An Unusual Case Report of Primary Intraosseous Carcinoma Impersonating as Missing Mandible
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Abstract
Primary Intraosseous Carcinoma is a rare malignant neoplasm of the jaws. It is a locally aggressive lesion with poor prognosis. We report an unusual case of Primary Intraosseous Carcinoma in a 45 year old female patient that resulted in almost complete destruction of mandible mimicking as of a missing mandible.

Keywords: Intraosseous; Carcinoma; Odontogenic; Mandible; Neoplasm; Squamous Cell Carcinoma.

Introduction
Primary Intraosseous Carcinoma (PIOC) is a carcinoma arising within the jaw. It was first described by Loos (1913) as Central Epidermoid Carcinoma of the jaw. The term PIOC was coined by Pindborg in 1971.1 About 150 cases of PIOC have been documented till now. It is derived either from the remnants of odontogenic epithelium, epithelial rests of Malassez or remnants of dental lamina.2, 3 WHO defines PIOC 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”. Hence the tumor is also referred as odontogenic carcinoma.4 The present an unusual case report of aggressive PIOC invading and destroying almost entire mandible and overlying skin. Our case was unique in that there was almost total absence of mandible except for few remnants giving the first impression as of a missing mandible.

Case report
A 45 year old female patient reported to the department of Oral and Maxillofacial Surgery with a chief complaint of swelling and pus discharge in the lower part of the face and upper part of the neck since two years. The swelling started as a small nodule over left side of the face and gradually increased in size and was associated with pus discharge. The sinus opening at the initial site of pus discharge healed gradually and later there was formation of multiple sinuses in adjacent areas. Healed areas showed severe scarring, involving lower part of face and neck (Figure 1a). She had pain in the same region since six months, which was dull aching, continuous and non-radiating in nature that subsided after taking analgesics. There was difficulty in chewing and paresthesia over the lower lip. She revealed that there was a gradual receding of her chin, decreased mouth opening and mobile teeth which later got exfoliated. There was no history of development of ulcers or any other soft tissue lesion in the oral cavity related to the findings and also no history of trauma. Her past medical, habitual and family history were non significant.

Extra oral examination revealed multiple discharging sinuses extending from both the preauricular regions, inferiorly involving the anterior part of the neck. On palpation, areas were tender, firm in consistency and without localized rise in temperature. Bilaterally cervical lymph nodes were palpable and fixed. Intraoral examination revealed limited mouth opening. Mandible was totally edentulous as the resorption started in the lower jaw with exfoliation of mobile teeth.

Orthopantomograph showed complete absence of the mandible except for the remnants seen over left side (Figure 1b). 3D computed tomography scan also showed complete destruction of the mandible with remnants of the body and ramus on the left side (Figure 1c). Osteolytic lesions were seen over lateral surface of maxillary sinus bilaterally and there was involvement of left side of nasal cavity, right temporal, mastoid and zygomatic regions. A provisional diagnosis of carcinoma of the mandible and
differential diagnosis of chronic suppurative osteomyelitis, cervicofacial actinomycosis, multiple myeloma, metastatic lesion of jaw and squamous cell carcinoma was made. Hematological investigations were within normal limits except for reduced hemoglobin [9 gm%] and WBC count (6500/cu mm). PA chest radiograph and ultrasound abdomen did not show distant metastasis or presence of any other primary sites of carcinoma. Pus culture was negative for sulphur granules, fungal hyphae or any other pathological organisms. Urine test was negative for Bence-Jones proteins. With this report a differential diagnosis of chronic suppurative osteomyelitis, cervicofacial actinomycosis, multiple myeloma was ruled out.

Incisional biopsy of the lesion was performed at the lesion over chin and neck region and histopathological examination of hematoxylin and eosin stained section revealed stratified squamous epithelium with focal hyperkeratosis, irregularly elongated rete pegs. Moderate dysplastic changes like nuclear pleomorphism, high mitotic activity and coarse chromatin clumping. Focal areas of infiltrative lesions consisting of multiple clusters and sheets of keratin pearl formation were seen associated with dense mixed inflammatory infiltration extending into deeper connective tissue stroma (Figure 1d). As the histopathological features were clearly pointing towards squamous cell carcinoma and there were no histopathological features favoring salivary gland involvement, we did not carry out any immunohistochemical studies to rule out salivary gland involvement.

Based on history, clinical, radiological findings and histopathological features, a final diagnosis of PIOC of mandible was made. Since the involvement and destruction was wide spread and the lesion extended to upper part of the face also, oncologist opinion was taken. As the lesion was extensive, only palliative medical treatment was carried out with chemotherapy and radiotherapy. Follow-up after six months revealed that the patient had mild complications like pain, dry mouth, trismus and anemia. Presently the patient is under the care of oncologist.

Discussion
The diagnosis of PIOC is often difficult as the lesion must be differentiated from
Jaws are the only skeletal bones affected primarily by intraosseous carcinoma, which suggests its origin from epithelial cells only present in them. Complicated tooth extraction or trauma may result in proliferation of these epithelial remnants which transform into odontogenic carcinoma.\(^5,10\) PIOC is seen in patients of age ranging from 4 to 80 years, with mean age of 50 years and has a male predilection.\(^10,12\) However, the present case was of a female patient. It is most commonly seen in the mandible and pain due to mandibular nerve infiltration is the most common complaint in them. Swelling, Paresthesia and trismus because of muscle infiltration are also common symptoms. The same features were seen in our case also. These patients are initially treated for presumed dental problems, which leads to delay in the diagnosis of PIOC.\(^12,13\)

Radiographically PIOC usually show an irregular pattern of bone destruction with ill-defined margins.\(^12\) Some cases may show mixed radiolucent-radiopaque appearance. Even though the lesion is rare, it shares radiological features with odontogenic cysts and tumors.\(^14\) Radiographical differential diagnosis must include periapical lesions, radicular cysts. In periapical lesions caries or trauma will be present whereas radicular cysts will display sclerotic margin.

Histopathological features of PIOC are similar to squamous cell carcinoma with or without keratinization. Ameloblastic carcinoma, intraosseous mucoepidermoid carcinoma, clear cell odontogenic carcinoma, and malignant variant of calcifying epithelial odontogenic tumour (CEOT) should be included in differential diagnosis of this lesion. Ameloblastic carcinoma shows prominent peripheral palisading and reverse nuclear polarization, which are absent in PIOC. The absence of a mucous component, clear cells, ghost cells and calcified material in PIOC serves to distinguish it from intraosseous mucoepidermoid carcinoma, clear cell odontogenic carcinoma, and malignant variant of CEOT respectively.\(^1,15\)

In the present case, the patient is having chronic extraoral discharging sinuses, severe scarring leading to a diagnosis of chronic suppurative osteomyelitis or cervicofacial actinomycosis. To our surprise, radiographs and CT scan revealed massive destruction of entire mandible with almost complete loss of mandible leading us to diagnose the lesion as a carcinoma. Histopathological reports showed moderately differentiated invasive squamous cell carcinoma. There is also no metastasis seen and no conclusive positive history to determine the cause of the condition. This seems to be a unique case which is puzzling to both the surgeon and the pathologist.

Treatment for all intraosseous carcinomas generally includes radical surgery with adequate resection.\(^11,12\) In cases where nerve infiltration is diagnosed and where the lesion cannot be surgically controlled radiotherapy and chemotherapy are used as palliative or adjuvant therapy.\(^12\) Prophylactic neck dissection is carried out if there is involvement of cervical lymph nodes.\(^3,11\) The prognosis of PIOC is poor and hence importance should be given to early diagnosis so that suitable treatment can be given at the earliest.

**Conclusion**

We present a case of PIOC of mandible which resulted in nearly complete destruction of mandible. It presented as an infectious disease but was diagnosed as a relatively rapidly destructive tumor. Even though the diagnosis of PIOC of jaws is considered rare, we suggest that it should be included in the differential diagnosis of jaw radiolucencies and should be biopsed at an early stage and should be treated immediately with a long term follow-up. We hope that presentation of such rare cases will contribute for better understanding about the various aspects of this entity.

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