Primary Intraosseous Carcinoma with Nodal Metastasis Masquerading as Ameloblastom—A Rare Case Report

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Introduction

Squamous cell carcinoma (SCC) of oral cavity usually presents as a non-healing ulcer or a growth in oral cavity and arises from the dysplastic oral mucosa that turns into invasive cancer. Most of the times it involves the mandible either by direct invasion of tumor, or through the alveolar ridge into the canal of inferior alveolar nerve [1].

Rarely SCC can arise primarily in the mandible, either de-novo or in preexisting benign cyst. Primary intraosseous carcinoma (PIOC) of the jaws is a rare tumor, defined as the squamous carcinoma which develops in the mandible without any initial connection with the oral mucous membrane, adjoining skin or nasal or antral mucous membranes, and which presumably develops from the residue of odontogenic epithelium deriving embryologically from the dental ledge (Malassez' remnants) [2].

Here we are presenting a rare case that clinically and radiologically seems to be a unilocular ameloblastoma of

mandible but actually it is primary intraosseous carcinoma probably arising from a cyst.

Case Report

A 47-year-old male patient, presented with h/o painless swelling on chin for 3.5 months (Figs. 1 and 2). After 2 months of appearance of swelling, he started having pus discharge from a small sinus opening at left lower alveolus. A dentist without any prior imaging removed his two teeth. The pus discharge continued. He had undergone biopsy twice. The superficial biopsy from sinus opening showed dysplastic squamous epithelium only. The deep biopsy taken from same site after incising the mucosa showed well differentiated squamous cell carcinoma. The ulcer healed after biopsy.

On clinical examination there was swelling of the chin. The oral mucosa was smooth, no ulcer was visible. There were palpable lymph nodes present at left cervical level Ib and II. The orthopantomogram showed a well defined, large lytic lesion in anterior arch of mandible (Fig. 1). The possibility of metastasis was ruled out by means of careful clinical examination and available radiological tests.

Patient was posted for surgery, and wide excision of the lesion with central arch mandibulectomy was done along with modified radical neck dissection (MRND) on left side and supra-omohyoid neck dissection (SOHND) on right side (Fig. 3). The reconstruction should ideally be done by free fibular flap, but here it was done by PMMC flap because patient did not agree for free fibular graft. The final histopathology report said it to be squamous cell carcinoma. Small area of the anterior cortex of mandible at left side was eroded involving soft tissue. Lymphatic invasion was seen. All the margins and cut ends of mandible were free from malignancy. Multiple sections examined from the part of cyst

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Fig. 1 orthopantomogram showing a large unilocular cystic lesion in the anterior arch of mandible, with preserved walls all around

wall that was away from the tumor, showed no evidence of malignancy, but the exact typing of cyst was not possible. In the left MRND specimen, out of 16 lymph nodes, 2 showed metastatic tumor. In the right SOHND specimen, all dissected 5 lymph nodes were free from tumor. There was no extra-capsular spread. Post-operatively patient did well. Adjuvant radiotherapy was given to patient.

Discussion

Primary malignant tumor of mandible was first described by Loos in 1913 as “Central Epidermoid Carcinoma” of the jaw. The World Health Organization (WHO) in 1972 suggested the term primary intraosseous carcinoma and classified the lesion as an odontogenic carcinoma.

The classification currently used is the one modified by Waldron and Mustoe [3]. They classified such neoplasms as:

- TYPE 1: PIOC ex odontogenic cyst.
- TYPE 2A: Malignant ameloblastoma.
- TYPE 2B: Ameloblastic carcinoma (de novo, ex ameloblastoma or ex odontogenic cyst).
- TYPE 3: PIOC developed de novo:
  - a. keratinising type.
  - b. non-keratinising type.
- TYPE 4: Central intraosseous mucoepidermoid carcinoma.

The jaws are the only bones of the skeleton affected primarily by intraosseous carcinoma, which suggests that it originates from epithelial cells present only in them. These epithelial remnants can proliferate and transform into odontogenic carcinoma, a process that could be potentially triggered by an inflammatory process, as a complicated tooth extraction [4]. PIOC arising de novo must be considered when no cystic component or other odontogenic tumor cells are demonstrated in the lesion. The etiology is still not clear,

Fig. 2 pre-operative photograph of patient with swelling at chin

probably PIOC derives from the remnants of odontogenic epithelium, epithelial rests of Malassez, or the remnants of the dental lamina [5].

These patients are frequently treated initially for presumed dental problems and this fact often delays the diagnosis of intraosseous carcinomas. In the absence of initial biopsy, the radiolucent lesion may simulate an odontogenic cyst and this situation could lead the surgeon to enucleate the lesion without adequate free margins. Thus, histological diagnosis during the surgical intervention is essential for a correct treatment [6].

Radiographically, PIOC shows great variation, but usually show a fully enclosed, irregular pattern of bone destruction with ill-defined margins [7]. The absence of a clear definition of the lesion, the sudden growth in size detected over successive radiographs and significant displacement of the inferior dental canal, are the signs that lead us to suspect the presence of such malignancy.

Fig. 3 specimen, excised with left MRND specimen. Notice that smooth alveolar mucosa (without any ulcer/growth) is visible, with absence of left canine and first premolar

Panoramic radiography can be useful in cases of gross disease, but is of limited value in evaluating margins, extension and invasion of the tumor mass; for that 'Denta scan' is needed.

Metastasis in the lymph nodes and hematogenous dissemination take time to occur even when the invasion of the bone and soft tissue is quite advanced [2].

Recommended treatment for this type of tumor involves surgery including large-scale surgical extirpation and the removal of the cervical nodes. Suspected diagnosis is mainly based on radiological study, while definitive diagnosis normally comes as a result of the anatomicopathological study performed [2]. The prognosis of PIOC is surprisingly good, [3] being more favorable when they originate from odontogenic cysts than when they originate de novo [5].

The histopathologic features of PIOC are consistent with a diagnosis of squamous cell carcinoma with or without keratinisation. The microscopic differential diagnosis should, therefore, include any lesion that produces squamous epithelium, such as acanthomatous ameloblastoma, squamous odontogenic tumor, metastatic carcinomas and central mucoepidermoid carcinoma. Microscopic examination of serial specimen sections is usually required to rule out the presence of odontogenic cyst lining epithelium or any other odontogenic tumor [8]. Radiotherapy and chemotherapy are used only as palliative therapy or in cases where nerve infiltration is diagnosed (adjuvant therapy) [8]. Prognosis is directly related to adequate and complete resection, which obviously requires early detection before the extensive involvement of adjacent tissues or the presence of metastasis to lymph nodes.

Summary

The PIOC's are rare malignant tumors of jaw bones that clinically and radiologically simulate benign neoplasms or cysts. This could lead the surgeon to enucleate the lesion without adequate free margins. Thus, histological diagnosis before the surgical intervention is essential for a correct treatment.

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