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Case Report

Unusual Case of Primary Intraosseous Carcinoma Mimics Periapical Cyst in the Mandible: A Case Report and Review of Literature

Ahmed Metwally Abdelmoghny Metwally[1], Youmna Alaa Yehia[2], Heba Ahmed Saleh[1]*
[1] Department of Oral and Maxillofacial Pathology Department, Faculty of Dentistry, Cairo University, Egypt
[2] Department of Oral and Maxillofacial Surgery Department, Faculty of Dentistry, Cairo University, Egypt

Email: heba_saleh@dentsitry.cu.edu.eg

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Abstract

Aim: Primary intraosseous carcinoma is a rare neoplasm of odontogenic origin. The World Health Organization defines it as a squamous cell carcinoma (SCC), usually arises inside the jaw bones and having no connection with the oral mucosa. This lesion is documented with low incidence and unfavourable prognosis. PIOC occurred mainly in older age group with potent preference to males. In this study we presented this case to add that unusual clinical presentation of this lesion to the current literature. This case showed this malignant tumour commonly known for its unfavourable behaviour and aggressiveness documented with in unusual radiographic presentation that mimics periapical cyst

Subjects and Methods: Case Presentation of an old-aged heavy smoker male patient was admitted to Cairo University dental hospital with a complaint of painful swelling in his right posterior area of the mandible, started 4 months ago. Intra-oral examination revealed a swelling in the lower right posterior area in relation to the non-vital lower molars. A panoramic x-ray showed a well-defined radiolucent lesion apically to roots of lower right posterior teeth.

Results: An incisional biopsy was performed and the microscopic examination was done. The final diagnosis was confirmed to be primary intraosseous carcinoma.

Conclusion: Reporting this case could add to the existing literature about adding primary intraosseous carcinoma in list of D.D of any intrabony lesion regardless its size, extension and radiographic margins. This would help clinicians to early detection and improve survival rate of such affected patients.

Keywords: Primary Intraosseous Carcinoma, PIOC, Jaw Malignancy, Malignant odontogenic Neoplasm, Intrabony SCC

Introduction

Primary intraosseous carcinoma (PIOC) is a rare tumour arising within the jaw bones. Intraosseous carcinomas of the jaws can be classified according to their histological subtype into carcinomas of salivary gland, carcinomas of odontogenic origin, and also primary intraosseous squamous cell carcinomas. Loos first described PIOC in 1913 as a central epidermoid carcinoma inside the bones of the jaw (Yang et al., 2019). Although the etiopathogensis of this carcinoma is not determined yet and remains unclear, this may be related to low incidence of reported cases. Regarding histogenesis of PIOC, it is documented to arise from remnants of any odontogenic epithelium, epithelial rests of Malassez or remnants of dental lamina and that can be associated or preceded by any odontogenic cyst or

benign odontogenic neoplasm (Dungarwalla et al., 2022).

According to the World Health Organization (WHO), PIOC is defined as "a squamous cell carcinoma (SCC) that occurs in the jaw, initially unrelated to the oral mucosa, and is thought to develop from remnants of the odontogenic epithelium". The WHO classified the lesion as odontogenic carcinoma (Doroy et al., 2020). Epidemiologically, PIOC are more observed in the 6th to 7th decade with potent male predilection (Doroy et al., 2020). Although, in this case report, we present the case for patient with PIOC in fifth decade. The clinical and imaging features are nonspecific, and inconclusive. Alveolar or gingival squamous cell carcinoma (SCC) with bone invasion and metastatic SCC are usually included on top of the list of PIOC differential diagnosis (D.D). There is a known diagnostic criteria usually existing in PIOC cases as the absence of any clinical changes in the overlying oral mucosa and any other primary tumour presented at the diagnosis time. In addition, the diagnostic features of PIOC detected by the histological examination, which usually diverges from well to poorly differentiated intrabony carcinoma (Negrello et al., 2020)

The differential diagnosis (D.D) is generally made based on the clinical and radiographic presentation of the lesion, especially with other lesions that present in the same internal structures (de Morais et al., 2021). The radiographic presentation of the few reported cases of PIOC documented ill-defined radiolucency of a rapidly growing lesion (de Morais et al., 2021). Cortical bony expansion and perforation with destruction of the surrounding laminal dura are highly reported as radiographic signs in such cases, denoting tumor aggressive biological behavior. Understanding of the clinical, radiographic, and histopathological features of this tumour allows accurate diagnosis and appropriate treatment of this rare malignancy (Negrello et al., 2020; de Morais et al., 2021). No definitive diagnostic molecular pathology was conclusive. Immunohistochemical analysis of the reported cases of PIOC showed potent CK19 positive expression, endorsing its odontogenic origin. According to the latest WHO reports the staging of this tumour is no determined yet by UICC staging guidance (the use of International Collaboration on Cancer) (de Morais et al., 2021).

The overall prognosis of PIOC is poor with elevated rates of recurrence and potentiality to metastasize especially to lungs, as well as low survival rates. Those reported outcomes are closely alike to case of stage IV oral SCC with overall five year survival of around 45%. Any delay in the diagnosis of such types of carcinomas will worsen its prognosis and treatment (Deshmukh et al., 2017). This presented case report of PIOC considered to be unique as it reports a solid form of carcinoma (not associated with cyst lining) but radiographically, it was a well-defined radiolucent lesion apical to roots of lower posterior teeth, thus it mimics the periapical cyst describes a rare case of.

Case Report

A 56-year-old Egyptian worker male was heading to the Department of Oral and Maxillofacial Surgery, Faculty of Dentistry, Cairo University suffering from large painful swelling in his right posterior area of the mandible, which started 4 months ago. The pain was relieved by analgesics. The patient is considered to be a heavy smoker (40 cigarettes per day). Otherwise, he had an unremarkable medical and family history.

Clinical examination revealed that the right submandibular lymph node was fixed upon palpation. Intra-oral examination showed a diffuse large yellowish pink friable buccal swelling in relation to the lower right first, second molars. Clinically, the lesion was 2.2 x 1.9cm in size and caused obliteration of the buccal right vestibule. Some areas of necrotic tissue in the centre of the lesion were obviously seen. The performed vitality test revealed that the two lower right molars (Deshmukh et al., 2017; Thakur et al., 2017) were non-vital. The patient has poor oral hygiene; thus, he suffered from periodontitis and gingival recession in the lower right posterior teeth.

The radiographic investigation was precisely performed through variable techniques. The panoramic radiograph revealed a well-defined radiolucent (RL) lesion extending periapically from the distal root of lower right 5 to the distal root of lower right 7, and from the apical one third of roots of lower right 6 to slightly above inferior alveolar canal. Slight resorption of the mesial root of lower right 6 were obviously detected (Figure 1). Cone beam computed tomography (CBCT) detected the perforation of the buccal plate of bone by the lesion, which extended buccally into surrounded soft tissue (Figure 2).

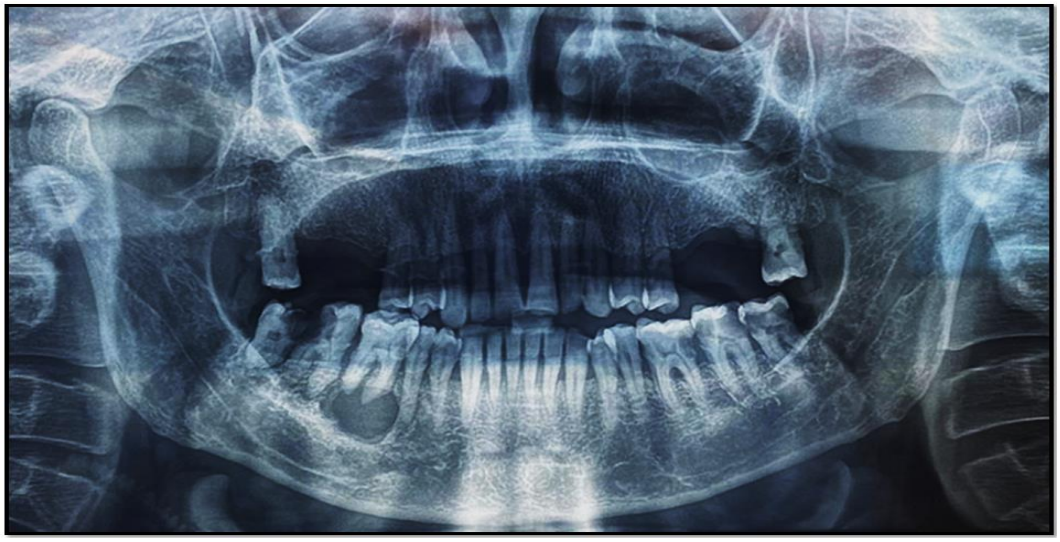

Figure 1: The panoramic radiograph showing a well-defined RL lesion periapical to the lower right first molar and the badly decayed second molar

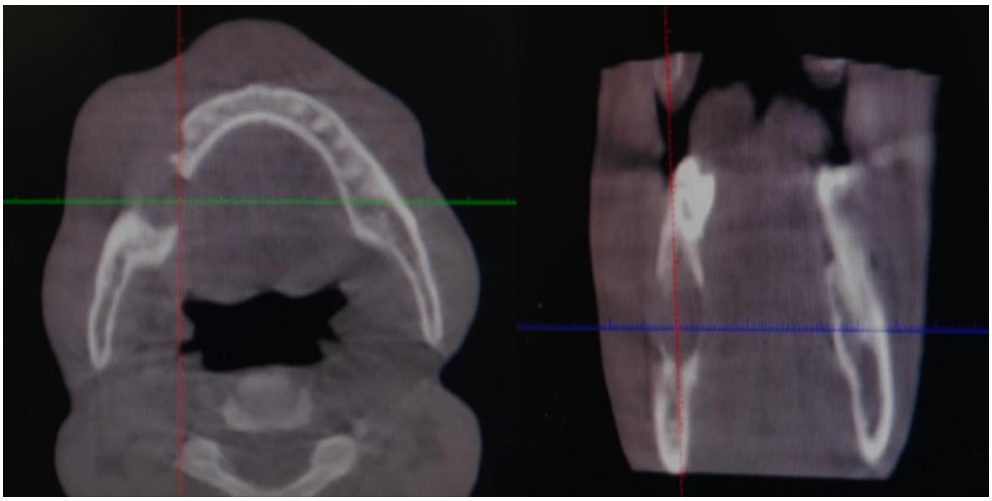

Figure 2: CBCT showing that the lesion perforated the buccal plate of bone and extended buccally into the surrounding tissue

The aspiration biopsy was negative; thus, the tumor was confirmed to be solid mass. Based on all the previous investigations, exclusion of tumors of cystic and inflammatory nature was done from the list of D.D for this case. Aggressive odontogenic neoplastic lesions were on top of our D.D list as keratocystic odontogenic tumour, pindborg tumour, PIOC and also intra osseous mucoepidermoid carcinoma were also included in this list. Other aggressive odontogenic neoplasms as ameloblastoma, ameloblastic carcinoma and myxoma were excluded from this list owing to their multilocular radiographic presentation.

A wedge shape incisional biopsy including normal and abnormal tissues was mandatory to reach final diagnosis of this case. Therefore a sulcular incision was made using Bard Parker blade no 15 and an envelope flap was made from the midline of the ipsilateral side and extended posteriorly; the flap was elevated using mucoperiosteal elevator. A soft tissue friable mass was obviously seen in the molar region; blunt dissection was made to separate the flap from the lining of the lesion and the flap was extended anteriorly and posteriorly until we found a sound bone for clinical determination of the extension of the lesion (figure 3b). Suturing was done using 3.0 silk sutures (figure 3c) and an anti-inflammatory medication was prescribed to the patient. The taken specimen was stored in 10 % formalin, and it was sent for microscopic examination at the Oral and Pathology laboratory.

After that, the macroscopic examination of the submitted specimen was done and showed multiple soft reddish-white solid pieces presented with an irregular surface and all of them were solid in the cut sections. The submitted parts were found to be of variable sizes, ranging from (2cm_0.5cm) in their measurement (Figure 3d).

The histopathological examination of H&E-stained sections revealed that the tumour was composed of sheets and strands of dysplastic squamous cells invading the surrounding stroma (figure 4a). The squamous tumour cells showed obvious signs of epithelial dysplasia as abnormal mitotic figures, nuclear hyperchromatism, pleomorphism and increase in the N/C ratio (figure 4b). Cellular pleomorphism and loss of cellular cohesion were also observed as signs of invasion of tumour cells. Intravascular invasion of dysplastic tumour cells and intense chronic inflammatory cell infiltrate and blood vessels were also seen in the surrounding fibrous stroma (Figure 4c). Dyskeratosis was clearly seen in this tumour in a form of keratin pearls inside cell nests and also individual cell keratinization of the tumour cells (figure 4d). Correlation between the clinical, radiographic and also the histopathological findings, the final diagnosis was confirmed to be PIOC and the patient was referred to National Cancer Institute to complete further investigations before starting the treatment. Adjuvant radiotherapy was administrated to this case after complete surgical removal of lesion with lymph node dissection in the neck. Performing periapical radiographs and clinical examination in the close follow-up visits every 3 months revealed no clinical recurrence until time of preparation of this manuscript.

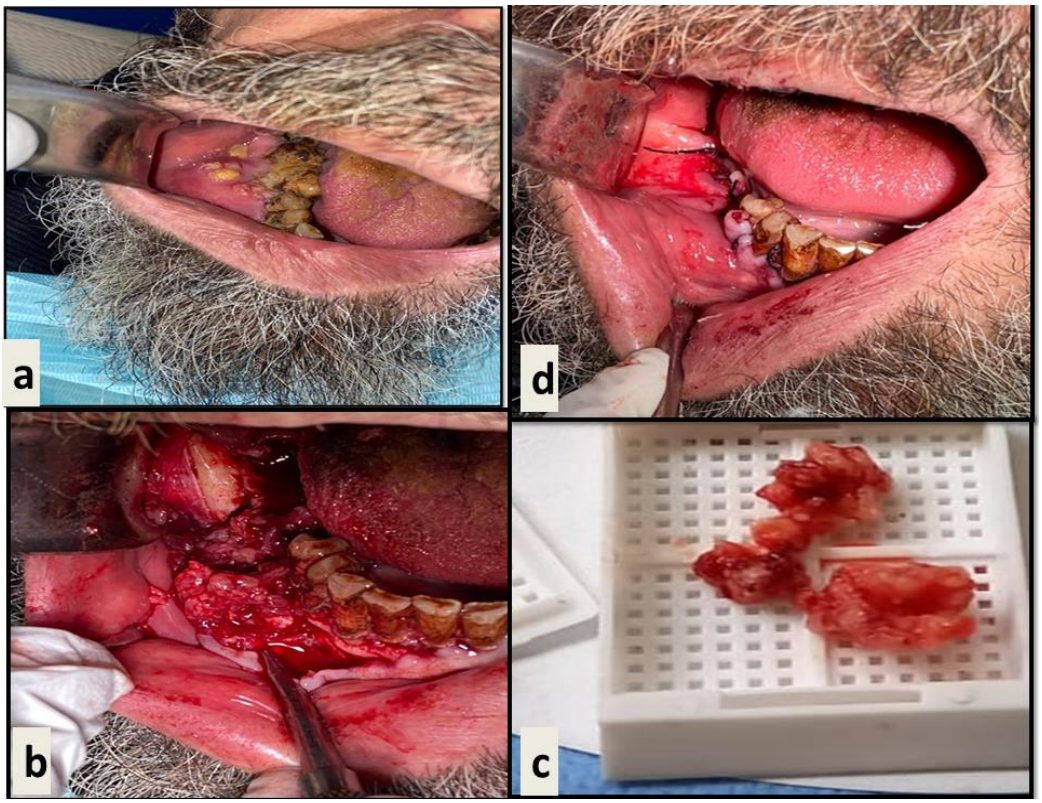

Figure 3: (a) preoperative clinical presentation of this patient with a diffuse buccal swelling with central yellow discolored areas related to lower right molars (b) illustrating the incisional biopsy steps, which started with the sulcular incision and gross examination of multiple soft masses. (c) Finally, the postoperative photomicrograph presents the sutured area after removal of the incisional biopsy. (d) Macroscopic examination of taken biopsy

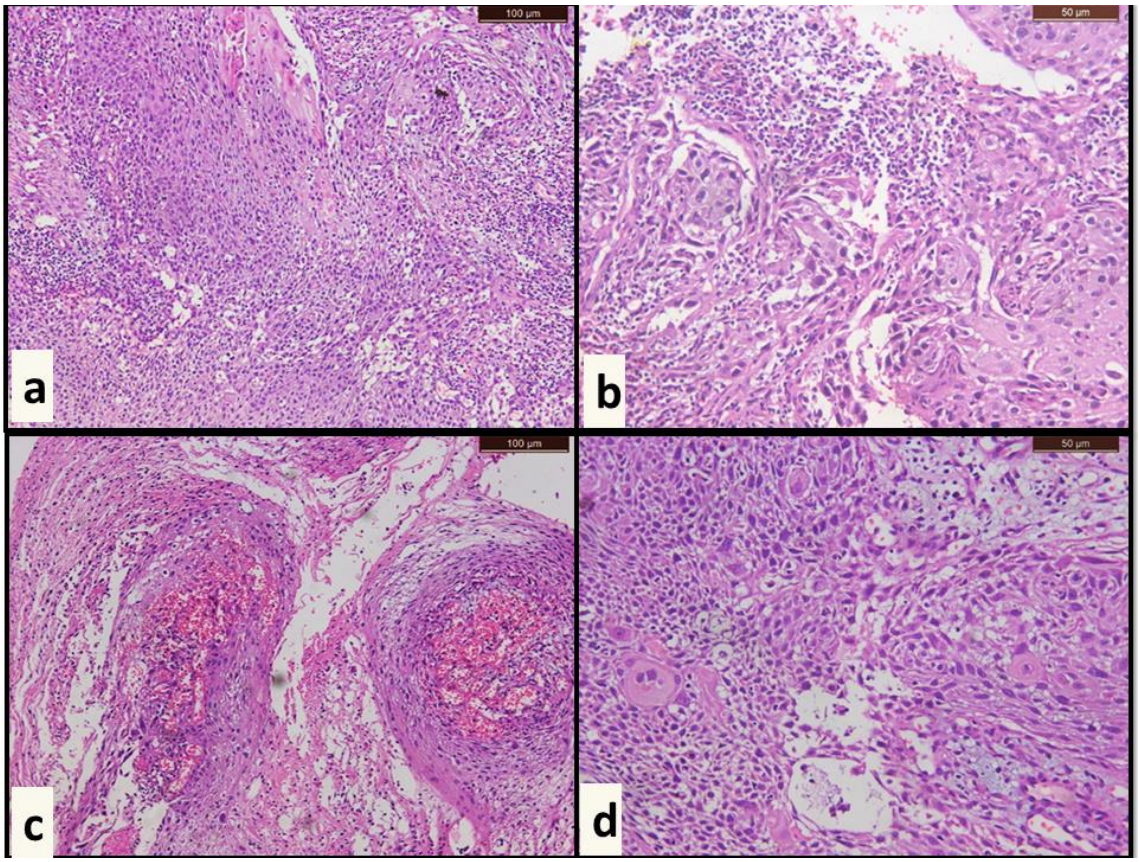

Figure 4: Photomicrographs of H&E-stained sections (a) presenting the invasion of dysplastic squamous cells into the connective tissue (x100), (b) squamous cells with several signs of dysplasia and infiltration of inflammatory cells (x200), (c) intravascular invasion of tumour cells (x100), and (d) dyskeratosis with dysplastic invasive cells (x200).

Discussion

PIOC is an uncommon malignant odontogenic tumour that accounts for approximately 1%-2.5% of all odontogenic tumours (Thakur et al., 2017). It occurs exclusively in the jaw bones (Speight, and Takata, 2018). This is known to be of odontogenic origin, and it may arise de novo or from the preceding odontogenic tumour (Deepthi et al., 2018).

The tumour is clinically found more frequently in males with male to female ratio (2:1) (Dungarwalla et al., 2022). Although it can occur at any age, it is most frequently discovered in the seventh decade of life (Yang et al., 2019). The tumour is more often intrabony located posteriorly in the lower jaw than in the upper jaw (Abdelkarim et al., 2019). Our presented and reported case was about an old male patient with the age of 56 years, presented with a diffuse painful swelling in the lower posterior tooth bearing area.

PIOC is mainly associated with different symptoms depending on its location and size. These symptoms may include pain, asymmetrical swelling, sensory disturbances and general dental disorders (Abdelkarim et al., 2019). The patient described here experienced painful swelling for about 4 months.

Radiographic examination is an additional method to diagnose PIOC that usually exhibits poorly defined, ragged borders of unilocular or multilocular radiolucency of rapidly growing tumours (Negrello et al., 2020). Due to such variations in radiological presentation of PIOC's margins, it is difficult to clinically differentiate them from other tumours especially of inflammatory or malignant nature (Abdelkarim et al., 2019).

The radiological and clinical characteristics of PIOC mimic many other odontogenic tumours. In some instances, early-stage PIOC may mimic routine dental disorders, such as periapical and periodontal disease, which may lead to delay in its diagnosis (Thakur et al., 2017). Trying to list the D.D of this case starting with the radiograph, we excluded the multilocular aggressive odontogenic lesions as ameloblastoma, ameloblastic carcinoma and myxoma based on radiographic unilocular presentation of our case. Discovering a very

important radiographic clue as the perforation of the buccal plate of bone, this raised our suspicion towards the aggressiveness of this lesion. The performed aspiration was negative denoting the solid component of this intrabony compact mass; this was confirmed during the macroscopic examination as the tumour presented with a solid cut section. This finding guided us to exclude many indolent benign or cystic lesions from our list of D.D.

Correlating all the findings from the previously mentioned investigations, guided us to put a final D.D list. In this list we included keratocystic odontogenic tumour, pindborg tumour and PIOC. Finally, we also included the intraosseous mucoepidermoid carcinoma which may arise in this location from walls of odontogenic cyst or from a pseudocyst known as static bone cyst.

The 5-year survival rate of PIOC was less than 45% of the documented cases. Owing to the elevation in the recurrence and the mortality rates, PIOC should be aggressively treated with radical surgery. In addition, concurrent chemoradiation therapy is strongly recommended for treatment of PIOC after tumour resection. This recommended line of treatment aiming to improve mortality rate and to reduce chances of recurrence and metastases (Negrello et al., 2020; de Morais et al., 2021). Finally, for documentation of the strengths of this presented case, this lesion originated de novo (not associated with any other odontogenic cyst) as well defined radiolucency in radiograph (apical to lower posterior teeth). But due to rarity of this lesion in the scientific we cannot ensure that is a constant finding for this lesion. Highlighting to specific staging and therapeutic protocols are recommended to be elaborated for such rare and aggressive intrabony carcinomas.

Conclusion

PIOC is an unusually presented malignant odontogenic neoplasm that can be misdiagnosed with any other odontogenic lesion because it occasionally presents a well-defined border on radiography. Careful examination and correlation between all the investigations are mandatory to reach quick and accurate diagnosis. Documentation of additional cases of unusual presentation is critical to provide detailed information about such malignancies and improve the understanding of the biologic behavior of this neoplasm.

List of Abbreviations

PIOC: Primary intraosseous carcinoma
WHO: World health organization
SCC: Squamous cell carcinoma
D.D: Differential diagnosis

Conflict of Interest:

All the authors declare that they have no conflict of interest regarding this submitted manuscript.

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This study proposal was revised and approved by Research Ethics Committee, Faculty of Dentistry, Cairo University (no.74 7 23). Patients' name included in this reports were kept confidential and were not utilized in this study.

Data Availability:

Data will be available upon request.

Credit Statement:

Author 1: Data curation, Writing-review & editing, Writing-original draft, Methodology, Conceptualization, Resources

Author 2: Data curation, Conceptualization, Project administration, Supervision, Methodology, Writing - review & editing, Writing - original draft

Author 3: Methodology, Writing - original draft, Writing - review & editing, Investigation, Formal analysis, Supervision, Data curation.

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