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Case Report

Primary Intraosseous Carcinoma of the Maxilla

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ABSTRACT

A primary intraosseous carcinoma (PIOC) is a squamous cell carcinoma arising within the jaw-bone and should be differentiated from a malignant ameloblastoma. It is not due to arising from an odontogenic cyst or tumor. The possibility of the lesion being a metastasis from another primary site should be considered, and excluded by a careful history and examination. The diagnosis of a PIOC is rare, but it is often worth considering in any differential diagnosis of jaw radiolucency. The prognosis associated with primary intraosseous carcinoma of the jaws is poor and needs for aggressive treatment. It is common for these patients to present with apparent routine dental.This paper reports a case PIOC of the maxilla. The patient was a 68-year-old woman with a chief complaint of swelling of her hard palate.

Keywords: Squamous Cell Carcinoma, Maxillae

Introduction

Primary intraosseous carcinoma is a rare tumor that has been infrequently reported (1). Some diagnostic criteria have been proposed to consider a lesion as PIOC: 1) absence of ulcer in the oral mucosa overlying the tumor, 2) absence of another primary tumor at the time of diagnosis and for at least 6 months during the follow-up and 3) histological evidence of squamous cell carcinoma (1). Waldron and Mustoe (2) proposed a classification for PIOC. According to this classification, PIOC may have different origins. Total absence of cystic component or other odontogenic tumor cells such as ameloblastoma are mandatory to diagnose PIOC type3 (de novo). In contrast, PIOC type 1 can be identified by the presence of odontogenic cyst. Similarly, PIOC type 2 can be distinguished by the presence of malignant ameloblastoma or ameloblastic carcinoma arising de novo. Thus, PIOC arising de novo should be differentiated from a malignant ameloblastoma, which is not due to arising from an odontogenic cyst or tumor (3). The possibility of the lesion being a metastasis from another

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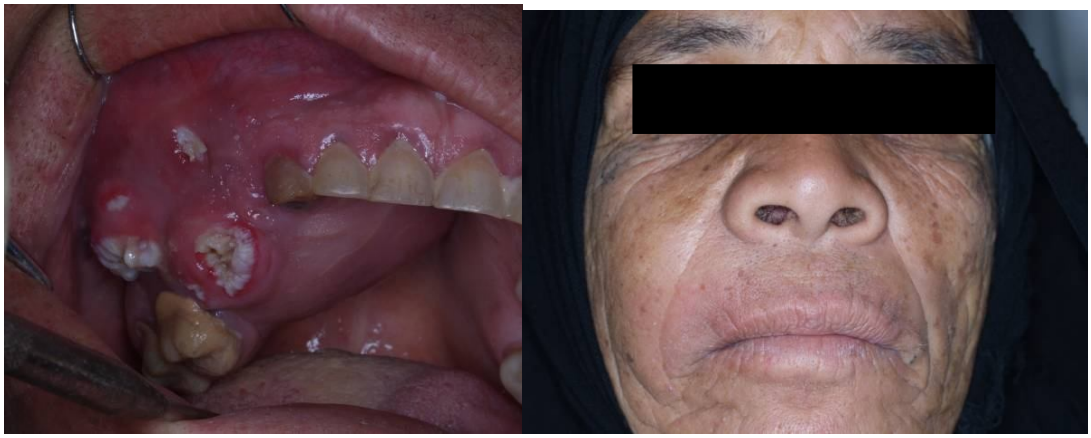
primary site should be considered, and excluded by a careful and complete examination include a chest radiography and the patient should be followed for at least 6 months (2, 3). According to some studies, the etiology PIOC is not clear (4).

Although several cases of malignant transformation of odontogenic cysts have been reported in the literatures (5, 6), but there are few reports of PIOC arising de novo (7-9). This paper reports a rare case of maxillary intra-osseous squamous cell carcinoma, which is probably of de-nova.

Case report

A 68-year-old Iranian woman was referred to the Department of Oral Medicine, Kerman Dental School, Kerman University of Medical Sciences, Iran by her dentist for evaluation of a painful swelling on the right side of the maxilla, which had been noticed 4 months previously. It had slowly increased in size. The patient did not have systemic disease. In addition, patient had a smoking habit of 2 to 4 cigarettes a week but no alcohol consumption.

The lesion expanded the medial and lateral corticalplates, but no ulceration of the overlying mucosa was demonstrated. On examination there was a tend firm, non-fluctuant mass measuring 6 cm by 6 cm at the right of the manxilla (Fig.1). In addition, swelling occurred at the side of the face and no associated lymphadenopathy. The over lining mucosa had a normal pinkish color (Fig. 1). The maxillary teeth had been extracted 5 years previously. Radiographs showed radiolucency with measuring 4.5 cm by 5.5 cm in the right side of the maxilla, with an ill defined and irregular border. Moreover, the tumor was found to have perforated the bottom maxillary sinus (Fig. 2). A diagnosis of primary intraosseous carcinoma with differential diagnosis of a maxillary sinus carcinoma, and cystic ameloblastoma was made. Uunder local anesthesia, simple enucleating of friable tissue was performed. Histological examination of the excised soft tissue showed features of a poorly differentiated squamous cell carcinoma with some cell degeneration, nests and cords of malignant epithelial cells with wide cytoplasm, round or oval nuclei, with prominent nucleoli and mitotic figures (Fig. 3). Oral mucosal epithelium covering the tumor was normal and any reason, which shows the malignancy and displasia, was not seen. In view of the diagnosis of squamous cell carcinoma, the following further investigations were carried out but all proved negative: chest radiograph, hematological and biochemical blood tests. The laboratory tests that requested for patient was WBC, RBC, Hb, HCT, MCV, MCH, MCHC, PLT, ESR, serum calcium, urine analysis, urine culture, liver function tests SGOT, SGPT, billirubin (direct, indirect), alp. Results were normal. Surgical ablation consisted of a hemi-manxillectomy in continuity with a radical neck dissection. Further histological examination of the main specimen confirmed the presence of an intraosseous carcinoma with no ameloblastomatous component. The patient has been under regular review for 6 months with no sign recurrent.



A **B**

Fig. 1-
A: Clinical examination reveals a mucosa membrane with a normal pinkish color and in some regions was keratotic and verrucos
B: The nasolabial fold was disappeared at the right of patient face

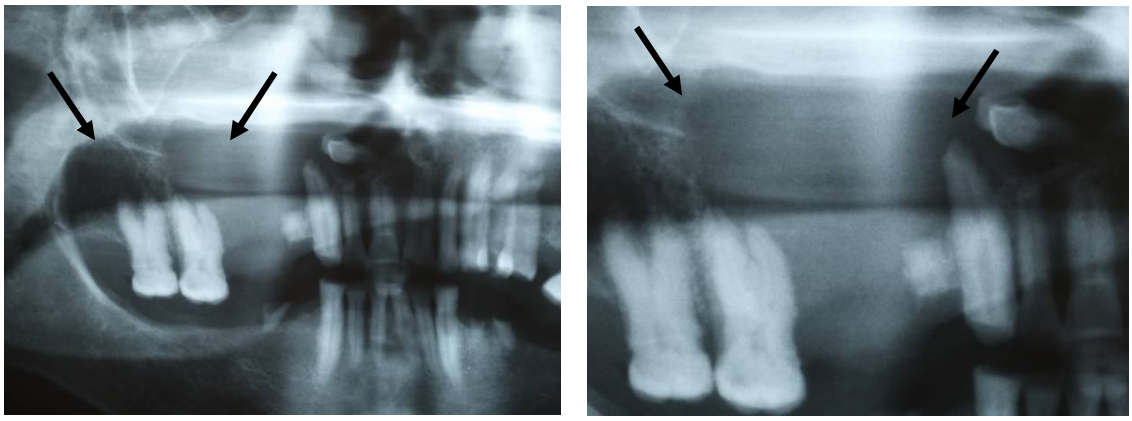


Fig. 2- Panoramic radiograph showing a radiolucent lesion in the right maxilla

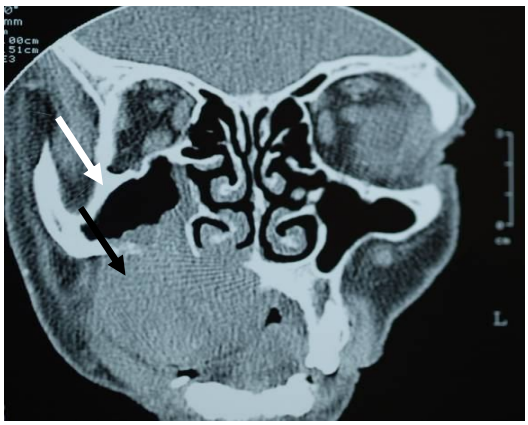


Fig. 3- CT Scan showing a lesion in the right maxilla and sinus maxillary

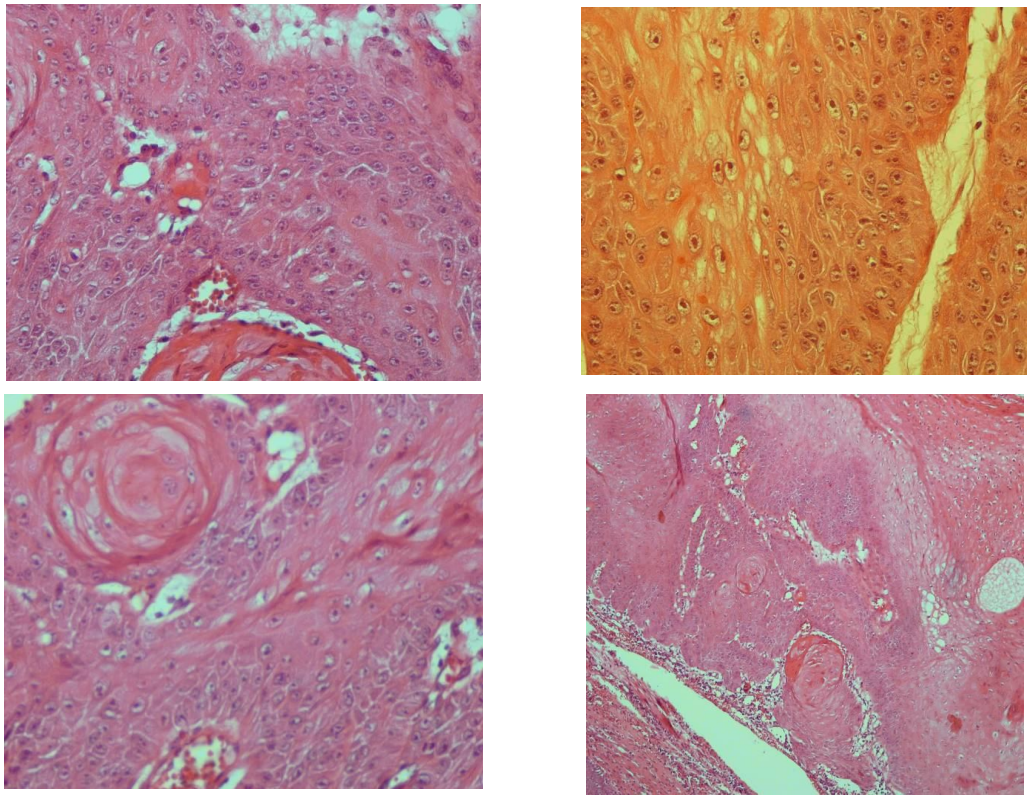


Fig. 4- A,B,C&D: Infiltrative atypical squamous cell nests and keratin pearls. (X100 H&E stain)

Discussion

PIOC is a rare neoplasia, which is very difficult to diagnose. It is necessary 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 (2). This tumor is most frequently located in the mandible and most patients had age 50 year or older. The male to female ratio reported from 2.80 to 3.2 in different studies (3). After an extensive search of the English language literature, only 12 cases of intra-osseous carcinoma of the maxilla were properly documented.

The most commonly associated signs and symptoms, pain and swelling, should be taken into consideration, as persistence may be a sign of malignant osseous tumor. The present case was a maxillary intraosseous squamous cell carcinoma that not originated from epithelial components of a preexisting cyst. It is suggested that maxillary intraosseous squamous cell carcinomas rarely arise from the epithelial. The absence of mucosal ulcers is one of the main characteristics for a diagnosis, which differentiates cases of PIOC from superficial squamous carcinoma arising in other parts of the oral cavity. The over lining mucosa in the present case had a normal pinkish color. The possibility of a metastatic lesion from a distant primary site has to be excluded. The most common primary sites likely to metastasis to the jaws are the breast, lung, and kidney. Therefore, a successful diagnosis of the primary disease was made by correlation of clinical findings with laboratory studies, diagnostic radiographs, and biopsies. Biopsy should be taken from lesion oral. Metastasis in the lymph nodes and hematogenous dissemination take time to occur even when the invasion of the bone and soft tissue is quite advanced. Local recurrence has been observed in some patients who have not been subjected to radical primary resection (10). Although a wide range of shapes and sizes are revealed, a ra-

diological study is one of the most effective ways of detecting tumors of this kind and PIOCs have vaguer, more irregular edges (10). PIOC must be identified from malignant ameloblastoma, ameloblastic carcinoma, and maxillary sinus carcinoma. Malignant ameloblastoma is a tumor that, in spite of a histologically typical or classic ameloblastoma (epithelial islands with peripheral, palisade, columnar basal cells, polarization of nuclei away from the basement membrane; and central areas of satellite reticulumlike cells), has given rise to pulmonary or lymph nodes metastasis while retaining the microscopic features of the primary tumor. The term ameloblastic carcinoma is characterized as a tumor that has some features of the ameloblastoma, but shows cytological features of malignancy in the primary or the recurrent tumor. It is a carcinoma arising within a pre-existing intraosseous or peripheral ameloblastoma and acts much more aggressively than ameloblastoma. Histologically, this tumor showed spindle shaped cells invading the normal bone. The tumors cells were arranged in an ameloblastic pattern with cellular atypia, marked nuclear pleomorphism, high mitotic rate, hyperchromatic nuclei, focal necrosis, perineural invasion, and increase nuclear/cytoplasmic ratio with or without evidence of metastatic disease (11-16).

Primary malignancies affecting the maxillary sinus include squamous-cell carcinoma, adenoid cystic carcinoma, and adenocarcinoma (17). The pathologic features of these tumors shows islands of basaloid epithelial cells that contain multiple cylindrical, cyst like space resemble Swiss cheese. The tumors cells are small and cuboidal, exhibiting deeply basophilic nuclei and little cytoplasm (18).

There is a useful feature for helping to differentiate between a tumor that originated in the maxillary sinus and one that has developed on the maxillary bone. If the maxillary sinus has been involved with a malignant tumor, the sinus walls are less well defined, and perhaps one or more are destroyed. The sinus itself shows density increasing and it is filled with tumor, entirely (19). In addition, PIOC must be considered in the presence of any symptom-free radiolucent lesion. This entity may be identified in a routine dental radiograph. It usually shows poorly defined margins and irregular pattern of bone destruction (20). Radiographs the present patients showed radiolucency an ill defined and irregular border. In addition, the tumor was found to have perforated the bottom maxillary sinus. The differential diagnosis of these radiological findings is metastatic tumors to the jaws, malignant minor salivary glands, osteogenic sarcoma, malignant ameloblastoma and ameloblastic carcinoma (19).

Prognosis is quite poor, with 5-year survival rates ranging from 30% to 40% (21, 22). Moreover in a recent , survival rates of 1, 2, and 3 years were 75.7%, 62.1%, and 37.8%, respectively (23). This has prompted some authors to speculate that it might be the delayed diagnosis that contributes to the poor prognosis (3). These findings advise the surgeon about the elevated aggressiveness of this tumor despite of adequate surgical treatment (3, 21, 22).

The study of Raul *et al.* showed that, 50% of the patients with PIOC presented with persistent symptoms following routine dental disorders(3). Also in previous study, it was showed that chief compliant patients are teeth removed, dental pain, periodontal disease (3). In the present case, the major sign and symptom were pain and swelling.

PIOC is a rare tumor that behaves like a locally advanced carcinoma of the oral cavity. Strict diagnostic criteria must be applied, with primary emphasis on the no connection of the tumor with the oral mucosa and on the histological features. The prognosis associated with PIOC of the jaws is poor and suggests the need for aggressive treatment.

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The authors declare that there is no conflict of interests.

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