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PRIMARY INTRAOSSEOUS CARCINOMA OF MANDIBLE: A RARE CASE REPORT

Oral Pathology

Dr. Latha Mary Cherian Professor and HOD, Department of Oral Pathology and Microbiology, Government Dental College Kottayam.

Dr. M. S. Aparna* Junior resident, Department of Oral Pathology and Microbiology, Government Dental College Kottayam. *Corresponding Author

Dr. Niva P Junior resident Department of Oral Pathology and Microbiology, Government Dental College Kottayam.

Dr. Aparna Thomas Junior resident, Department of Oral Pathology and Microbiology, Government Dental College Kottayam.

ABSTRACT

Primary intraosseous carcinoma (PIOC) is a rare type of squamous cell carcinoma arising within the jaws, having no initial connection with the oral mucosa. It usually develops from the residues of the odontogenic epithelium. The diagnosis of PIOC is challenging as it has to be differentiated from other odontogenic carcinoma or metastatic tumor arising from a primary tumor at different site. In this article, we are reporting a diagnostically challenging case of primary intraosseous carcinoma involving the left mandible.

KEYWORDS

Jaw bone, Primary intraosseous carcinoma, Squamous cell carcinoma

INTRODUCTION

Primary intraosseous carcinoma (PIOC) is a rare central carcinoma accounting for approximately 1–2% of all cases of odontogenic tumors.[1] In 1913, Loos first identified and described PIOC as a central epidermoid carcinoma of the jaw. Later in 1972, Pindborg came up with the term primary intraosseous odontogenic carcinoma.[2] According to WHO classification of head and neck tumors 2022, PIOC is defined as a central jaw carcinoma that cannot be categorized as any other type of carcinoma.[1] It is believed to originate from the malignant transformation of odontogenic cysts or odontogenic embryonic remnants, including the epithelial rests of Malassez, dental lamina, and dental follicles within the jaw.[3]

Definitive diagnosis of PIOC is often challenging as the lesion must be differentiated from other odontogenic carcinomas such as ameloblastic carcinoma, squamous cell carcinoma (SCC) arising from overlying oral mucosa and tumors that could have metastasized to the jaw from other primary sites.[3] In this article, we are reporting a long standing case of primary intraosseous carcinoma involving the left mandible.

CASE REPORT

A 46-year-old male presented to the outpatient department with a chief complaint of pain and swelling on the left side of face for the past 4 months. On extra-oral examination, there was a diffuse swelling on the left ramus and body of mandible. On palpation, swelling was firm and tender. Bilateral submandibular lymph nodes were palpable, tender and fixed.

On intra-oral examination, diffuse swelling was present over the left alveolus and ramus of mandible (Figure 1). There was no ulcer or any other soft tissue changes. Pus discharge was noted concerning left and right posterior teeth. Deranged occlusion with open bite was noted. Grade II mobility was noted from 33 to 37. His medical history and family history were non-contributory. Patient reported a habit history of smoking and tobacco chewing for 30 years. Upon routine blood investigations, all values were within normal limits.

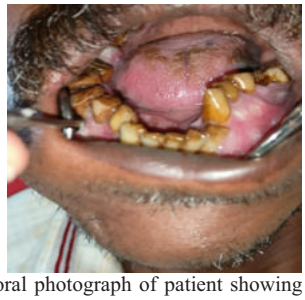

Figure 1: Intraoral photograph of patient showing diffuse palpable swelling in the left alveolus and ramus of mandible

On radiographic examination, areas of cortical disruption and pathological fracture were noted within the left mandibular body (Figure 2). Due to extensive involvement of the mandible, a provisional diagnosis of primary intraosseous carcinoma, ameloblastic carcinoma and metastatic tumor of jaw were considered. Patient underwent Contrast-enhanced CT (CECT) of thorax and abdomen to determine whether the origin of tumor was primary or metastatic. CECT reports were normal indicating that the lesion was indeed a primary tumor of the jaw.

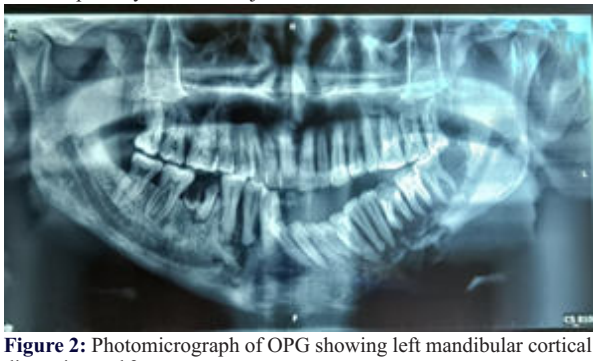

Figure 2: Photomicrograph of OPG showing left mandibular cortical disruption and fracture.

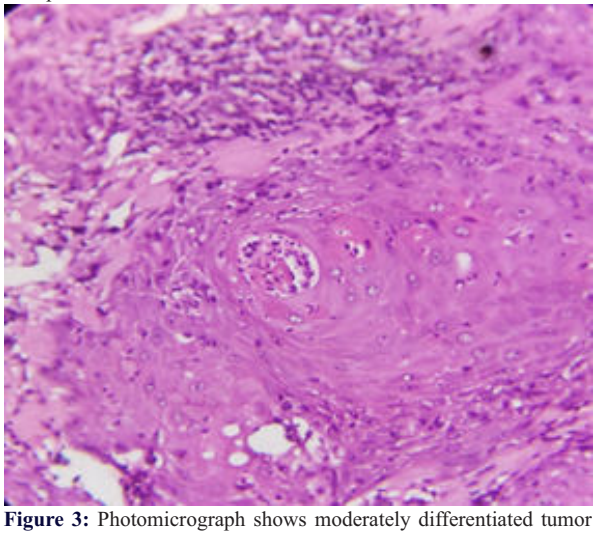

Figure 3: Photomicrograph shows moderately differentiated tumor

cells with cellular and nuclear atypia (H & E Stain, x 400)

An incisional biopsy was done and the specimen was sent for histopathological examination. The hematoxylin and eosin-stained sections revealed dysplastic parakeratinized stratified squamous epithelium. Connective tissue stroma was infiltrated by tumor epithelial cells in the form of numerous islands, nests and cords. The tumor cells were moderately differentiated and showed atypia including cellular and nuclear pleomorphism, multiple prominent nucleoli and increased mitosis. The infiltrative islands and cellular atypia were indicative of a moderately differentiated SCC (Figure 3). There were no dysplastic or malignant changes in the overlying mucosal epithelium. With these microscopic findings, a final diagnosis of primary intraosseous carcinoma was given.

The patient underwent left segmental mandibulectomy with left selective neck dissection. The left mandible was subsequently reconstructed using iliac bone graft. Patient will be followed up regularly for any evidence of recurrence.

DISCUSSION

Primary intraosseous carcinoma (PIOC) is a rare carcinoma that develops from odontogenic epithelial remnants retained in the jaw and unrelated to the mucous membranes of the oral cavity.[5] Epidemiologically, PIOC mostly affects adults, and the male-to-female ratio is 2.5:1, suggesting a predilection for males.[6] The prevalence rises with age and risk factors include a history of alcohol and tobacco use, as well as co-morbid conditions like diabetes and high blood pressure. PIOC is most common in the fifth to seventh decade of life with an average age of 54 years at diagnosis.[7]

The underlying mechanism of malignant transformation of odontogenic epithelium is poorly understood. The pathogenesis is most likely related to the unique anatomical and histological nature of the jaw as well as inflammation-induced epithelial damage. Inflammation-induced carcinogenesis has been suggested as a possible mechanism in pathogenesis. Inflammation promotes generation of reactive oxygen metabolites which can damage cell membranes, proteins, and DNA. In response, a compensatory proliferative process occurs in neoplastic cells, bypassing the normal apoptotic mechanism and ultimately leading to uncontrolled cell growth and proliferation. Given the potential role of chronic inflammation, alcohol and tobacco use have been investigated as risk factors due to their contribution to increased cellular oxidative stress. However, a definitive link between these substances and PIOC pathogenesis remains unconfirmed.[3]

Clinically, PIOC presents as a diffuse jaw enlargement, accompanied by pain, tooth mobility and non-healing extraction sites. Most cases occur in the mandible. Slow-growing tumors typically have well-defined borders, while rapidly expanding lesions may display a ragged periphery. Other findings include paresthesia or numbness of the mandibular nerve.[8,9]

Radiographically, most PIOC appear as radiolucent or mixed radiopaque- radiolucent lesions, displaying significant variability in size, shape, and border definition. They may be well-defined or poorly-defined with a characteristic moth-eaten appearance. Most cases present with root resorption and erosion of the buccal and lingual cortical plates. Patients may even present with pathological fractures if there is significant delay in seeking treatment, as seen in our case.[10] In contrast to metastatic tumors of jaw, PIOC usually arise above the inferior alveolar canal.[11]

Microscopically, PIOC consists of well to poorly differentiated SCC, which can be either keratinized or non-keratinized. It presents as sheets, islands or cords of malignant epithelial cells which are dispersed within a moderate to dense fibrous stroma.[12] The tumor cells exhibit cellular and nuclear pleomorphism, nuclear hyperchromatism and increased mitotic activity.[13] Additionally, PIOC lacks palisading columnar cells and stellate reticulum-like cells, distinguishing it from ameloblastic carcinoma.[14]

PIOC requires aggressive management with radical surgery. This includes surgical excision with marginal resection and mandibulectomy. Additionally, postoperative management should include radiotherapy, chemotherapy or a combination of both.[15] The prognosis of PIOC is poor. The 2-year and 5-year survival rates are around 60 and 40 percent, respectively.[16,17]

CONCLUSION

Primary intraosseous carcinoma is a rare central carcinoma accounting for less than 2 percent of all malignant odontogenic tumors. An incisional biopsy before initiation of treatment is always advisable for radiolucent lesions of jaw bone as this helps in excluding malignancies. PIOC possess considerable diagnostic challenges as it has to be differentiated from SCC of oral mucosa and other metastatic carcinoma of the jaws. Hence, clinicians should be aware of the various manifestations of PIOC to arrive at a conclusive diagnosis.

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