CASE REPORT

Primary intraosseous mucoepidermoid carcinoma of maxilla

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ABSTRACT

Primary intraosseous mucoepidermoid carcinoma (PIOC) of the jaw bones is an extremely rare malignant salivary gland tumor, comprising 2–3% of all mucoepidermoid carcinomas reported. It is commonly seen in the posterior part of the mandible; its occurrence in the maxilla is rare. They have been reported in patients of all ages, ranging from 1 to 78 years, with the overwhelming majority occurring in the 4th and 5th decades of life. They are histologically low-grade cancers and radiographically seen as uniocular or multiocular lesions. We report a rare case of PIOC in posterior palatal region in 18-year-old male.

Key words: Intraosseous carcinoma, mucoepidermoid carcinoma, salivary gland neoplasms

INTRODUCTION

Mucoepidermoid carcinoma (MEC) is a malignant epithelial tumor first studied and described by Stewart, Foote and Baker in 1945. Rarely, it may occur intraosseously from the epithelial lining of the odontogenic cyst and/or epithelial remnants of ectopic salivary glands.[1]

MEC is the most commonly occurring malignant salivary gland neoplasm, comprising 2.8–15% of all salivary gland tumors.[2] Aberrant salivary gland neoplasms arising within jaws as primary central bony lesions are extremely rare, comprising 2–4.3% of all MECs reported.[3,4] Central MEC is a rare but well-known entity affecting the jaw bones. Its pathogenesis, radiological and histopathological aspects have been extensively discussed.[4-8] Most primary central MEC lesions occur in the mandible, but are rare in the maxilla.[4-9] Here, we report a rare case of a primary intraosseous mucoepidermoid carcinoma (PIOC) in the posterior palatine region in a 18-year-old male patient, which was challenging because of the clinical presentation as a cystic lesion.

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CASE REPORT

An 18-year-old male patient reported to the Outpatient Department (OPD) of the hospital with a chief complaint of swelling in the left maxillary posterior region since 2 years. Swelling was small in size initially and asymptomatic but gradually increased in size and was painful. Also complains of pus drainage for which antibiotics were prescribed, which temporarily relieved symptoms.

On examination, soft, fluctuant, tender swelling of approximately 3 × 2 cm in the left palatal region opposite the permanent maxillary premolar molar region was observed [Figure 1]. The overlying mucosa was non-ulcerated and appeared normal. Permanent maxillary first and second maxillary molar were carious. Left submandibular nodes were palpable and tender. Two milliliter straw-colored fluid was aspirated from the swelling. Occlusal view radiograph revealed a well-defined unilocular radiolucency in relation to premolar molar region with sclerotic margins [Figure 2a]. Reformatted computed tomography (CT) image revealed palatal bone perforation extending to the alveolar process between permanent maxillary first and second maxillary molar on the left side [Figure 2b].

On evaluation of clinical and radiographic features, a provisional diagnosis of a cystic lesion mainly a radicular cyst was considered. Subsequently, an incisional biopsy was done which was inconclusive. This was followed by a complete surgical curettage of the lesion.

Histopathological examination revealed large number of haphazardly dispersed cystic (duct like) spaces filled with eosinophilic material; surrounded by tumor nests; composing of mucous and squamoid (epidermoid) cells in variable combinations; scanty and hyalinized stroma; and in few areas extravasated mucin pools [Figure 3a]. Cystic spaces were lined by mucus cells in single or double rows and contained eosinophilic material which was Periodic Acid Schiff PAS-positive [Figure 3b]. Stratified squamous epithelium of varying thickness with intraepithelial clefts and PAS mucous cells in superficial layer could also be appreciated [Figure 3c] Histopathological diagnosis of a central low-grade MEC of the left posterior palatal region was made.

A planned treatment of the lesion was done, which included right maxillectomy with neck dissection. Further histopathological examination confirmed the diagnosis of a low-grade MEC. Metastasis was not observed in the dissected lymph nodes. The patient has been on regular follow-up for more than a year without any evidence of recurrence.

DISCUSSION

Primary intraosseous MEC is an uncommon lesion which was first reported and described by Leep[10] in 1939. Waldron and Mustoe[11] suggested that intraosseous MEC be included in the primary intraosseous carcinomas of the jaws as type 4.

Primary central MEC has been reported to be present in the first to seventh decade; however, cases occurring in the fourth and fifth decades are most common,[12] but in the present case, the patient was of 18 years. In children, gender ratio and the mandible to maxilla ratio is 1:1, whereas in adults, MEC is slightly more common in females and occurs in the posterior mandible,[12] but in this case the patient was a male and the site was the posterior palate. MEC usually presents as a painless swelling. Pain, paresthesia, numbness and tooth mobility are usually occasional and late findings.[13]

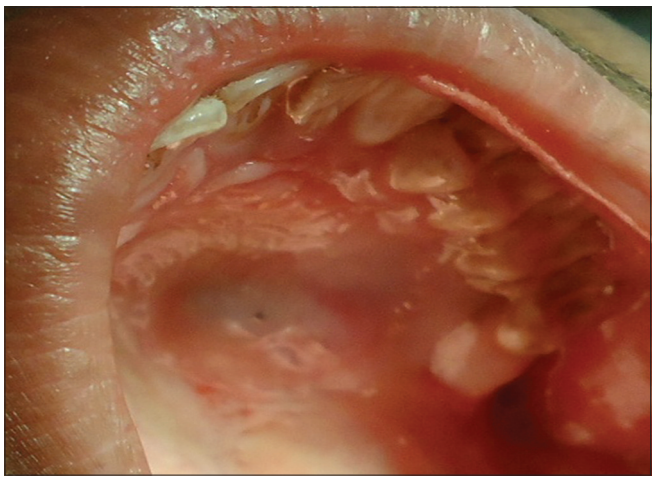

Figure 1: Preoperative view showing swelling of about 3 × 2 cm in left palatal region opposite premolar molar region

According to the criteria of Seifert and Sobin[14,15] and Auclair *et al*.,[16] the tumors are graded for malignancy as follows: (1) Low grade: Highly differentiated neoplasia with a predominance of macro and microcysts. Presence of intermediate and mucin-producing cells, (2) Intermediate grade: Predominance of intermediate cells and a few cysts. Presence of mucin-producing cells and islands of epidermoid cells, (3) High grade: Poorly differentiated neoplasia with predominance of intermediate and epidermoid cells in solid blocks. Mucin producing cells are present.

Histopathologically, low-grade MEC s are composed of cuboidal to columnar mucus secreting cells arranged around microcystic structure with intermingling of epithelial or intermediate cells with few epidermoid cells. Coalescence of small cysts into large cystic spaces is typical of a low-grade malignancy.

The pathogenesis of the central MEC has been discussed extensively and various possible origins have been considered, including:[17,18] (1) Entrapment of the retromolar mucous

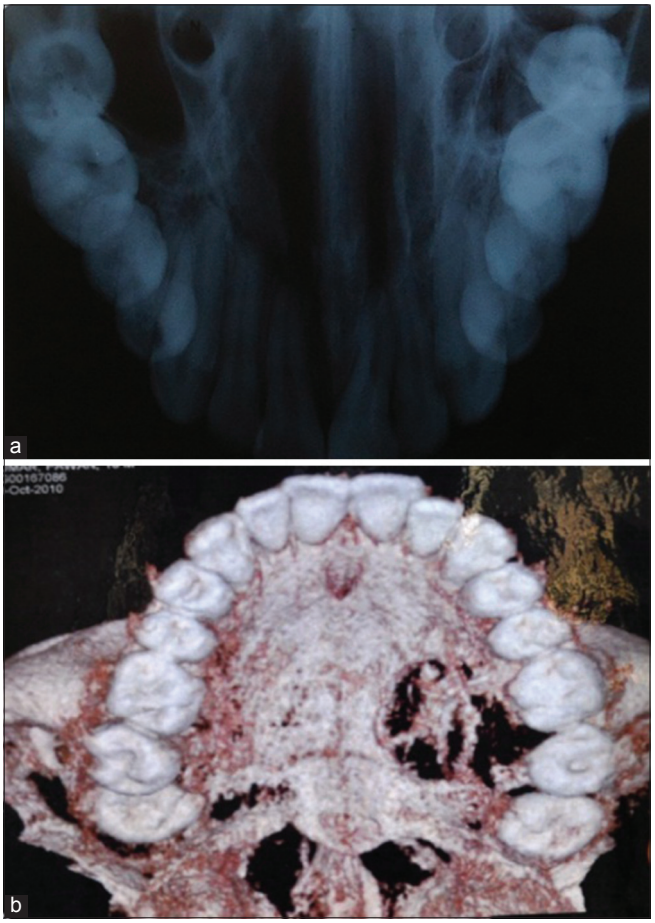

Figure 2: (a) Occlusal view showing a well-defined unilocular radiolucency in the left posterior palatal region extending from left first premolar to molar, with smooth, corticated margins (b) Reformatted computed tomography (CT) image axial view showing palatal bone perforation extending to alveolar process between first and second molar on left side

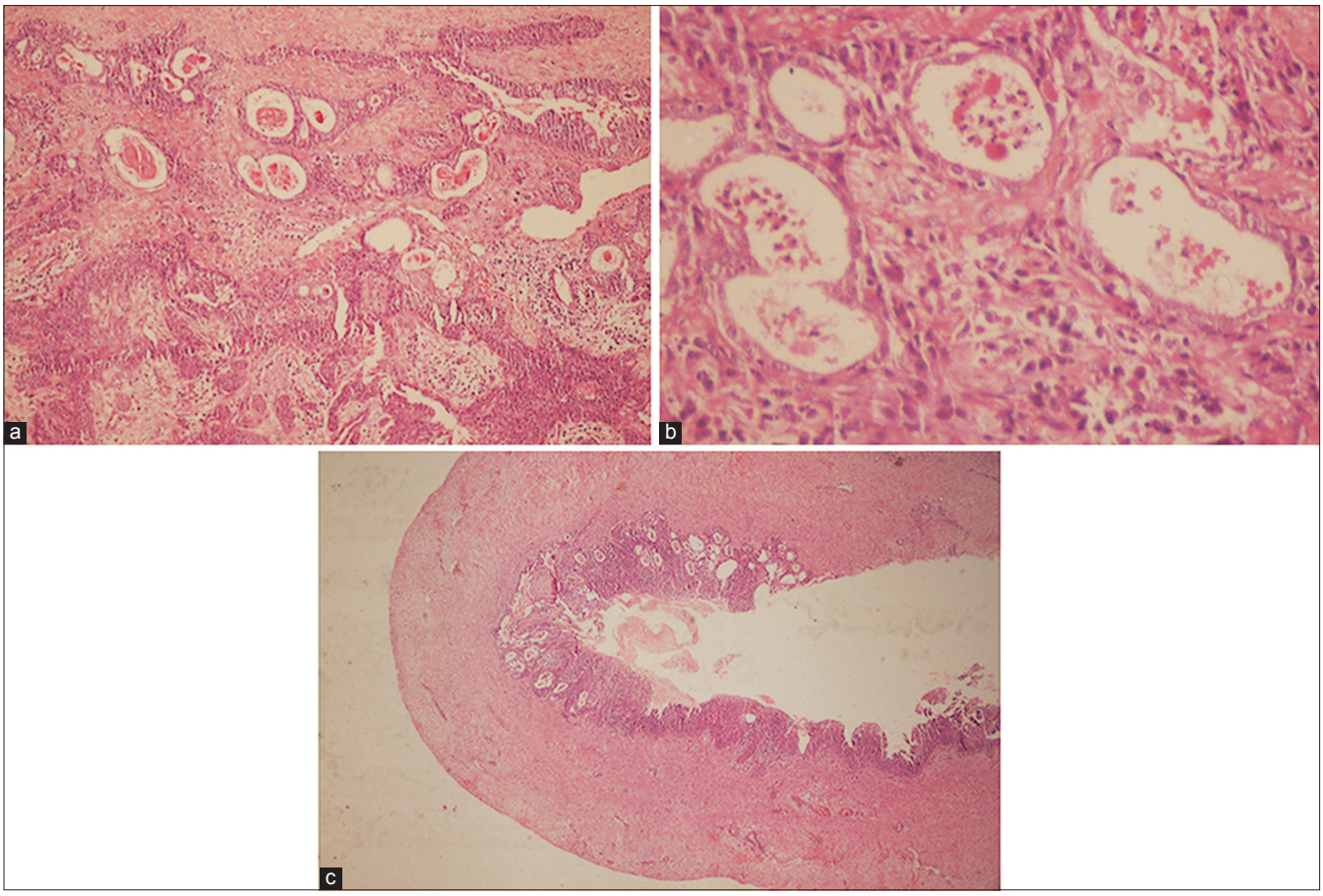

Figure 3: (a) Photomicrograph showing sheets of epidermoid cells and mucus-secreting cells, a few areas of cystic cavities lined with mucus-secreting cells (H&E stain, x100) (b) Photomicrograph showing cystic spaces that are lined by mucus cells in single or double row and were containing eosinophilic material which was PAS positive (H&E stain, x400) (c) Photomicrograph showing cystic cavity lined by stratified squamous epithelium of varying thickness with intraepithelial clefts and superficial layer of mucus cells (H&E stain, x40)

glands within the mandible, which later undergo neoplastic transformation, (2) embryonic remnants of the submandibular and sublingual glands trapped within the mandible during development, (3) neoplastic transformation and invasion from the lining of the maxillary sinus, (4) neoplastic transformation of the mucus-secreting cells from the epithelial lining of the dentigerous cyst associated with impacted third molars, (5) neoplastic transformation of entrapped minor salivary glands within the maxilla.

However, central MEC of the maxilla is a rare lesion and the pathogenesis has not been extensively discussed. The possibilities of the origin of the central MEC in this case may be: (1) Neoplastic transformation and invasion from the lining of the maxillary sinus, (2) neoplastic transformation of entrapped minor salivary glands within the maxilla.

Radiographic examination is important to develop a differential diagnosis, evaluate categorization as a central jaw lesion,[19] the extent of the lesion and involvement or encroachment of adjacent vital structures. Brookstone and Huvos[13] suggested a three-grade classification for intraosseous MEC: Grade 1, without expansion and rupture of cortical plate; Grade 2, with expansion but without rupture of cortical plate; and Grade 3, with rupture of cortical plates or presence of regional metastasis. Our case fit into Grade 3 with rupture of cortical plates.

The primary treatment modality for patients with central intraosseous MEC is surgery, including curettage, enucleation, marsupialization and wide local excision.[7,8,13] As a rule, even as low-grade tumors, MEC should be managed by wide local resection, *en bloc* resection, or hemimandibulectomy,[6,13,20] or hemimaxillectomy. Neck dissection is usually part of the treatment in cases where metastasis to the cervical nodes is suspected.[20] Radiotherapy is recommended for high-grade MEC cases.[20,21]

These tumors usually show a good overall prognosis, but central MEC cases should be followed up for a longer period up to 10 years, due to the possibility of late recurrence or regional metastasis.[13,18,20] However, death may occur as a result of extension into vital structures such as the base of the brain.[5,8] Very little information is available in the literature about the recurrence rate of central MEC.[3]

Due to rare occurrence of central MEC, we lack systematic data and criteria for diagnosis. In 1974, Alexander *et al*.[22] introduced the following criteria for diagnosing central MECs include: (a) Presence of a radiographic distinct osteolytic lesion; (b) positive mucicarmine staining; (c) absence of rupture of one or more cortical plates; (d) clinical and histological exclusion of a metastasis or an odontogenic lesion; (e) exclusion of the origin from a soft tissue salivary gland; (f) histologic confirmation, which was similar to Brookstone and Huves criteria.[13] There are still some contradictions in these criteria as cases have been diagnosed with cortical plate perforation and invasion of nearby tissue.

In conclusion, central MEC in the jaws is a rare entity. The posterior part of the mandible is the common site. However, occurrence in the maxilla cannot be overlooked. Clinical and radiological presentation are challenging especially on low-grade variants. Treated cases of central MEC should be followed up for a longer duration to detect late local recurrence and regional metastasis.

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