



Correspondence

Primary intraosseous carcinoma arising in orthokeratinized odontogenic cyst

**KEYWORDS**

Primary intraosseous carcinoma;
Orthokeratinized odontogenic cyst;
Malignant transformation

Primary intraosseous carcinoma, NOS (PIOC) is a rare central jaw carcinoma with male predilection. The mean age of diagnosis of PIOC is 55–60 years. PIOC occurs most frequently in the posterior mandibular body and ramus. When presenting in the maxilla, these tumors have a predilection for the anterior segment. For cases arising in cysts, radicular/residual cysts are the most common precursors, following by dentigerous cysts and odontogenic keratocyst (OKC). Clinically, most cases of PIOC were identified as incidental radiographic findings; the advanced cases might cause swelling, pain, ulceration, loosening of teeth, pathological fracture and neural sign. Radiographically, the tumors show ill-defined osteolytic lesion, often with cortical perforation. Radical resection is the treatment modality, and neck dissection might be performed for clinically suspicious lymph node. The 5-year survival rate of cases arising in cysts is at 40%.¹

A 76-year-old male patient suffered from lower lip paresthesia with pain and swelling over his left lower cheek for 2 months, whilst the oral mucosa and skin were intact (Fig. 1A and B). Reviewing the history of the patient, he had injury of head 10 years ago, when he was sent to our emergency department and the radiographs of skull were taken. It showed a well-defined oval shaped pericoronal radiolucent lesion of horizontally impacted tooth 38, and

the lower border of left mandible was intact (Fig. 1C). The most recent panoramic radiograph revealed a poorly defined osteolytic lesion with cortical perforation over the left mandibular body (Fig. 1D). A large amount of pus discharge was seen after the incision and drainage performed at left submandibular area. The clinical diagnosis was an odontogenic cyst with secondary infection and enucleation under general anesthesia was arranged. During surgery, extensive proliferation of the lesion to the adjacent soft tissue of mandible was noted. The specimen from enucleation revealed a cystic lesion lined by orthokeratinized stratified squamous epithelium with prominent proliferation (Fig. 1E–G). Down growing epithelium invading to the fibrous wall was noted, and the tumor cells were arranged in sheets and islands (Fig. 1H and I). Immunohistochemically, the cystic epithelial lining and tumor cells were positive for CK19 (Fig. 1J). The final diagnosis was a PIOC arising in orthokeratinized odontogenic cyst (OOC). Then, the second surgery for radical resection with flap reconstruction was subsequently performed. No recurrent lesion was found in post-operative 6-month follow-up.

The diagnosis of PIOC should fulfill these criteria: first, no ulcerative lesion on the overlying oral mucosa; second, excluding the possibility of other odontogenic carcinoma and central salivary gland malignancy; third, rule out a

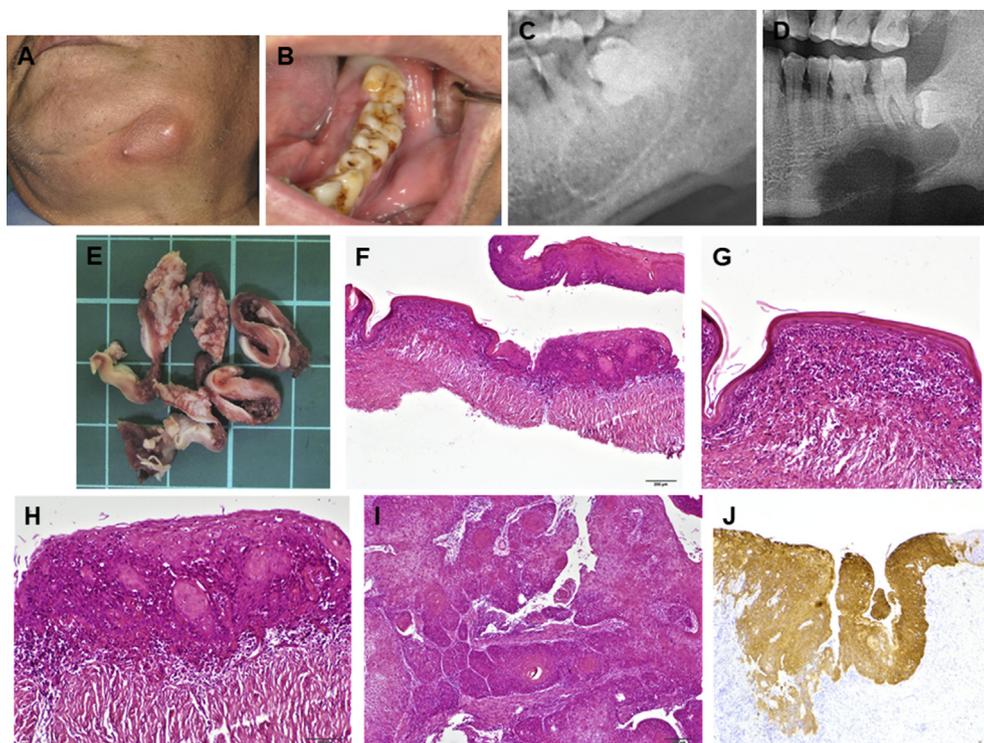


Figure 1 Clinical, radiographic and microscopic pictures of the current case of primary intraosseous carcinoma arising in orthokeratinized odontogenic cyst. (A and B) Swelling and redness over his left submandibular area, and the oral mucosa and skin were intact. (C) Cropped lateral view of skull radiograph displayed a well-defined oval-shaped pericoronal radiolucent lesion of horizontally impacted tooth 38; the lower border of the left mandible was intact (D) Current panoramic radiograph showed a poorly-defined osteolytic lesion with cortical perforation over the left mandibular body. (E) Gross examination of the specimen from enucleation showed a cystic lesion with focal verrucoid proliferation. (F) Cystic lesion lined by orthokeratinized stratified squamous epithelium with prominent proliferation (H&E; magnification, 40 \times). (G) The lining epithelium showed orthokeratinized stratified squamous epithelium (H&E; magnification, 100 \times). (H) Proliferation of the lining epithelium with tumor nests invading to subjacent fibrous wall (H&E; magnification, 100 \times). (I) The tumor cells were arranged in sheets and islands infiltrating in the fibrous stroma (H&E; magnification, 40 \times). (J) The cystic lining epithelium and tumor cells were focally positive for CK19 (magnification, 40 \times).

metastatic cancer.² Reviewing English literature, to the best of our knowledge, there are only 3 cases of PIOC arising in OOC to date.^{3–5} OOC accounts for about 1% of all odontogenic cyst, and it is considered as less aggressive than OKC with less than 2% in recurrent rate. Although malignant transformation of odontogenic cyst is rare, the pathologists should carefully examine the differentiation pattern of cystic epithelial lining. When dysplasia or carcinoma in situ is identified incidentally after excision, close follow-up is strongly recommended.

Declaration of Competing Interest

The authors have no conflicts of interest relevant to this article.

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Chih-Huang Tseng

Division of Oral Pathology & Maxillofacial Radiology,
Kaohsiung Medical University Hospital, Kaohsiung, Taiwan

Wen-Chen Wang

Division of Oral Pathology & Maxillofacial Radiology,
Kaohsiung Medical University Hospital, Kaohsiung, Taiwan
School of Dentistry, College of Dental Medicine, Kaohsiung
Medical University, Kaohsiung, Taiwan

*Oral & Maxillofacial Imaging Center, College of Dental
Medicine, Kaohsiung Medical University, Kaohsiung, Taiwan*

Chung-Ho Chen
*Division of Oral & Maxillofacial Surgery, Kaohsiung Medical
University Hospital, Kaohsiung, Taiwan*

Yuk-Kwan Chen*
*Division of Oral Pathology & Maxillofacial Radiology,
Kaohsiung Medical University Hospital, Kaohsiung, Taiwan
School of Dentistry, College of Dental Medicine, Kaohsiung
Medical University, Kaohsiung, Taiwan*

*Oral & Maxillofacial Imaging Center, College of Dental
Medicine, Kaohsiung Medical University, Kaohsiung, Taiwan*

*Corresponding author. 100 Shih-Chuan 1st School of
Dentistry, College of Dental Medicine, Kaohsiung Medical
University, Kaohsiung, Taiwan. Fax: +886 7 3210637.
E-mail address: k0285@ms22.hinet.net (Y.-K. Chen)

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