CASE REPORT

Clear cell variant of intraosseous mucoepidermoid carcinoma: Report of a rare entity

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ABSTRACT

Intraosseous mucoepidermoid carcinoma of jaw bones is a rare lesion. Abundance of clear cells in an intraosseous mucoepidermoid carcinoma may complicate its histopathologic diagnosis. It becomes extremely important to distinguish this lesion from other clear cell lesions of jaw region. Here, we report a case of clear cell variant of intraosseous mucoepidermoid carcinoma in the mandible.

Key words: Clear cells, intraosseous mucoepidermoid carcinoma, mandible, mucicarmine staining, PAS

INTRODUCTION

Mucoepidermoid carcinoma (MEC) is a common malignant salivary gland neoplasm originating in both major and minor salivary glands. It occurs mainly in the parotid gland (89.6%), followed by submandibular gland (8.4%)[1] Intraorally it shows a strong predilection for palate. As the name implies, MEC is composed of a mixture of cell types - mucus secreting, epidermoid and intermediate cells. Clear cells are a rare finding in MECs. Clear cells may occur in focal areas or may predominate large areas of the tumor, thus complicating the diagnosis.[2-5]

Though rare, MEC occurs in the jaw bones[6,7] (intraosseous mucoepidermoid carcinoma) Waldron and Mustoe has classified primary intraosseous carcinomas (PIOC) in which intraosseous MEC is included as Type 4 [Table 1].[8] When MEC occurring in jaw bones demonstrate predominantly clear cells, the diagnosis becomes difficult.[9] It is important to distinguish them from other clear cell lesions of the jaw region.

We report here a case of intraosseous MEC of mandible, which showed abundant clear cells.

CASE REPORT

A 50 year old male patient reported at the Department of Oral Pathology, Government Dental College, Calicut, with a painless slow growing swelling on left side of mandible at the angle – ramus region of 4 years duration. He gave a history of a similar swelling at the same location 12 years ago, which was diagnosed as dentigerous cyst associated with an impacted third molar. It was treated by cyst enucleation and removal of the impacted tooth. The patient remained symptom free for about 7 years after the procedure, following which he developed a painless swelling, which reached the present size.

Clinical examination showed the presence of a diffuse, nontender swelling of approximately 6 × 4 centimeters over the left angle–ramus region of mandible [Figure 1]. Intraorally the swelling extended from 34 to retromolar region, obliterating the buccal vestibule [Figure 2]. The mucosa overlying the swelling was intact with normal color and smooth texture. A panoramic radiograph was taken which showed a multilocular radiolucency, which extended from 34 region toward coronoid and condyle, involving both [Figure 3]. With these features a provisional diagnosis of ameloblastoma was made. The patient underwent an intraoral incision biopsy from the lesion.

H and E stained sections of the biopsy specimen showed cystic spaces filled with eosinophilic material, surrounded by epidermoid cells and sheets of large polygonal cells with centrally placed nuclei, clear cytoplasm, and sharply defined

Table 1: Classification of primary intraosseous carcinomas (Waldron and Mustoe[8])

Type 1	PIOC ex odontogenic cyst
Type 2A	Malignant Ameloblastoma
Type 2B	Ameloblastic Carcinoma
Type 3	PIOC developed *de novo*
	a. keratinising type
	b. non keratinizing type
Type 4	Central intraosseous Mucoepidermoid carcinoma

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cytoplasmic borders [Figure 4]. The intervening connective tissue stroma was scanty. The sections were stained with mucicarmine and Periodic acid Schiff's reagent (PAS) to assess the nature of clear cells. The eosinophilic material in cyst like spaces was PAS and mucicarmine positive. Mucus-secreting cells were visualized through mucicarmine staining [Figure 5]. The clear cells retained PAS positivity after diastase digestion [Figure 6] with a focal positivity for mucicarmine [Figure 7]. Diagnosis of clear cell variant of intraosseous MEC was confirmed on this basis.

Figure 1: Clinical photograph - extra oral

Figure 2: Clinical photograph - intraoral

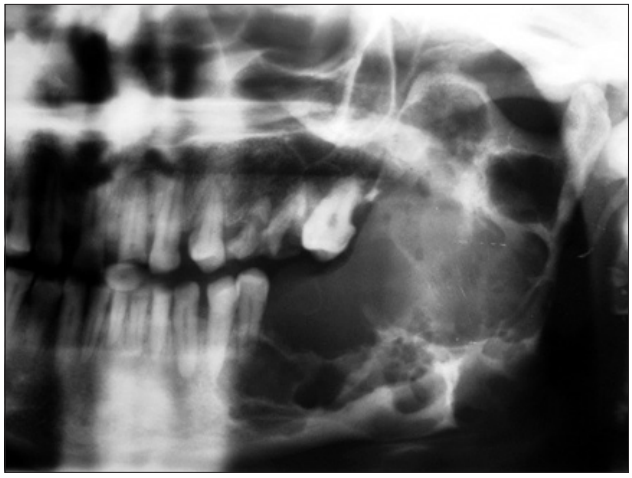

Figure 3: Panoramic view

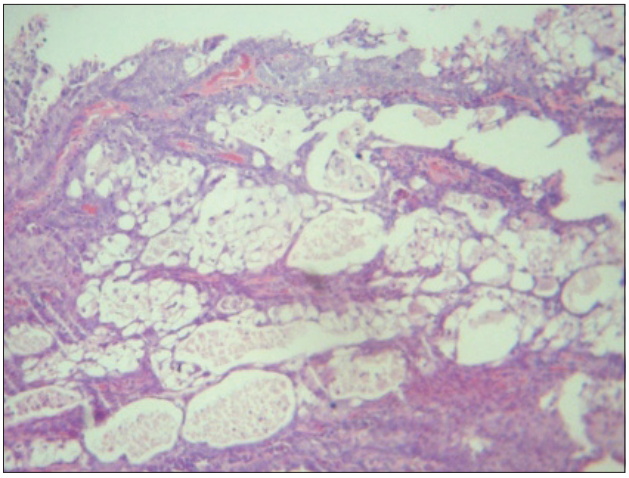

Figure 4: Cyst like areas, epidermoid, and clear cells – H and E, ×100

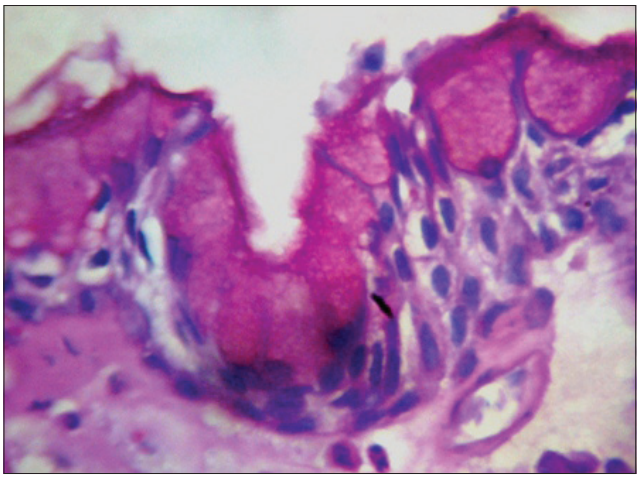

Figure 5: Mucus cells - mucicarmine stain, ×400

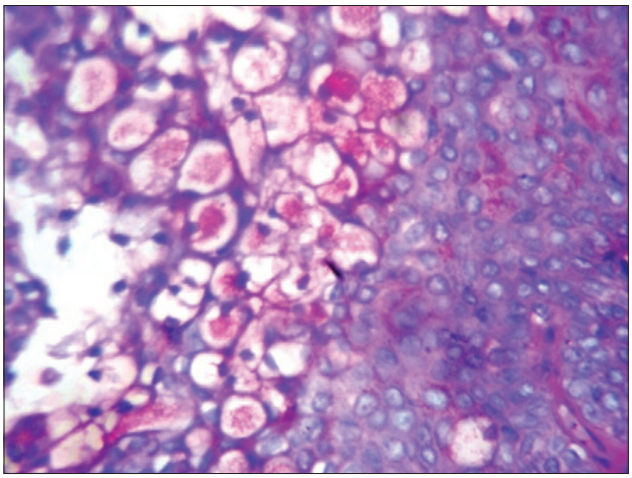

Figure 6: Clear cells - PAS with diastase resistance, ×400

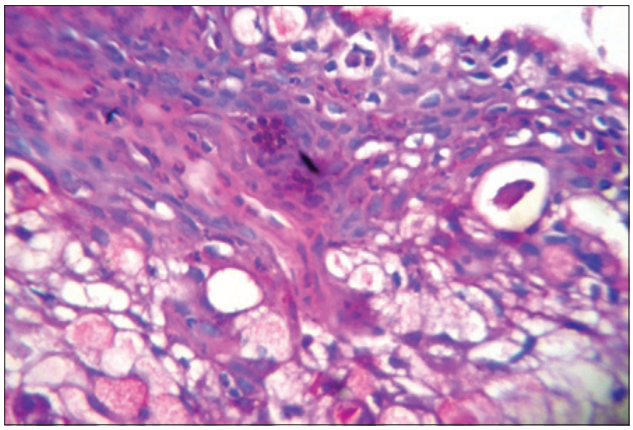

Figure 7: Clear cells - mucicarmine stain, ×400

DISCUSSION

Intraosseous MECs though rare, tend to occur in jaw bones. Mandible is thrice more affected than maxilla.[10,11] Majority of cases occur in the 4th to 5th decades of life.[7,11] The clinical presentation of our case showed the classic features of intraosseous MEC.

The pathogenesis of intraosseous MEC is much debated. It may originate from[11]

- Entrapment of retromolar mucus glands within the mandible which undergo neoplastic transformation
- Neoplastic transformation of mucus secreting cells found in the pleuripotent epithelial lining of dentigerous cysts associated with impacted third molars.
- Developmentally induced embryonic remnants of the submaxillary gland within the mandible.
- Neoplastic transformation and invasion from the lining of maxillary sinus

Diagnostic criteria for intraosseous MEC proposed by Alexander and modified by Browand and Waldron are given in Table 2.[12,13]

In addition to the typical features of MEC such as cystic spaces lined by mucus cells and epidermoid cells, an unusual finding in our case was the presence of clear cells. Clear cells may be a predominant component or rare finding in salivary gland tumors. Clear cells appear as large, polygonal cells with distinct outlines and a hydropic, water clear cytoplasm[9,14] The nuclei are small, vesicular or pyknotic, and centrally placed. The presence of clear cytoplasm can be due to three basic factors. First, due to intracellular accumulation of nonstaining components like glycogen, lipid, or mucin. Second, due to a true scarcity of cytoplasmic organelles, and thirdly due to a fixation artifact.[9,15]

The predominating presence of clear cells in otherwise definable lesions like MECs may lead to histologic misinterpretation.[9,15] This necessitates the consideration of various intraosseous lesions with a clear cell component.

Table 2: Diagnostic criteria for intraosseous MEC (Alexander, modified by Browand and Waldron[12,13])

Intact cortical plate
Radiologic evidence of bone destruction
Histologic confirmation
Positive mucin staining
Absence of primary lesion in the salivary gland
Exclusion of an odontogenic tumor

Differential diagnosis should include metastatic renal cell carcinoma, clear cell odontogenic carcinoma, and clear cell variant of calcifying epithelial odontogenic tumor.[9] Clear cells in metastatic renal cell carcinoma are positive for glycogen and lipid.[16] A diagnosis of renal cell carcinoma can be made only by clinical evaluation of a renal primary tumor.[15] Clear cell odontogenic carcinomas are made up of clear cells of uniform size with a delicate, but well-defined cell membrane. MECs do not contain such a majority of clear cells as in clear cell odontogenic carcinoma.[17] Intraosseous MEC should also be distinguished from cystic primary intraosseous carcinoma where it is a squamous cell carcinoma that demonstrates a cystic component with a lumen-containing fluid or keratin and a stratified squamous epithelium showing cytological atypia.[18] Clear cell variant of calcifying epithelial odontogenic tumor shows a pleomorphic cellular picture with typical calcifications and amyloid formation which are not found in MEC.[19]

Our case fulfilled the diagnostic criteria for intraosseous MECs proposed by Alexander, modified by Browand and Waldron. The histopathologic examination established the presence of PAS positive material in the clear cells and focal positivity for mucin. The patient in this case had a positive history of cyst enucleation in relation to an impacted third molar, 12 years back. In our view, remnants of odontogenic epithelium in the area might have undergone neoplastic transformation giving rise to an intraosseous MEC.

CONCLUSION

The clinical significance of malignant nonodontogenic tumors arising from odontogenic cysts should not be underestimated as demonstrated by the present case. This rare case of intraosseous MEC with abundant clear cells also emphasizes the need for establishment of definitive diagnostic criteria to distinguish the clear cell lesions of oral cavity and jaw bones. The use of special stains also plays an important role in diagnosis of rare lesions like intraosseous MECs showing abundant clear cells.

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