Primary Intraosseous Carcinoma of Mandible: A Case Report

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INTRODUCTION

Primary intra-osseous carcinoma (PIOC) is an uncommon but well-known entity. Histopathologically it is defined as an SCC arising primarily from the jaw bone having no initial connection with oral mucosa. The etiology is unclear, although cellular source of origin come from epithelial rests of Malassez present within the periodontal ligament, reduced enamel epithelium around the crown of unerupted tooth, rests of dental lamina in the gingival tissue or within the bone.1 It may also arise from previous odontogenic cyst. Malignant transformation of odontogenic cyst or odontogenic tumor have been frequently reported in the literature, but PIOC arising de novo has been less commonly reported. We hereby report a case of PIOC in a 52-year-old male patient with gross destruction of left mandible.

CASE REPORT

A 52-year-old male reported to Department of Oral Medicine and Radiology, Manipal with complaint of pain and swelling on the left side of the lower jaw since 40 days (Figure 1). Patient reported that the swelling had developed after he was hit on the left side of the face during domestic violence. He visited a local dentist thereafter, where he was diagnosed with a fracture of left body of mandible. His lower left posterior teeth were extracted as they were mobile. However, no surgical intervention was done for the fracture. Numbness of lower lip was initially present, which has recovered over the time to normal. He did not report of any difficulty in mouth opening, pus discharge or bleeding. Medical and family history was non-contributory. However the patient had a history of cigarette smoking 1 packet/day since 10 years.

Extra-oral examination revealed a firm, large swelling present in left mandibular region measuring approximately 4 cm × 3 cm causing slight facial asymmetry. The swelling was diffuse involving middle and lower third of the left side of the face extending from ala tragal line to the inferior border of the mandible. The swelling extended from angle of the mouth to the posterior border of the ramus of the mandible antero-posteriorly. Mouth opening was adequate (5 cm) without deflection or deviation of mandible. Step deformity was palpable in the lower left border of mandible 1 cm anterior to the angle of mandible. Lymph nodes were not palpable. Intraoral examination revealed Class I molar relation with no occlusal derangement. Reduced alveolar ridge height evident posterior to 36. No obliteration of vestibule was seen. The alveolar socket of 37, 38 were completely healed with overlying mucosa normal in appearance (Figure 2). No cortical expansion and no mobility of any tooth in the quadrant were elicited. However, a step defect was palpable distal to 36 with sharp bony margins of buccal and lingual cortical plates. A provisional diagnosis of fracture of left body of mandible was given.

Keywords: Carcinoma, Jaw, Mandible
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**Differential Diagnosis**

Based on the history and the findings obtained on the conventional radiographs, the differential diagnosis of chronic suppurative osteomyelitis, primary malignancy (Epithelial in origin – SCC, intra-osseous carcinoma, Connective tissue in origin – Sarcomas), secondary malignancy (metastasis) was given. Chronic suppurative osteomyelitis is an inflammatory condition of the bone marrow that initially produces pus and later affects the calcified components. Etiology can be odontogenic infections and fractures following trauma. It is more common in men and more commonly occurs in mandibular premolar region. Radiographically, there is poorly defined irregular radiolucency (moth-eaten appearance) and the presence of sequestrum. In our case, there was a history of extraction of teeth following trauma that could be the cause of infection that could lead to suppurative osteomyelitis. However, absence of pus discharge or sinus opening in the patient and absence of sequestrum in radiograph, helped in ruling out this condition.

SCC is the most common malignant tumor of the oral cavity. It can arise from the epithelium of oral cavity or de novo from the bone from the epithelial cell rests or from epithelium of odontogenic lesions like cysts and ameloblastoma. PIOC is however a rare entity.

Sarcomas arising in jaw bones include osteosarcoma and chondrosarcoma. Chondrosarcomas rarely found in jaws and occurs in anterior alveolar process of maxilla and
at angle and alveolar ridge of premolar-molar region of mandible. However, it is slowing growing and painless in early stages. Osteosarcoma is most common primary malignant tumor of bone and among the jaw bones occur most commonly in the body of mandible. However, it is rare in jaw bones accounting for only 7% among all osteosarcomas with most frequent association in long bones. It grows rapidly giving a moth-eaten appearance in initial osteolytic stage. The mean age of occurrence is 33 years.

Since the patient had a history of smoking and the most common metastasis to jaw bones is from lungs, a chest radiograph was taken, which came out to be normal. Ideally a bone scan should have been performed, but due to financial constraints of the patient, it was not carried out. A bone biopsy was performed from 36, 37 region and was sent for histopathological examination. Biopsy revealed presence of dysplastic cells invading the underlying connective tissue stroma, marked pleomorphic cells and nuclei, altered nuclear-cytoplasmic ratio, numerous keratin pearl formation, individual cell keratinization, loss of cohesion and occasional mitotic figures. The histopathological diagnosis came out to be moderately differentiated SCC (Figure 5).

The origin of squamous cells can be from the oral mucosa or from within the bone. The absence of any defect, proliferative growth or mass intraorally suggested that the origin of the squamous cells was de novo from within the bone. Hence, the final diagnosis was “PIOC.”

Hemi-mandibulectomy was planned with supra-omohyoid resection after the status of lymph node involvement is assessed, with reconstruction of mandible. However, due to unaffordable financial conditions of the patient, he was lost for follow-up.

**DISCUSSION**

PIOC describes the SCC that develops likely from the residues of the odontogenic epithelium entrapped within the jaw with no connection with the surface oral mucosa. This tumor was first labeled by Loos in 1913. The World Health Organization in 1972 proposed the term PIOC and classified this entity as an odontogenic carcinoma.1

PIOC is more common in adults, in sixth to seventh decade of life with a male to female ratio of 3:1. It is usually situated in the posterior mandible.2 In a systematic collective analysis of world literature, the mean age of the patients at the time of diagnosis was 52.3 years with male to female ratio 2.5:1.3 Cases with the anterior maxillary involvement have also been stated.4

Its etiology is not clear however the most common factor may be a reactive inflammatory stimulus with or without genetic predisposition.5 Since SCCs may appear within the bone thus the diagnosis of PIOC is by exclusion.

Suei et al.6 anticipated few diagnostic criteria for PIOC:
1. To differentiate PIOC from SCCs of surface mucosal origin, no ulcer formation must be present on the overlying oral mucosa except due to trauma or tooth extractions.
2. To exclude the possibility of other odontogenic carcinomas, several sections of the histological specimens should demonstrate SCC without any cystic components or other odontogenic tumor cells.
3. To discard a distant metastasis from a primary tumor, chest radiographs must be evaluated at the time of diagnosis and throughout the follow-up period of more than 6 months. In our case, there was no continuity of the tumor with the overlying mucosa and the overlying, and the surrounding mucosa were quite normal.

The first and the most common clinical features may be pain and swelling of the affected area. In a study conducted by Thomas et al.3 on 33 patients pain was the most common presenting feature in 17 (54.8%) patients followed by swelling of the jaw in 16 (51.6%) and sensory disturbances were reported in five cases (16.1%).

The PIOC of the jaws are classified based on their possible origins into four types:
Type 1: PIOC from odontogenic cysts.
Type 2:
A: Malignant ameloblastoma
B: Ameloblastic carcinoma

![Figure 5: Histopathological picture of moderately differentiated squamous cell carcinoma](image-url)
Type 3: PIOC arising de novo
- Keratinizing
- Nonkeratinizing

Type 4: Intra osseous mucoepidermoid carcinoma.

The radiological investigations provide valuable information in diagnosing these clinically incomprehensible conditions. PIOC exhibit radiolucency with a wide variation in size and shape. Thomas et al. in his study reported varied radiographic presentations like small radiolucent loculations, well-defined lesions, cup or dish-shaped patterns and poorly defined moth-eaten appearance. Slowly growing tumors often display well-defined peripheries, whereas rapidly expanding lesions exhibit poorly defined, ragged borders with permissive type of destruction. Degree of raggedness of the border may reflect the aggressiveness of the lesion. 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 center of the lesion. Sometimes in small lesions the bucal and lingual cortical plates may mimic the appearance of trabecular bone. These lesions are capable of destroying the floor of the maxillary antrum, nasal cavity, the cortical outlines of inferior alveolar canal of the 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, 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 myelomas, fibrosarcoma and carcinoma arising in a dental cyst must be ruled out.

Histologically, they vary from well-differentiated tumors exhibiting keratinization to nonkeratinized poorly differentiated carcinomas. Yamada et al. in his clinicopathologic 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 lesion was a moderately differentiated SCC with no evidence of the cystic component. Our case was in accordance with almost all the criteria proposed by Suei et al. as there was the absence of any cystic component histologically thus suggesting this case a primary de novo intraosseous SCC.

Around 66% of patients with PIOC have either clinical or histological evidence of regional metastasis, which manifests initially or during the course of the disease. According to Thomas et al. metastasis to the regional lymph nodes was seen in 31.4% of cases. Metastatic spread to cervical lymph nodes has been discussed by Elzay and Muller and Waldron. It is important to rule out metastasis to the jaws in cases of suspected PIOC. Since the majority of metastatic SCCs 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 financial constraints of the patient, we went ahead with only chest radiograph.

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

The prognosis of this tumor is generally poor. Elzay reported 12 cases of PIOC among which 40% of patients had successfully completed 2 years survival. Similarly, among 28 cases of this prototype reported by Thomas et al. 46% of the patients subsisted for a period ranging from 6 months to 5 years. Early diagnosis and management eventually yields a better prognosis of these rare tumors.

CONCLUSION

This case presented to us with a trauma and turned out to be a carcinomatous lesion. Thus, the purpose of reporting this rare tumor is to add existing database that will further service in acquiring knowledge about the origin, behavior, diagnostic imaging, and treatment modalities of this truly uncommon neoplasm.

REFERENCES


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