A 50-year-old female patient reported with a chief complaint of dull continuous pain, numbness, loosening of teeth and swelling in lower left back region of the jaw since two months. General physical examination revealed sign of anemia with pale conjunctiva, and with noncontributory medical history. There was no history of trauma.

Extra-oral examination revealed gross facial asymmetry with a GLIIXVH¿UPDQGWHQGHUVZHOOLQJRIîFPLQYROYLQJWKHOHIW posterior lower jaw (Figure 1a). Left submandibular and cervical %DVHGRQWKHFOLQLFDODQGUDGLRORJLFDO¿QGLQJVDGLIIHUHQWLDOGL sections of the specimen were taken to exclude the presence of odontogenic cystic lining epithelium or any other odontogenic tumor. To rule out metastatic tumor to the jaw, chest X-ray, whole-body CT scan and bone scintigraphy were preformed, but were noncontributory.

Based on the clinical and radiological findings, a differential diagnosis of odontogenic pathology and osteomyelitis was made. Microscopic examination revealed numerous malignant squamous epithelial tumor islands (keratinized-type) of different sizes infiltrating the fibrous connective tissue stoma (Figure 3). Serial sections of the specimen were taken to exclude the presence of odontogenic cystic lining epithelium or any other odontogenic tumor. To rule out metastatic tumor to the jaw, chest X-ray, whole-body CT scan and bone scintigraphy were preformed, but were noncontributory.

**What is your diagnosis?**

See the next page for diagnosis.
Primary intraosseous carcinoma (PIOC) was first described by Loos in 1913, and the term was suggested by the WHO in 1972. Later, Waldron and Mustoe’s classification of PIOC is widely accepted and frequently cited.

- Type 1: PIOC arising from odontogenic cyst
- Type 2
  - Type 2A: Malignant ameloblastomas
  - Type 2B: Ameloblastic carcinoma arising de novo from ameloblastoma or from odontogenic cyst
- Type 3
  - Type 3A: PIOC arising de novo: keratinizing
  - Type 3B: PIOC arising de novo: nonkeratinizing
- Type 4: Intraosseous (central) mucoepidermoid carcinoma

POIC arising de novo has been infrequently reported. It arises principally within the jaw bone, with posterior mandible being the most common site. It is usually noticed during the sixth to seventh decade, with a mean age of 50 years, having male predominance.1

The diagnostic criteria proposed for PIOC are as follows1,3:

- Absence of initial connection with the overlying mucosa or skin
- Microscopic evidence of squamous cell carcinoma without a cystic component or other odontogenic tumor cells
- Absence of another primary tumor on chest radiographs obtained at the time of diagnosis and during a follow-up period of more than 6 months

Various types of metastatic carcinoma (thyroid, kidney, prostate, lungs, etc.) to the jawbone should be ruled out by carefully reviewing the history and comprehensive systemic evaluation of the patient. PIOC usually exhibits marked variation in radiography and is worth considering as a differential diagnosis of jaw radiolucency. Confirmation of PIOC is only through histopathology.2

The most accepted treatment for PIOC is radical surgery with adequate resection. Radiotherapy and chemotherapy are used as palliative therapy/adjuvant therapy in cases of nerve infiltration. Metastasis to cervical lymph nodes has been described more frequently in de novo PIOC, therefore prophylactic neck dissection is recommended.1

Lin, et al., reported a 2-year survival rate of 53% for the type-1 entity, whereas Elazy demonstrated a 2-year survival rate of 40% for the de novo lesion. These findings indicate that PIOC’s originating from odontogenic cysts have a better prognosis than the de novo lesions.2

The present case of PIOC is reported in a 50-year-old female patient causing pathological fracture of mandible, and meets all the above mentioned diagnostic criteria of de novo PIOC.

References