Case Report

Central Mucoepidermoid Carcinoma of Mandible. A Case Report and Literature Review

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ABSTRACT

Central mucoepidermoid carcinoma (CMC) is a rare lesion seen in the maxillofacial region. It is frequently seen in the mandible, especially the posterior than in the anterior region. Since the pathogenesis is not well explored in the past and the variations in the clinical presentation, the diagnosis of this lesion can be challenging. We report an extremely unusual case of a CMC in the premolar region of the mandible of a 47-year-old male patient. The clinical, radiological presentation and the surgical management of the tumor along with the reconstructive techniques utilized to restore the mandible. Although CMC is an unusual tumor of the jaws, it can be locally aggressive, but if treated with wide surgical excision it has a good prognosis. The patient was under follow-up and had not shown any signs of recurrence 2 years after surgery.

Key words: *Mucoepidermoid Carcinoma, Mandible, Oral Cavity, Salivary Gland, Intra-Osseous.*

The most common type of salivary gland malignancy, a mucoepidermoid carcinoma, represents between 2.8%–15% of all cancers of the salivary glands [1]. The condition usually occurs in the parotid and minor salivary glands, rarely intraosseous [1]. Since Lepp published the first central mucoepidermoid carcinoma in the mandible in 1939, fewer than 120 cases have been reported [2]. Only about 2 to 4.3% of the cases involve central involvement of the mandible or maxilla [4].The posterior part of the mandible is usually affected more than the maxilla [2-4]. In a study conducted by Bouquot et al. [4], intraosseous salivary tissue was found in 0.3% of upper jawbones [5].

Consequently, the World Health Organization (WHO) recommended changing the diagnosis of mucoepidermoid tumor to mucoepidermoid carcinoma in 1991 based on the cancer's propensity to metastasize, clinical behavior, and treatment [5]. The name mucoepidermoid carcinoma implies that it is an epithelial mucin-producing neoplasm. Even though low-grade mucoepidermoid carcinomas can metastasize, they predominantly have a non-aggressive nature [3]. The origin of the central mucoepidermoid carcinoma (CMC) of the mandible can be one of the following: (i) metaplasia of odontogenic cysts epithelium; (ii) entrapment of salivary tissues from the submandibular, sublingual, or minor salivary glands, during embryonic development; (iii) entrapment of minor salivary glands from the retromolar area; (iv) maxillary sinus epithelium; (v) iatrogenic entrapment of minor salivary glands; (vi) odontogenic remnants of the dental lamina [4,6,7]. In clinical terms, CMC manifests as a long-standing painful

lesion, movement of teeth, swelling, and, on rare occasions, altered sense of the inferior alveolar nerve [3]. The condition can occasionally be confused with benign odontogenic tumors or cysts and presents as asymptomatic. As clinical symptoms are typically unrepresentative of CMC, the diagnosis is typically determined by a histopathological examination.

CASE DESCRIPTION

This is a case report of a 47-year-old male patient who presented to the Department of Oral and Maxillofacial Surgery, Ali Omar Askar Neurosurgery Centre in Sbea Tripoli, Libya, with pain throughout the left posterior mandible. One of his primary concerns was mobility, especially of lower left incisors, canines, and molars. The swelling began four months ago and gradually increased in size. The patient had previously been seen by a dentist for several loose teeth in the left mandible and had his lower left second premolar extracted. The socket on his lower jaw never healed, so he developed lower lip paraesthesia in the post-extraction period. He was prescribed multiple courses of antibiotics and analgesics by the dentist. No significant medical history existed for his jaws. There was no history of trauma or surgery.

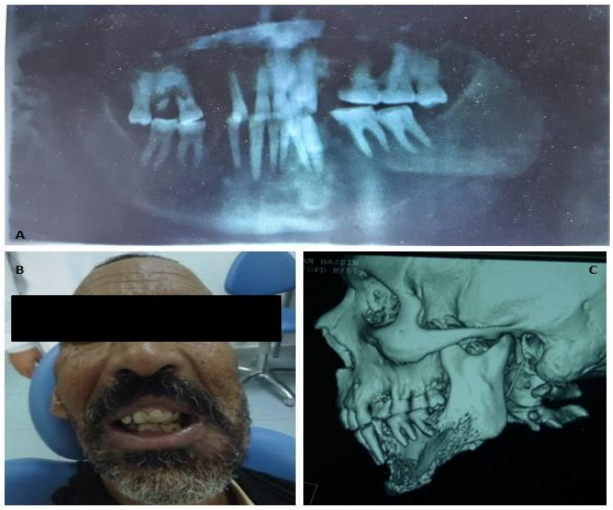

Figure 1: Orthopantomogram showing non-healing sockets of left mandibular premolars and irregular radiolucencies in the symphyseal region extending to the molars [A]. Clinical pictures of the patient showing the asymmetry of face and swelling in left side of the mandible[B]. Three-dimensional CT scan showing the extent of the resorption of symphyseal and body of left mandible[C].

On clinical examination, the patient developed left mandible swelling extending from the midline to the angle of the mandible (Figure 1A-C). The skin overlying the lymph nodes was normal. An occurrence of paraesthesia was noted on the lower lip left as well as a submandibular lymph node. There is an intra-oral swelling that involves the buccal and lingual mucosa, showing signs of expansion in both cortices. Several lower incisors, canines, and molar teeth in the left mandible showed grade 2 mobility. The mobility of these teeth contrasted drastically with their periodontal health and teeth in other quadrants. A pan-tomogram showed an indefinite radiolucent region encompassing the symphysis and the angle of the mandible. On three-dimensional computed tomography, the right posterior mandible displayed an ill-defined lesion. Microscopic examination revealed mucoepidermoid carcinoma of the mandible after an incisional biopsy.

Excision of the lesion was performed to the extent of the left angle of the mandible and the right parasymphysis (segmental mandibulectomy), followed by plate reconstruction to restore the defect (Figure 2). The patient recovered without incident and continued to be monitored.

On microscopic inspection, the tumor was characterized by several cysts lined with columns, mucous cells, and epidermal cells. In figure 3-5, histopathological findings are presented, and explanatory legends discuss these findings in detail. Based on the clinical presentation, along with radiographic findings (osteolytic lesion with signs of periapical erosion), a diagnosis of peripheral mucoepidermoid carcinoma of low-grade malignancy was made.

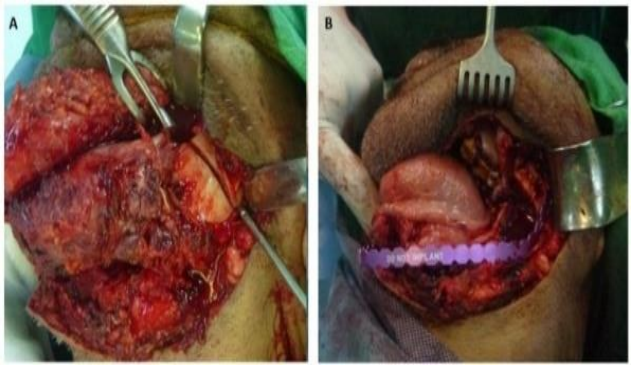

Figure 2: Segmental mandibulectomy extending from left angle of the mandible and right parasymphysis[A]. Reconstruction with plate extending from left ramus and right parasymphysis of the mandible[B].

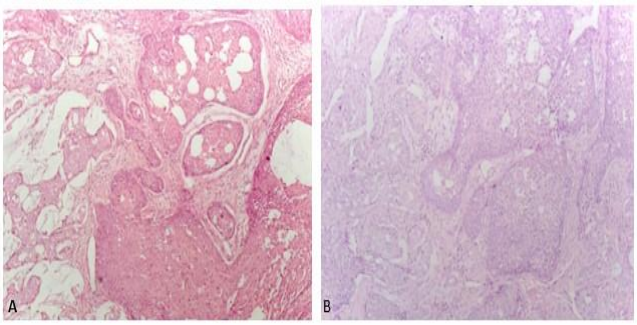

Figure 3: H&E stained section shows solid pattern punctuated by occasional cystic spaces composed predominantly of sheets of malignant squamous cells intermixed with intermediate cells, few mucinous cells seen [A] and shows predominantly of sheets of malignant squamous cells intermixed with intermediate cells, few mucinous cells seen[B].

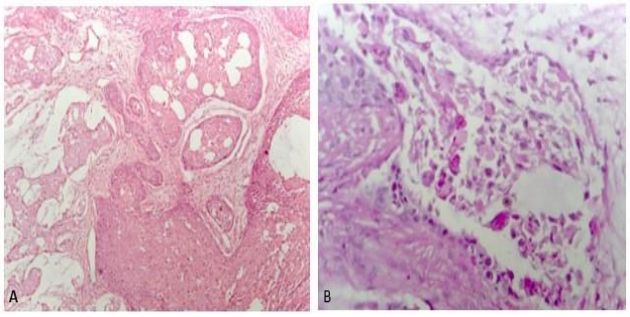

Figure 4: H&E stained section [X20] shows solid pattern punctuated by occasional cystic spaces composed predominantly of sheets of malignant squamous cells intermixed with intermediate cellsshowing desmoplastic reaction [A] and showsmucinous cells show positivity with PAS-PAS diastase and a part of solid tumor appears on the top left [B].

DISCUSSION

Central salivary gland tumors are extremely rare in the jaws, but the CMC represents the largest percentage with 64% to 75% of the cases [4]. Waldron and Mustoe [7] proposed the mucoepidermoid intraosseous carcinoma as an independent entity within primary intra-osseous carcinoma (PIOC). As CMC of the jaws was histologically identical to salivary mucoepidermoid carcinoma and was thought to arise from epithelial remnants of the odontogenic cyst, CMC could be considered an odontoid tumor [7,8].

The diagnosis of CMC was always difficult in the absence of characteristic radiological findings and clinical manifestations. In an instance of CMC diagnosis based on radiological, clinical, and histopathological manifestations, Waldron and Koh [1990] set out diagnostic criteria. Later in 2004, Kochaji et al proposed revised those guidelines, which include:(i) intact cortical plates; (ii) radiographic evidence of bony destruction; (ii) exclusion of another primary tumor that in its metastasis could histologically mimic the central tumor; (iv) exclusion of an odontogenic tumor; (v) histopathological confirmation; (vi) detectable intracellular mucin [10]. Several authors agree that intact cortical plates do not have to be present to diagnose CMC [12-15].

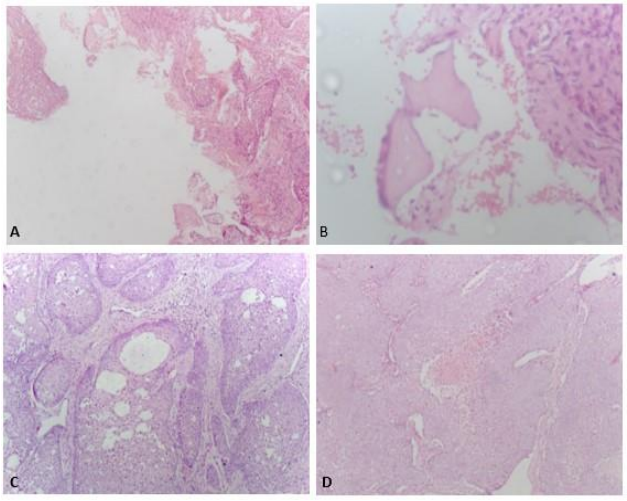

Figure 5: M/E Section shows sheets of malignant squamous epithelial cellsinvading bony tissue with associated desmoplastic stromal reaction [A], sheets of malignant squamous epithelial cells associated desmoplastic stromal reaction [B], solid pattern punctuated by cystic spaces [C], sheets of malignant squamous epithelium with areas of coagulative tumor necrosis [D].

Since the first case of CMC was reported in 1939, the etiology of CMC has remained unclear until now. Several theories regarding the origin and pathogenesis were postulated, some of them include (a) entrapment of retromolar mucous glands within the mandible with subsequent neoplastic transformation of these glands, (b) developmentally included embryonic remnants of the submandibular and sublingual glands within the mandible, (c) neoplastic transformation of the mucous-secreting cells commonly found in the pluripotential epithelial lining of

dentigerous cysts associated with impacted third molars, (d) neoplastic transformation and invasion from the lining of the maxillary sinus [6,11].

Female predilections are slightly more common in the 4th and 5th decades of life when it comes to CMC [11]. According to a review of 147 cases conducted by de Souza et al., the female:male ratio was only 1.07:1.00 and female patients suffered twice the morbidity rate of male patients [13]. A study of 24 patients with CMC by He et al. found that 15 men and 9 women had a predisposition to CMC [14]. The case under report is a 47-year-old male patient who has CMC.

The incidence of CMC is two times higher in the mandible than the maxilla in adults, while in children, the incidence is almost identical in the mandible and maxilla [12]. He et al.,[14] found higher rates of CMC in the maxilla than in the mandible, and they did not see a significant difference between the maxilla and mandible. Our patient's case is consistent with clinical findings that premolar–molar–angle/molars are the most common sites of occurrence [13-16]. The most common symptoms of CMC are painless swelling in the oral cavity, trismus, pain, paresthesia, numbness, craniofacial asymmetry, and loose teeth [10-17]. The patient presented with a history of pain, swelling, trismus and loose teeth that are all consistent with previous published literature.

According to He et al. [14], swelling of the jaws may be an early stage of cancer and a sign of well differentiated tumors, which may lead to patients with localized swelling being mislabeled with benign tumors or cysts. Lesions can actively invade the inferior alveolar nerve, causing pain or paresthesia without swelling. These symptoms suggest a poor prognosis for patients with active tumors and may be indicative of their disease's active phase [12]. In most cases, CMC occurs with invasion of local tissue without causing distant metastasis [7-9]. It was reported by Brookstone and Huvos that only 9% of patients with CMC suffered from metastatic disease [17].

Reviewing treatment options as well as prognostic factors, de Souza et al. recommended excision of the tumor and margin without dissection of the lymph nodes [12]. Most metastatic diseases are found in the local lymph nodes, while the cervix, breast, lung, and skin are rarely affected [17]. There were no lymph nodes involved in the present case. Abu-Karaky and colleagues found that only 4% of CMCs recurred after radical methods despite some authors having done neck dissections where lymph nodes were involved [19].

In the past, some authors used conservative treatment modalities, such as enucleation and curettage. However, these treatments are no longer recommended since they can lead to recurrence rates of up to 40%-45% [7-9]. Bell et al. found that patients with lymph node metastasis had a fourfold greater risk of dying than patients without lymph node metastasis [20]. Metastatic disease is not uncommon in CMC patients [2], but it can occur at any time since treatment, so regular follow up is recommended for such patients [10,17,20]. The patient has been placed on a regular follow-up program after having been informed about the risks of recurrence.

CONCLUSION

Preoperative diagnoses of CMC are still challenging since it is a rare malignant neoplasm of the jaw. There are several pathologies that may mimic CMC, including ameloblastoma, odontogenic cysts, and GOC. Histopathological and imaging studies are usually needed to determine the definitive diagnosis. The majority of CMCs exhibit low-grade malignancy. There should be radical surgery performed to enhance the prognosis. It is imperative that a long-term follow-up system be used since CMC is prone to recurring long after surgery. To fully understand the CMC, additional studies are required due to its rarity and controversial aspects.

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