PRIMARY INTRAOSSEOUS ADENOID CYSTIC CARCINOMA OF THE MANDIBLE – A CASE REPORT

V.Sadhish Valavan1*, C.R.Murali2, S.Soundarya3, N.V.Vani3

Abstract
Primary intraosseous adenoid cystic carcinoma (PIACC) of mandible is a rare malignant tumour of the salivary tissue with only 47 cases being reported in the literature. This article reports a case of PIACC in a 34 year old male patient in left mandibular region presenting as an irregular lytic expansile lesion on Computer tomography. Microscopic examination of the lesion showed basaloid cells arranged mostly in solid islands with few cribriform areas suggesting solid subtype of intraosseous adenoid cystic carcinoma. Solid growth pattern of the tumour indicates poor prognosis, thus warranting regular follow up of the patient. This case implies the importance of considering malignant salivary gland tumors in the differential diagnosis of poorly defined radiolucencies of the mandible. Early detection of such lesions will benefit the patient and reduces the associated mortality and co-morbidity.

Author Affiliations:
1,2,3,4Department of Oral Pathology and Microbiology, Best Dental Science College and Hospital, Madurai, Tamilnadu, India.

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*Corresponding Author:
Dr. V.Sadhish Valavan,
Postgraduate Student,
Department of Oral Pathology and Microbiology,
Best Dental Science College,
Madurai-625 104,
Tamilnadu. India.
Mobile: 9894518123,
Email: sadhish.v@gmail.com

INTRODUCTION
Malignant salivary gland tumors are uncommon neoplasm attributing to 3-5% of head and neck tumors. [1] Adenoid cystic carcinoma (ACC) is considered as the most common malignant salivary gland tumors. However, primary intraosseous ACC (PIACC) of the mandible is
of trauma and any other associated symptoms. His past medical and dental history were not contributory except for the uneventful extraction of left mandibular third molar. Extra oral examination showed facial asymmetry in left cheek region with enlarged, firm, non-tender left submandibular lymph nodes. On intraoral examination, a diffuse swelling measuring 2.5 × 2 cm was seen involving the alveolar ridge in relation to mandibular left third molar extending buccally from distal aspect of tooth 37 to the retro molar area, lingually it extends from distal aspect of 36 to retro molar area antero-posteriorly; from marginal gingiva to mucolinguval fold supero-inferiorly. The mucosa overlying the swelling is apparently normal with no ulcerations. On palpation, the swelling was tender, firm in consistency with slight lingual cortical expansion (Figure 1).

**Case Report:**
A 34-year-old male patient presented at the outpatient department of Best Dental Science College and Hospital, Madurai with a painful swelling in lower left back teeth region since 2 months. The pain was intermittent, localized and dull to moderate in intensity. There was no history
Computed tomography (CT) showed an irregular expansile lytic lesion measuring 3× 1.5 × 2 cm involving left ramus of the mandible [Figure 2a, 2b and 2c]. Based on the clinical and radiographic findings, a provisional diagnosis of ameloblastoma was given. However, a differential diagnosis of Keratocystic Odontogenic tumour (KCOT), intraosseous salivary gland neoplasm, central squamous cell carcinoma, metastatic lesion of the jaw were considered.

An incisional biopsy was done in the left mandibular second and third molar region and subjected for histopathological examination. Microscopically, the soft tissue specimen showed glandular epithelial cells within the fibrous connective tissue stroma. The glandular epithelial cells were uniform basaloid cells with round to ovoid nucleus arranged mostly in the form of solid islands with few cribriform areas. The cribriform areas showed micro cystic spaces with angular cells arranged in Swiss-cheese pattern. A histopathologic diagnosis of primary intraosseous adenoid cystic carcinoma (Solid subtype) of the mandible was rendered (Figure 3A, 3B and 3C).

To rule out distant metastasis, CT thorax and lung was done which doesn’t reveal any
positive findings. The patient underwent left hemimandibulectomy with radicular neck dissection which was followed by mandibular reconstruction with free fibular flap. Later, he was treated with external beam radiotherapy at a dose of 66 Gy/33 fractions for 33 days. The patient was put on regular follow-up and showed positive response to treatment. Histopathological examination of the complete excised lesion correlated with the pre-operative diagnosis of Primary intra-osseous adenoid cystic carcinoma.

DISCUSSION:

ACC is most commonly found in the head and neck region that originates from the major and minor salivary glands. However PIACC is a rare entity, and their origin and pathogenesis have not been well reported in the literature.\(^8\) The origin of PIACC remains controversial, and two hypotheses have been proposed regarding its histogenesis. Some authors have suggested that ACC of the mandible is derived from ectopic retro molar region mucus glands or the remnant submandibular gland in the mandibular lacuna or fissures during development.\(^9\) Others believe that it originates from metaplasia and malignant transformation of epithelial cells in the mandible, such as the epithelial cell rests of Malassez and reduced enamel epithelium.\(^10\) Although more scholars support the former theory, strong evidence is absent for either theory. Primary intraosseous ACC (PIACC) of the mandible is very rare, with only about 47 cases being reported in the literature till now.\(^12\) The age of patients with PIACC ranges from 24 to 82 years and equally distributed in both the sexes.\(^11\) The present case was reported in a 34 year old male patient correlating with clinicopathologic characteristics of this entity. A similar case was reported in the same site in a 49 year male patient by Iyengar et al in 2016.\(^12\) The clinical and imaging findings of PIACC are similar to other malignant or borderline-malignant mandibular tumors, thereby necessitating thorough histopathologic examination to arrive at a definitive diagnosis.\(^13\) Histopathologically, ACC is identified as a tumour with biphasic differentiation of epithelial and myoepithelial cell (MEC), which often shows mixed patterns of cribriform, tubular and solid types. It is usually classified according to the predominant growth pattern.\(^14\) The cribriform pattern is most frequent, showing nests of cells with micro cystic spaces and the tubular pattern consists of tubules with central lumina lined by inner epithelial and outer MECs. Generally, the tubular and cribriform patterns are known for their indolent behaviour, while the solid subtype, formed of sheets of basaloïd cells lacking tubular or micro cystic formation, is considered high grade with more aggressiveness and poor prognosis.\(^15\) Although ACC is known as a biphasic tumour composed of ductal and MECs, the amount and
distribution of the MECs in different subtypes and their relation to the grading and prognosis remain controversial.

As early as 1958, Patey and Thackray\cite{16} proposed that a solid growth pattern reveals a poor prognosis. Subsequently, this tumor has been graded into 3 types with increasing aggressiveness based on predominant growth pattern: tubular as Grade I, cribriform as Grade II, and solid type as Grade III. Regarding the histopathological grading of ACC, several systems have been proposed. In Perzin/Szanto system,\cite{17, 18} ACC is considered high grade if a solid component represents more than 30% of the tumor. Meanwhile, Spiro\cite{19} recommended that it is considered high grade only when the solid part account for more than 50% of the tumor. Recently, Van Weert et al. proposed that the presence of a solid pattern regardless of its quantity is a poor prognosticator.\cite{20} The cribriform and tubular subtypes of ACC consist of biphasic differentiation with MECs, however the solid growth pattern has gland differentiation with loss of MEC differentiation causing higher aggressiveness and proliferation rate. This lends to hypothesize that the loss of MEC differentiation might contribute to the poor prognosis of the solid subtype of ACC.\cite{2, 21} The reported case showed glandular epithelial cells mostly in solid growth pattern indicating poor prognosis, thus warranting regular follow up of the patient.

In 1979, Batsakis proposed diagnostic criteria for primary intraosseous salivary gland neoplasm’s\cite{22} which includes radiographic evidence of osteolysis, presence of intact cortical plates, intact mucous membrane overlying the lesion, absence of any primary tumours within major or minor salivary gland and histological confirmation of the typical architecture and morphological features of a salivary gland tumour. The present case satisfied the proposed diagnostic criteria except for the presence of intact cortical plates which is probably due to the chronicity of the lesion. Based on the destruction of bone and cortex, Brookstone and Huvos\cite{23} established a staging system for central salivary gland malignancies: 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 due to the presence of massive osteolysis in the left side of the mandible on Computed Tomography (CT) examination. The increased tendency of this tumor for local recurrence and distant metastasis after adequate loco regional control requires lifelong follow-up of these patients.\cite{24}

**CONCLUSION**

Primary intraosseous adenoid cystic carcinoma of mandible is a relatively uncommon tumor and their significance should not be ignored due to their biologic behaviour, hence it should be considered in the differential diagnosis of aggressive lesions of the mandible. A
Multidisciplinary approach should be adopted for management of such lesions and long-term follow-up is necessary to detect metastasis and recurrence. The present case emphasizes the importance of considering intraosseous malignant salivary gland tumors in the differential diagnosis of ill-defined radiolucencies of the mandible.

**Conflict of Interest Statement**

There is no conflict of interest.

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