Dental Research Journal

Case Report

Central unilocular mucoepidermoid carcinoma of the mandible: A case report and literature review

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Received: April 2018
Accepted: May 2018

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ABSTRACT

Mucoepidermoid carcinoma (MEC) is the most common salivary gland malignancy. Central MEC (CMEC) is a rare tumor which affects women more than men and is more common in the mandible. Most cases are histologically classified as low-grade tumor and radiographically appear as well-defined unilocular or multilocular radiolucent lesion, although this tumor causes destruction and metastasis to other organs. In this article, we present a rare case of CMEC in a 47-year-old woman with unilocular radiolucent lesion of the mandible. Early and accurate diagnostic approach in all lesions with clinical/radiographical bland appearance is important, and all possibilities such as a malignant lesion should be considered.

Key Words: Mandible, mucoepidermoid carcinoma, salivary gland

INTRODUCTION

Mucoepidermoid carcinoma (MEC) of the jaw is the most common malignant salivary gland neoplasm which comprises 2.8%–15% of all salivary gland tumors.[1,2]

This malignant tumor composed a mixture of the epithelial, mucin-producing, and intermediate cells. this malignancy may occur in all ages, but its prevalence increases in the fourth and fifth decades of life.[1]

Primary central MEC (CMEC) is a rare intraosseous tumor with a widely discussed pathogenesis. Various possible origins have been considered, as follows:

a. Entrapment of the retromolar mucous glands within the mandibular jaw, which shows neoplastic transformation thereafter
b. Embryonic remnants of the submandibular and sublingual glands trapped within the mandible during development
c. Neoplastic transformation and invasion from the lining of the maxillary sinus
d. Neoplastic transformation of the mucous-secreting cells from the epithelial lining of the dentigerous cyst associated with impacted third molars
e. Neoplastic transformation of entrapped minor salivary glands within the maxilla.[3]

The first case of CMECs was reported in the mandible of a 66-year-old woman by Lepp in 1939.[4]

From that time, the criteria for confirming the diagnosis of CMEC were determined and the cases were published in the literature.[5-9]

Access this article online

Website: www.drj.ir
www.drjjournal.net
www.ncbi.nlm.nih.gov/pmc/journals/1480

How to cite this article: Aghakouchakzadeh A, Kargahi N, Shahnaseri S, Shakeri S. Central unilocular mucoepidermoid carcinoma of the mandible: A case report and literature review. Dent Res J 2019;16:127-30.

This case report presents CMEC with unilocular radiographic feature. Based on our collected data, <10 similar studies from 2010 were published in English.

CASE REPORT

A 47-year-old woman referred to the Department of Oral and Maxillofacial Surgery at Dental School of Isfahan University of Medical Sciences, exhibiting a lesion in the right posterior region of the mandible, which had been present for the previous 4 months. There were lower lip paresthesia and pain in the right ear [Figure 1a].

In oral examination, a fairly firm mass in the edentulous area on the right posterior region of mandible was seen. The lesion was not wounded with dark or black color and was covered with intact oral mucosa [Figure 1b].

The teeth were not mobile, and on medical history, she is a well-known hypothyroidism patient.

In the patient's radiographic examination in the panoramic view, an oral and maxillofacial radiologist reported an ill-defined unilocular radiolucent lesion in the right posterior mandible, and the radiologic report from computed tomography scan showed an ill-defined lytic lesion in the right posterior mandible. There were no bone fractures, and there were some lymph nodes with maximum diameter of 5.5 mm in right submandibular space [Figure 2].

With respect to ominous criteria such as lip parasites and ill-defined radiolucency, the malignant odontogenic and nonodontogenic tumors were suggested as differential diagnosis.

Incisional biopsy was performed, and microscopic examination revealed MEC.

The oromaxillofacial surgeon removed the soft tissue from the retromolar area and bone tissue from the posterior lingual area of the mandible with 1 cm safe margin around the tumoral area. Two pieces of tissue were sent to the Oral Pathology Laboratory. In the gross examination, the diameters of the brown-gray soft tissue sample were 30 mm × 25 mm × 7 mm, and the diameters of other bony pieces from the posterior part of the lower jaw were 40 mm × 12 mm × 12 mm [Figure 3].

In microscopic examination, the neoplastic proliferation of mucinous, intermediate, epidermoid, and clear cells was observed in the form of cellular sheets and cystic spaces. Some areas were affected by dysplastic changes, including hyperchromatism and pleomorphism nuclei, especially in the epidermoid and intermediate cells. The ultimate diagnosis was reported to be CMEC [Figure 4a and b].

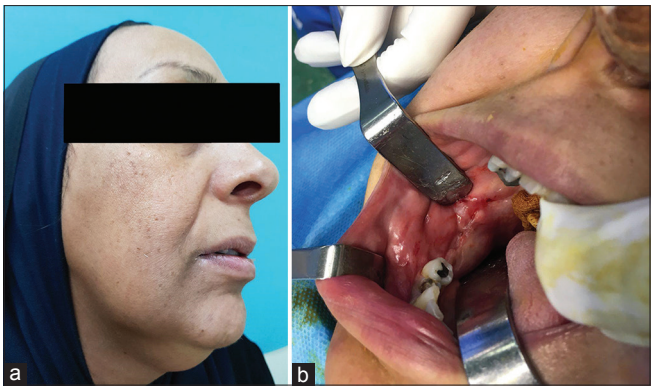

Figure 1: (a) Extraoral examination showed swelling in the right posterior region of the mandible, (b) intraoral examination revealed that the lesion was covered with intact oral mucosa.

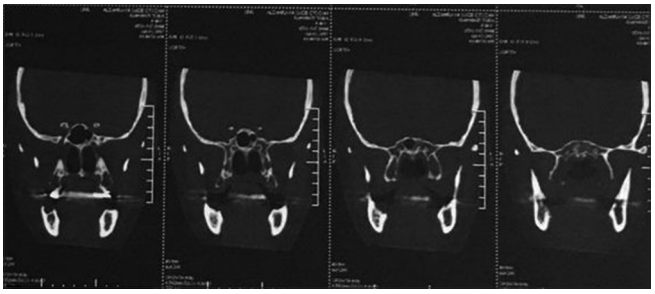

Figure 2: Computed tomography scan showed an ill-defined unilocular radiolucent lesion in the right posterior region of the mandible.

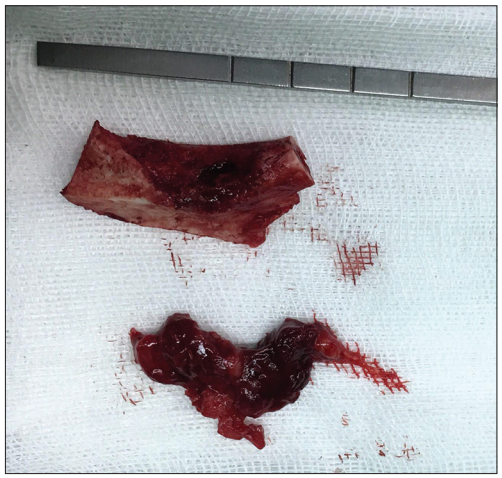

Figure 3: Two pieces of tissue were sent to the Oral Pathology Laboratory, the diameters of the soft tissue sample were 30 mm × 25 mm × 7 mm, and the diameters of other bony pieces from the posterior part of the lower jaw were 40 mm × 12 mm × 12 mm.

After that, the patient underwent adjuvant radiotherapy and chemotherapy. She received 64 Gy of radiation in 33 fractions and ERBITUX (cetuximab) at the same time. Six months after radiotherapy and chemotherapy, there was no evidence of tumor [Figure 5a and b].

DISCUSSION

CMEC affects women twice more than men, mostly occurring in the fourth and fifth decades of life, and involves the mandible (the molar angle region) twice as much as the maxilla.[4] The main symptoms are swelling and pain, the others such as trismus, paresthesia, and tooth mobility being noted occasionally.[6] However, these symptoms are not always necessarily present.[7]

The following criteria for diagnosis of CMEC were introduced:

1. Absence of any primary lesion in the salivary glands
2. Absence of any odontogenic tumors
3. Radiographic evidence of bone destruction
4. Retention of cortical plate integrity
5. Positive mucin staining
6. Microscopic confirmation of diagnosis.[3]

Radiographic features usually appear as unilocular or multilocular radiolucent lesions with sclerotic and well-defined margins. These same characteristics are found in some cystic lesions and tumors with odontogenic origin such as ameloblastoma and keratocystic odontogenic tumor.[7,8] However, it should not exclude less common, but more serious conditions as metastatic tumors, malignant osseous tumors, primary intraosseous carcinoma, and malignant salivary gland tumors.[9]

The present case showed a unilocular radiographic lesion, and for a better comparison, a literature review was conducted in the PubMed database to survey the published case reports of unilocular CMEC after 2010, as described in Table 1.

On microscopic examination, CMEC revealed an infiltrative neoplastic lesion characterized by proliferation of nests, islands, and cystic structures that

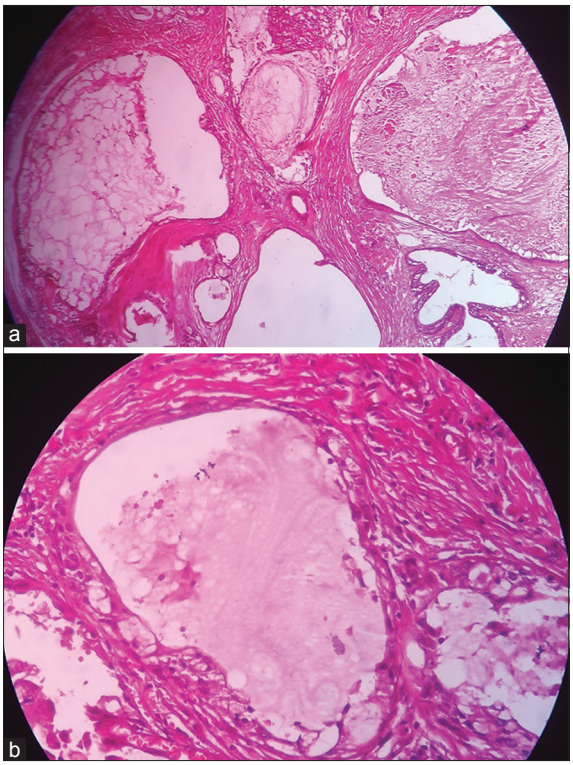

Figure 4: In microscopic examination, the neoplastic proliferation of mucinous, intermediate, epidermoid, and clear cells was observed in the form of cellular sheets and cystic spaces. (a) H and E, ×100, (b) H and E, × 400.

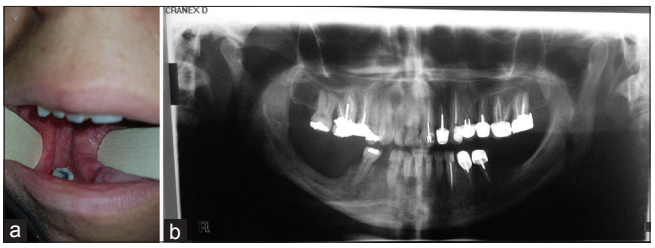

Figure 5: Six-month follow-up after radiotherapy and chemotherapy, there was no evidence of tumor. (a) Intraoral view, (b) radiographical view.

Table 1: Worldwide distribution of unilocular central mucoepidermoid carcinoma from 2010 to date

Authors	Age	Gender	Symptoms	Anatomic site	Radiographic findings	Diagnosis hypotheses
Aggarwal and Saxena, 2011[10]	8	Female	Pain	Mandible	Unilocular radiolucent	Dental granuloma, keratocystic odontogenic tumor, ameloblastoma
Chiu *et al.*, 2012[11]	55	Male	Pain	Mandible	Unilocular radiolucent	Cystic lesion
Rathore *et al.*, 2014[12]	18	Male	Pain	Maxilla	Unilocular radiolucent	Cystic lesion
Nallamilli *et al.*, 2015[13]	36	Male	Pain	Maxilla	Unilocular radiolucent	Odontogenic tumor
Sruthi *et al.*, 2017[9]	42	Male	Pain	Mandible	Unilocular radiolucent	Cystic lesion
Present case (2017)	47	Female	Pain	Mandible	Unilocular radiolucent	Cystic lesion

are composed of epidermoid, mucous, intermediate, clear, and occasionally oncocytic cells.[3,7]

It often demonstrated prominent cystic growth and was divided into low-, intermediate-, and high-grade types on the basis of morphological and cytological features. CMEC represented 2%–4% of mucoepidermoid carcinomas.[14]

The treatment of CMEC included surgical removed of the tumor and postoperation radiotherapy. More conservative surgical approaches such as curettage, enucleation, and marginal resection of the jaw, with or without postoperative radiation, led to the relapse of lesion in 40% of cases, while the recurrence was seen in 4% of the cases in segmented resection of the mandible.[15]

Gradation of the tumor was one of the most important predicting factors for the survival of patients suffering from CMEC. High grade was characterized by larger bone destruction, enlarged frequency of relapses, and presence of regional and distant metastases.[16]

CONCLUSION

CMEC is rare in the jaws; however, since the lesion's behavior is different clinically and radiographically, it is important that the clinician should be aware of the various presentations of a particular disease process and try to achieve the diagnosis accurate and fast. As we observed in this case, a unilocular lesion could be a major concern, and all such lesions should be carefully examined. Surgery such as *en bloc* resection is the treatment of choice with adjuvant radiotherapy in high-grade cases.

Financial support and sponsorship
Nil.

Conflicts of interest
The authors of this manuscript declare that they have no conflicts of interest, real or perceived, financial or nonfinancial in this article.

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