

Primary Intraosseous Carcinoma of Mandible: A Case Report

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ABSTRACT

Primary de novo intraosseous carcinoma is a rare neoplastic lesion which commonly occurs in the jaws. It is an epithelial odontogenic malignancy arising from odontogenic epithelial residues in the bone rather than from a preexisting epithelial lesion. In the present case report, the clinical, radiological and histological features of primary de novo intraosseous carcinoma are discussed and its aggressiveness and local invasiveness are highlighted.

Keywords: Mandible, Primary intraosseous carcinoma, Squamous cell carcinoma.

Received: May 2010

Accepted: October 2010

Dent Res J 2011; 8(2): 102-107

Introduction

Primary intraosseous carcinoma describes a squamous cell carcinoma arising primarily from the jaw bone having no initial connection with oral mucosa, and presumably developing from residues of the odontogenic epithelium.¹ It may arise within the jaws either from a previous odontogenic cyst or de novo rather than from a pre-existing epithelial lesion.² Several cases of malignant transformation of odontogenic cysts or odontogenic tumors have appeared in the literature, while primary intraosseous carcinoma arising de novo has been infrequently reported. Here, we report a case of primary intraosseous carcinoma in a middle aged female patient with gross destruction of hemimandible with local metastasis.

Case Report

A 54 year old female patient reported to our department with the chief complaint of pain and swelling in the right lower back region of the jaw since three months ago. Initially, the patient developed pain in the teeth in the fourth quadrant of the jaw, followed by a localized swelling. Within one month, the teeth became mobile and were extracted, however the swelling persisted and grew progressively.

The patient had no contributing medical history however she had a habit of quid chewing two to three times per day since 20 years ago.

General physical examination of the patient revealed signs of anemia with pale conjunctiva and nail beds. There was a gross facial asymmetry with diffuse swelling involving middle and lower third of the right side of the face extending from ala-tragus line to approximately four centimeters beyond the inferior border of the mandible (Figures 1A and B). The swelling extended anteriorly to the angle of the mouth and posteriorly to the posterior border of the ramus of the mandible. On palpation, two swellings were appreciated. One swelling which was extending from ala-tragus line to the inferior border of the mandible was fixed. The other swelling extended below the inferior cortex, ovoid in shape and measured approximately four centimeters in diameter. Both of the swellings were tender and firm in consistency. The condyle of the mandible on the same side was not palpable, while the jaw was deviated to the right on opening and closing the mouth (Figure 2A). The bilateral submandibular lymph nodes were palpable, tender, firm in consistency and not fixed. Intraorally, vestibular obliteration was seen extend-

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ing from the canine to the anterior border of the ramus, posteriorly. The extraction socket distal to the second premolar showed erythematous granular mass covered partly with necrotic slough (Figure 2B). There was no obvious relation between the mandibular mass and the overlying mucosa. On palpation, the findings of inspection were confirmed, while the swelling was tender and indurated with no cortical expansion. First and second premolars were vertically compressible into their sockets. Considering the aggressive nature and the extension of the growth, carcinoma of the right mandibular alveolus and intraosseous carcinoma were considered in the differential diagnosis.



Figure 1. (A) It shows a diffuse swelling involving middle and lower thirds of the right side of the face. (B) It shows the swelling extending beyond the inferior border of the mandible.



Figure 2. (A) It shows the deviation of the mandible towards the right side. (B) It shows the erythematous granular mass partially covered with necrotic slough within the extraction socket in the first molar region.

Panoramic radiograph revealed gross destruction of the right body and ramus of the mandible extending from the right lower canine to the mandibular notch with bay and promontory appearance of the margins. Right lower premolars showed floating tooth appearance with few radiopaque flecks of bone scattered within the lesion (Figure 3). Post-

erior-anterior radiograph of mandible showed involvement of both medial and lateral cortical plates (Figure 4). A screening chest radiograph was made to rule out any primary lesion in the lungs however, nothing abnormal was found.



Figure 3. Orthopantomograph showing gross destruction of right body and ramus of the mandible with flecks of bone within the lesion.

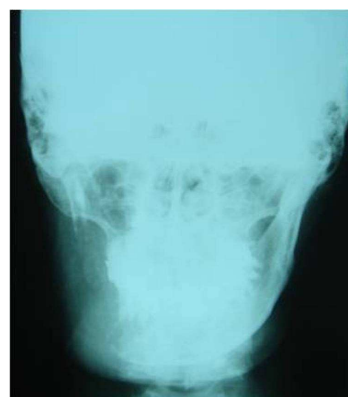


Figure 4. Postero-anterior mandibular radiograph showing the involvement of medial and lateral cortical plates of mandible.

CT scan was performed to know the extension of the lesion. It revealed a destructive lesion in the right half of the mandible extending from subcondylar to right parasymphiseal region. It showed a unicentric lesion with inside out growth and permeative type of destruction. The margins were indistinct with wide zone of transition suggesting an intraosseous malignant neoplasm. Masseter and part of pterygoid muscle was bulky suggesting infiltration. CT scan also showed a large oval well-defined submandibular mass with central necrosis suggestive of metastatic submandibular lymph node (Figure 5).

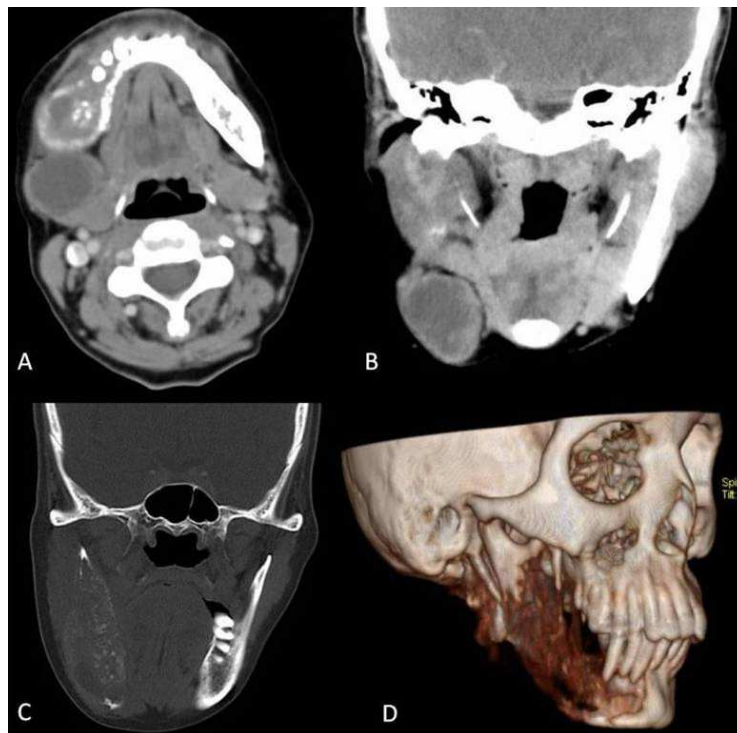


Figure 5. Axial (A) and coronal (B) postcontrast CT showing the unicentric destructive lesion involving the body and ramus of the right half of the mandible, respectively. A submandibular mass with central necrosis is also seen in both images. Coronal CT section-bony window (C) showing destruction of ramus with bony flecks within the lesion. 3D reconstructed CT image (D) showing gross destruction of mandible.

High resolution ultrasonography was performed to know the lymph node involvement. It revealed a well defined nodular mass inseparable from the right submandibular salivary gland with central cavitation and papillary projections into the cavity suggesting a neoplastic process involving the right submandibular gland (Figure 6).

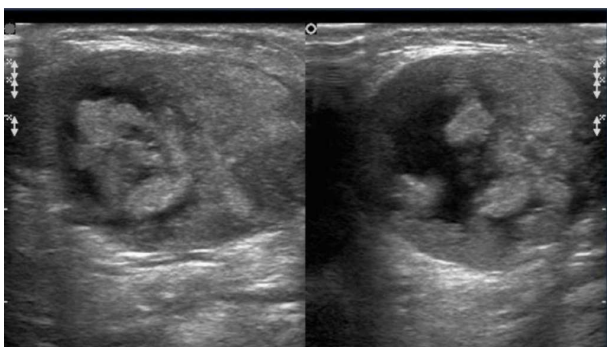


Figure 6. Ultrasonographic image of submandibular mass showing central cavitation and papillary projection into the cavity.

Aspiration of the submandibular mass was done which revealed a clear straw colored fluid (Figure

7). Cytological smear showed squamous cells with mild pleomorphism and large number of inflammatory cells spread over a background of eosinophilic acellular material. Histopathological report revealed islands of tumor epithelial cells with abundant keratin formation scattered over a densely fibrous stroma with dense chronic inflammatory infiltrate suggesting a well- differentiated squamous cell carcinoma (Figure 8).



Figure 7. Clear straw colored fluid aspirated from the submandibular swelling.

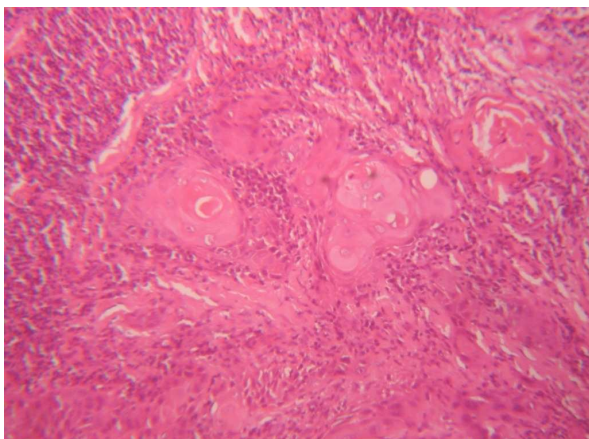


Figure 8. Histopathological photomicrograph revealing islands of tumor epithelial cells with abundant keratin formation.

Patient was subjected to wide surgical excision of the lesion (hemimandibulectomy). Radical neck dissection was performed using Macaffe incision sacrificing internal jugular vein, sternocleidomastoid muscle, spinal accessory nerve, omohyoid muscle, facial artery and vein, occipital artery and submandibular gland and its duct. Reconstruction was done with pectoralis major myocutaneous flap. Follow up of the patient showed satisfactory healing of the wound with mild disfigurement of the face due to scar contracture (Figure 9).



Figure 9. Follow up photograph showing satisfactory wound healing with mild disfigurement of the face.

Discussion

Primary intraosseous carcinoma (PIOC) describes the squamous cell carcinoma that develops presumably from the residues of the odontogenic epithelium entrapped within the jaw with no initial connection with the surface oral mucosa. This tumor was first described by Loos in 1913. The World Health Organization (WHO) in 1972 suggested the term primary intraosseous carcinoma and classified

the lesion as an odontogenic carcinoma.³ PIOC occurs mainly in adults, in sixth to seventh decade and have a male to female ratio of 3:1. It is situated usually in the posterior mandible.⁴ In a pooled analysis of world literature, the mean age of the patients at the time of diagnosis was 52.3 years with male to female ratio being 2.5:1.⁵ Cases with anterior maxillary involvement have also been reported.⁶ The etiology of PIOC is not clear however the most common factor may be a reactive inflammatory stimulus with or without a predisposing genetic cofactor.⁷ Since squamous cell carcinomas may appear within the bone from other sources, the diagnosis of PIOC is by exclusion.

Suei et al.,⁸ proposed few diagnostic criteria for PIOC; they were as follow: (1) to differentiate PIOC from squamous cell carcinomas of surface mucosal origin, no ulcer formation must be present on the overlying oral mucosa except when due to such causes as trauma or tooth extractions; (2) to rule out the possibility of other odontogenic carcinomas, serial sections of the histological specimens must demonstrate squamous cell carcinoma without cystic components or other odontogenic tumor cells; and (3) to rule out a distant primary tumors, chest radiographs must be clear at the time of diagnosis and throughout a follow-up period of more than six months. In our case, a tooth-extraction socket was present. However, there was no continuity of the tumor with the overlying mucosa and the overlying and the surrounding mucosae were quite normal.

The common clinical features in PIOC include pain and swelling of the affected area. In a pooled analysis of 33 cases recorded in world literature, performed by Thomas et al.,⁵ pain was the commonest presenting feature in 17 (54.8%) patients followed by swelling of the jaw in 16 (51.6%) and sensory disturbances in five (16.1%). In many instances, the nonspecific clinical findings may mimic inflammatory dental processes. In few cases, patients have a history of prior dental procedures (e.g., extractions and denture adjustments) attempting to resolve the symptoms associated with the neoplasm.⁹

The primary intraosseous carcinomas of the jaws are classified based on their possible origins into four types:

- Type 1: PIOC such as odontogenic cysts.
- Type 2 A: Malignant ameloblastoma
- B: Ameloblastic carcinoma
- Type 3: PIOC arising de novo

- Keratinizing
- Nonkeratinizing

Type 4: Intraosseous mucoepidermoid carcinoma¹⁰

The radiological investigations provide valuable information in diagnosing these clinically bewildering conditions. Primary intraosseous carcinomas exhibit radiolucencies with a wide variation in size and shape. Thomas et al.⁵, in his study found that PIOC's have varied radiographic presentations like cup- or dish-shaped patterns, well defined lesions, small radiolucent loculations and poorly defined moth eaten appearance. Slowly growing tumors often exhibit well defined peripheries, whereas rapidly expanding lesions typically demonstrate poorly defined and ragged borders with permeative type of destruction. The degree of raggedness of the border may reflect the aggressiveness of the lesion. If enough in size, pathological fracture occurs due to cortical plate thinning with subsequent step deformity. The internal structure is totally radiolucent with no evidence of bone production and very little residual bone left within the centre of the lesion. Sometimes in small lesions, the buccal and lingual cortical plates may cast a shadow that may mimic the appearance of trabecular bone. These lesions are capable of destroying the floor of maxillary antrum and nasal cavity, the cortical outlines of inferior alveolar canal of mandible and effacement of lamina dura. Root resorption is unusual. Teeth that lose both lamina dura and the supporting bone appear to be 'floating' in space. If the lesions are not aggressive with smooth borders and centered about the apex of a tooth, they may be mistaken for periapical cysts and granulomas. If the lesions are infiltrative with extensive bone destruction, a metastatic lesion must be excluded as well as multiple myeloma, fibrosarcoma and carcinoma arising in a dental cyst must be ruled out.

Histologically, they vary from well-differentiated tumors exhibiting significant keratinization to nonkeratinizing poorly differentiated carcinomas. Yamada et al.,¹¹ in his clinico-pathological study of five cases of PIOC found three cases of well-differentiated carcinoma, one moderately differentiated carcinoma, all those three arising de novo and the one arising from an odontogenic cyst. In our case, the tumor was a well differentiated keratinizing squamous cell carcinoma with no evidence of odontogenic cystic component. Thomas et al.,⁵ in his study observed that keratinization is present in 31.4% of all PIOC's

included in the study. Our case fulfilled almost all the criteria suggested by Swei et al.,⁸ and moreover there was no evidence of any cystic component histologically suggesting our case is a primary de novo intraosseous keratinizing squamous cell carcinoma.

Around 66% of patients with de novo PIOC have clinical and/or histological evidence of regional metastasis, either initially or during the course of the disease.⁶ In our patient, we could find local invasion into the pterygoids and the masseter and metastasis to ipsilateral submandibular gland. Metastasis to the regional lymph nodes at the time of presentation was seen in 31.4% of cases in the analysis made by Thomas et al.⁵ Metastatic spread to cervical lymph nodes has been discussed by Elzay¹² and Muller and Waldron.¹³ It is important to rule out metastasis to the jaws in cases of suspected PIOC. Since the majority of metastatic squamous cell carcinomas to the jaws arise within the lungs, it is necessary to evaluate the patient thoroughly including chest radiography in an attempt to detect any occult primary tumor. Ideally bone scan should be performed to locate the primary or the metastatic lesions, but because of the lack of facilities and unaffordable financial position of the patient, we had to rely on chest radiograph alone.

PIOC are currently managed by wide surgical resection. Other treatment modalities, such as radiotherapy or chemotherapy, should be considered only for lesions that cannot be surgically controlled. In cases of advanced operable cancers, preoperative chemo-radiotherapy and radical surgery may be effective.¹⁴ However, the effectiveness of these modalities is unclear because of less number of cases and documented follow-up.

The prognosis of de novo PIOC is generally poor.¹⁵ In the 12 cases of de novo PIOC reported by Elzay,¹² a 40% two-year survival was noted. Similarly, in the review of 28 cases of de novo carcinoma reported by Thomas et al.,⁵ 46% of the patients survived for a period varying from six months to five years. At the time of writing, the survival time for the presented de novo PIOC patient was 13 months after the initial diagnosis. Early diagnosis and management eventually yields a better prognosis of these rare tumors.

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