CASE REPORT

An Unusual Case of Primary Intraosseous Carcinoma of the Mandible

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Abstract

Primary de novo intraosseous carcinoma (PIOC) is a rare and unique neoplastic lesion commonly occurring in the jaws. It is locally aggressive with very poor prognosis. Hereby we are presenting a case of proliferative growth from the non-healing extraction socket in a 45 year old female patient with eroded coronoid process and ramus of the mandible radiographically.


Key words: primary intraosseous carcinoma, non-healing socket, mandible, squamous cell carcinoma.

Introduction

Primary intraosseous carcinoma (PIOC) was first described by Loos in 1913 as a central epidermoid carcinoma of the jaw.\(^1\)\(^2\) It has been referred to by a variety of names such as primary carcinoma of the mandible,\(^3\) primary epithelial tumor of the jaw,\(^4\) intra alveolar carcinoma of the jaw,\(^5\) primary intra alveolar epidermoid carcinoma,\(^6\) primary intra osseous carcinoma,\(^7\) primary intra alveolar squamous cell carcinoma (SCC) of the mandible,\(^8\) malignant primary intra osseous carcinoma\(^9\) and central SCC of the mandible.\(^10\) PIOC term was coined by Pindborg in 1971.\(^10\)

According to World Health Organization, PIOC is defined as “A Squamous cell carcinoma arising within the jaw, having no initial connection with the oral mucosa and presumably developing from residues of the odontogenic epithelium”.\(^10\)

To define a lesion in the jaws as PIOC, 3 specific criteria may be present:\(^5\)\(^11\)\(^12\)

1. Histological evidence of squamous cell carcinoma,
2. Absence of ulcer formation on the overlying mucosa, and
3. Absence of a distant primary tumor at the time of diagnosis and at least 6 months during the follow-up period.

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 coronoid process and ramus of the mandible with local metastasis.

**Case Report**

A 45 year old female patient came to the Department of Oral Medicine with complaints of swelling in the lower left back tooth region of the face since 1 week. Swelling was insidious in onset and gradually progressed to the present size. It was associated with mild, dull and intermittent type of pain. She also gave history of non-healing extraction socket since 1 year. Patient gives no history of major illness, hospitalization, prolonged medication. No history of reported drug allergies. Past dental history revealed extraction of the tooth in the same region one year back. No positive history of any deleterious habits. The patient was well oriented and her vital signs were stable.

On Extra Oral Examination, a diffuse swelling noticed in the lower left back tooth region of the face measuring 3X3cms. On palpation, it was soft in consistency, non-tender on palpation. Left submandibular lymphadenopathy was noted.

On Intra oral Examination, a well-defined solitary, proliferative growth noticed arising from the extraction socket of left mandibular third molar (Fig. 1). On palpation, it was soft in consistency, non-tender on palpation. The surface appeared to be covered by necrotic slough due to trauma from the opposing tooth. It was associated with bleeding. On hard tissue examination, multiple root stumps and missing teeth noticed.

Intra oral periapical radiograph in relation to the left mandibular third molar (Fig. 2) revealed multilocular radiolucency with alveolar bony erosions and remnant internal septae. Panoramic Radiograph showed (Fig. 3) complete destruction of left coronoid process and the erosion of the anterior border of the ramus and the alveolar bone in the molar region.

The Provisional diagnosis of intraosseous carcinoma of the mandible with the differential diagnosis of intraosseous mucoepidermoid carcinoma was considered. Incisional biopsy was done and the histopathological report showed round to spindle shaped epithelial cells proliferating in an organoid pattern. Epithelial cells were showing dysplastic features like altered nuclear cytoplasmic ratio, prominent and multiple nucleoli, pleomorphism, mitotic figures. The final diagnosis of intraosseous carcinoma of the left mandible was given. The patient was referred to the regional cancer centre for the further management.

**Discussion**

PIOC of the jaws is a rare tumor presumably developing from residues of the odontogenic epithelium. 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. There are several classifications but Waldron and Mustoe’s classification is widely accepted and frequently
cited according to which PIOC may have different origins.

Type 1: PIOC ex odontogenic cyst

Type 2a: Malignant ameloblastoma

Type 2b: Ameloblastic carcinoma arising denovo, ex ameloblastoma or ex odontogenic cyst

Type 3: PIOC arising denovo
  (a) Keratinizing type
  (b) Non keratinizing type

Type 4: Intraosseous mucoepidermoid carcinoma.

Approximately more than 150 cases of PIOC have been documented till now; consisting of more than 90 cases of PIOC type 1 and rest are PIOC type 3.13 Present case falls in the category of type 3 as it meets all the criteria explained earlier.

Only a few cases have been reported in the literature. Some reports include an exhaustive review of the published cases to define the diagnostic criteria.14 However, information about treatment modalities, patient outcomes, and comparisons with other oral cavity primary sites of squamous cell carcinoma are rarely available.

PIOC affects patients ranging from 4-90 years of age with mean age of 57 years. It affects men more than women. Majority of the cases arise in the posterior mandible where remnants of the dental lamina are most likely to be the source of epithelium. Thomas et al.10 have reported that 77.14% cases occur in the posterior mandible. Only a few occur in midline (anterior mandible), indicating that some lesions may have arisen from epithelial remnants in line of fusion of facial processes.15

Most of the carcinomas in the mandible appear as direct extensions from the oral mucosa, occasionally from intramedullary salivary gland tissue from malignant transformation of the epithelium of odontogenic cysts or possibly from ameloblastoma. Metastases to the mandible from the breast, lung, thyroid, and kidney also can occur.16 None of the features indicating any of the above tumors was encountered in our case.

The common clinical features in PIOC include pain and swelling of the affected area.17 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,7 as seen in our case.

The radiological investigations provide valuable information in diagnosing these clinically bewildering conditions. PIOC exhibit radiolucencies with a wide variation in size and shape. In a study by Thomas et al.,10 had found that PIOC 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 which was a similar finding in our case. The degree of raggedness of the border may reflect the aggressiveness of the lesion.17

Histologically, they vary from well-differentiated tumors exhibiting significant keratinization to nonkeratinizing poorly differentiated carcinomas. In our case, the tumor was a well differentiated keratinizing squamous cell carcinoma with no evidence of odontogenic cystic component.

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.18 In our patient, we could find metastasis to ipsilateral submandibular lymph nodes.

Surgery is the treatment of choice and in most cases consisted of enbloc excision or radical resection of involved bone. Distraction osteogenesis of mandibular segmental defect may be a valid alternative in those patients who are not candidates for more aggressive surgical procedures. Radiotherapy and chemotherapy should be considered only in lesions that cannot be surgically controlled.6,10,11 In cases of advanced operable cancers, preoperative chemo-radiotherapy and radical surgery may be effective.19 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. Elzay, reported a 40% two-year survival rate in the 12 cases of de novo PIOC. Similarly in the study of Thomas et al., 46% of the patients survived for a period varying from six months to five years out of 28 cases of de novo carcinoma.

Conclusion

Our case highlights that clinical and radiographic examination is one of the most effective methods for detecting early lesions of PIOC. So if a patient is reporting clinically with non healing extraction socket of the jaw for long duration and radiographically an ill-defined osteolytic lesion is seen, PIOC should be ruled out before moving forward as it can prolong the life of a patient which is the main commitment of the dentistry to each of its individual. Being an oral diagnostician, the sole responsibility we can perform in case of PIOC is the early diagnosis. Hence, accurate knowledge of this rare entity is must to prevent delayed diagnosis.

References