Case Report

Primary intraosseous odontogenic squamous cell carcinoma of the mandible

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Abstract

Primary intraosseous odontogenic squamous cell carcinoma (PIOSCC) is a rare tumor. The incidence is low, and approximately 200 cases are reported in literature. The etiology is associated with the malignant degeneration of embryological remains. Differential diagnosis includes alveolar carcinomas, jaw metastases from other locations, odontogenic tumors, and tumors of the maxillary sinus. However, the diagnosis could be delayed due to the absence of symptoms in early stages. Surgery represents the first choice treatment. Whereas, postoperative radiotherapy could be helpful to improve the overall survival. The prognosis is generally poor. In this report, we describe the case of a 77-year-old man accidentally diagnosed PIOSCC. The patient referred a tooth extraction and cystectomy 3 years before. The pathological examination of the cyst tissue evidenced an inflammatory cyst. However, no patient follow-up was performed. Hence, is important to stress that routine monitoring of patients affected by large inflammatory cysts of the jaw bones should be mandatory.

Keywords: Diagnostic delay, extirpative surgery, postoperative radiotherapy, primary intraosseous squamous cell carcinoma

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INTRODUCTION

Primary intraosseous odontogenic squamous cell carcinoma (PIOSCC) is a rare and malignant tumor. The incidence is low and approximately 200 cases are reported in literature.[1] It affects the jaws, regardless of the interest of oral mucosa.[2] The etiology seems to be related to the malignant degeneration of embryological remains. In this line, epithelial rests of Malassez, dental lamina and epithelium of the dental follicle represent potential suspects.[3]

According to the 2005, the World Health Organization Classification of tumors, three subtypes of PIOSCC exist:[4]

1. Solid tumor that invades the marrow spaces and provokes osseous absorption
2. Squamous cell carcinoma (SCC) originated from the lining of an odontogenic cyst. Specifically, this subgroup includes two variants: keratocystic odontogenic tumor and odontogenic cysts
3. SCC associated with other benign epithelial odontogenic tumors.

The diagnostic criteria of PIOSCC included undamaged oral mucosa and absence of distant metastasis during the follow-up. Moreover, the switch from the epithelium

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of the cyst to SCC is essential for a proper diagnosis.[5] Differential diagnosis includes alveolar carcinomas, jaw metastases from other locations, odontogenic tumors and tumors of the maxillary sinus. In this sense, diagnosis is often delayed by the absence of symptoms in early disease.[6] Fortunately, neck metastases from PIOSCC are uncommon. Two-thirds of the cases do not present ganglion involvement at presentation. However, patients show a poor prognosis.[7] In fact, due to the rarity of this disease, there are no established treatment protocols. The adequate surgical resection remains the gold standard for the treating this tumor.[8]

In this report, we describe the case of a 77-year-old man accidentally diagnosed PIOSCC. The patient referred a tooth extraction and cystectomy 3 years before. The pathological examination of the cyst tissue evidenced an inflammatory cyst. However, no patient follow-up was performed. Hence, this case underlines the importance of routine monitoring of patients presenting large inflammatory cyst. Although rarely this lesions might experience a neoplastic transformation.

CASE REPORT

The article describes a case of PIOSCC of the mandible affecting a 77-year-old man. More in detail, the patient was referred at maxillofacial outpatient department of Granada University Hospital by his general practitioner. In fact, the patient reported a progressive painless swelling in the left side of mandible. Thus, a careful anamnesis and oral inspection were carried out. Oral and extraoral examination evidenced an abnormal mandibular ballooning in the left side. In view of that, an orthopantomography and a computed tomography (CT) scan of the cervicofacial area were performed for allowing the proper diagnosis. Panoramic radiography evidenced the presence of a large cyst in the left molar region and the lack of several tooth. CT scan evidenced an aggressive lesion invading body and ascendant branch of the jaw [Figures 1-3]. After analyzing this data, anamnesis and physical examination were repeated. Patient referred a tooth extraction of tooth 38, 3 years ago because of the presence of a pericoronal cyst. The histopathological analysis of the cyst tissue revealed an inflammatory cyst. However, no patient follow-up was carried out. Bearing this in mind, we carried out an incisional biopsy of the lesion. The result showed a poorly differentiated PIOSCC. After due consideration of all data, we staged cancer as cT3N0M0. In the oncological committee of our hospital, we decided to treat the case with a wide surgical resection, neck dissection and postoperative radiotherapy. Postoperative examination confirmed the diagnosis of PIOSCC poorly differentiated [Figure 4]

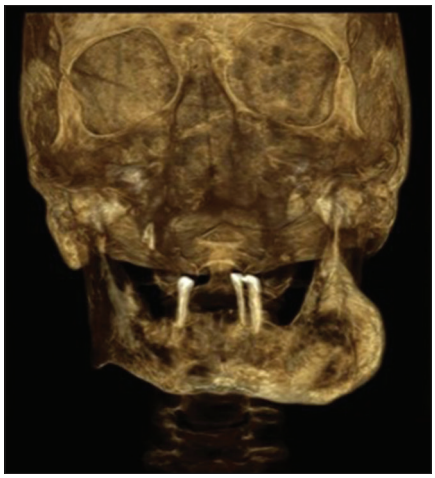

Figure 1: Three-dimensional computed tomography image showing an intraosseous odontogenic squamous cell carcinoma of the mandible. At the physical examination, the patient presented a bone consistency with swelling of vestibular region in the left side of the jaw

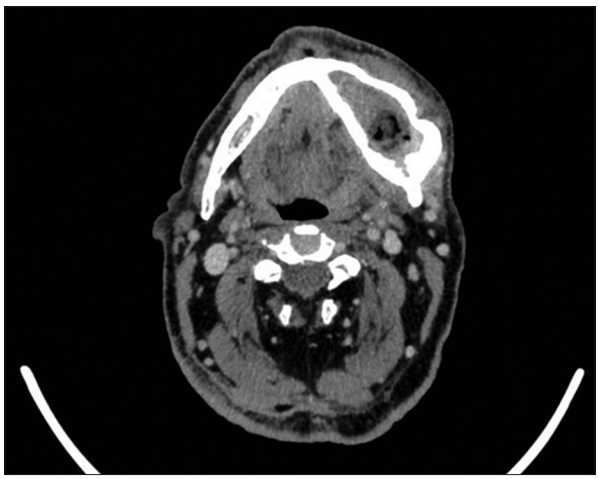

Figure 2: Axial computed tomography image of mandibular intraosseous odontogenic squamous cell carcinoma. The lesion affects the body and the ascendant branch of the mandible in the left side

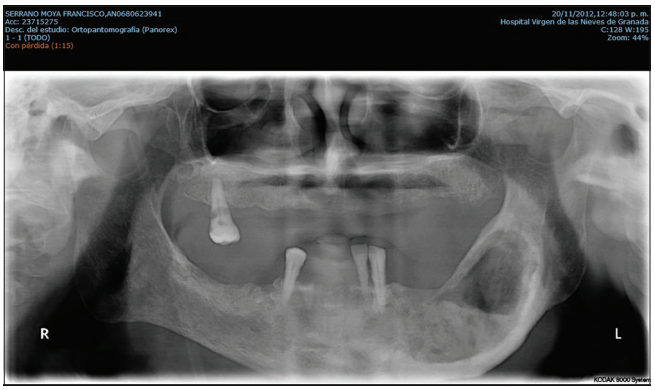

Figure 3: Orthopantomogram of mandibular intraosseous odontogenic squamous cell carcinoma

with resection margins free. The patient has no clinical signs of recurrence 27 months after surgery.

DISCUSSION

The incidence of PIOSCC is rare. Only approximately 200 cases have been reported to date. The clinical behavior is variable. However, the majority (50%) are well-grade malignant tumors. The median age of presentation is between the sixth and eight decade of life and it occurs more often in males.[5]

According to the literature, the time diagnosis is often delayed due to the absence of symptoms provoked by pathology. In this line, a review of 30 cases evidenced that PIOSCC showed the following forms of presentation: asymptomatic (26,7%), asymptomatic mass (26.7%), mass (16.7%), pain (13.3%), nerve involvement (10%) and expansion (6.7%). Moreover, PIOSCC might be confused with other inflammatory pathologies.[9] In our case, a patient referred a tooth extraction and cystectomy 3 years before the diagnosis. The pathological examination of cyst evidenced an inflammatory cyst. Unfortunately, any follow-up was performed. Thus, it is possible that the tumor could have been confused with a pericoronal cyst.

Panoramic X-ray and CT scan are essentials for a proper diagnosis and analysis of the tumor extension. In addition, an incisional biopsy is needed to complete the diagnosis.

A number of other lesions should be considered in the differential diagnosis such as alveolar carcinomas, jaw metastases, odontogenic tumors and maxillary sinus tumors.[6]

Wide surgical resection is currently the gold standard for treating PIOSCC. Moreover, several reports suggest that postoperative radiotherapy could be helpful to improve the overall survival. The role of postoperative chemotherapy is questionable. Survival rate of 3 years is described to be 37.8%.[10]

We treat the case with surgical resection, neck dissection and postoperative radiotherapy. Considering the wide surgical margins and lack of evidence about the effectiveness of chemotherapy, we decided to treat the patient only with surgery and postoperative radiotherapy. No signs of recurrence or metastases were evidenced 27 months after surgery. Despite this a close follow-up is mandatory.

Resuming, this case underlines the importance of routine follow-up of patients presenting large inflammatory cyst. Although rarely, this lesions might experience a neoplastic transformation. This practice allows early diagnosis. This is essential for increasing the overall survival of these patients.

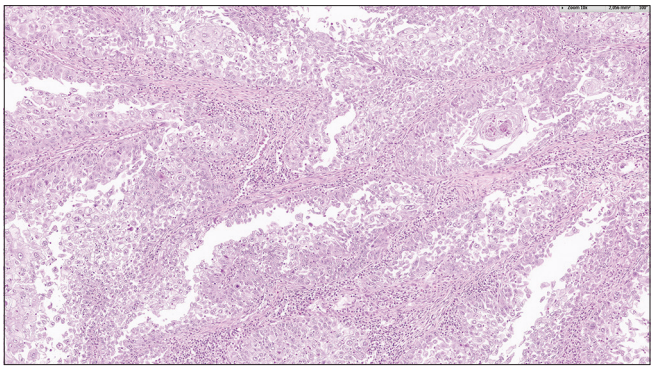

Figure 4: Histopathological image of mandibular intraosseous odontogenic squamous cell carcinoma with H&E stain

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Conflicts of interest
There are no conflicts of interest.

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