Case Report

Central mucoepidermoid carcinoma: Case report with review of literature

ABSTRACT

Occurrence of central mucoepidermoid carcinoma within the jaw bones from aberrant salivary tissues is extremely rare and accounts for 2%–4% of all central mucoepidermoid carcinoma. Mandible is more commonly affected than maxilla in a ratio of 2:1. The origin of mucoepidermoid carcinoma is controversial and questionable. Central mucoepidermoid carcinoma is frequently misdiagnosed radiographically and clinically as a benign odontogenic tumor or cyst. Hence, it is important to detect malignant signs and histopathological feature of central mucoepidermoid carcinoma of the mandible to confirm the diagnosis. This paper reports a case of central mucoepidermoid carcinoma of the mandible in a 56-year-old patient with review of literature.

Keywords: Carcinoma, central mucoepidermoid carcinoma, salivary gland tumor

INTRODUCTION

Mucoepidermoid carcinoma is most commonly occurring salivary gland tumor, which was first described by Stewart *et al*. in 1945 as epidermal cellular element and mucus secreting.[1] Eighty-six percent of these types of salivary gland tumor occur in parotid gland, followed by submandibular gland (8%), sublingual gland (0.4%), and minor salivary glands of palate.[2]

Occurrence of central mucoepidermoid carcinoma within the jaw bones from aberrant salivary tissues is extremely rare and accounts for 2%–4% of all central mucoepidermoid carcinoma. Mandible is more commonly affected than maxilla in a ratio of 2:1.[2,3] One-half of these carcinomas of mandible are associated with impacted teeth.[2] Lepp in 1939 reported the first case of central mucoepidermoid carcinoma of the mandible in a 66-year-old woman.[4]

The origin of mucoepidermoid carcinoma is controversial and questionable, but there are several possibilities have been considered, including: (1) metaplasia of odontogenic cysts epithelium; (2) entrapment of salivary tissues from the submandibular, sublingual, or minor salivary glands, during embryonic development; (3) entrapment of minor salivary glands from the retromolar area; (4) maxillary sinus epithelium; (5) iatrogenic entrapment of minor salivary glands (e.g. chronic osteomyelitis and sinusitis); and (6) odontogenic remnants of the dental lamina.[5-7] More recently, intraosseous salivary tissue was demonstrated in 0.3% of all bony specimens of jaw bones studied by Bouquot *et al*.,[6] providing new evidence for the origin of intraosseous salivary carcinomas.

Tumor shows varied degree of clinical presentation, which may include pain, movement of teeth, swelling, and

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Received: 02-12-2016, **Revised:** 19-12-2016, **Accepted:** 28-12-2016

Access this article online	
Website: www.njms.in	Quick Response Code
DOI: 10.4103/0975-5950.259843	

How to cite this article: Singh H, Yadav AK, Chand S, Singh A, Shukla B. Central mucoepidermoid carcinoma: Case report with review of literature. Natl J Maxillofac Surg 2019;10:109-13.

even altered sensation of the inferior alveolar nerve for a long-standing lesion. However, most of the time, it remained asymptomatic and diagnosed as an incidental finding on a dental radiograph.[8]

Central mucoepidermoid carcinoma is frequently misdiagnosed radiographically and clinically as a benign odontogenic tumor or cyst. Hence, it is important to detect malignant signs and histopathological feature of central mucoepidermoid carcinoma of the mandible to confirm the diagnosis.[9]

This paper reports a case of central mucoepidermoid carcinoma of the mandible in a 56-year-old patient with review of literature.

CASE REPORT

A 56-year-old patient reported to a private dental clinic, Lucknow, with a chief complaint of pain and swelling in the lower left region of the jaw since 1 year. Pain was dull and intermittent in nature and was localized. The patient was apparently asymptomatic 1 year back; then, he noticed bleeding from the lower left back tooth region.

Patient's personal history revealed that he had a habit of tobacco chewing 5–6 times a day since 30 years. On extraoral examination, a firm swelling was present on the left middle third of face, round measuring about 3 cm × 3 cm. The swelling was nontender, with no change of temperature of the overlying skin. The left submandibular lymph nodes were palpable, slightly mobile, and tender.

Intraoral examination revealed that patient had a poor oral hygiene. A healing biopsy wound was present in relation to lower left permanent third molar region [Figure 1]. Slight obliteration of left buccal vestibule was also present in relation to lower left permanent first, second, and third molar region. Orthopantomogram [Figure 2] and computed tomography (CT) scan with three-dimensional reconstruction [Figure 3] followed by excisional biopsy were advised. On the basis of clinical, radiological, and histopathological findings, diagnosis of mucoepidermoid carcinoma was made, and under general anesthesia, partial mandibulectomy of the left side was done [Figure 4] and specimen [Figure 5] was sent for histopathological examination which confirms the diagnosis and reconstruction was done with a recon plate.

DISCUSSION

Central mucoepidermoid carcinoma of the mandible is a rare lesion, but it is well recognized, with more than 110 reported cases in the literature. Most commonly, it occurs in the fourth to sixth decades of life with a slight female predilection.[10] It involves the mandible two times more often than the maxilla.[11] In children, the incidence is almost equal in both maxilla and mandible. The most common site of occurrence is the premolar–molar–angle region of the mandible. It is associated with mandibular cysts in approximately 50% cases.[2,12] As the tumor shows a tendency to crop up at puberty, hormonal influence of the salivary glands was suggested as an etiological factor.[13] Painless swelling of the jaw is the most common presentation and occasionally presents with pain, paresthesia, numbness, and loosening of a tooth.[12]

Although rare, intraosseous carcinoma arising in the jaw bones is a well-known clinical entity and was first described as a central epidermoid carcinoma by Loos in 1913.[14] Later, Pindborg, in the first edition of WHO classification of the odontogenic tumors based on histopathological differentiation, coined the name "primary intraosseous carcinoma" (PIOC).[15] The WHO defines PIOC of the jaw as a "squamous cell carcinoma arising within the jaw," having no initial connection with the oral mucosa and presumably developing from residues of the odontogenic epithelium.[15]

Later, Waldron and Mustoe[16] suggested that intraosseous mucoepidermoid carcinoma is included in the classification of PIOCs as type 4 [Table 1]. This was based on the fact that mucoepidermoid carcinoma of the jaws was similar to salivary mucoepidermoid carcinoma histologically and it was thought to arise from epithelial remnants of the odontogenic cyst.

Clinically, it exhibits rapid onset, pain, movement of teeth, swelling, destruction of local structure, and even altered

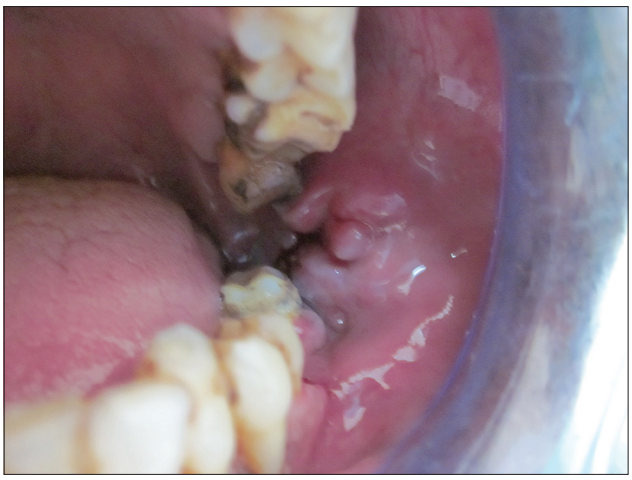

Figure 1: Healing biopsy wound

sensation of the inferior alveolar nerve for a long-standing lesion. Most of the time, it remains asymptomatic and diagnoses as an incidental finding on a dental radiograph. Metastases can occur in regional lymph nodes.[17]

Radiographically, lesion is well-defined unilocular, lobulated or multilocular cyst-like radiolucency. Displacement of teeth and root resorption also occur commonly. Cortical perforation usually occurs in long-standing cases.[8] For knowing the extent of the lesion and its relation to vital structures, panoramic radiography is a simple and cost-effective imaging modality. For exact extent of lesion, nodal status, and to evaluate bony destruction, CT scan is useful diagnostic tool.[18] In 1992, Brookstone and Huvos based on radiology proposed a staging system based on the condition of the overlying bone [Table 2].[12]

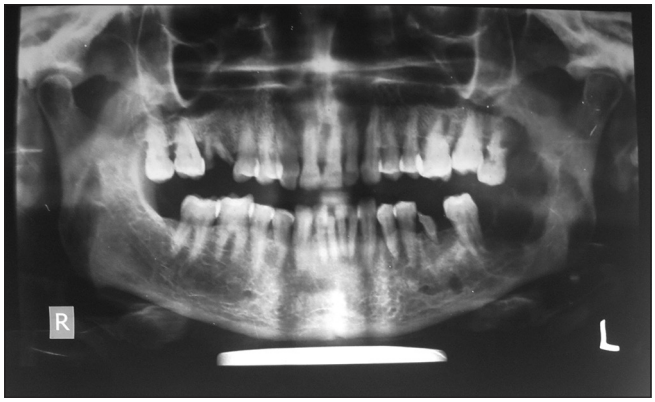

Figure 2: Orthopantomogram

Besides clinical and radiographic examination, biopsy is also necessary for the final diagnosis and treatment planning in all cases of central mucoepidermoid carcinoma. Literature review considers the origin of central mucoepidermoid carcinoma from odontogenic cysts and tumors. Most of the cases are associated with dentigerous cysts, but there is evidence of their association with periodontal apical cysts, residual cysts, glandular odontogenic cysts, and odontogenic tumors.[7,12] Most of the reported central mucoepidermoid carcinoma are

Table 1: Waldron and Mustoe modification of WHO classification of primary intraosseous carcinoma

Type	Modified classification of PIOC (WHO 2005)
Type 1	PIOC ex odontogenic cyst
Type 2a	Malignant ameloblastoma
Type 2b	Ameloblastic carcinoma arising *de novo*, ex-ameloblastoma or ex-odontogenic cyst
Type 3	PIOC arising *de novo*
Type 3a	Keratinizing type
Type 3b	Nonkeratinizing type
Type 4	Intraosseous mucoepidermoid carcinoma

PIOC: Primary intraosseous carcinoma

Table 2: Clinical staging of central salivary gland tumor including central mucoepidermoid carcinoma

Stage	Condition of overlying bone
Stage I	Without bony expansion and rupture of cortical plate
Stage II	Rupture of cortical plate with bony expansion
Stage III	Rupture of cortical plate or nodal involvement

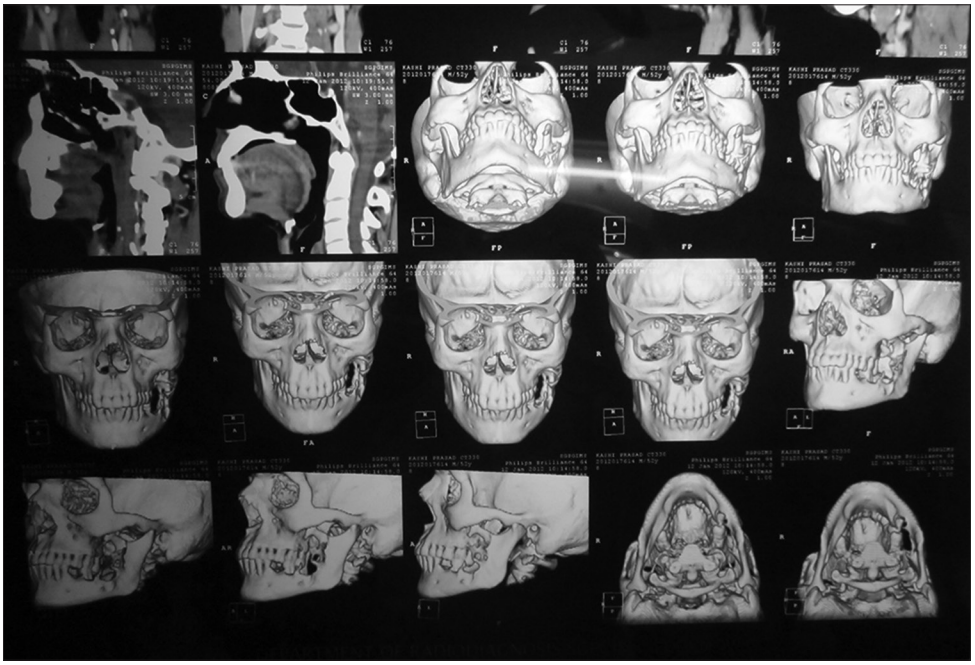

Figure 3: Three-dimensional computed tomography scan

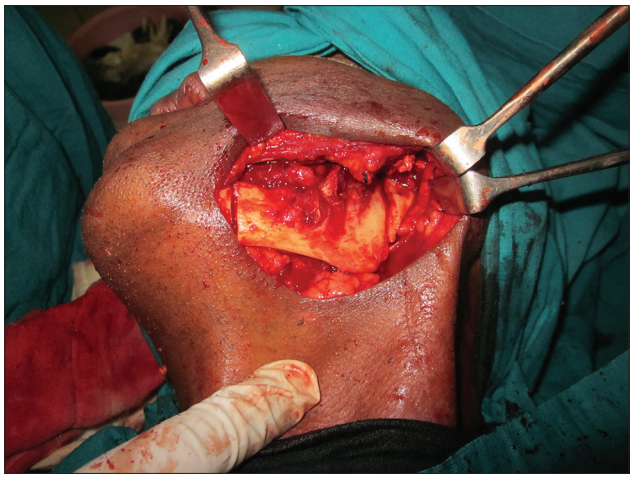

Figure 4: Partial mandibulectomy

histologically as low-grade tumors and usually carry a favorable prognosis[7,13,19]

Kochaji *et al*.[3] published guidelines for the diagnosis of primary mucoepidermoid carcinoma of jaws. The most important criteria for diagnosis of primary mucoepidermoid carcinoma of jaws are to exclude primary lesion in the salivary glands.

- Intact cortical plates (however, cortical perforation does not exclude PIOC type 4)
- Radiographic evidence of bony destruction
- Exclusion of another primary tumor that in its metastasis could histologically mimic the central tumor
- Exclusion of an odontogenic tumor
- Histopathological confirmation
- Detectable intracellular mucin.

Most surgeons agree that surgical extirpation of these lesions is the most appropriate treatment, but the extent of resection and treatment of regional lymphatic involvement varies from surgeon to surgeon. Surgical approaches include curettage, enucleation, marginal *en bloc* resection, and segmental mandibulectomy (for more extensive lesions).[19] Regional lymph node metastasis occurs in approximately 10% cases and for these palpable neck diseases. For high-grade tumor, postoperative radiotherapy recommended.[20]

In the present case, after confirmation of diagnosis, partial mandibulectomy was done, followed by reconstruction with recon plate being performed depending on the extent of lesion. Postoperative healing was uneventful, and the patient was kept under regular follow-up of 3 years till date.

CONCLUSION

Central mucoepidermoid carcinoma of the mandible occurs rarely and can be misdiagnosed radiographically. Therefore, any lesion in the posterior aspect of the mandible suspected as cystic lesion than differential diagnosis of central mucoepidermoid carcinoma should be considered. Late recurrences and metastases are common in such type of lesion; hence, prolong follow-up is required.

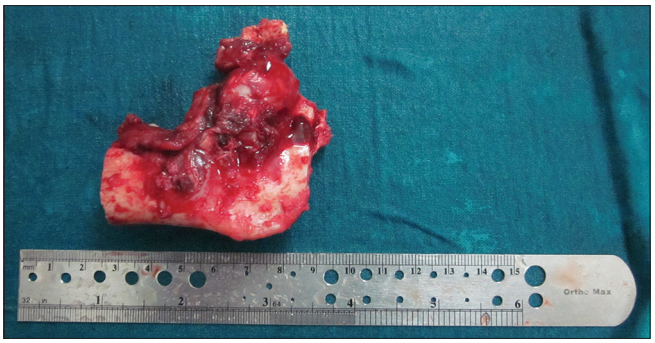

Figure 5: Resected specimen

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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