinicians.

CASE REPORT

A 55-year-old male was referred to the Department of Oral and Maxillofacial Surgery complaining of swelling and pain in the left lower jaw for 2 months. The patient had noticed swelling along the left side of the jaw approximately a year before. Clinical examination revealed a large, noninflamed swelling on the buccal aspect of the left side of the mandible causing facial deformity. The regional lymph nodes were not palpable. Intraorally, a hard swelling extending from the region of the left canine

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to the first molar was observed. The overlying mucosa appeared normal [Figures 1 and 2]. Orthopantomogram of the mandible showed a radiolucent lesion with irregular borders which extended from the first molar to the canine region [Figure 3]. The possibility of a malignant tumor was considered based on the clinical findings. A biopsy was performed in the central region of the swelling. Microscopically, the lesion showed overlying mucosa with no abnormality. Tumor cells were seen deep in the tissue. The tumor consisted of solid pattern with basaloid-type tumor cells in varying sized islands of cells arranged in cords and sheets in a fibrous connective tissue stroma. In some areas, islands of epithelial cells containing numerous spherical spaces showing a classical "Swiss cheese" pattern were evident. Based on these findings, a diagnosis of intraosseous ACC was made. In this case, the histological pattern predominantly seen was solid pattern with few areas showing cribriform pattern [Figure 4a-c]. A whole body scan and bone scan excluded distant metastasis to the bone. Surgical treatment consisted of hemimandibulectomy followed by mandibular reconstruction using titanium plates and free fibular flaps. Histopathological analysis of the resected specimen showed intraosseous involvement of ACC and the assessment of surgical margins did not show evidences of residual tumor neither in the lymph nodes. The patient was succumbed to the disease before completion of the treatment.

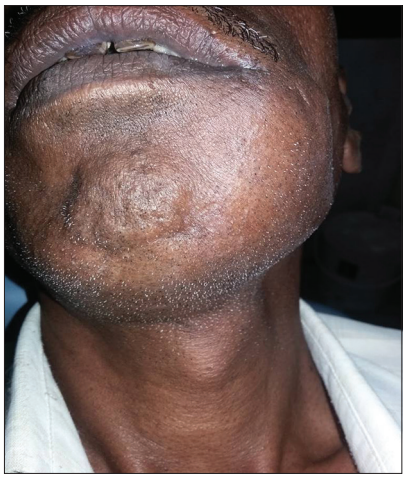

Figure 1: Photograph showing facial asymmetry with swelling on the left side of the lower third of the face

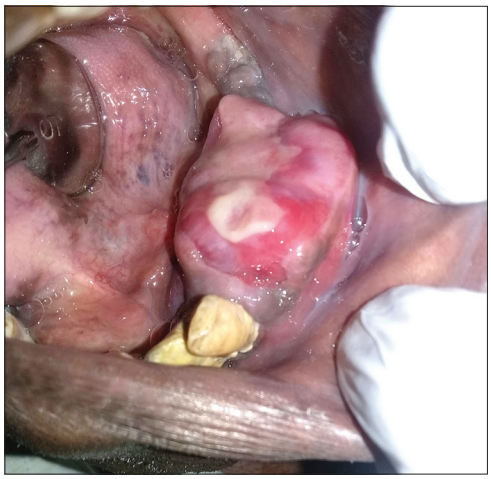

Figure 2: Intraoral photograph showing swelling in the left mandibular region extending from 33 to 36

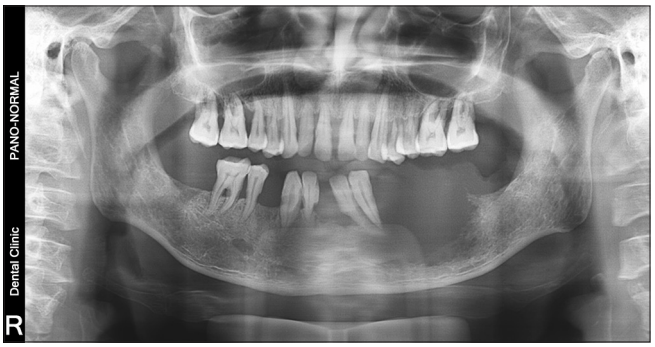

Figure 3: Panoramic radiograph showing a lytic lesion with irregular margins involving the left mandibular region

DISCUSSION

Salivary gland carcinomas located centrally within the mandible are rare comprising <0.4% of all salivary gland carcinomas. Mucoepidermoid carcinomas are the most frequently reported types of primary central salivary gland carcinomas of the mandible followed by ACCs, adenocarcinomas and acinic cell carcinomas.[7]

A total of 47 cases of primary ACC of the mandible have been reported in the literature until date.[8] The present case is the 48th case added to the literature. It generally occurs in the fifth decade of life with slight male predominance. The tumor is located in the posterior region of the mandibular body and in the angle (88.9%).[9] In the cases previously reported, swelling (51%) and pain (46.7%) were the most common clinical findings.[10]

In the present case, the tumor presented with associated pain and swelling in the left region of the mandible, in a 55-year-old male patient. These features are similar to those reported in the literature.[5] Radiographically, the lesions in general are poorly defined and have infiltrative margins.[4] Pulmonary metastases were detected in one-third of the patients during the follow-up of 1–14 years.[11] The current case is within these characteristics, but did not reveal any distant metastases.

Intraosseous salivary gland tumors do not differ microscopically from their soft-tissue counterpart. Histologically, there appear

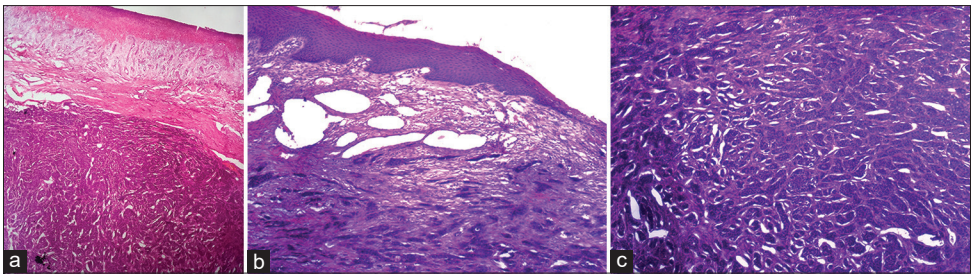

Figure 4: (a) Tumor cells seen deep in the tissue (H and E, ×40), (b) Solid pattern of basaloid type tumor cells in varying sized islands, chords and sheets (H and E, ×100), (c) Islands of epithelial cells showing "Swiss cheese" pattern (H and E, ×400)

to be three recognizable patterns of growth: solid, cribriform and tubulo-ductal.[12] Usually, combinations of the above patterns are seen and the tumor is classified based on the predominant pattern.[13] In the present case, histopathology of the surgical specimen revealed solid and a few areas cribriform pattern suggestive of a poor prognosis.

Central ACC is often misdiagnosed clinically as osteomyelitis, odontogenic cyst or ameloblastoma. To make a diagnosis of central ACC, metastatic lesions from other organs must be excluded.[12] In this case, no other malignant lesion was found.

Batsakis proposed diagnostic criteria for primary intraosseous salivary gland neoplasms, which include (1) radiographic evidence of osteolysis, (2) presence of intact cortical plates, (3) presence of intact mucous membrane overlying the lesion, (4) absence of any primary tumor within major or minor salivary glands and (5) histological confirmation of the typical architecture and morphological features of a salivary gland tumor.[14] For the case reported here, all these diagnostic criteria were satisfied, except by a focal erosive area of the cortical bone.

According to Brookstone *et al*., lesions that are located within undisturbed, intact cortical bone and overlying periosteum and that show no signs of cortical expansion offer the best prognosis and, therefore, suggest Stage I disease. Stage II disease is characterized by lesions surrounded by intact cortical bone that has undergone some degree of expansion. Cortical perforation, breakdown of the overlying periosteum or nodal metastatic spread is categorized as Stage III disease.[15] The present case can be categorized as being a Stage II disease, which has a bad prognosis.

The pathogenesis of central salivary gland neoplasms is unknown. The retromolar mucous gland and submandibular/sublingual salivary glands may get embedded in the lingual cortex during embryological development of the jaw bones. Some theories propose that the neoplastic transformation of these glands leads to intraosseous ACC. Furthermore, neoplastic transformation of odontogenic cyst epithelium and sinus epithelium has been suggested as the reason for the occurrence of these neoplasms.[16] In the present case, we can relate the tumor with remanescent of salivary gland tissue or oral ectoderm with potential salivary tissue differentiation, as the tumor is located in the posterior mandible.

The main treatment for central ACC ranges from enucleation or curettage to *en bloc* or radical excision, but no method of treatment reduces the potential for recurrence.[17] A wide surgical resection paying particular attention to obtain clear margins around regional nerves was the suggested treatment as this tumor demonstrates a propensity for perineural growth.[12]

However, long-term follow-up is indispensable regardless of the site because of the tumor's susceptibility for late recurrence and metastasis.[18] In the present case, the patient underwent wide surgical excision of the tumor, and no metastasis was discovered, which otherwise is a common finding in such cases.

The survival rate is estimated at 77% at 5 years and 57% at 10 years;[19] in our case, the patient died within 3 years after initial diagnosis, possibly due to the insidious course that characterizes ACC as it infiltrates nerves, perineural spaces and tissue planes.

CONCLUSION

Although salivary gland tumors arising within the mandible are uncommon, their significance should not be ignored. The primary treatment objective in ACC patients is local control, normal functionality and distant metastases prevention. For this purpose, early detection by the team of dental specialists is a prerequisite, in order to enable a more favorable prognosis and better quality of life. The therapy involving combination of surgery and radiotherapy remains the modality of choice in most cases. The early

detection will benefit the patient and reduces the morbidity associated with these lesions.

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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