Primary intraosseous carcinoma of the mandible

Eui-Hwan Hwang, Yong-Suk Choi, Sang-Rae Lee

Department of Oral and Maxillofacial Radiology, College of Dentistry, Kyung Hee University

ABSTRACT

A very uncommon tumor, primary intraosseous carcinoma (PIOC), is a carcinoma arising within the jaw. The definite diagnosis of PIOC is often difficult as the lesion must be distinguished from alveolar carcinoma that may invade the bone from the overlying soft tissues or from the tumors that have metastasized to the jaw from a distant site. A case of PIOC arising in the mandible is presented. The clinical, radiologic, and histologic features are described. This rare lesion should be considered in any differential diagnosis of a jaw radiolucency. (***Korean J Oral Maxillofac Radiol 2005; 35 : 235-9***)

KEY WORDS : Jaw Tumors; Intraosseous Carcinoma; Odontogenic Tumor; Mandible

Primary intraossous carcinoma (PIOC) is a very rare but well-recognized entity. 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.[1] Primary intraosseous squamous cell carcinoma include the two different entities of PIOC and malignant change in odontogenic cysts.[1-3] Until Elzay's reivew in 1982,[4] these odontogenic carcinomas were considered together. PIOC is now critically defined and clearly separate from a carcinoma arising from an odontogenic cyst.[1]

The tumor was probably first described by Loos in 1913 as a central epidermoid carcinoma.[5] Several other terms have also been used, though these have been somewhat misleading; they include intra-alveolar epidermoid carcinoma[6] and primary intra-alveolar epidermoid carcinoma.[7] Pindborg et al.[8] in 1971 were the first to use the term PIOC.

The definite diagnosis of PIOC is often difficult as the lesion must be distinguished from alveolar carcinoma that may invade the bone from the overlying soft tissues or from the tumors that have metastasized to the jaw from a distant site and from the primary tumors of maxillary sinus origin.[9] Since the most common symptom of PIOC is swelling and persistent pain in the mandible, the diagnosis is often difficult and an infectious etiology is most likely to be considered.[10] Furthermore, as the diagnostic criteria of PIOC are obscure, the cases, which have been reported as PIOC in the literature, are quite few.

The present report illustrates an PIOC arising in the posterior mandible in a 50-year-old woman, as well as to review the existing literature.

Case report

A 50-year-old female was referred to the Dental Hospital of Kyung Hee University from a local clinic for the extraction of partially impacted right third molar. This patient was feeling the discomfort to mastication and dull pain on right posterior mandible about 1 week earlier and complained of increasing numbness of right lower lip. Oral examination revealed an intact, normal-appearing overlying mucosa and no fluctuation to palpation without pus discharge. There was no associated cervical lymphadenopathy. Her medical history was noncontributory.

Conventional radiograph revealed unilocular radiolucency with poorly defined margin and infiltrative osteolysis in the right second and third molar region of the mandible (Fig. 1). And also, the cortical bones of buccal and inferior alveolar border were eroded by the lesion (Fig. 2). In intraoral radiograph, the alveolar lamina dura adjacent to the lesion was lost (Fig. 3). Computed tomography showed perforation of labial cortical plate of the mandible and a soft tissue mass with contrast enhancement extending through the labial cortical plate of the mandible into adjacent soft tissue (Fig. 4). Radiographic impression was concluded as metaststic malig-

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Correspondence to : Prof. Eui-Hwan Hwang
Department of Oral and Maxillofacial Radiology, College of Dentistry, Kyung Hee University 1 Hoigi-dong, Dongdaemun-gu, Seoul 130-701, Korea
Tel) 82-2-958-9405, Fax) 82-2-965-1256, E-mail) hehan@khu.ac.kr

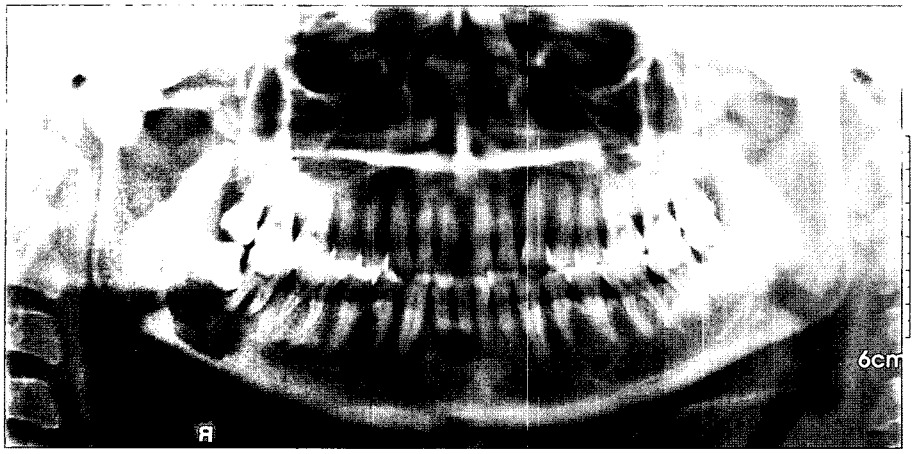

Fig. 1. Panoramic radiograph shows unilocular radiolucency with poorly defined margin and infiltrative osteolysis in the right second and third molar region of the mandible.

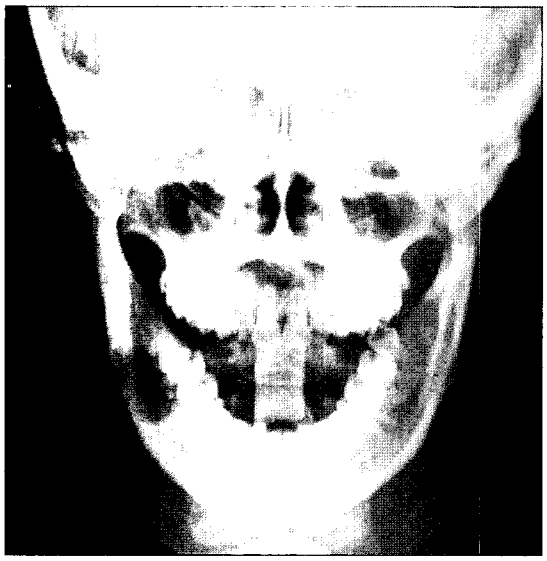

Fig. 2. Mandibular P-A radiograph shows the cortical bones of buccal and inferior alveolar border eroded by the lesion.

nant tumor or primary intraosseous malignant tumor. Preoperatively, a bone scan revealed a strong uptake at the peripheral site of the lesion (Fig. 5). A full medical examination did not reveal any evidence of a separate primary site and metastases.

An incisional biopsy was performed after extraction of right mandibular second and third molar. At the biopsy the tumor was solid and contained no fluid. Histopathologic feature was reported as squamous cell carcinoma (Fig. 6). Preoperative chemotherapy and partial mandiblectomy from the right premolar region to the right mandibular ramus with regional neck dissection was performed.

Histologically, the surgical specimen consisted of a well differentiated squamous cell carcinoma with no evidence of a cystic component. The tumor tissue consisted of irregular nests of abundant keratinization with a few dyskeratotic cells and foreign body ginat cell reaction. The tumor nests were superficially no contact with the normal-appearing overlying mucosa. Sections from right submandibular gland and regional lymphnodes revealed no tumor invasion.

Discussion

PIOC is a rare malignant neoplasm of the jaw. This tumor is believed to arise from the odontogenic epithelium and hence is also referred to as odontogenic carcinoma.[11] The reason why the odontogenic epithelial rests begin to proliferate is not properly understood. The most common factor may be a reactive inflammatory stimulus with or without a predisposing genetic cofactor, inducing neoplastic formation.[12]

The incidence of this lesion is very low. Review of all published cases of the English-language literature by To et al.[13] revealed only 21 well-documented cases, to which the authors proposed addition of three cases. Age, sex, and location predilection reported by Shear, McGowan, and Elzay indicate that the tumor occurs mainly in adults, in sixth to seventh decade, has a male to female ratio of 3 : 1, and is situated usually in the posterior mandible.[14] The case reported by Coonar,[15] which occurred in the anterior maxilla, was the exception.

The most common reported symptom is pain and swelling and sometimes sensory disturbance like paresthesia and numbness.[11] PIOC may also mimic routine dental disorders resulting in delayed diagnosis.[3] To et al.[13] cited a delay in correct diagnosis ranging from a few weeks to as long as 18 months. Obviously this delay can contribute to a poor prognosis.

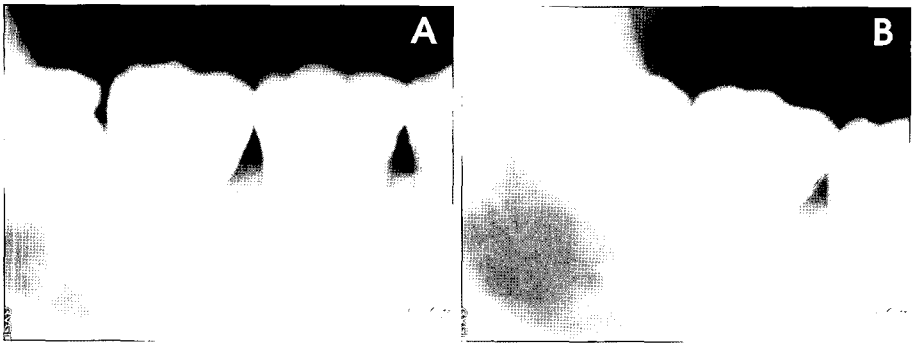

Fig. 3. Intraoral radiographs show loss of the alveolar lamina dura adjacent to the lesion.

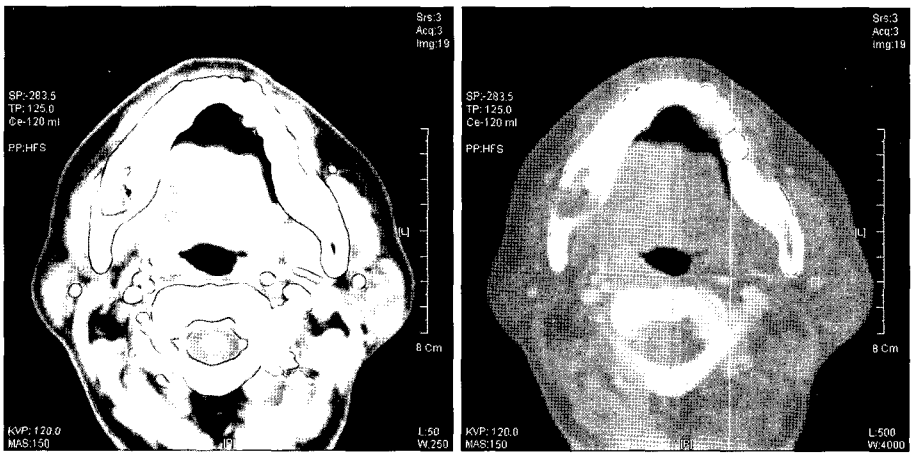

Fig. 4. Contrast-enhanced axial CT scans show the perforation of labial cortical plate of the mandible and the contrast-enhanced soft tissue mass extending through the labial cortical plate of the mandible into adjacent soft tissue.

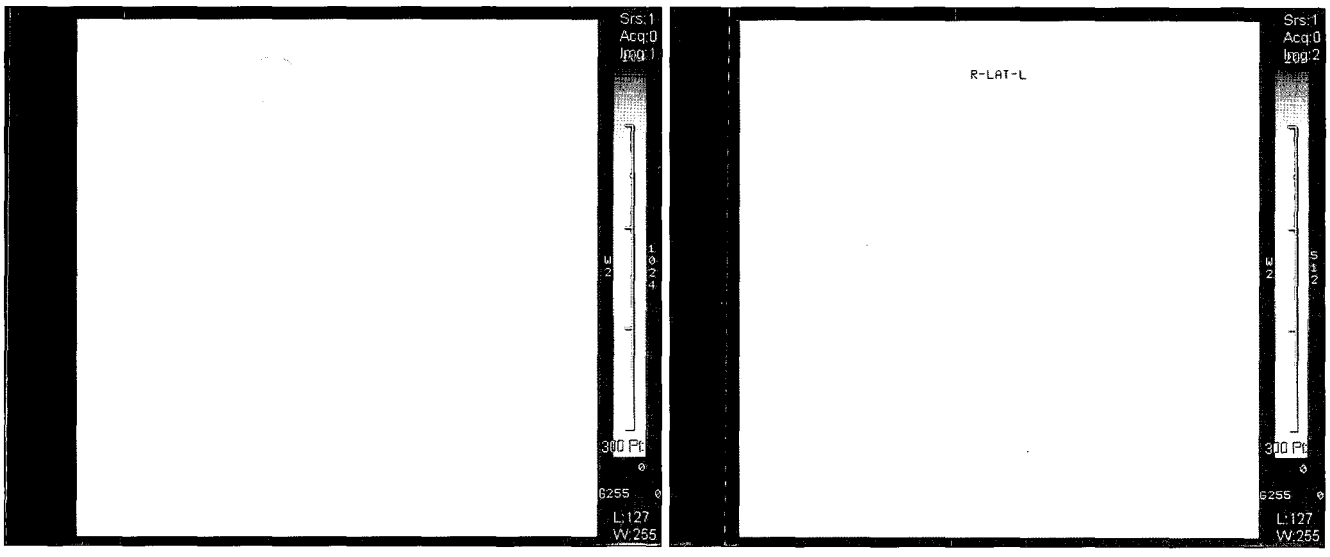

Fig. 5. RI scintigraph using ^{99m}Tc-MDP shows a strong uptake at the peripheral site of the lesion and otherwise no active bony lesion.

The diagnosis of PIOC is difficult, partly because the initial symptoms are often thought to be of dental origin.[16] In several cases, teeth have been extracted as the initial treatment.[7,17] However, an important indication of a more serious disease is the involvement of the inferior alveolar nerve with reduced sensation in the skin. This disturbance is, in fact, often found in association with PIOC.[16-18] In our case, the first symptoms were a pain of the posterior mandible and numbness of the

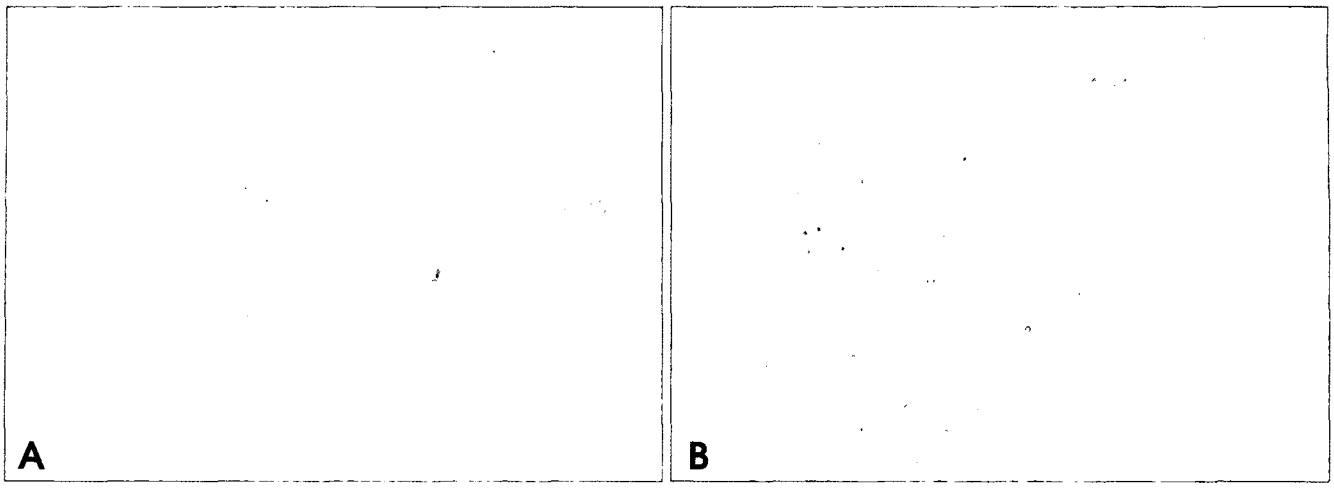

Fig. 6. Photomicrographs of the lesion show marked hyperplasia of well differentiated squamous cells and solid tumor tissue, consisted of irregular proliferation of atypical squamous cells (H&E stain, A. ×40, B. ×100).

lower right lip.

Radiographic examination is one of the most effective means of detecting PIOC. However, PIOC shows great variation in size and shape, and in the appearance of their border.[9] Nolan[19] reported that PIOCs that grew slowly usually had a well-defined, rather smoothly contoured border, while those that grew more rapidly showed a poorly defined, ragged border. Because the margins of PIOCs show great variation, they are usually indistinguishable from other benign or malignant tumors.[9] Our case had an ill-defined, unilocuar radiolucency around the partially impacted third molar area. and also, CT images revealed extensive bone destruction within endosteum and relatively small cortical bone perforation.

The following strict criteria for diagnosing a lesion as PIOC have been proposed: 1) an intact oral mucosa prior to diagnosis; 2) no histological evidence of associated odontogenic cyst; and 3) ruling out a metastatic deposit from a distant primary.[4,9,13,20,21] Since most PIOCs have been referred for further evaluation of an impaired wound or obvious tumor growth following extraction, there seems insufficient proof to accept such cases as true PIOC that have no initial connection with the overlying mucosa.[22] In this patient, the possibility of a metastasis was rule out, since there was no history of a malignant disease and clinical and radiological examinations did not reveal anything pathological elsewhere in body. On the other hand, the mucosa in the oral cavity and the skin of the face were normal. A continuous growth from a carcinoma of the surface epithelium can thus ruled out. Although the definitive diagnosis is always difficult,[9,13,21] our present case indeed fulfils these criteria.

The reported histologic appearances of PIOC is not pathognomonic. Several features indicative of odontogenic origin, such as peripheral palisading, plexiform or alveolar growth, central stellate reticulum-like areas and stromal hyalinization around the epithelial islands, while not specific, are frequently present and are helpful in reaching a diagnosis.[1,4,9,22] Thus, histologic features should be assessed together with supporting clinical and radiographic information in approaching a definite diagnosis.[1] The diagnosis must be made after it is confirmed that there is no evidence of lining epithelium of odontogenic cysts or other odontogenic tumor cell, which requires the careful examination of serial section of the specimen.[4] In this case, the carcinoma showed an intimate relationship with the impacted tooth but multiple histological sections did not show it to be continuous with the reduced enalmel epithelium surrounding the crown. There was no indication of a pre-existing odontogenic cyst in this patient. Furthermore, clinically and histologically there was no evidence that the lesion had any connection with the oral mucosa.

The prognosis of PIOC is quite poor and importance should be given to early diagnosis so that suitable treatment can be given at the earliest.[11] In the 12 cases of PIOC reported by Elzay,[4] a 40% 2-year survival was noted. 66% of these patients had regional metastasis. Similarly, in the review of 28 cases of PIOC reported by To et al.[13] 46% of the patients survived for a period varying 6 months to 5 years.

The importance of this case is that it illustrates the central origin of the tumor, that it present as a dental problem and that it is a relatively rapidly growing tumor.[14] The diagnosis of a PIOC is rare, but it is often worth considering in any differen-

tial diagnosis of a jaw radiolucency. We would add that any radiolucency should either be biosied at an early stage or followed closely with regular radiographs.[13] The prognosis of PIOC is quite poor and importance should be given to early diagnosis so that suitable treatment can be given at the earliest.[11]

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