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## Squamous cell carcinoma arising from an odontogenic keratocyst: A case report

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### Abstract

Squamous cell carcinoma (SCC) derived from keratocystic odontogenic tumor is an extremely rare tumor that is limited to the jaws. Most intraosseous carcinomas originate from the epithelial lining of odontogenic cysts, so they are called odontogenic carcinomas. They occur more frequently in men and the mean age of patients is 57 years.

The following report describes an extremely rare case of an odontogenic carcinoma derived from a keratocystic odontogenic tumor in a 20-year-old man. The patient presented with an exophytic lesion in the retromolar region of the mandible which was first noticed by the patient 25 days earlier. In panoramic radiograph a well-defined radiolucency around the impacted 3rd molar was observed. Clinical and radiographic diagnosis was odontogenic SCC. Surgical resection was performed and histopathologic examination of the lesion confirmed the diagnosis of SCC in the wall of a keratocystic odontogenic tumor.

**Key words:** *Odontogenic squamous cell carcinoma, primary intraosseous odontogenic carcinoma, odontogenic cyst, oral squamous cell carcinoma.*

### Introduction

Squamous cell carcinoma (SCC) arising from the wall of an odontogenic cyst (also called a primary intraosseous carcinoma) is a rare tumor which occurs only in jaw bones (1). Since the source of this lesion is epithelium involved in the odontogenesis, the World Health Organization (WHO) in 1972 suggested the term primary intraosseous carcinoma and classified the lesion as an odontogenic carcinoma (2,3). The most recent WHO classification of Odontogenic Tumors categorizes primary intraosseous carcinomas (PIOC) as: solid type carcinomas, carcinomas originating from keratocystic

odontogenic tumor (odontogenic keratocyst), and carcinomas arising from odontogenic cysts other than keratocystic odontogenic tumors (3).

PIOC may theoretically arise from the lining of an odontogenic cyst or de novo from presumed odontogenic cell rests (e.g. reduced enamel epithelium) (4,5).

Various odontogenic cysts have been associated with odontogenic SCC, including residual cyst, dentigerous cyst, calcifying odontogenic cyst, and lateral periodontal cyst. The most common associated cyst is a residual cyst, followed by dentigerous cyst (6).

According to the new histological classification of tu-

mors of the WHO, odontogenic keratocyst is nowadays considered a specific odontogenic tumor and the PIOC derived from it is considered as a specific entity which is different to other PIOC's derived from the odontogenic cysts (3).

In this article we report a rare case of SCC which originated from a keratocystic odontogenic tumor in a 20-year-old patient.

### Case Reports

A 20-year-old man was admitted to the Department of Oral Medicine in the Mashhad Dental Faculty with a chief complaint of a painful lesion and swelling in the right retro-molar region of the mandible which was first noticed by the patient 25 days earlier with gradual increase in size and occasional bleeding.

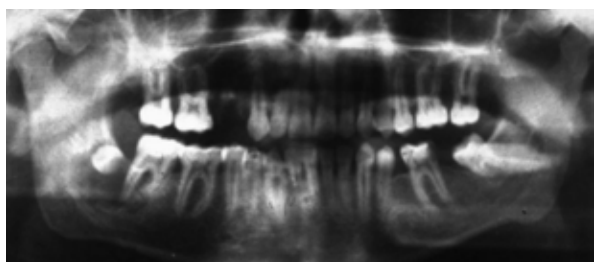
Intra-oral examination revealed a painful sessile exophytic lesion with a verrucous surface with an approximate size of 2× 3 cm (Fig.1).

It was firm on palpation and bleeding was noticed from the posterior gingival sulcus of the second molar of the same side. There were not associated anaesthesia or paraesthesia in the area. No palpable local or regional lymphadenopathy was observed and the patient was otherwise healthy.

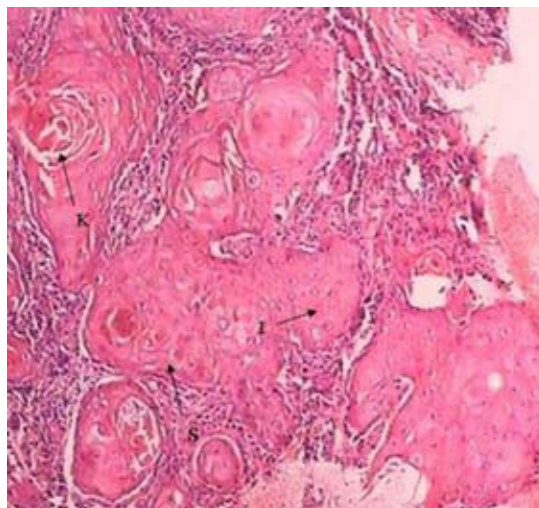
A well defined unilocular radiolucency around the impacted 3rd molar was seen on panoramic radiograph, and there was no complete radicular development (Fig.2).



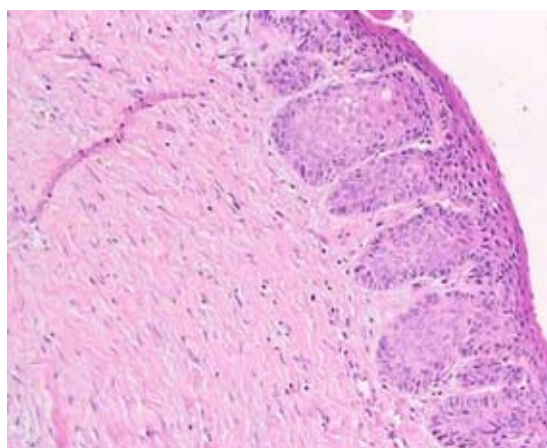
**Fig. 1.** Clinical intra-oral view: Exophytic lesion with verrucous surface.



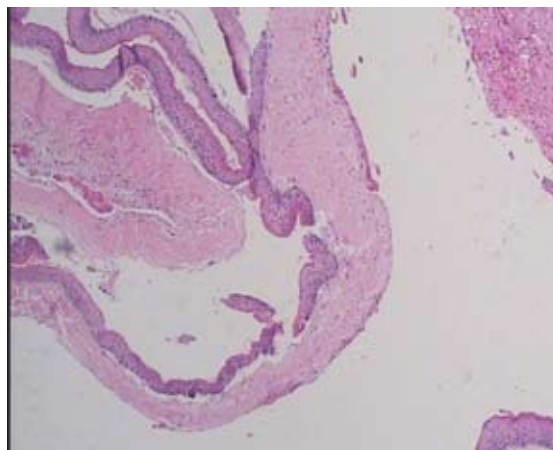
**Fig. 2.** Panoramic radiography: Well-defined unilocular radiolucency around unerupted 3rd molar is observed.



**Fig. 3.** Microscopic view of the lesion: Malignant squamous epithelial islands (S) with keratin pearl formation (K) and individual cell keratinization (I) [H&E, Original magnification×1000].



**Fig. 4.** Microscopic view of the central lesion: Dysplastic changes in epithelial lining of the cyst [H&E, Original magnification×1000].



**Fig. 5.** Another microscopic view of the central lesion: An Odontogenic Keratocyst. [H&E, Original magnification×400].

Considering clinical features, history and radiographic findings our diagnosis was squamous cell carcinoma in the wall of an odontogenic cyst. Incisional biopsy was performed and the specimen was sent to the Pathology Department.

Microscopic examination revealed a malignant neoplastic proliferation of stratified squamous epithelium forming sheets or islands of cells that invaded the connective tissue. Individual cell keratinization and keratin pearl formation was observed (Fig.3).

We searched for a probable primary tumor by careful clinical examination, chest radiograph and ultrasound examination of various organs and the possibility of metastasis was ruled out.

The patient was referred to an oncology center, where a radical surgery using the Commando approach was performed. Histopathologic findings of the surgical specimen reconfirmed our initial diagnosis of SCC in the wall of an OKC (Fig.4 and 5).

The patient was followed-up for 6 consecutive months. There was no sign of recurrence of the lesion in that period. Later the patient moved to another province and did not return for future control examination.

## Discussion

Odontogenic carcinomas are extremely rare tumors that were first described by Loos in 1913 as a "Central Epidermoid Carcinoma" of the jaw (7).

It is difficult to determine the total number of reported cases because of insufficient data about published cases and different classifications suggested for these tumors, although it appears to be about 82 cases reported in the world literature (6,8-12).

Similar studies conducted by Aboul-hosn Centenero et al. (8), Mosqueda-Taylor et al. (10), Scheer et al. (11) and others (6,13-16) revealed that odontogenic carcinoma is more common in men than women (2:1) and the mean age of the patients is 57 years (6,10,11,13,17), although one case in a 16-month-old girl (18) and another in a 5-year-old girl (19) have also been reported.

The mandible is affected more frequently than the maxilla (8,11,13) and the most common symptoms in these malignant tumors are pain and swelling. Paresthesia and numbness can also occur; however, the patient may be asymptomatic with the lesion being found on a routine panoramic radiograph (6,11,13,16).

Long-standing chronic inflammation has been suggested as a predisposing factor. Gardner (20) and Yu et al. (21) proposed that possibility of malignant changes in an odontogenic cyst is very low when there is not inflammation.

Van der Wal et al. (22) suggested that the presence of keratinization in the cyst lining is a risk factor for malignant changes and Browne et al. (23) indicated that odontogenic cysts with keratinization are more prone

for malignant changes than non-keratinizing types.

Anneroth et al. (24) believe that the most common factor may be an inflammatory reaction with or without a genetic background.

Ward and Cohen pointed to three possible mechanisms: 1) a pre-existing cyst becomes secondarily involved in a carcinoma of unrelated origin arising either from an adjacent epithelial structure or as a distant primary tumor; 2) the lesion is a carcinoma from the outset, a part of which has undergone cystic transformation; 3) the initial lesion is a cyst, and malignant changes have subsequently taken place in the epithelial lining (25).

Radiographically, a unilocular or multilocular lesion with an ill-defined or well-defined border is observed in odontogenic SCC (26,27).

Malignant changes in an odontogenic cyst should be considered if the radiolucent area has jagged or irregular margins with indentations and indistinct borders (6).

The histopathologic features are consistent with a diagnosis of squamous cell carcinoma. The majority of cases represent well differentiated keratinizing carcinoma (6,28).

PIOC must be considered in the differential diagnosis of malignant tumors of odontogenic epithelium, including ameloblastic carcinoma, intraosseous mucoepidermoid carcinoma, clear cell odontogenic carcinoma, and a malignant variant of CEOT. In addition, metastatic SCC must be ruled out (6,21).

Treatment of choice for these lesions is surgery and/or radiation therapy (29). In general, the prognosis is poor and metastasis to cervical lymph nodes is observed in up to 50% of cases (11,21). Two year survival rate of patients has been reported in 53% (29).

Although at the early stage of the lesion there is no connection between the cyst and overlying mucosa, but as the lesion progresses, it finally perforates the cortical bone and is present in the oral cavity.

In our case PIOC occurred in a 20-year-old patient, which is unusual for SCC because of the young age of the patient and the lesion perforated the cortex of the bone and appeared in the oral cavity, which has not been reported in the literature yet.

We considered that it was possible that two different co-existing lesions, an overlying mucosal squamous cell carcinoma and an odontogenic keratocystic, could have merged together with time, but SCC in a 20-year-old healthy person is not probable and also malignant histopathologic changes observed in the wall of the cyst confirmed diagnosis of odontogenic SCC.

In summary, we emphasize the importance of careful examination and regular follow up of patients with impacted teeth. Careful histopathological examination of apparently innocuous odontogenic cysts is also recommended because of the possibility of carcinomatous changes in their epithelial lining.

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