CASE REPORT

Primary intraosseous carcinoma in the parasymphseal region of mandible: A case report and review

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Abstract

Primary intraosseous carcinoma (PIOC) or primary intraosseous squamous cell carcinoma is a rare and unique tumor, which occurs within the jawbones without any connection with the oral mucosa. Etiology of PIOCs is not clear, although they are presumed to be arising from the odontogenic epithelial rests or from direct transformation of odontogenic epithelium. The present case report is of a 56-year-old male patient with the complaint of swelling in the edentulous region of 44, which on radiographic evaluation and histopathological examination is diagnosed as de novo PIOC.

Keywords:
Intra alveolar epidermoid carcinoma, primary intraosseous carcinoma, Squamous cell carcinoma

Introduction

Primary intraosseous carcinoma (PIOC) is a rare and unique tumor which is exclusive to the jaws which are derived from the odontogenic epithelium without connection with the oral mucosa. It was first described by Loos in 1913 as central epidermoid carcinoma of the jaws.[1] It was given a variety of names such as primary carcinoma of the mandible, primary epithelial tumor of the jaw, intra-alveolar carcinoma of the jaw, primary intra-alveolar epidermoid carcinoma, PIOC, primary intra-alveolar squamous cell carcinoma (SCC) of the mandible, malignant PIOC, and central SCC of the mandible.[2]

According to the World Health Organization (WHO), PIOC is defined as “an SCC arising within the jaw, having no initial connection with the oral mucosa and presumably developing from residues of the odontogenic epithelium.”[3] The etiology of PIOC is not clear, and it is presumed to be derived from the remnants of the odontogenic epithelium, epithelial rests of Malassez, or remnants of dental lamina. It may arise from the previous odontogenic cyst or tumor or de novo.[4] The cellular sources of PIOC may be derived from the direct transformation of the odontogenic epithelium, particularly the odontogenic epithelial rests, such as epithelial rests of Malassez, from within the alveolar bone following tooth loss or from the remnants of the dental lamina inside the fibrous tissue of the gingiva or within the bone and the reduced enamel epithelium surrounding an unerupted or impacted tooth.[1,5]

Classification of PIOC

Waldron and Mustoe’s classification (1989)

- This 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 de novo, ex ameloblastoma, or ex odontogenic cyst
  - Type 3: PIOC arising de novo (a) keratinizing type (b) non-keratinizing type
  - Type 4: Intraosseous mucoepidermoid carcinoma.[6]

According to the WHO (2005)

- PIOC may be categorized into three types:
  i. A solid tumor invading the bone marrow spaces and inducing osseous resorption
  ii. An SCC arising from the epithelial lining of an odontogenic cyst and
  iii. An SCC that is associated with other benign epithelial odontogenic tumors.[7]
WHO 2017

PIOC appears as a single diagnostic entity in 2017. There was an attempt to divide PIOC into three subtypes according to their putative origin from odontogenic keratocysts or from other odontogenic cysts in 2005. The new edition recognizes that though some PIOC may arise from pre-existing cysts, designation as specific subtypes was not necessary nor justified on clinicopathological grounds.\[8\]

The present report describes a case of PIOC of anterior mandible arising de novo in a 56-year-old male patient.

Case Report

A 56-year-old male patient came with the chief complaint of pain in the right back tooth region of the jaw from past 4 months. The patient gave a history of mobility of 44 with associated pain 4 months back, for which he got the tooth extracted in a private clinic. Extraoral examination revealed a well-defined swelling present on the right side of mandible which is extending anteroposteriorly from parasymphysis to the body of the mandible and superioinferiorly from the corner of the mouth till the lower border of the mandible. Intraoral examination revealed an unhealed socket in relation to extracted 44 and diffuse swelling in the vestibular region of size 2 cm * 1 cm, oval in shape extending from the distal aspect of 43 to mesial aspect of 46 with partially edentulous in relation to 44 [Figure 1].

Orthopantomogram revealed a solitary well-defined radiolucency of approximately of size 2.5 cm * 1 cm extending from mesial side of 41 till mesial side of 46 and superior-inferiorly from alveolar crest till 2 cm above the lower border of mandible [Figure 2].

Based on the clinical history and well-defined radiolucency on radiograph, a differential diagnosis of central giant cell granuloma, central fibroma, and intraosseous carcinoma was considered.

An excisional biopsy was done and sent for histopathological examination. H and E stained tissue section showed the presence of numerous small and large epithelial islands in the connective tissue stroma. These epithelial islands showed dysplastic features such as nuclear and cellular pleomorphism, altered nuclear-cytoplasmic ratio, numerous atypical mitotic figures, nuclear hyperchromatism, and few bizarre cells. Few of these epithelial islands showed forming epithelial pearls and focal areas of peripherally placed tall columnar cells with centrally placed squamous cells. There is evidence of necrosis within the epithelial islands. The connective tissue stroma shows bundles of collagen fibers, fibroblasts, and endothelium lined blood vessels [Figures 3-5]. Hence, based on clinical, radiological, and histopathological features, the present case was diagnosed as PIOC.

Further investigations were followed by multidetector computed tomography (MDCT) which revealed a destructive lesion involving the parasymphysial region of the mandible

Figure 1: Intraoral photograph showing a swelling in relation to unhealed extraction socket of 44 and diffuse swelling in the vestibular region

Figure 2: Orthopantomogram showing well-defined radiolucency extending from mesial aspect of 41 to mesial aspect of 46

Figure 3: Low-power view showing islands of epithelial cells in the connective tissue stroma
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with involvement into mandibular canal and evidence of soft tissue thickening on both buccal and lingual aspects of right side of mandible with extension into lingual space and floor of the mouth. [Figure 6]. Further, treatment plan considered as surgical resection, for which a general examination was done, which revealed multiple calcific nodules in lungs, leading to pulmonary hypertension is noticed. As the general health status of the patient was compromised, surgical intervention was not performed, and hence, the patient was referred to radiotherapy.

Discussion

PIOC is an uncommon neoplasm. PIOC is seen more commonly in males than females (M:F=3:2) and is more frequent in the sixth and seventh decades of life. It occurs more frequently in the mandible (especially the posterior section) than in the maxilla. In the classification proposed by Waldron and Mustoe, the present case was a Type-3a PIOC, based on the representative histological findings of the individual cell keratosis.

Suei et al. proposed three diagnostic criteria for PIOC

1. To differentiate it from SCC of surface mucosal origin, no ulcer formation must be present on the overlying mucosa except that due to causes such as trauma or tooth extraction
2. To rule out the possibility of another odontogenic carcinoma, serial sections of histological specimens must demonstrate SCC without cystic component or other odontogenic tumor cells
3. To rule out distant primary tumor, the chest radiograph must be clear at the time of diagnosis and for a follow-up period of more than 6 months.[9]

Thomas et al. pooled analysis of PIOCs show varied radiographic presentations such as cup- or dish-shaped patterns, well-defined lesions, small radiolucent loculations, and poorly defined moth-eaten appearance.[5] According to Zwetyenga et al., osteolytic bone changes with irregular or regular margins and pathological fractures are seen in PIOC.[10] 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.[11]

Most of the cases of PIOC arise in the posterior mandible where the remnants of the dental lamina are most likely to be the source of epithelium. Thomas et al. have reported that 77.14% cases occur in the posterior mandible and a few of them occur in midline of mandible, indicating that some lesions may have arisen from epithelial remnants in line of fusion of facial processes. They have reported that sensory disturbances such as paraesthesia and numbness, mimicking facial neurological problems were the principal manifestations of de novo PIOC.[3]

Histopathologically, PIOC is characterized by the islands of malignant epithelium resembling SCC. Hence, the neoplasms that originate from squamous epithelium are considered under differential diagnosis.[12] Shear proposed that the histologic picture of PIOC will that be of SCC with the absence of keratinization, but Elzay has contradicted that it is not necessary that PIOCs are non-keratinized SCCs, but rather they can also be keratinized.[13,14] The present case is characterized by keratinizing type of PIOC. Thomas et al. reported a majority of solid PIOC are said to be keratinizing and that there was no evidence of odontogenic components in the cases.[3] Hence, to confirm the diagnosis of solid de novo PIOC, the specimen should
be thoroughly examined to confirm that there is no evidence of odontogenic cysts or other odontogenic tumor components.\[12\]

In the present case, the histopathological sections revealed islands of tumor cells with dysplastic features with no contact with the above mucosal/surface epithelium and carcinoma also showed an intimate connection to the surgical extracted site. There was no pre-existing odontogenic cyst in the patient, suggesting that this case was a primary de novo intraosseous keratinizing SCC.

Most of the cases of PIOCs are managed by wide surgical resection, and other modalities such as radiotherapy or chemotherapy are considered for the lesions which cannot be surgically treated. In the present case, as the general health status of the patient is compromised, the patient is referred to regional cancer institute for radiotherapy and a 6-month follow-up period will be done. Actually, the prognosis is hard to estimate as the disease occurs considerably rare. Elzay et al. reported that de novo PIOC has 40% 2-year survival rate and a study conducted by Thomas et al. showed 46% for a period varying from 6 months to 5 years.\[3,14\]

References