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PRIMARY INTRAOSSEOUS CARCINOMAS OF THE JAW: A REPORT OF TWO RARE CASES

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Abstract

Primary intraosseous carcinoma of the jaw is a rare tumour presumably developing from residues of the odontogenic epithelium. The epithelial lining of odontogenic cysts has the potential to transform into various types of odontogenic tumour. Only a few cases have been reported in the literature with malignant changes in odontogenic cysts. In most cases, the diagnosis is delayed because of nonspecific clinical and radiologic findings, when present, they should alert the clinician to consider potential neoplastic growth within the cystic lining. The frequency of carcinomatous transformation within odontogenic cysts is reported to range from 0.13 to 3%. Knowledge of the clinical, radiographic, and histopathologic features of PIOC allows accurate diagnosis and appropriate treatment of this rare malignancy. Here we report two cases of primary intraosseous carcinoma of the jaw.

Key words: primary intraosseous carcinoma, odontogenic cysts, Residual cyst, Odontogenic keratocyst

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Introduction

Primary intraosseous carcinoma (PIOC) is defined as a squamous cell carcinoma arising within the jawbones; it has no initial connection with the oral mucosa and develops from remnants of odontogenic epithelium [1]. According to WHO, PIOC may be categorized into three types: i) A solid tumour invading the bone marrow spaces and inducing osseous resorption; ii) a squamous cell carcinoma arising from the epithelial lining of an odontogenic cyst; and iii) a squamous cell carcinoma that is associated with other benign epithelial odontogenic tumors. Although the classification has improved, the etiology of PIOC remains unclear. PIOC may be derived from the direct transformation of the odontogenic epithelium, particularly the odontogenic epithelial rests, such as Malassez's epithelial rest, from within the alveolar bone following tooth loss or from the remnants of the dental lamina and the reduced enamel epithelium surrounding an unerupted or impacted tooth [2]. The process of how benign cysts and tumours undergo malignant transformation is yet to be fully understood, yet it is hypothesised that chronic inflammation-induced carcinogenesis is responsible[3]. Two rare cases of Primary intraosseous squamous cell carcinoma (PIOSCC) ex‐odontogenic cyst is being presented here.The histopathology findings in the cases we present also corroborate an inflammatory-type reaction.

Case Report 1

A 57-year-old male suffered from pain and swelling with pus discharge on anterior maxillary edentulous ridge for one week. Reviewing the history of the patient, he had an injury to anterior maxilla 42 years back. He developed radicular cyst associated with non-vital 22 after a period of forty years. Cystic enucleation with multiple teeth extraction was done and the microscopic examination of cystic lining was consistent with inflammatory odontogenic cyst.

Current examination revealed a diffuse swelling along the edentulous ridge extending mesiodistally from the distal aspect of 12 to 26 region and superio inferiorly from the depth of the vestibule to alveolar crest. Swelling was soft to firm in consistency with tenderness on palpation. Mucosa over the swelling was intact with normal colour and texture [Figure:1]. Cone beam computed tomography (CBCT) revealed a large solitary expansile osteolytic lesion of size 39.65 x 12.56 x18.20 on maxillary anterior alveolus extending mesiodistally from 12 to 26 region with extensive bone destruction of the buccal cortex. The overall margins of the lesion were ill-defined and non-corticated [Figure:2]. However corticated well-defined margins were appreciated distal and superior to 11. Following an incisional biopsy of the lesion, histopathology revealed dysplastic epithelial islands within connective tissue. Cells exhibited cellular and nuclear pleomorphism, individual cell keratinisation, abnormal mitotic figures and prominent nucleoli leading to a diagnosis of poorly differentiated squamous cell carcinoma [Figure: 3].

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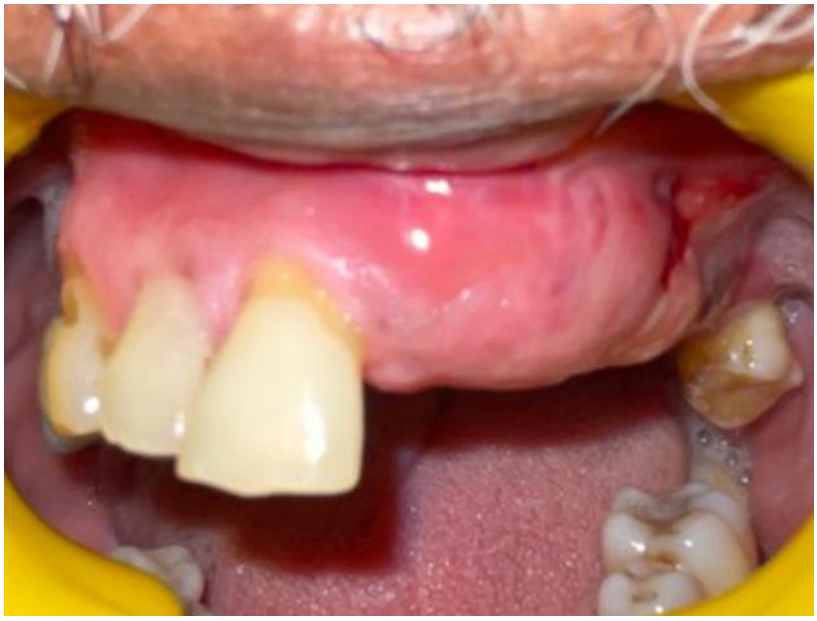

Fig 1: Diffuse swelling along the edentulous ridge extending mesiodistally from the distal aspect of 12 to 26 region

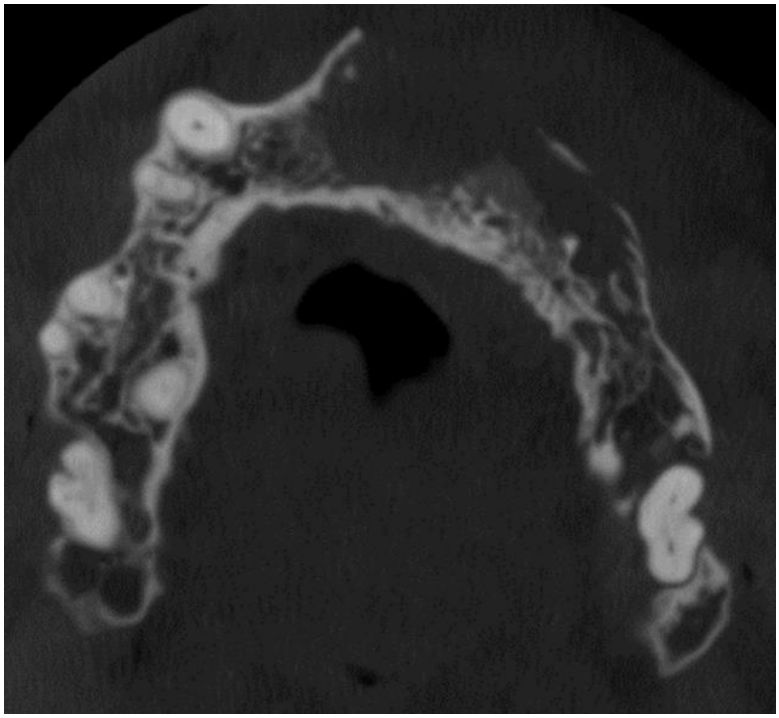

Fig 2: A large solitary expansile osteolytic lesion on maxillary anterior alveolus extending mesiodistally from 12 to 26 region

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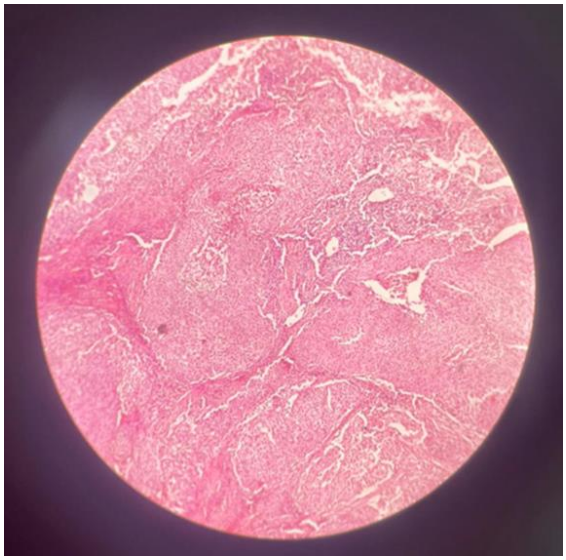

Fig 3: Cells exhibited cellular and nuclear pleomorphism, individual cell keratinisation, abnormal mitotic figures

Case report 2

A 50-year-old woman reported with pain and intermittent pus discharge on lower left back tooth region since 3 weeks. She had undergone Surgical intervention under LA three times within a span of 1 year for the recurrence of OKC in the same site. She experienced paraesthesia for 5-6 months. Thus, a careful anamnesis and oral inspection were carried out. Intraoral examination evidenced pus discharge from 34 region without any abnormal mandibular ballooning in the left side [Figure 4]. CBCT scan evidenced a surgical defect with possible healing new bone formation irt 35,36 region and possible recurrence of okc irt anterio-inferior aspect of mental foramen and posteriorly 36 ,37 region along the inferior alveolar nerve canal [Figure 5]. Incisional biopsy of the lesion revealed focal areas of dysplastic epithelium with keratin pearl formation, mitotic figures and individual keratinisation [Figure 6]. A final diagnosis of well differentiated PIOSCC arising from the remnants of OKC was established.

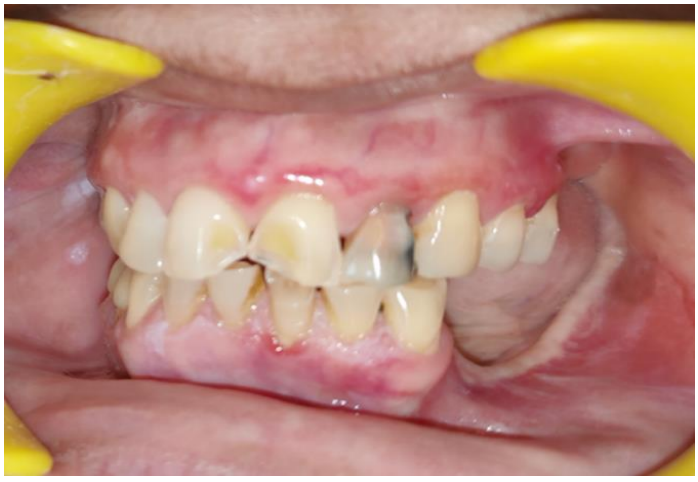

Fig 4: Left mandibular posterior edentulous region without superficial ulceration

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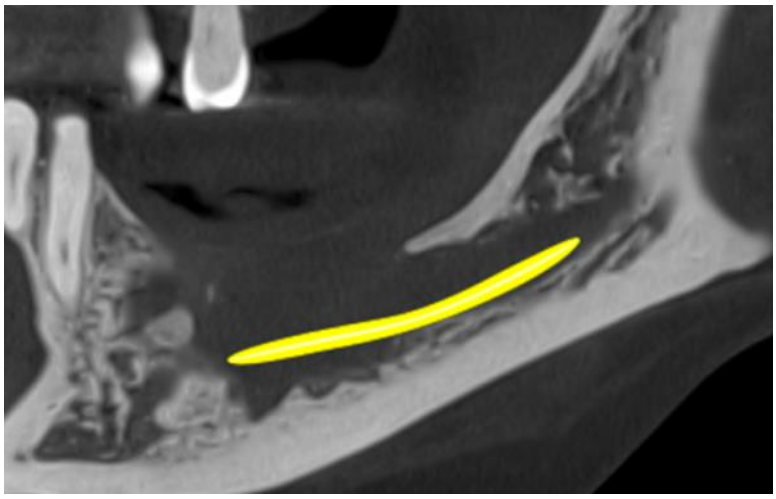

Fig 5: CBCT scan evidenced a surgical defect with possible healing new bone formation

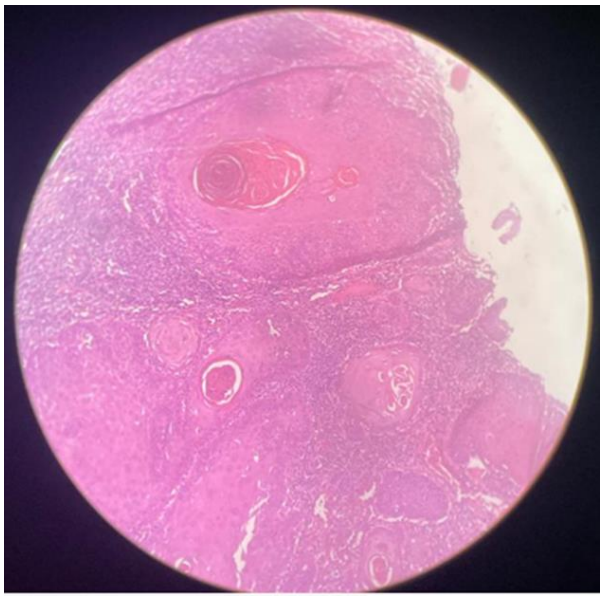

Fig 6: Focal areas of dysplastic epithelium with keratin pearl formation, mitotic figures and individual keratinisation

Discussion

Primary intraosseous carcinoma (PIOC), NOS has no specific features and can be definitively diagnosed only upon fulfilment of various criteria after careful evaluation to rule out a distant primary tumour or invasion of a surface lesion or other odontogenic tumours. According to the literature, the time diagnosis is often delayed due to the absence of symptoms provoked by pathology. The peak incidence of PIOSCC is in the fifth-sixth decade (mean 54 years), ranging from 4 to 81 years, although it is exceptional in the first two decades.our patients were in their fifties. Male:female ratio of 2.5:1 has been observed by many authors. The mandible (79%) is affected more frequently than the maxilla (21%), especially the posterior portion, while the anterior locations are very rare. Therefore, the unusual location of the lesion in the anterior maxilla in our second patient could have led to misdiagnosis and delay in the proper management [4]. Due to timely intervention, we were able to save the patient.

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Suei et al.,proposed few diagnostic criteria for PIOC;they were as follow: (1) to differentiate PIOCs from squamous cell carcinomas of surface mucosal origin, no ulcer formation must be present on the overlying oral mucosa except when due to such causes as trauma or tooth extractions; (2) to rule out the possibility of other odontogenic carcinomas, serial sections of the histological specimens must demonstrate squamous cell carcinoma without cystic components or other odontogenic tumour cells; and (3) to rule out a distant primary tumors, chest radiographs must be clear at the time of diagnosis and throughout a follow-up period of more than six months.[5]

Despite classification of the disease improving throughout the years, the etiology and pathogenesis remain to be fully understood, and the factors underlying the malignant transformation of the benign cystic lining of odontogenic cysts are not known[6].It has been suggested that chronic inflammation from infection of an odontogenic cyst may serve as a key factor in carcinogenesis .This is based on the observation that chronic infiltration of plasma cells and lymphocytes in the connective tissue of the cyst wall accompanied the malignant transformation of cyst epithelium [1]. The present cases presented with a persistent infection. It is likely that the discharge and recurrent infections were caused by the underlying malignancy. The persistent infections associated with odontogenic cysts should encourage the clinician to consider the possibility of an underlying malignancy.

The radiological appearance of PIOSCC is variable, ranging from well-defined benign-like massesto ill-defined osteolytic lesions. The most common imaging presentation is a radiolucent cupor dish-shaped osteolytic bone lesion, followed by diffuse and poorly defined borders appearanceknown as ‘moth-eaten’ radiographic pattern[7].Kaffe et al. proposed that the presence of indistinct margins without a sclerotic outline may be an important peculiarity of PIOSCC. Radiopaque foci corresponding to calcifications or dentinoid structures and ground-glass opacity (possibly mimicking fibrous dysplasia or ossifying fibroma) have also been reported by some authors.[8]

PIOSCC is not included in the AJCC classification, and there is not an internationally approved therapeutic protocol. The staging of PIOSCC is difficult. Firstly, PIOSCC cannot be classified as a primary bone tumor despite the intraosseous occurrence; secondly, the international oral squamous cell carcinoma classification is not applicable because all PIOSCCs should be classified T4 regardless of the size, due to the intimate contact with the bone marrow.[9]

Conclusion

Primary intraosseous squamous cell carcinoma is a rare and hardly recognizable tumour. The diagnostic delay could range from a few weeks to 18 months which may affect the survival rate. Lesions which may seem inoffensive and harmless both clinically and radiographically may conceivably be malignancy. With that recurrent cyst should be contemplated with foreseeable malignancies when examining clinically and radiographically.

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