CASE REPORT

Solid-type primary intraosseous squamous-cell carcinoma in the mandible: Report of a rare case

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Abstract
Primary intraosseous squamous cell carcinoma (PIOSCC) is a rare malignant neoplasm that has an exquisitely exclusive affection to the jawbone. It is defined as squamous cell carcinoma arising within the jaw and developing from residual odontogenic epithelium or from a preexisting odontogenic cyst or tumor. The solid-type of this tumor is a central jaw carcinoma arising de novo and has no initial connection with the oral mucosa. Herein, we report a case of solid-type PIOSCC involving the mandible in a 37-year-old male patient elucidating its histopathological and imaging findings. The patient underwent surgical resection followed by post-operative adjuvant radiotherapy. The close 2-year follow up of the patient revealed neither locoregional nor distant metastasis.

Introduction

Primary intraosseous squamous-cell carcinoma (PIOSCC) is a rare jawbone malignant neoplasm uniquely exclusive to the mandible and the maxilla. It was first designated by Loos [1] in 1913 as central epidermoid carcinoma. Willis [2], in 1948,
used the term *intra-alveolar epidermoid carcinoma* for this lesion. Later, in 1971, Pindborg et al. [3] regarded it as a primary intraosseous carcinoma. It was classified as an odontogenic carcinoma and defined as “a squamous cell carcinoma that arises within the jaw, with no initial connection with the oral mucosa and presumably develop from residues of the odontogenic epithelium” [3]. In the latest World Health Organization classification, PIOSSC is subdivided according to its potential origin into three types: solid-type carcinoma, carcinoma arising from a keratocystic odontogenic tumor, and carcinoma arising from an odontogenic cyst [4]. The diagnosis of PIOSSC is often challenging, as other lesions need to be excluded, such as neoplasms that metastasize to the jaws, gingival carcinomas that have invaded the bone from the oral mucosa, and neoplasms that originate from the maxillary sinus.

The solid-type PIOSSC is a central jaw carcinoma arising de novo and has no initial connection with the oral mucosa. It has been hypothesized that this lesion originates from odontogenic epithelial rests, including rests of Malassez, rests of Serres, and the reduced enamel epithelium surrounding a tooth prior to eruption [5]. The solid-type PIOSSC has been occasionally reported in the literature [6], as it is the rarest type among PIOSSCs [7]. This report describes a solid-type PIOSSC as a rare lesion arising from the posterior area of the mandible, and reviews the literature.

**Case report**

A 37-year-old male patient presented to the dental clinic at the College of Dentistry, King Saud University, Riyadh, Saudi Arabia, with a 3-month history of a left-cheek swelling that has recently been associated with pain and impairment of the sensation of his lower lip. Prior to his visit to our clinic, the patient had been seen by a dentist elsewhere who thought that the jaw swelling was due to infection, and was managed with a tooth extraction without improvement. The patient’s past medical history was otherwise unremarkable. His intraoral examination showed a firm, nonfluctuant swelling in the left retromolar area extending to the ramus of the left mandible. No mucosal lesion or abnormalities were present. The Orthopantomograph panoramic radiograph revealed an ill-defined irregular radiolucency at the left mandibular ramus (Figure 1). The differential diagnoses of the lesion included osteomyelitis, osteosarcoma, intraosseous carcinoma, and ameloblastic carcinoma of the mandible. Incisional biopsy from the ramus was performed, which demonstrated features of squamous-cell carcinoma. Based on the clinical, radiographic, and histological correlations, the diagnosis of intraosseous squamous-cell carcinoma was established.

The patient was referred to a tertiary-care center that specializes in head and neck cancer treatment, King Faisal Specialist Hospital & Research Centre in Riyadh, for further management. His head and neck computed-tomography scan confirmed the presence of an irregular, destructive soft-tissue mass lesion centered at the ramus of the left mandible, which extended from the distal aspect of the second molar to the posterior border of the ramus and from the sigmoid notch to the angle of the mandible (Figure 2). The computed-tomography scan of the chest, abdomen, and pelvis showed no distant metastasis. There was a positive uptake in the mass lesion and bilateral upper jugulodiagnostic lymphadenopathy on positron-emission-tomography scan. The patient underwent tracheostomy, total radical parotidectomy, left hemimandibulectomy, zygomatic-arch excision, and manipulating fixation and wide local excision of tumor in the left infratemporal and pterygoid fossae. The histopathological assessment of the surgical specimen confirmed that the tumor was an intraosseous squamous-cell carcinoma, well to moderately differentiated (Figure 3). The islands of the tumor cells extended into the bone (Figure 4), and perineural invasion was noted. The tumor size was 5.5 cm in the maximum dimension. The dissected surgical specimen of the neck showed reactive lymph nodes with no evidence of malignancy. Postoperative adjuvant radiotherapy was administered, and the close follow-up of the patient for nearly 2 years by the multidisciplinary team revealed neither locoregional nor distant metastasis. The patient is presently scheduled for facial reanimation.

**Discussion**

Solid-type PIOSSC is a rare malignant disease that arises from remnants of the odontogenic apparatus in the jaw [5]. The diagnosis of such lesion is challenging, as precise clinical and pathological details are important in eliminating an oral

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**Figure 1** Panoramic radiograph showing a radiolucent area with ill-defined margins in the left mandibular ramus.
mucosal origin. Additionally, the histological sections should be thoroughly examined to exclude the presence of any pre-existing odontogenic lesions. Furthermore, a careful review of the history and radiographic investigations is mandatory.

Figure 2  Computed-tomography images: (A) axial computed-tomography slice; (B) coronal computed-tomography slice. An ill-defined osteolytic lesion is observed at the center of the left mandibular ramus.

Figure 3  Photomicrograph of the lesion shows well to moderately differentiated squamous cells with cellular atypia and pleomorphism. Keratin-pearl formation is seen (hematoxylin-and-eosin stain).

Figure 4  Microscopic examination showing infiltrative squamous-cell nests invading the bone (hematoxylin-and-eosin stain).
in excluding metastatic lesion from a distant site. Sui et al. [8] proposed widely used criteria for diagnosing this type of PIOSCC: (a) an intact overlying oral mucosa preceding diagnosis, except due to tooth extraction or trauma; (b) squamous-cell carcinoma with no histological evidence of cystic components or other odontogenic tumor cells; and (c) no metastatic deposit from a distant primary at the time of diagnosis and throughout a follow-up period of more than 6 months. The present case fulfilled these criteria, and hence, was classified as solid-type PIOSCC.

It is difficult to determine the real incidence of the solid-type PIOSCC, owing to the paucity of well-documented cases; however, it is the rarest type among PIOSCCs [7]. Lugakingira et al. [6] reviewed the literature between 1996 and 2010, and found 32 cases were of the solid-type PIOSCC, 29 of mandibular, and four of maxillary types. The most commonly affected site is the posterior mandible. It has been reported that males were more commonly affected than females [6]. Our patient had the expected findings of male gender and involvement of the posterior mandible. The age of affected patients ranged from 24 years to 76 years, where most of the cases occurred in patients older than 50 years [6]. The patient who is the subject of this report was 37 years old at the time of diagnosis, which is not a common finding. Few reports have described solid-type PIOSCC in patients younger than 40 years [9–11].

The lesion at early stages is usually asymptomatic; however, the most common presenting features of advanced cases may include pain, swelling, numbness, and trismus [6,8,11,12]. Our patient experienced a persistent pain and paresthesia of the lower lip. He reported that he had a tooth extraction at a private clinic that suspected his initial symptoms to be due to a dental infection rather than a tumor. It is worth mentioning that, because odontogenic infections occur more frequently, few clinicians consider other entities; therefore, misdiagnosis and wrong treatment may occur. It has been reported that interossous malignancy of the maxillofacial region can mimic dental infection, and hence, gets misdiagnosed [13,14]. Yamada et al. [15] described cases of PIOSCC where they were initially diagnosed as dental-infection-related diseases.

The radiographic and histological investigations are essential in diagnosing solid-type PIOSCC. Radiographically, the lesion can show numerous destructive effects on the bone. It usually exhibits an osteolytic appearance with ill-defined and noncorticated borders [16,17], and our case followed this pattern. The microscopic features of the solid-type PIOSCC are not specific, as all neoplasms that originate from squamous epithelium need to be considered [8]. It is characterized by islands of epithelial malignant neoplasm showing the features of squamous-cell carcinoma [4]. The specimen should be thoroughly examined to confirm that there is no evidence of odontogenic cysts or other odontogenic tumor components [8].

The mainstay of treatment for oral squamous-cell carcinoma is usually surgery. External beam radiotherapy with or without chemotherapy is generally employed in three situations: (a) adjuvant to primary surgery to enhance locoregional control for cases with unfavorable pathological features, (b) primary treatment for cases unable to tolerate or unsuited for surgery, and (c) salvage treatment in the persistent or recurrent disease setting [18,19]. The treatment of choice for PIOSCC, in particular, is radical surgical resection [20]. The prognosis is generally poor, supporting the need for an initial aggressive surgical intervention to decrease the local recurrence rate [21]. The overall survival rates at 1 year, 2 years, and 3 years have been reported as 75.7%, 62.1%, and 37.8%, respectively [11]. Although organ preservation has become a prevalent treatment approach for managing locoregionally advanced head and neck cancer of the hypopharynx, the oropharynx, and the larynx, this approach has not been widely applied to patients with oral-cavity squamous-cell carcinoma. There are no clinical-trial data comparing extensive primary surgical approaches to organ preservation because of the paucity of PIOSCC cases. All reports on PIOSCC strongly recommend aggressive surgical intervention [6,22–24]. The need for adjuvant chemotherapy and/or radiotherapy is controversial [12,25]. However, several studies recommend postoperative radiotherapy, which significantly improves the rates of local and regional control and disease-free survival [20,26]. Zwetyenga et al. [21] concluded that surgery and postoperative radiation therapy might have the best results.

In conclusion, we presented a case of solid-type PIOSCC affecting the mandible, which may contribute to the expanding database of this rare neoplasm. Although PIOSCC is rare, it is important to be considered in the differential diagnoses of any jaw radiolucency.

Conflicts of interest

The authors have no conflict of interest to declare.

References


