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Primary Intra-Osseous Carcinoma Of The Maxilla With Advanced Local Disease And Extensive Nodal Metastasis: A Rare Case Report

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Abstract:
Primary intraosseous carcinoma (PIOC) is a rare malignant neoplasm arising within the jaw bones, derived from odontogenic epithelial remnants without initial connection to the oral mucosa. We report a case of PIOC in a 65-year-old male who presented with a progressive swelling in the right maxillary alveolar region a year after tooth extraction. Imaging revealed an expansile osteolytic mass involving the right maxilla with extension into the maxillary sinus and palate. Histopathological diagnosis from core biopsy confirmed PIOC. The patient underwent subtotal maxillectomy with modified radical neck dissection. Histology demonstrated lymphovascular invasion and nodal metastasis in 11 out of 18 lymph nodes, with extranodal extension and salivary gland involvement. Such a presentation with advanced local disease and extensive nodal metastasis is rare and holds clinical significance.

Keywords*: Maxilla, jaw, tumor*

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I. Introduction

Primary intraosseous squamous cell carcinoma (PIOSCC) is a rare malignant epithelial tumor arising within the jaw bones without any initial continuity with the overlying oral mucosa, skin, or sinonasal lining [1,2]. According to the World Health Organization Classification of Head and Neck Tumours, 5th edition (2022), PIOSCC is categorized under malignant odontogenic tumors and is believed to originate from odontogenic epithelial remnants such as the dental lamina, epithelial rests of Malassez, or through malignant transformation of odontogenic cysts and benign odontogenic tumors [1,3]. PIOSCC constitutes less than 1–2% of all oral squamous cell carcinomas, making it a distinctly uncommon entity [2,3].

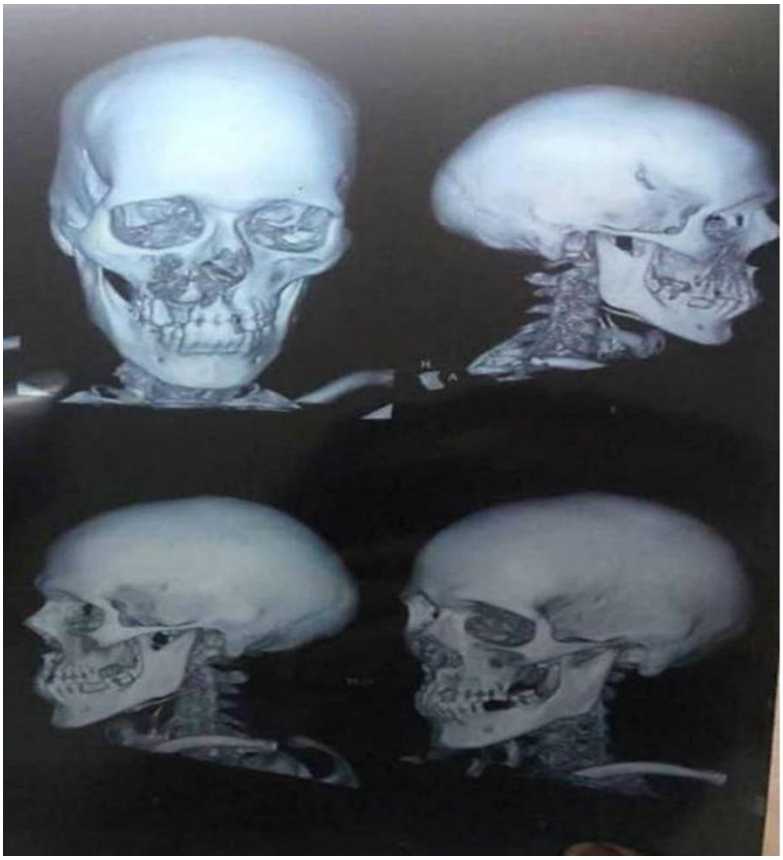
The mandible is the most frequently involved site, particularly the posterior region, whereas maxillary involvement is rare [2,4]. Despite its lower incidence, maxillary PIOSCC poses significant diagnostic and therapeutic challenges due to its close proximity to the maxillary sinus, nasal cavity, orbit, and skull base [4,5] The previously reported cases describe a limited metastatic potential of this lesion. [4] We report one rare case of maxillary PIOSCC with advanced local disease and extensive nodal metastasis.

II. Case Presentation

A 65-year-old male presented with a gradually progressive swelling in the right anterior maxillary region. The swelling was first noticed approximately one year after extraction of a right upper anterior tooth and gradually increased in size. There was no associated pain, ulceration, discharge, or sensory disturbance.

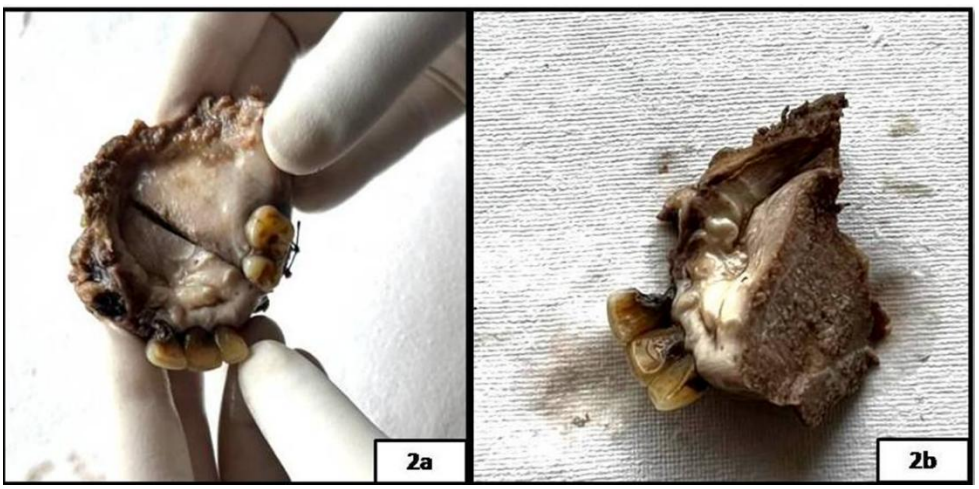
Intraoral examination revealed a solitary, diffuse swelling measuring approximately 3 × 2 cm in the right maxillary alveolar region, extending into the buccal vestibule and palatal aspect. The overlying mucosa appeared normal in color and texture, with no evidence of ulceration. On palpation, the lesion was bony hard and non-tender. Extraoral examination showed mild fullness over the right maxilla, with freely mobile overlying skin and no facial nerve involvement.

Contrast-enhanced computed tomography of the face and neck demonstrated an ill-defined, heterogeneously enhancing expansile osteolytic lesion measuring approximately 29 × 19 × 28 mm involving the right maxilla (Figure 1).

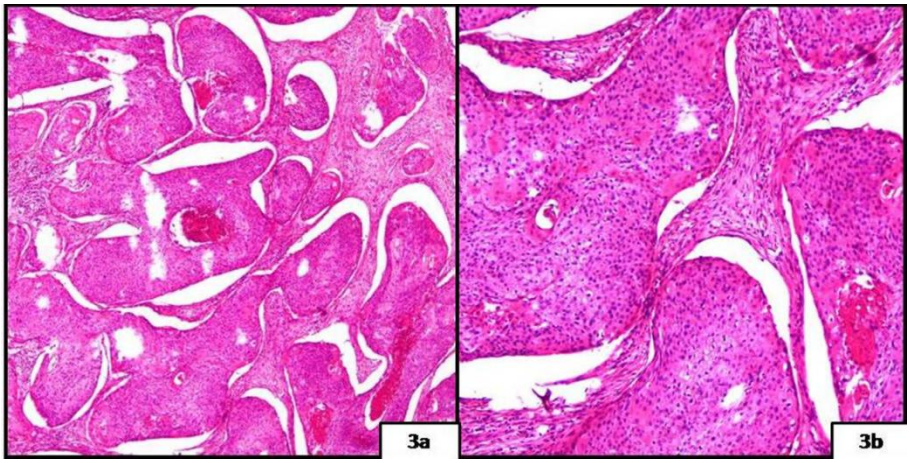


There was destruction of the hard palate and medial and inferior walls of the maxillary sinus, with extension into the sinus cavity. No obvious primary mucosal lesion was identified. A core needle biopsy was performed, and histopathological examination was consistent with primary intraosseous squamous cell carcinoma. The patient subsequently underwent subtotal maxillectomy with modified radical neck dissection to achieve oncological clearance.

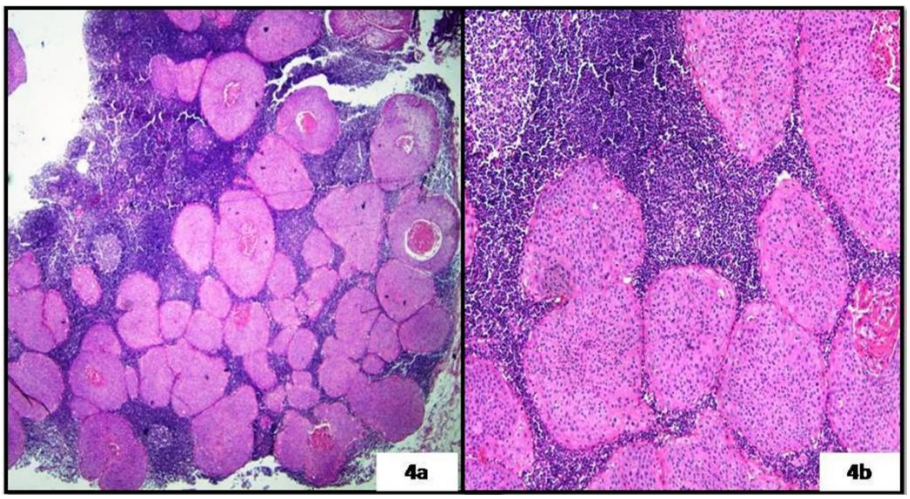
Gross examination of the resected specimen revealed an ill-circumscribed infiltrative tumor involving the maxillary bone, with extension into adjacent soft tissue and salivary gland structures. The cut surface was firm and grey-white, with areas of bone destruction (Figure 2).



Microscopically, the tumor was composed of invasive nests, islands, and sheets of atypical squamous epithelial cells infiltrating the bone and surrounding soft tissues. The tumor cells exhibited marked nuclear pleomorphism, hyperchromasia, prominent nucleoli, and increased mitotic activity. Keratin pearl formation and individual cell keratinization were evident, consistent with squamous cell carcinoma (Figure 3). Lymphovascular invasion and perineural invasion were identified .



A total of eighteen cervical lymph nodes were dissected, of which eleven showed metastatic squamous cell carcinoma (Figure 4).



Several involved lymph nodes demonstrated extranodal extension. All surgical resection margins were free of tumor. In conjunction with the clinico radiologic findings, final diagnosis of PIOC was rendered.

III. Discussion

The WHO 2022 classification recognizes PIOC under malignant odontogenic tumors with three categories: type 1 solid de novo, type 2 associated with odontogenic cysts, and type 3 with odontogenic tumors. Strict diagnosis requires:evidence of intraosseous tumor origin without mucosal involvement, exclusion of metastasis from other head and neck or distant sites and histopathological confirmation as squamous cell carcinoma. This case fulfilled these criteria supported by absence of mucosal involvement or remote primary tumors.

Most published reports of primary intraosseous carcinoma (PIOC) of the maxilla describe lesions detected at an early or intermediate stage, frequently following dental extraction or during evaluation of presumed odontogenic cysts [4,5,6]. Early maxillary cases reported by Ohba et al. and Lukinmaa et al. presented as small cyst-like radiolucencies with limited cortical destruction and minimal cervical nodal involvement [4,5]. Similarly, Takahashi et al. described a dentigerous cyst–associated maxillary PIOC detected incidentally and managed successfully without nodal metastasis [6]. In contrast, the present case demonstrated advanced local disease at initial presentation, with extensive osteolytic destruction of the maxilla and invasion into the hard palate and maxillary sinus, indicating a more aggressive biological behavior than that seen in most previously reported cases.

Another major point of contrast is the extent of cervical lymph node metastasis. Earlier literature suggested that PIOC, particularly tumors arising from odontogenic cysts, may exhibit relatively limited metastatic potential [7]. Bodner et al., in their large clinicopathologic review, reported nodal metastasis in a subset of cases, often involving fewer lymph nodes [2]. However, more recent studies, including those by Yamada et al. and Oriyama et al., have highlighted that advanced-stage and maxillary tumors show a

significantly higher propensity for nodal spread [8,9]. The present case is distinguished by extensive nodal involvement with extranodal extension (ENE), with metastasis in 11 of 18 lymph nodes—an extent of disease that is rarely documented in earlier maxillary PIOSCC reports and represents a clear indicator of poor prognosis [2,8].

Regarding etiopathogenesis, several reported maxillary PIOCs demonstrated a clear association with odontogenic cysts, supported by histologic transition from cyst lining to invasive carcinoma [6,10]. Molecular studies, such as those by Zhang et al., have emphasized the role of chronic inflammation and activation of oncogenic pathways in cyst-associated tumors [11]. In contrast, the present case did not reveal a recognizable cystic precursor, suggesting a probable de novo intraosseous origin. This distinction is clinically relevant, as de novo PIOC has been reported to exhibit more aggressive behavior than cyst-derived tumors [3,9].

Radiologically, many published cases initially showed well-defined or cystic lesions, contributing to diagnostic delay [4,6]. In contrast, imaging in the present case revealed a frankly destructive, ill-defined osteolytic lesion, consistent with advanced malignancy rather than benign odontogenic pathology.

Overall, compared with previously reported maxillary PIOCs, the present case is notable for advanced local invasion, extensive nodal metastasis with ENE, and probable de novo origin, reinforcing that maxillary PIOC can behave as a highly aggressive malignancy requiring early recognition and radical management [2,8].

IV. Conclusion

Primary intraosseous squamous cell carcinoma of the maxilla is a rare and aggressive odontogenic malignancy that often presents with nonspecific clinical features, leading to delayed diagnosis [2,4]. Persistent or progressive osteolytic lesions of the jaw, particularly following dental extraction, should raise suspicion for this entity. Accurate diagnosis relies on careful clinicoradiological correlation and strict application of diagnostic criteria [1,3]. Aggressive surgical management combined with appropriate neck dissection and adjuvant therapy offers the best chance for disease control [2,8]. Lifelong follow-up is recommended due to the high risk of recurrence and metastatic spread.

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Legends For Figures

Figure 1: CECT Showing Locally Advanced Lesion: Ill-Defined, Heterogeneously Enhancing Expansile Osteolytic Lesion Extending Into Hard Palate And Maxillary Sinus

Figure 2: Gross Image Showing An Ill Defined Invasive Mass Lesion Within The Maxillary Bone With Unremarkable Overlying Mucosa.

Figure 3a: Microscopic Image Showing Nests Of Tumor With Squamous Diffrentiation, H&E, 200x
Figure 3b:.Higher Power View Showing Intracellular Keratin And Intercellular Bridges, H&E, 400x

Figure 4a: Microscopic Image Showing Metastasis To Lymphnode, H&E, 200x
Figure 4b: Microscopic Image Showing Metastasis To Lymphnode, H&E, 400x