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Metastatic PIOSCC of the mandible arising from recurrent OKC

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Abstract

Primary intraosseous squamous cell carcinoma (PIOSCC) is a rare malignant neoplasm derived from odontogenic epithelial remnants in the central jaw bone. Most PIOSCCs originate from odontogenic cysts with a nonkeratinized epithelial lining, especially from radicular/residual and dentigerous cysts. There have been few reports of PIOSCCs derived from the odontogenic keratocyst (OKC), the diagnosis of PIOSCC is difficult and based on exclusion of other carcinomas, including metastatic tumors from other primary sites. Here, we report an extremely rare case of PIOSCC derived from the recurrent OKC of the mandible.

Introduction

Primary intraosseous squamous cell carcinoma (PIOSCC) is a carcinoma arising from the central bone without any initial connection to various epithelia **[**1**]**. In 2017, in the World Health Organization (WHO) classification of tumors, the name was changed from PIOSCC to primary intraosseous carcinoma (PIOC), not otherwise specified (NOS) **[**2**]**. The diagnosis of PIOSCC requires specific criteria to be met, including the absence of oral ulceration or communication with the overlying mucosa, the absence of a distant primary tumor at the time of diagnosis, and histologic evidence of squamous cell carcinoma **[**1, 2**]**. The diagnosis of PIOSCC can be difficult, and it must be differentiated from other malignancies such as ameloblastic carcinoma and metastatic carcinomas; when it causes destruction of the cortical bone and invades adjacent soft tissues, it may be confused with a carcinoma of the oral mucosa. Most PIOSCCs originate from the epithelial lining of odontogenic cysts, especially radicular, residual, and dentigerous cysts **[**1**]**. There are about 120 reported cases of PIOSCCs arising from cysts, with 25 of them being derived from odontogenic keratocysts (OKCs) **[**3, 4**]**. PIOSCCs arising from OKCs are extremely rare, and fewer than 30 have been reported so far. Few reports have presented clear evidence of histological transition between squamous cell carcinoma and the odontogenic epithelial lining of the cyst wall. Here, we present a very rare case of initial-stage PIOSCC derived from the Recurrent OKC of the mandible.

Case Report

A 32-year-old lady reported with a complaint of mild, dull pain and growth in the left lower jaw for two weeks (Fig.1). The patient had history of swelling in the left lower gingiva and was evaluated at a local hospital and incision and drainage was done in 2014. She had a swelling in the left lower haw in 2017 and was evaluated with OPG (Fig.2), which showed a diffuse multilocular radiolucent lesion extending from the distal root of the lower left premolar to the ascending ramus of the left mandible. The computerized tomography (CT) scan also suggested expansion and thinning of both the buccal and lingual cortical plates. Based on the history and clinical as well as radiographic examination, a provisional diagnosis of OKC, ameloblastoma, ameloblastic fibroma, intraosseous mucoepidermoid carcinoma was given.

Biopsy showed OKC and Enucleation was done under GA in 2017. She reported extra oral sinus opening (Fig.3), with pus discharge in 2018, where she underwent multiple curettage and biopsy showed recurrent OKC with secondary infection. The lesion recurred in 2019, CT scan showed radiolucent lesion in the left ramus and body of mandible with erosion of both buccal and lingual cortical plates and she underwent Left hemimandibulectomy with Recon plate fixation. The final HPR showed PIOSCC. Then she was referred to our institute. When she presented to our institution, intraoral soft tissue examination revealed verrucous proliferative growth in the left floor of the mouth and previous surgical site (Fig.4). Biopsy showed Squamous cell carcinoma and CT scan revealed residual lesion in the left floor of the mouth adjacent to the reconstruction plate with no cervical lymphadenopathy or pulmonary metastasis.

Her case was discussed in the tumor board and Surgery with Adjuvant CRT was planned. She underwent Re-wide excision of the left floor of mouth lesion (Fig.5), with recon plate removal and Left Modified radical neck dissection preserving IJV and SAN and reconstruction with PMMC flap. Her Final HPR showed pT4aN0 (Fig.6). In view of multiple recurrences and the aggressive nature of the disease, she underwent 30 fractions of 60 Gy Radiotherapy with 5 cycles of concurrent Chemotherapy with Cisplatin, completed in May 2019.

On follow-up, the patient presented with subdermal nodule in the right arm, on biopsy it was a metastatic nodule(Fig.7). A PETCT scan revealed multiple distant metastases including both lungs, lytic lesions in the right 5th rib and left iliac bone along with a subcutaneous nodular lesion in the right proximal forearm and intramuscular lesion in left adductor magnus. At present, she is on Palliative chemotherapy and regular follow-up.

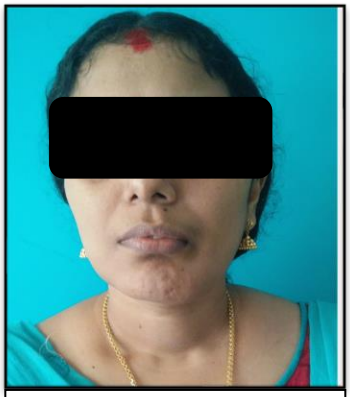

Fig.1 Frontal view

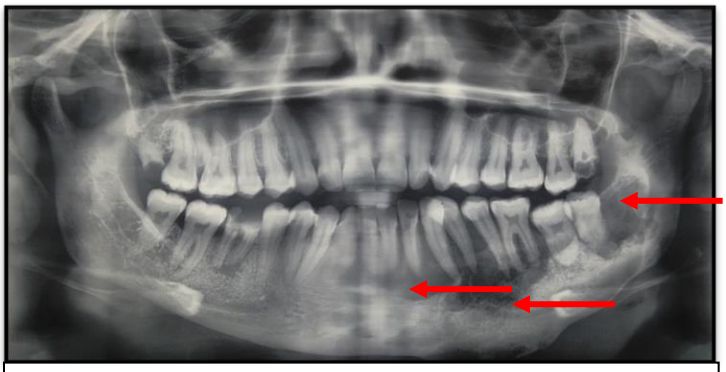

Fig.2 Orthopantomogram

Arrows showing radiolucency suggesting OKC

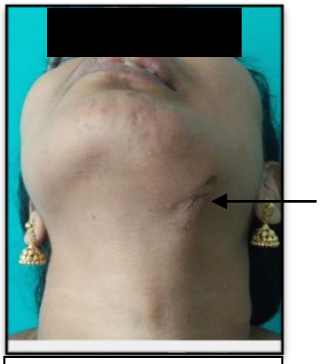

Fig.3 Extra oral sinus opening

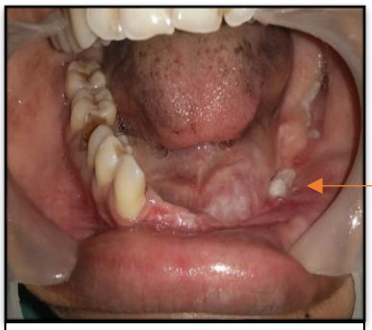

Fig.4 Intra oral photograph showing

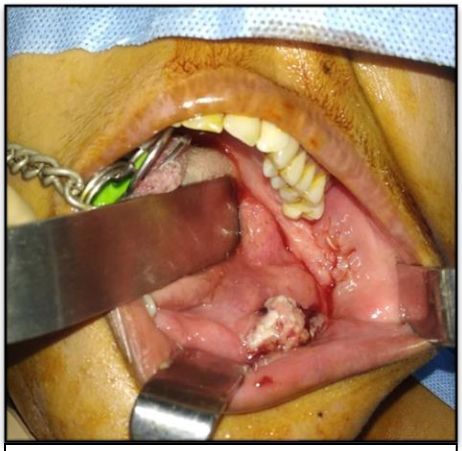

Fig.5 Intra operative photograph showing tumor

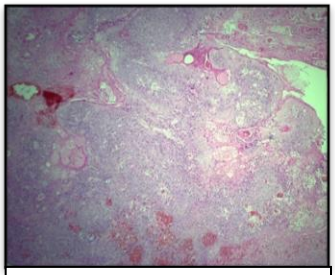

Fig.6 Histopathology of Resected mandible showing SCC

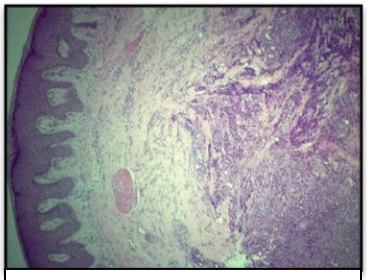

Fig.7 Histopathology of Skin nodule showing metastases

Discussion

Malignant changes in the epithelial lining of odontogenic cysts have been described previously [1]. Although the exact number of documented cases is difficult to determine, Gardner [5] reviewed all cases documented between 1889 and 1967 and considered 25 of them to be acceptable examples of malignant transformation of the epithelial lining of an odontogenic cyst. Gardner [5] proposed the following definitive criteria for identifying a lesion as PIOSCC derived from the odontogenic cyst:

(i) a microscopic area of transition from a benign cystic epithelial lining to SCC

(ii) an intact overlying oral mucosa

(iii) the absence of carcinoma in adjacent structures

(iv) the absence of metastatic carcinoma from a distant tumor [6].

In a review of 81 documented cases in the world literature, Woldron and Mustoe [6] considered the incidence of carcinoma arising from the odontogenic cyst to be approximately 1-2 per 1000 [7]. In a recent retrospective study of 116 cases of PIOSCC between 1938 and 2010, Bodner et al. in 2011 [3] found only 16 confirmed cases of PIOSCC arising from the OKC. It has been reported that keratin metaplasia followed by hyperplasia and dysplasia of the cyst epithelium are signature events in the development of SCC in the OKC [8]. Also, the presence of keratinization in the cyst lining suggests a greater risk of malignant change [9]. Furthermore, Gardner [5] and Tamgadge et al. [10] have suggested that longstanding chronic inflammation is a factor related to malignant transformation of benign epithelium.

In general, 50% of cases of PIOSCC show metastasis to the cervical lymph nodes. Therefore, radiation and chemotherapy may also be performed as adjunctive treatments along with the surgical excision. According to the literature, its prognosis is relatively poor, with a five-year survival rate ranging from 30% to 40%.

In our case, the patient had multiple recurrences of OKC, which may support the theory of PIOSCC arising from the chronic inflammatory response. Though she never had nodal disease, there was metastases at distant sites in spite of delivering adjuvant CRT, which is rare and points at the aggressive nature and poor prognosis of the disease.

Conclusion

PIOSCCs arising from OKCs are extremely rare, and fewer than 20 have been reported so far. Few reports have presented clear evidence of histological transition between squamous cell carcinoma and the odontogenic epithelial lining of the cyst wall. This highlights the importance of careful histopathological examination of apparently innocuous odontogenic cysts because of the possibility of carcinomatous change in their epithelial lining.

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