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De Novo Primary Intraosseous Carcinoma: Case Report

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Abstract : *Primary intraosseous carcinoma (PIOC) arising as a de novo is a very rare malignant neoplasm, which is locally aggressive with poor prognosis. According to WHO, PIOC is defined as a squamous cell carcinoma arising within the jawbone without connection to the oral mucosa, probably from odontogenic epithelial residues. The definitive diagnosis of PIOC is very difficult because the lesion must be distinguished from tumors that have metastasized to the jaw from distant sites, from alveolar carcinoma that have invaded the bone from the surface, and from tumors of maxillary sinus origin. Because of this, the case reports of primary intraosseous carcinoma of jaw are rare. This case report is intended to discuss a case of primary intraosseous carcinoma of jaw along with a review of literature.*

Keywords: *Malignant Jaw Neoplasms ;Squamous Cell Carcinoma*

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I. Introduction

Primary intraosseous carcinoma arising *de novo* is very rare, but well known neoplasm which occurs exclusively in the jaw bones. It is locally aggressive with poor prognosis, with only 40 percent of the patients surviving more than 2 years.[1] Subcategories of PIOSCC include (1) a solid tumour that invades marrow spaces and induces osseous resorption, (2) squamous cancer arising from the lining of an odontogenic cyst and (3) a squamous cell carcinoma in association with other benign epithelial odontogenic tumours.[2] The clinical, radiological, histopathological hallmarks have been extensively discussed in the literature[1].

Clinical and imaging features are non-specific and the first impression of both clinicians and radiologists usually favours the diagnosis of SCC with bone invasion, or metastatic SCC.[3] We report a PIOSCC with atypical imaging findings on Panoramic Radiograph CT and Histopathology. the absence of an ulcer in the oral mucosa overlaying the tumor; and histopathological evidence of transition of the epithelial lining into squamous cell carcinoma. Absence of another primary tumor, as metastatic carcinoma is the most common malignancy of the jaw and thus, the diagnosis of PIOC must always be confirmed by the exclusion of a metastasis. A discussion on the relevant imaging and histological features is also provided.

II. Case Report

A 62-year-old male presented to the Department of Oral Medicine & Radiology, Government Dental College Calicut (Kozhikode) North Kerala. Due to swelling and pain in the left side of face noticed since 3 weeks. He also reported paraesthesia in the lower lip. No history of bleeding or discharge. No history of trauma .The patient was otherwise healthy with no significant past medical history, has habit of betel quid and tobacco chewing for the past 10-20 years. 4-5 times per day.

A complete head and neck examination revealed Facial asymmetric due to a diffuse swelling in the left side of face extending anteroposteriorly from the corner of mouth to the angle of mandible and supero-inferiorly from a horizontal line joining the commissure of mouth to the submandibular region on right side. The skin

overlying the swelling appears normal. No bleeding, discharge or visible pulsations noted. Mouth opening was restricted- 20mm. The inspection findings were confirmed. The swelling was tender and firm to hard on palpation. No local rise in temperature. No bleeding or discharge present and non pulsatile. Submandibular lymphnodes on right side was palpable and was enlarged, non-tender and fixed.

Panoramic radiograph showed a lytic lesion on the right side of mandible, extending anteriorly from mesial aspect of left mandibular first molar to the distal aspect of left mandibular third molar and supero-inferiorly from alveolar crest i.r.t left mandibular second molar, left mandibular third molar region to the inferior border of mandible.No root resorption of adjacent teeth seen. Destruction of the alveolar crest i.r.t left mandibular second molar and left mandibular third molar region and pathologic fracture of lower border of mandible seen. Lamina dura of adjacent teeth were lost. Displaced fracture with body of mandible left side. Primary intraosseuos carcinoma, Squamous cell carcinoma – stage IV, Metastatic lesion, Myeloma were considered the differential diagnosis. Vitality test with electrical pulp testing showed delayed response of left mandibular first molar.

Plain and contrast CT sections of mandible revealed an expensile lytic lesion with erosion noted involving body of mandible on left side with soft tissue compartment extending medially and laterally. Laterally with the lesion is noted extending to buccal musosa and bucal cavity. Posteriorly noted to cause destruction of posterior alveolar process. No evidence of extension into submandibular space, submandibular gland appears normal. Heterogenous contrast enhancement noted involving the soft tissue compartment. Bilaterally maxillary ethomoid sinus appears normal. No mucosal thickening. Nasopharynx shows no mass. Visualised orbit appears normal.

Impression: CT study of mandible shows expensile lytic lesion with soft tissue component involving body of mandible possible malignant neoplasm.

Biopsy and histopathology: Incisional biopsy of the lesion was done. Submitted section shows tissue composed of of hyperplastic hyper parakeratinized stratified squamous epithelium arranged in islands and sheets with intervening moderately collagenous connective tissue stroma. Epithelium shows keratin pearl formation , prominent inter cellular bridges with minimal dysplastic features. Area of necrosed bone and degenerated muscle bundles also present. The histopathological diagnosis came out to be moderately differentiated SCC. And Final diagnosis was made as Primary Intraosseous carcinoma Left body of mandible

III. Discussion

Primary intraosseous carcinoma (PIOC) was first described by Loos in 1913 as a central epidermoid carcinoma of the jaw.[4] Willis in 1948 renamed it as an intraalveolar epidermoid carcinoma. It was Pindborg who coined the term PIOC in 1971.[5] Swelling(32%) and pain(16%) most common, Sensory alterations(3%), Site : mandible –posterior region and maxilla - anterior region.[6]

According to World Health Organisation (WHO) PIOC is defined as "A Squamous cell carcinoma arising with in the jaw, having no initial connection with the oral mucosa and presumably developing from residues of the odontogenic epithelium". WHO classified the lesion as odontogenic carcinoma.[7] There are several classifications but Waldron and Mustoe's [8]classification is widely accepted and frequently cited according to which PIOC may have different origins.[7, 8]

- 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 According to this classification, PIOC may have different origins. Total absence of cystic component or other odontogenic tumour cells such as ameloblastoma is mandatory to diagnose PIOC type 3 (de novo). But PIOC type 1 can be identified by the presence of odontogenic cyst. Similarly, PIOC type 2 can be distinguished by the presence of malignant ameloblastoma or ameloblastic carcinoma arising de novo. Discrimination between type-3a and 3b PIOCs is based on the former lesion possessing keratin pearls and/or individual keratoses, whereas these features are absent in the latter. PIOC arising de novo must be considered if no cystic component of other odontogenic tumour cells is demonstrated. Although several cases of malignant transformation of odontogenic cysts have been reported in the literature while PIOC occurring denovo is rare. To define a lesion in the jaws as PIOC, 3 specific criteria may be present.[9]

(1) Histological evidence of squamous cell carcinoma,
(2) Absence of ulcer formation on the overlying mucosa, and

(3) Absence of a distant primary tumour at the time of diagnosis and at least 6 months during the follow-up period. To eliminate the possibility of distant primary tumour concurrence, chest radiographs, bone scintigram, and endoscopy of the gastrointestinal system and upper respiratory tract should be performed during the diagnostic phase and follow-up period. Etiology of PIOC is not clear probably it arises from the remnants of odontogenic tissues, either the epithelial rests of Malassez or the remnants of dental lamina.[7]

These epithelial remnants proliferate and transform into odontogenic carcinoma, a process that is potentially triggered by an inflammatory process. It does not have its origin from the epithelial lining of a pre-existing odontogenic cyst or the epithelial component of an odontogenic tumour. Tumour is locally aggressive and metastasis to lymph nodes. Prognosis is quite poor, with 5-year survival rates ranging from 30% to 40%. Prognosis further worsens with delayed diagnosis and treatment.[10] To *et al*. has reported delays in correct diagnosis, ranging from a few weeks to as long as 18 months.[11]

IV. Conclusion

Our case highlights that radiographic examination is one of the most effective methods for detecting early lesions of PIOCs. So if a patient is reporting clinically with asymptomatic swelling of jaw of 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.

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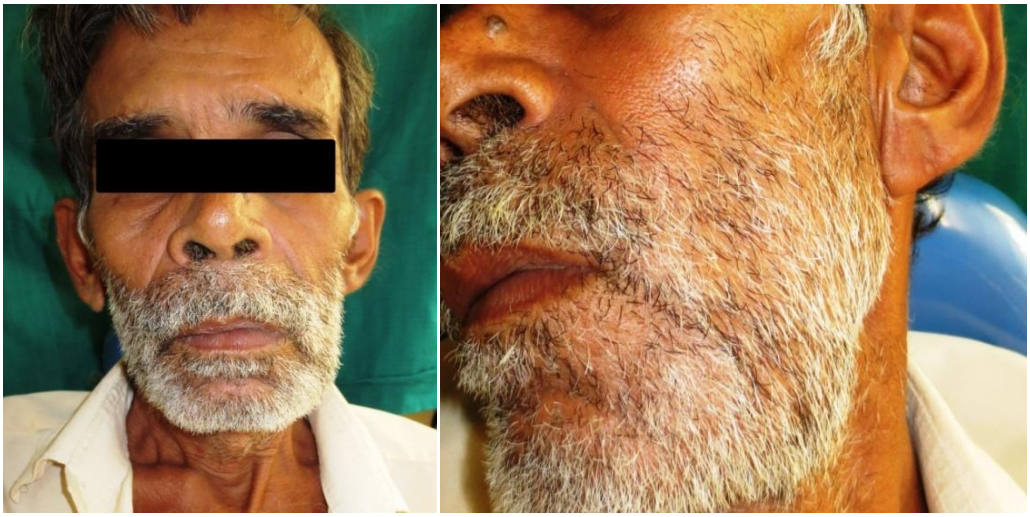
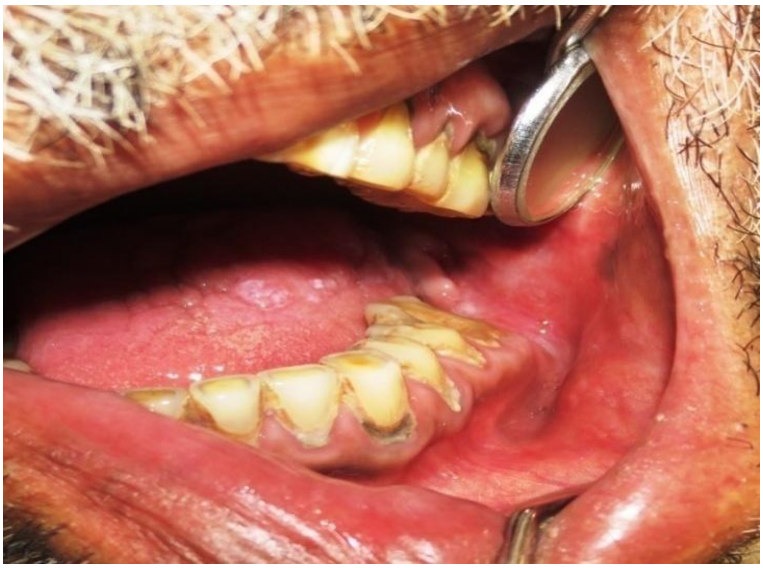
V. Figures

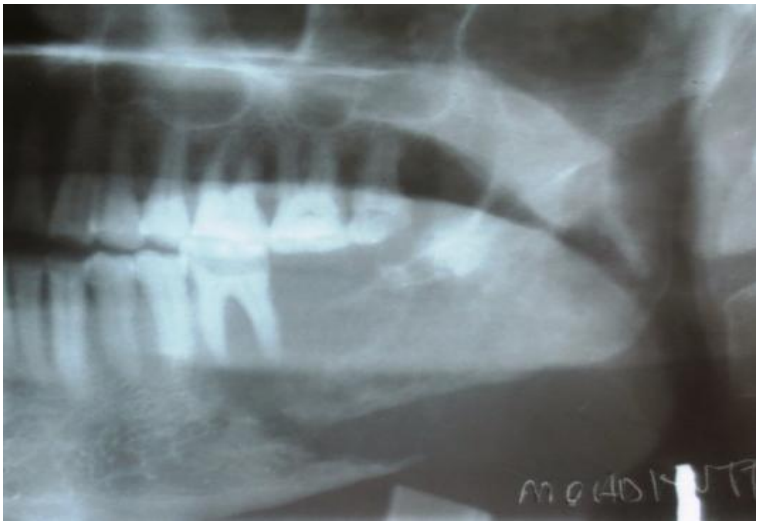
CLINICAL PHOTOGRAPHSEXTRA ORAL VIEW



INTRA-ORAL VIEW



CROPPED PANORAMIC RADIOGRAPH



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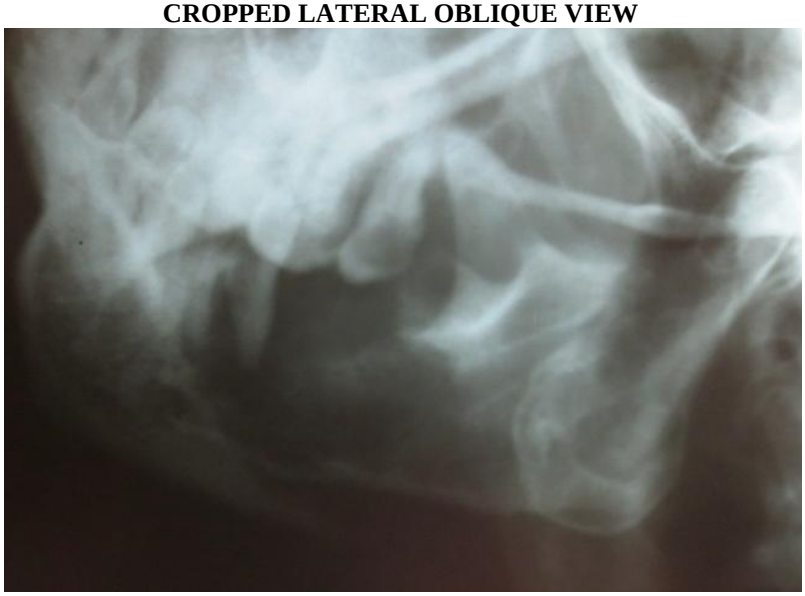

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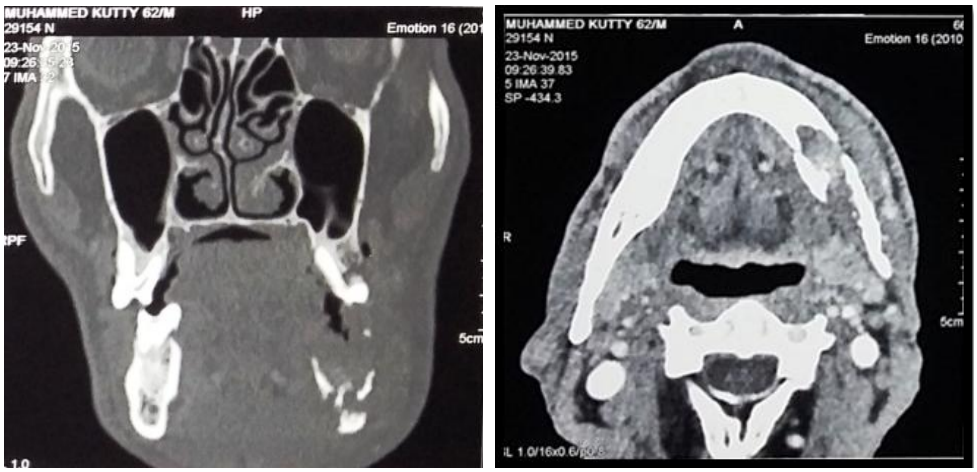
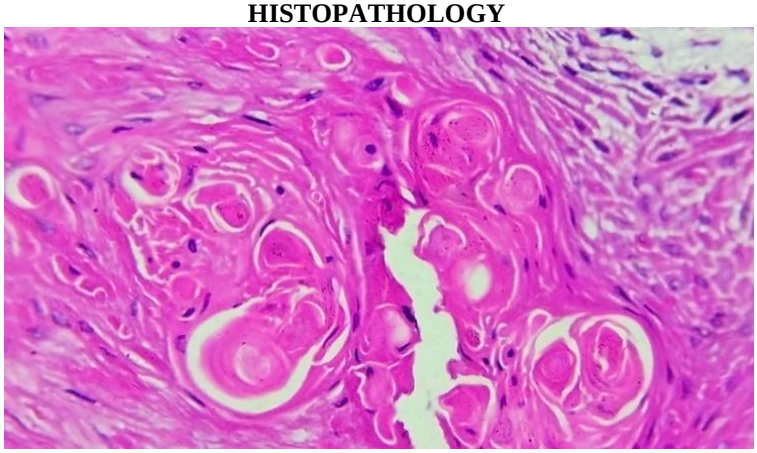


HISTOPATHOLOGY



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