

Primary Intraosseous Adenoid Cystic Carcinoma of the Mandible with Distant Metastasis: A Case Report

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Abstract: Intra-mandibular localization of adenoid cystic carcinoma is an extremely rare neoplasm. This tumor is characterized by progressive local, regional, and distant aggressiveness. We reviewed the latest data on this rare type of cancer with a small number of reported cases, a lack of consensus for its treatment, and its bad prognosis. A case of primary intraosseous adenoid cystic carcinoma of the mandible with lung and bone metastasis in a 52 years old man is reported.

Keywords: intraosseous adenoid cystic carcinoma, mandible, distant metastasis.

INTRODUCTION

Adenoid cystic carcinoma (ACC) is a rare malignant tumor of the salivary glands accounting for less than 1% of maxillofacial cancers [1]. The intra-osseous maxillo-mandibular location is very rare [1]. It most commonly affects adults; with a peak incidence in the 4th to 6th decades [2]. Alcohol consumption, smoking, and poor dental health are not predisposing factors for IACC [3, 4].

The most commonly involved sites are the parotid gland, submandibular gland and palate. Very rarely, ACC may arise centrally within the jawbones, usually located in the posterior region of the mandibular body and in the angle, but the parasymphiseal and symphiseal regions may also be involved [5].

Several hypotheses have been made on the mechanism underlying the atypical intra-osseous location [6], it could be due to either ectopic salivary glands or embryonic remnants of submandibular or sublingual glands trapped within the bone or to the malignant degeneration of mucus secreting cells normally found within the odontogenic epithelia [7]. About 40 -60 % of patients develop distant metastases (lung, bone, and soft tissues) despite local control of the tumor [7-8].

It often develops insidiously and its diagnosis is made at an advanced stage when management can be problematic [1, 2]. Moreover, the risk of distant metastases and its neurotropic nature worsen the prognosis [9]. There is no consensus on the therapeutic management due to the rarity of cases. We had for aim to review the latest data regarding intra-mandibular adenoid cystic carcinoma (IACC). We report a case of primary intraosseous ACC of the mandible with lung and bone metastasis in a 52 years old man.

CASE REPORT

A male patient aged 52 years complained of a right mandibular mass that gradually increased in size

for approximately 2 years and an occasional swelling sensation.

The extraoral examination revealed a firm tender swelling in the right cheek extending from tooth 35 to the right ramus of the mandible with a size of approximately 9x4 cm. the swelling was nonfluctuant and nonpulsatile (Figure A, B). The overlying skin was normal with a smooth surface. An enlarged lymph node with a diameter of 1.5 cm was palpable in the right submandibular region. Intraoral examination revealed a limited mouth opening and an intraoral mass. The overlying mucosa was normal but slightly erythematous (Figure C).

A panoramic radiograph revealed a large radiolucent area in the mandible (Figure D). The patient was scheduled for further investigations computed tomography (CT) images revealed extensive bone destruction in the mandible along the mandibular canal, a significant soft tissue mass, and multiple bilateral enlarged lymph nodes in level I and level II of the neck. The preliminary clinical diagnosis was a malignant tumor of the mandible.

The patient was subjected to incisional biopsy, and histopathological examination revealed an ACC. After diagnosis, Chest X-ray revealed a coin lesions lung. Bronscopic biopsy confirmed the diagnosis of adenoid cystic carcinoma (Figure E). A whole bone scintigraphy confirmed distant metastases (Figure F).

Consequently, the final diagnosis was primary intraosseous adenoid cystic carcinoma of the mandible with distant metastasis to lung and bone. The patient was referred to the adjuvant chemo- and radiotherapy. Unfortunately he died before the treatment had been achieved.



Fig-A –B: The swelling in the right cheek with a size of approximately 9x4 cm. the swelling was nonfluctuant and nonpulsatile



Fig-C: Limited mouth opening and an intraoral mass.



Fig-D: Panoramic radiograph revealed a large radiolucent area in the mandible



Fig-E: Chest X-ray revealed a coin lesions lung

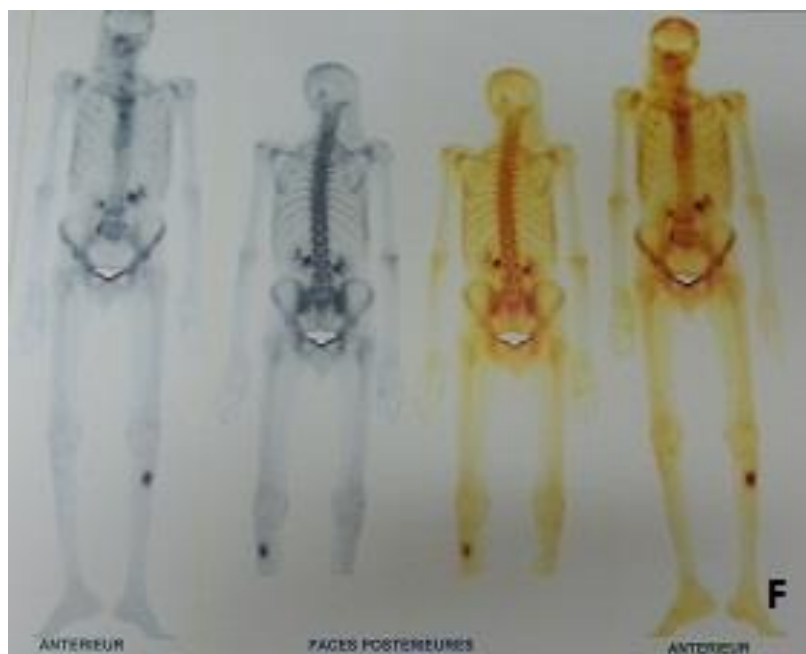


Fig-F: Bone Scintigraphy confirmed distant metastases

DISCUSSION

ACC is classified among malignant tumors of the salivary glands and was first described by Robin and Laboulbene in 1853. Theodor Biltroth called it cylindroma in 1856 [3]. It is most commonly found in the head and neck region and mainly originates from the salivary glands. IACC is very rare, and the origin and pathogenesis of IACC have not been well reported in the literature. The most common areas in which PIACC occurs include the body and the angle of the mandible. A literature review revealed that the age of patients with IACC ranges from 24 years to 82 years and there are no significant gender differences [5].

The most common clinical manifestations of ACC in the mandible are pain and swelling [10], but

patients can also present with lower lip numbness, loose teeth, toothache, or other symptoms. These Clinical signs are not specific because the evolution is slow and insidious these manifestations are similar to other malignant tumors originating in the jawbone. Weight loss may also be observed because of eating difficulties

IACC should be managed like cancer, given the clinical and radiological signs of malignancy and the impossibility of performing an extemporaneous examination. The diagnosis is confirmed by histology [5-9].

The panoramic radiograph is the initial examination showed multilocular hypodense areas with clear boundaries, and the roots of the teeth adjacent. CT

scan is currently the key imagery revealing the tumoral mass and whether the premandibular soft tissues are involved or not; it can also reveal cervical lymphnode involvement. The examination can also be extended to the thorax, abdomen, and pelvis. MRI can also reveal the lesion, and show the state of surrounding tissues, notably the involvement of the inferior alveolar nerve (an important prognostic criterion), and possible locoregional or distant metastases. Panendoscopy is not useful until surgery, because the tumor occurs in salivary glands.

Because IACC is rare and lacks specific clinical and imaging findings, this tumor is prone to misdiagnosis prior to surgery. For different tumor types, the corresponding treatments can be very different. Although the role of routine pretreatment biopsy for salivary tumors remains a controversial topic, incisional biopsy is an important procedure to establish a definitive diagnosis. However, it is difficult to carry out the incisional biopsy of the intraosseous tumors [11].

IACC cannot be staged according to the AJCC clinical staging criteria for salivary gland cancer and oral cancer. Some scholars have suggested that IACC should be staged based on damage to the mandible rather than tumor size as follows: no changes in the morphology and integrity of the bone cortex surrounding the lesions and the presence of surface periosteum (stage I); intact surface of the bone cortex with a certain degree of swelling (stage II); and damage to the bone cortex, periosteum rupture, or lymph node metastasis (stage III)[12].

We believe that this classification method has some significance. Tumors within the jawbone are often occult. During the early stages, patients often mistake the symptoms as a toothache and therefore do not pay attention to the condition. Tumors are often discovered after tooth extraction. Moreover, tooth extraction can cause tumors to break through the jawbone and invade the gums and surrounding soft tissues, leading to tumor progression. Therefore, patients are often already in an advanced stage of the disease when they seek clinical treatment.

Microscopic observations of ACC reveal a non-encapsulated well-defined solid tumor. In terms of histology, there are 2 cell lines: epithelial cells and myoepithelial cells with a hyperchromatic, angular nucleus and scant cytoplasm. Three architectural forms have been described [8]: the tubular form includes tubes lined by a double layer of cells, with epithelial cells making up the internal layer and basal cells the outer layer; the cribriform structure has a microcystic aspect with the presence of basophilic mucoid and/or hyaline material; the solid form includes uniform basal cells (without tubes or microcysts). We observed that IACC is similar to ACC of the salivary gland and that IACC tumors tend to invade the nerves and blood

vessels (3/4, 75%). The IHC results showed that IACC is composed of myoepithelial cells and lumencells; the cribriform structures primarily expressed P63, and the solid structures primarily expressed CK7, CK19 and other markers of lumen cells. These results indicate that the main components of solid structures in these cases are lumen cells, with a loss of myoepithelial cells that is similar to the previously reported phenotypes of salivary gland ACC [12].

We recommend a multidisciplinary meeting including surgeons, radiotherapists, dentists, and chemotherapists, as for any type of cancer. IACC is primarily treated with surgery, which can be supplemented with radiotherapy, although a few cases can be treated with chemotherapy or radiotherapy alone. The indication and the choice of therapy are always controversial [5]. Nonetheless, for several authors, surgery is the mainstay of management [3-4]. It consists in a radical resection of the lesion with wide resection margins (macroscopic margins of at least 1 cm) [3-8]. It often involves segmental mandibulectomy, extended to the underlying and overlying structures, depending on the tumor extension. Systematic cervical lymphadenectomy is recommended by most authors, whatever the lymph-node status, and should be bilateral if the tumor is symphyseal, or has reached or extended beyond the median line. It should be homolateral in other cases. Complementary postoperative radiotherapy is performed in accordance with the most recent recommendations, given the radio susceptibility, for better local control in terms of recurrence; it is indicated whatever the surgical margins and pathology results (histological grade) are. Its lack of susceptibility to chemotherapy can be explained by its slow evolution; this explains why chemotherapy has been abandoned as the first-line treatment. However, it is indicated for strictly palliative care in case of metastases.

Patients with IACC have a poor prognosis, and the common prognostic factors include local recurrence and distant metastases [12].

CONCLUSION

IACC is relatively rare. Clinical and imaging findings often lead to the misdiagnosis of odontogenic tumors or bone tumors. The present case calls attention to the possible occurrence of primary IACC with metastases showing a very aggressive course.

The histological features, immunophenotype, molecular changes and prognosis are similar to salivary gland ACC (SACC), indicating that they have the similar origins and pathogenic mechanisms. IACC treatment also requires surgical excision, postoperative radiotherapy and long-term follow-up. These results provide an understanding of the molecular mechanism of ACC in the mandible

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