Primary Intraosseous Carcinoma of Mandible: 
A Case Report 

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Abstract 

A primary intraosseous carcinoma (PIOC) arising de novo in the mandible is presented. A 50-year-old patient came for help with persistent pain and swelling over left mandibular area for months. After systemic evaluation, ablation and reconstruction surgery was performed. The final diagnosis of PIOC was made. This is an uncommon lesion and is seen essentially in the jawbones. While the majority of the reported cases of PIOC arise from the odontogenic cysts, those arising de novo are infrequently illustrated. The early lesions are either asymptomatic or painful swelling. Thus delay of diagnosis or treatment is quite often. Like other malignancies, metastasis to regional lymph nodes or distant organ is observed in advanced PIOC. 

Key words: Primary intraosseous carcinoma, Mandible. 

Introduction 

Primary Intraosseous Carcinoma (PIOC) is a very rare lesion. It was first described by Loos in 1913 and named as intra-alveolar epidermoid carcinoma by Wills in 1948[1,2]. The term primary intraosseous carcinoma (PIOC) was suggested by Pinborg et al in 1972[3]. According to the World Health Organization (WHO) classification, PIOC is an odontogenic carcinoma defined as “a squamous cell carcinoma arising within the jaw, having no initial connection with the oral mucosa, and presumably developing from residues of the odontogenic epithelium” [4]. Up to two-thirds of PIOC is malignant transformation within odontogenic cysts while PIOC arising de novo is relatively rare[5,6]. To the best of our knowledge, there were only 35 cases reported in English literature[6]. Asserts to its rarity, we herein present a case of PIOC de novo in the mandible. 

Case report 

A 50-year-old male first came to the Department of Oral and Maxillofacial Surgery of Mackay Memorial Hospital, Taipei in December, 2002. At that time, he had been suffering from painful swelling of left face for four months. According to his statement, he went to a local
dental clinic due to toothache of left mandibular area six months ago. His left mandibular first molar was extracted. Pain and progressive swelling of the extraction wound persisted (Fig 1A). So the patient was referred to our outpatient clinic. At that time his pan oral mucosa was intact in appearance (Fig 1B). He also denied recent development of ulcer or other soft tissue lesion in his oral cavity. Bony expansion was noted over left mandibular molar region. Panoramic radiography demonstrated a radiolucent bony destruction of left mandibular body area (Fig. 2A). The patient, however, refused biopsy or operation at that time. In April 2003, he presented again with progressive symptoms and signs. Clinical examination revealed the swelling over his left face with mild tenderness. The range of mouth opening remained normal and there was no palpable cervical lymph node. A sinus tract was noted at the lingual aspect of left mandible alveolar ridge with purulent discharge. Radiological finding showed the enlargement of the lesion (Fig. 2B). Under the impression of left mandibular osteomyelitis or metastatic malignancy, he was admitted for further management.

His family and medical histories were non-contributory. Habits of smoking have remained for years with alcohol consumption socially. He denied betel nuts chewing. Hematology investigation including serological titers of tumor markers (SCC, CEA, AFP) and biochemical profile were within normal limits. Preoperative assessments of chest x-ray (Fig. 3A), EKG, whole abdomen sonogram were all normal. Computed tomography demonstrated bony destruction in the left body of mandible with enhancement of adjacent soft tissue (Fig. 4). The nasopharynx, oropharynx and hypopharynx appeared symmetric and normal. Slightly enlarged lymph nodes were seen in the submental, bilateral submandibular regions and left internal jugular chain at level II. Gallium and Tc99m bone scan showed increased radiotracer uptake in the left mandible, which indicated probable active inflammation or tumor growth. Culture from sinus tract discharge showed no specific pathogen.

After initial biopsy, histopathologic examination revealed a pattern of squamous cell carcinoma. As confirmed of its malignancy, the commando surgery was performed under general anesthesia. The procedure included left side functional neck dissection, wide excision of the tumor and hemimandibulectomy. Then the defect was reconstructed by bone plate and free fibular osteoseptocutaneous flap. The postoperative course was uneventful, and he was discharged two weeks after the surgery.

The tumor involved large area of mandible bone grossly, and was totally enclosed within the jawbone except for the edge around the fistula. Microscopically, the tumor showed a picture of squamous cell carcinoma mainly well to focal moderately differentiated (Fig. 5). Frequent keratin pearl formation was noted. The overlying gingiva was erosive. Submandibular gland and all of the lymph nodes were free of tumor cell invasion, which was confirmed by immunohistochemical staining with cytokeratin. Perineural invasion was found at the periphery of the tumor.

Because the possibility of metastatic tumor could be ruled out and the lesion showed no connection with oral mucosa initially, the final diagnosis of primary intraosseous carcinoma was made. With respect to the progressive behavior of this disease, the patient received postoperative radiotherapy and is under periodic follow-up. Chest x-ray showed no abnormalities five months after the surgery (Fig. 3B).
Figure 1A: Facial appearance showed mild swelling of the left cheek.

Figure 1B: The oral mucosa was intact around the lesion.
Figure 2A: Panoramic radiograph showed an ill-defined radiolucency of the left mandibular body.

Figure 2B: The lesion enlarged after 4 months.
Figure 3A: Preoperative chest x-ray showed no abnormal lesion.

Figure 3B: Chest x-ray showed no abnormalities five months after surgery.

Figure 4: Computed tomograph revealed a large destructive lesion of the left mandibular body.
Figure 5A: Histopathologic appearance of primary intraosseous carcinoma (H & E stain, original magnification x 40)

Figure 5B: The tumor lesion showed no direct connection with oral mucosa (H & E stain, original magnification x 100)
Discussion

Primary intraosseous carcinoma (PIOC) occurs only in the jaw bones and predominantly in the posterior mandible\(^7-13\). It affects men more than women with the ratio about 2.2:1\(^6\), and mostly occurs in the elder patients aged above 60\(^6,11\).

To establish the diagnosis of PIOC, the lesion must be distinguished from the tumors that metastasize to the jaw from distant sites, from gingival carcinomas that have invaded the bone from the surface, and from tumors that originated from maxillary sinus\(^14\). Careful investigations of the systemic condition and inspection of the intraoral condition are necessary to exclude the possibilities. Diagnostic aids such as chest radiograph, abdomen echo and bone scan can be helpful for general evaluation.

These tumors are thought to originate from residues of the odontogenic epithelium and known as odontogenic carcinomas\(^15\). There are two possible pathogenesis origins: either by malignant transformation of the epithelial lining of the odontogenic cysts or the remnants of odontogenic tissue\(^5,9,12,13,16,17\). In the latter situation, they are referred to as PIOC arising de novo. According to the classification system modified by Waldron and Mustoe, this entity was separated as PIOC Type-3\(^13\).

In an analysis proposed by Thomas et al in 2001, there were totally 35 cases of PIOC arising de novo reported in the literature since 1964\(^6\). The clinical features were nonspecific while pain (54.8%), swelling (51.6%) and sometimes sensory disturbances (16.1%) presented in most of the patients. Radiological findings of PIOC could not offer much hints for differential diagnosis with its great variation. However, it may be one of the most effective methods for early detection of the bony lesions. Computed tomography also acts as a useful evaluation tool to show the extent of the lesion within jawbone and soft tissue involvement\(^18\).

The histological differential diagnosis should include the following lesions: acanthomatous ameloblastoma, ameloblastic carcinoma, squamous odontogenic tumor, and mucoepidermoid carcinoma as well as benign or malignant salivary gland tumor with squamous metaplasia\(^10\). The final diagnosis of PIOC must match with the clinical features and the results of general assessments.

The disease is rare; therefore the prognosis is hard to estimate. Most likely it may be affected by the treatment modalities. The incidence of lymphadenopathy was reported as 10 out of the 29 cases (34.8%)\(^11\). However, To et al. in 1991 reported that the presence of lymphadenopathy did not worsen the prognosis\(^19\). There is still no consensus in the treatment modalities but surgical intervention is generally accepted (71.4%)\(^6,7,9,10,19-21\). Radiation and/or chemotherapy were also taken in some cases without conclusion of their effectiveness yet\(^11,19,20,22-24\).

In our case there was a delay in diagnosis and treatment for possibly more than ten months since toothache was noted. This not only complicated the management but also worsened the prognosis. It reminds us the importance of careful evaluation and close observation of which seems to be a common dental disorder. Furthermore, more cases should be reported to get further information concerning this rare type of neoplasm.

References

Primary Intraosseous Carcinoma of Mandible: A Case Report


下顎骨內原發性上皮癌：一病例報告

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摘   要

骨內原發性上皮癌為一罕見且僅發生於顎骨之惡性病症。根據文獻回顧，大部分乃齒源性囊腫之惡性轉變，而原發病例則較為少見。發生時的早期症狀為疼痛及腫脹，通常表現並不明顯且無特異性。因此常造成診斷及治療上的延遲。部分病例亦出現局部淋巴結或遠處之轉移而影響預後。一名五十歲男性因下顎區域持續疼痛腫脹至本院求診，經診斷為骨內原發性上皮癌，就此本文提出其病例報告與相關文獻之討論。

關鍵語：骨內原發性上皮癌，下顎骨。

Received: July 17, 2003
Accepted: November 3, 2003
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