Central tumor of the Maxilla- A Diagnostic Challenge: A Rare Case Report

Shilpa Dutta Malik[1], Meghanand T Nayak[2], Upender Malik[3], Mohammad Zainul Abedeen[4]

Associate Professor [1], Professor & Head [2], Professor[3], Assistant Professor[4]

1,2,4: Department of Oral and Maxillofacial Pathology and Oral Microbiology, Teerthanker Mahaveer Dental College and Research Centre, Moradabad, Uttar Pradesh, India

3: Department of Oral Medicine and Radiology, Teerthanker Mahaveer Dental College and Research Centre, Moradabad, Uttar Pradesh, India

ABSTRACT:

Primary intraosseous carcinoma (PIOC)is an infrequent malignancy of epithelial origin frequently arising in the mandible and seldom in the anterior maxilla. It usually arises from a pre-existing odontogenic tumor or epithelial cell rests present in tooth-bearing region and is usually challenging to diagnose because of its infrequent prevalence hence, a confirmative diagnosis can only be given based on the clinical data, radiological assessment, and histopathological evaluation. Objective of the present paper is to discuss a rare case of primary intraosseous carcinoma in 55-year-old male. This case is unique as there are very few cases of such type reported so far.

Keywords: Intraosseous, carcinoma, maxilla; jaw neoplasm, solid type, novel diagnostic techniques; case report.

INTRODUCTION

The name “intra-alveolar epidermoid carcinoma” first coined by Loos in the year 1931, but it was Pindborg et al in 1971 who gave the term “intraosseous carcinoma”.[1,2] WHO, 2017 defines it as “central jaw carcinoma that cannot be characterized as any other type of carcinoma. It is assumed to arise from odontogenic epithelium. Primary intraosseous carcinoma (PIOC) is exclusively seen in jaws as a consequence of malignant transformation of epithelial cells derived from odontogenesis. Although some cases arise from an odontogenic cyst or other benign precursors few do not show any connection with odontogenic epithelium”. [3] Based on the origin, several classifications have been proposed. [4,5,6] The criteria for diagnosing PIOC incorporates nonattendance of association with overlying mucosa or skin and prohibition of metastasis with far off tumor.[7]

CASE REPORT

A 55-year-old male patient reported to the OPD of our institution and with a complaint of swelling in the upper front teeth region of three months’ duration. Detailed medical history of the patient revealed no signs of any systemic disease although patient was a chronic smoker but no signs related to tobacco smoking was observed on oral mucosa upon examination. Intra oral examination revealed multiple carious teeth with generalized periodontitis. The intra oral swelling was 2.5 cm × 2.5 cm × 2 cm in dimension and extending mediolaterally from the mesial aspect of 14 extending to the distal aspect of 22 with a buccopalatal expansion. The overlying mucosa associated was edematous, erythematous and ulcerated owing to traumatization by the mandibular teeth and the swelling was non-fluctuant and non-compressible with a firm to hard consistency. (Fig. 1. A & 1. B) The right and left submandibular lymph nodes were single, enlarged, firm, tender on palpation, and movable. Orthopantomogram revealed ill-defined radiolucency with diffuse margins involving 11, 12, 13, 21, 22, 23. Both 13 and 23 were impacted with ill-defined radiolucency and loss of cortication surrounding their coronal aspects. Loss of lamina dura was noted in relation to. 11, 12, 21, 22. (Fig. 2) Maxillary occlusal topographical view

revealed loss of lamina dura in relation to 11,12,21,22 and the teeth appeared to be hanging in air. Impacted 13 and 23 showed loss of cortication around the teeth all suggestive of a malignant rapidly destructive lesion. (Fig. 3) During incisional biopsy it was observed that the pathology was free from the surface mucosa. The tissue was processed for routine histochemistry and immunochemical study. Microscopic examination revealed infiltrative islands, strands and individual cells of squamous cell origin. Minimum to moderate keratinization was also noted in these squamous cells. Features of varying degrees of epithelial dysplasia like anisocytosis, hyperchromatism, increased N:C ratio, increased & abnormal mitotic figures, and individual cell keratinization were noted. Vesicular nucleus and increased nucleoli are also appreciated. Immunohistochemical analysis for cytokeratin revealed strong positivity in the epithelial islands. (Fig. 4. A & Fig. 4. B) Although the histopathological report was suggestive of malignancy but due to financial implications the patient can't undergo further investigatory procedures such as a chest and abdomen radiological investigations and also PET scanning.

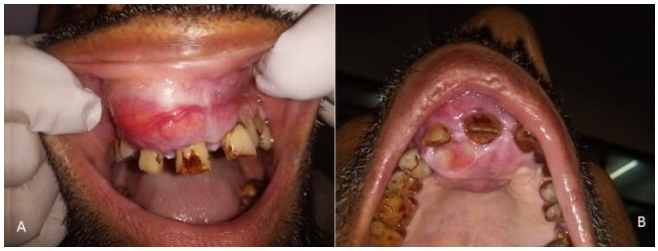

Fig. 1. Intraoral clinical photograph **s**howing an exophytic swelling in the anterior maxilla. **B.** The mucosa on palatal surface was erythematous showing indentations secondary to trauma

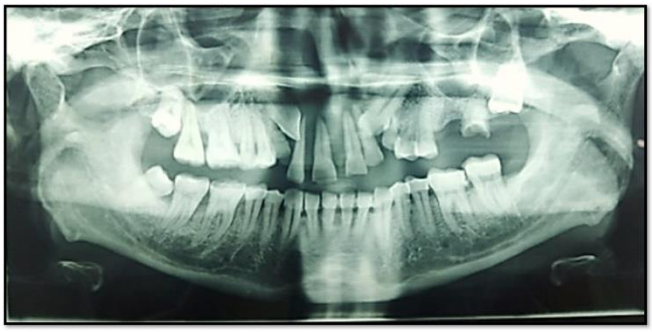

Fig. 2 Orthopantomogram depicting ill-defined radiolucency with diffuse margins and impacted permanent maxillary canines surrounded with radiolucency in their coronal aspect

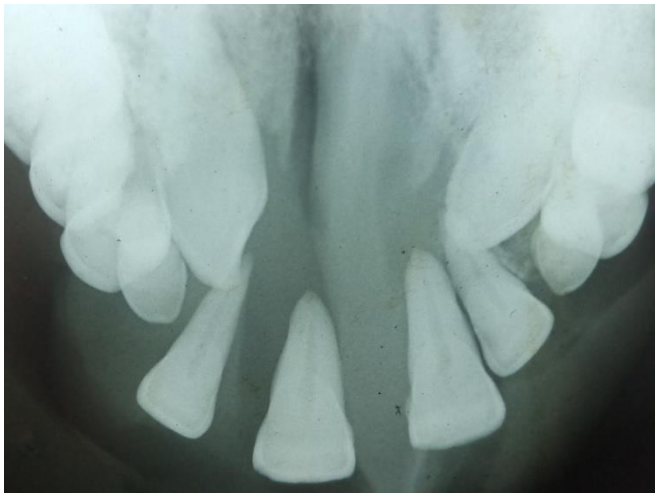

Fig 3 occlusal topographical view revealed loss of lamina dura in relation to 11,12,21,22 and the teeth appeared to be hanging in air.

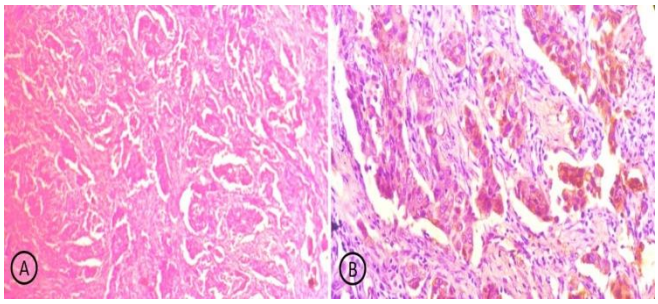

Fig. 4. A Dysplastic epithelium in the connective tissue stroma (H&E stain; ×100). **B.** Section showing positive immune reactivity (Pan-cytokeratin AE/AE3; ×400).

Discussion

PIOCS., arising solely in tooth-bearing areas of the jaws mainly in the posterior mandible and sparsely in the maxilla. The cases reported in maxilla till date have been reported only in the anterior region.[8]The majority of PIOCs are found to have male predilection (2:1).[3]and the mean age of occurrence is 51 years.[5] In the present case, the patient was male with 55 years of age, who presented with a swelling in the anterior maxilla. The clinical features in this condition run an unpredictable course and often imitate routine oro-dental diseases which could lead to delay in diagnosis, but usually the foremost frequent indications of patients withwith PIOSCC are sensory disturbances such as numbness and paresthesia.[9] Swelling and pain can also occur. The differential diagnoses include alveolar carcinomas, jaw metastases from other locations, odontogenic tumors, and tumors of the maxillary sinus.[10] On the basis of histopathology, differential diagnoses of adenoid cystic carcinoma, mucoepidermoid carcinoma, clear cell odontogenic carcinoma can be made. As PIOC is a rare, and very

difficult to diagnose and is essential to distinguish the lesion from tumors, which have metastasized in the jaws and alveolar carcinomas, which invade the bone from the surface, and maxillary sinus tumors. Suei Y, Tanimoto K, Taguchi A, Wada T[5]proposed few diagnostic criteria for PIOC [2]Radiographic investigation is one of the foremost compelling implies of identifying PIOC. To eliminate the plausibility of other odontogenic carcinomas, serial segments of the histological examples must illustrate SCC without cystic components or other odontogenic tumor cells; and moreover it can be differentiated based on IHC. To exclude distant primary tumor, chest radiographs must be clear at the time of determination and all through a follow-up period of over six months. PIOCS are classified into subtypes based on their origin.[2] Our case was of type 3A (PIOC arising de novo: keratinizing type) and showed no prime connection with the overlying mucosa or skin. Any metastasis from a distant primary tumor was ruled out and no cystic component was found. Radiolucency with respect to impacted teeth 13, 23 was also present with non-corticated borders and teeth had no lamina dura bounding them. Our case was similar in these regards to the cases reported by Bridgeman A, Wiesenfeid D, Buchanan M, Salvin J, Costello B.[2,8]As unlike other skeleton bones maxilla and mandible contains remnants of dental lamina, and reduced enamel epithelium, which is one of the possible sources of the tumor and the tumor to be called as PIOC must originate from residual odontogenic epithelium (e.g., dental lamina or Hertwig root sheath remnants), must be located within the jaws and should show no initial connection with the overlying oral, antral, or nasal mucosa or skin. The foremost suitable assignment for this injury may be a keratinizing PIOC of the front maxilla emerging de novo, sort 3A assortment.

On minuscule examination, PIOCs ordinarily display the same histopathologic characteristics as any squamous cell carcinoma (SCC). The tumor is composed of sheets, islands, and strands of squamous cells appearing stamped with cellular pleomorphism, atomic hyperchromatism, and mitotoic action. Few cases may display an unmistakably odontogenic design appearing basal sort cells orchestrated in alveoli or in a plexiform design with palisading of the fringe cells.[4] In any case, gauges of 5-year survival rates are approximately 30-40%. [2] With tumor size more than 4 cm and patients giving a positive history of smoking, indicate a poor prognosis lower survival rate [11,12].

CONCLUSION

In the present case, although the tumor size was less than 4 cm, a positive history of smoking may have caused inadvertent effects on the treatment. In the present case, among the most striking features was the loss of lamina dura around the roots of the involved teeth which when present should alert the clinician and radiologist of a possibility of malignancy and that's what alerted us all.

REFERENCES

1) Tiwari M. Primary intraosseous carcinoma of the mandible: A case report with literature review. J Oral Maxillofac Pathol. 2011 May;15(2):205-10. doi: 10.4103/0973-029X.84506. PMID: 22529582; PMCID: PMC3329686.

2) Bridgeman A, Wiesenfeid D, Buchanan M, Salvin J, Costello B. A primary intraosseous carcinoma of the anterior maxilla. J Oral Maxillofac Surg 1996; 25:279-81.

3) El-Naggar AK, Chan JKC, Grandis JR, Takata T, Slootweg PJ. WHO classification of head and neck tumors. 4th ed. Lyon, IARC;2017.

4) el-Naaj IA, Krausz AA, Ardekian L, Peled M. Primary intraosseous carcinoma of the anterior maxilla: report of a new case. J Oral Maxillofac Surg 2005;63(3):405-9.

5) Suei Y, Tanimoto K, Taguchi A, Wada T. Primary intraosseous carcinoma: Review of the literature and diagnostic criteria. J Oral Maxillofac Surg 1994; 52:580–3.

6) Elazy RP. Primary intraosseous carcinoma of the jaws. Review and update of odontogenic carcinomas. Oral Surg 1982; 54:299-303.

7) Waldron CA, Ustoe TA. Primary intraosseous carcinoma of the mandible with probable origin in an odontogenic cyst. Oral Surg 1989; 67:716-24.

8) Iino M, Ishikawa S, Ozaki H, Kobayashi T, Tachibana H, Sakurai H, Kikuchi N. Solid type primary intraosseous squamous cell carcinoma in the maxilla: report of a new case. BMC Ear Nose Throat Disord 2013; 13:13.

9) Zhang L, Liu SL, Ye WM, Zheng J. STAT3 is a key pathway in primary intraosseous squamous cell carcinoma arising from an odontogenic keratocyst. Am J Transl Res. 2015 Oct 15;7(10):1860-9. PMID: 26692930; PMCID: PMC4656763.

10) Cariati P, Fernandez AB, Tara MP, Solis JF, Lara IM. Primary intraosseous odontogenic squamous cell carcinoma of the mandible. J Oral Maxillofac Pathol 2017; 21:182.

11) Shen Q, Chen Y, Gokavarapu S, Cao W, Ji T. Primary intraosseous squamous cell carcinoma of the mandible: locoregional control and survival is significantly reduced if the tumour is more than 4cm in size. Br J Oral Maxillofac Surg 2018;56(1):48-53.

12) Lo Muzio L, Mangini F, De Falco V, Pennella A, Farronato G. Primary intraosseous carcinoma of the mandible: a case report. Oral Oncol 2000;36(3):305-7.

Corresponding Author-
Dr. Shilpa Dutta Malik
Associate Professor
Department of Oral and Maxillofacial Pathology and Oral Microbiology, TMDC&RC, Moradabad
Email- shilpa.dental@tmu.ac.in

How to cite this article: Malik SD, Nayak MT, Malik U, Abedeen MZ. Central tumor of the maxilla a diagnostic challenge: A rare case report. TMU J Dent. 2023; 10(1): 32-35.