

Case Report

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Central Mucoepidermoid carcinoma of mandible – A case report and review of the literature

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Abstract

Background: Primary central mucoepidermoid carcinoma of jaws is a rare lesion comprising 2–3% of all mucoepidermoid carcinomas reported in literature.

Case presentation: The case presented here illustrates the hypothesis that its specific pathogenesis is unknown.

Conclusions: Mucoepidermoid carcinoma of the jaw is a rare tumour of unknown aetiology. Although about a 100 cases has been reported in literature, the speculation on its aetiopathogenesis has mainly centred on the pluripotential capabilities of the epithelial lining of odontogenic cysts.

Background

Mucoepidermoid carcinoma is usually associated with salivary glands and comprises 5–10% of all salivary gland tumours [1–3]. In 1945, Stewart and associates described its mucous-secreting and epidermal cellular elements thus establishing it as a distinct pathologic entity [4]. Eversole reviewed 815 cases and found that of the major salivary gland tumours, 89.6% involved the parotid, 8.4% submandibular and 0.4% sublingual gland [1]. The palate was the most common site for minor salivary gland involvement, accounting for 41.1% of intraoral lesions [1].

Aberrant salivary gland neoplasms arising within the jaws as primary central bony lesions are extremely rare comprising 2–3% of all mucoepidermoid carcinomas reported [2]. Lepp in 1939 first reported an intraosseous mucoepidermoid carcinoma of the mandible in a 66-year-old woman [5] and Bhaskar [6] in 1963 reported two cases discussing the criteria for their origin, histological com-

position and possible explanations for tumour pathogenesis. In 1991, after a systematic review of its histology and degree of differentiation the WHO classification recommended that the term "mucoepidermoid tumour" be changed to "mucoepidermoid carcinoma" [7]. Waldron and Mustoe [8] suggested that intraosseous mucoepidermoid carcinoma be included in primary intraosseous carcinoma of jaw as type 4 (Table 1) [9]. A thorough review of the English literature revealed about 100 reported cases of mucoepidermoid carcinoma arising in mandible [2,3,10,11]. We report here another case of central mucoepidermoid carcinoma within the bony mandible with no initial connection to alveolus.

Case presentation

A 67-year-old hypertensive woman sought treatment at a local hospital for a painful swelling in the right posterior mandible of three weeks duration. 9-years previously she had undergone tooth extraction and cyst enucleation in

Table 1: Classification of Primary Intraosseous Carcinoma

Type 1: PIOC exodontogenic cyst
Type 2A: Malignant ameloblastoma
Type 2B: Ameloblastic carcinoma arising de novo, exameloblastoma or exodontogenic cyst
Type 3: PIOC arising de novo
a) Keratinising type
b) Nonkeratinising type
Type 4: Intraosseous mucoepidermoid carcinoma.

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the same region. The swelling was diffuse and bony hard involving the ramus and body from the region of the 2nd molar onwards. There was expansion of buccal and lingual cortices and paraesthesia of ipsilateral tongue. Cervical lymphadenopathy was absent. Panoramic radiograph of the mandible revealed an irregular, lobulated, expansile osteolytic lesion 5 cm in size, extending from the mesial aspect of the 2nd molar to the ramus and coronoid process posteriorly (Figure 1).

With a provisional diagnosis of ameloblastoma a hemimandibulectomy was performed. The gross examination of resected specimen revealed a brownish black soft tissue mass expanding both cortices and involving the ramus, coronoid process and body till the region of 2nd molar.

Microscopic examination revealed a neoplasm composed predominantly of cystic spaces and an epidermoid component in a fibrous stroma (Figure 2). The cystic spaces were of varying sizes, lined by mucous secreting cells and cells of intermediate type (figure 3). Nests and strands of squamous cells formed the epidermoid component. Mucinous material was seen within the cystic spaces and extravasating into the stroma (Figure 4).

With a diagnosis of mucoepidermoid carcinoma with predominant mucinous component she was referred to Regional Cancer Centre, Thiruvananthapuram for further management. A histopathological review reconfirmed the diagnosis and patient was treated with adjuvant external beam radiotherapy at a dose of 45 Gy over 20 fractions. She is on regular follow-up and is disease free after 2-years.

Discussion

Central mucoepidermoid carcinoma affects females twice more frequently than males and involves the mandible twice more often than the maxilla [3]. The most common site of occurrence is the premolar-molar-angle region of the mandible [2,3]. It has been reported in all ages ranging from 1 to 78-years, with the overwhelming majority occurring in 4th and 5th decades of life [3]. Eversole *et al.*, [1]

found approximately 50% of the mandibular tumours associated with dental cysts and/or impacted teeth, while Brookstone and Huvos [10] reported a rate of 32%.

In the children the sex ratio is similar to that in adults although the mandible to maxilla ratio is 1:1 [3]. As its occurrence in children is rare, it is unlikely to be a developmental disturbance or a teratoma [3]. The tumour shows a tendency to crop up at puberty, which might suggest a hormonal influence on salivary glands [3].

The main symptoms are swelling and pain with trismus, paraesthesia and tooth mobility being noted occasionally [10]. Previous history of a cyst or impacted tooth gives credence to the theory that odontogenic epithelium is capable of giving rise to mucous secretory cells which may undergo neoplastic transformation to mucoepidermoid carcinoma [12]. The radiographic features are usually a well circumscribed unilocular / multilocular radiolucency [13].

The authenticity regarding its central origin is greater when the tumour is in the mandible, since such lesions in maxilla may actually arise from the submucosal mucous secreting glands in the antrum, or represent intraosseous extension of minor salivary gland tumours of the sinus mucosa [14].

Speculation abounds regarding its pathogenesis and four possible origins have been described [4]. 1) entrapment of retromolar mucous glands within the mandible, which subsequently undergo neoplastic transformation; 2) developmentally included embryonic remnants of the submaxillary gland within the mandible; 3) neoplastic transformation of the mucous secreting cells commonly found in the pluripotential epithelial lining of dentigerous cysts associated with impacted third molars; and 4) neoplastic transformation and invasion from the lining of the maxillary sinus. Our patient gave a prior history of cyst enucleation in the same region as the tumour, which may indicate the possibility of neoplastic transformation of the cyst wall into a malignant nonodontogenic tumour.



Figure 1
OPG of the mandible showing the lytic lesion in the body and ramus of the mandible.

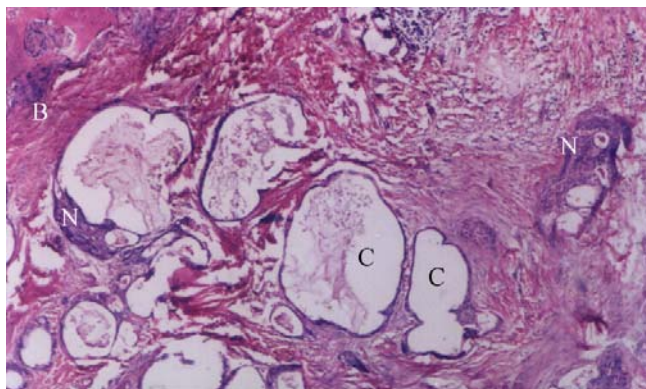


Figure 2
Mucoepidermoid carcinoma, mandible. Cystic spaces (C) and nests of neoplastic squamous (N) cells along with bone spicules at the periphery (B). (H&E \times 50)

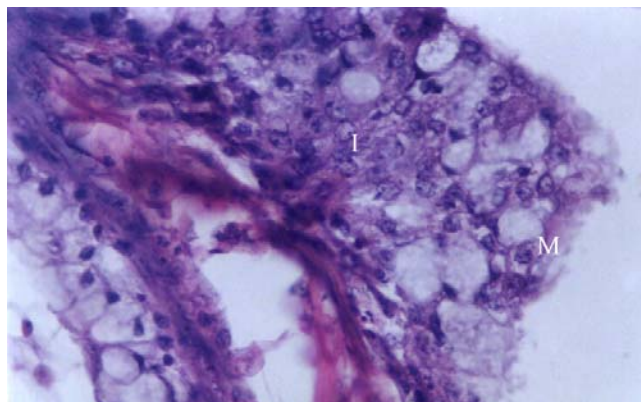


Figure 3
Mucoepidermoid carcinoma. Cystic spaces lined by mucous secreting cells (M) and intermediate cells (I). (H&E \times 400).

Table 2: Diagnostic Criteria

1. Intact cortical plates (However, cortical perforation does not exclude PIOC type 4)
2. Radiographic evidence of bony destruction
3. Exclusion of another primary tumour that in its metastasis could histologically mimic the central tumour
4. Exclusion of an odontogenic tumour
5. Histopathological confirmation
6. Detectable intracellular mucin.

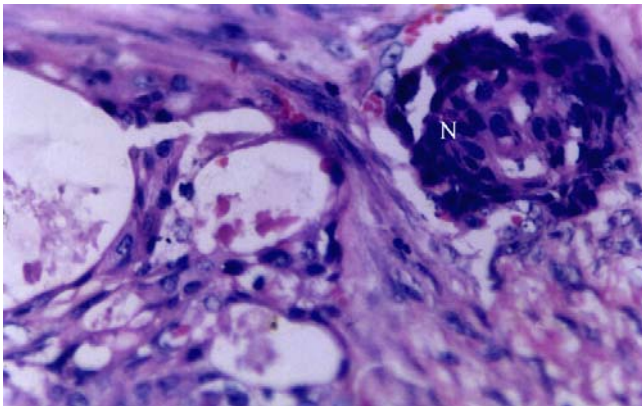


Figure 4
Mucoepidermoid carcinoma. Nest of neoplastic squamous cells (N) along with cystic spaces. (H&E × 400).

The most commonly accepted criteria for diagnosis proposed by Alexander *et al.*, [15] and modified by Browand and Waldron [16] is detailed in table 2. Our case had expanded, yet intact cortices without any radiographic evidence of bone destruction. A thorough search made for a primary tumour elsewhere by detailed clinical and other diagnostic methods proved futile. The histopathological picture was typical of the lesion and intracellular mucin could be detected.

Brookstone and Huvos [10] had put forward a staging system based on condition of the overlying bone. Lesions with intact cortical plates with no evidence of bony expansion offer the best prognosis and indicate stage I disease. Stage II disease is surrounded by intact cortical bone that has undergone some degree of expansion. Any instance of cortical perforation, breakdown of the overlying periosteum or nodal spread is best categorized clinically as stage III disease. The case presented here showed expanded, but intact buccal and lingual cortices and could be imputed as stage II disease. Metastases are reported in 9% of central mucoepidermoid carcinomas mainly to the regional

lymph nodes [10,13] and occasionally to the ipsilateral clavicle, [17] lung and brain.

Surgery is the mainstay of treatment. In a review of 64 patients, Brookstone and Huvos observed 40% recurrences after conservative surgical modalities like curettage, enucleation, marsupialization and marginal resection with or without adjuvant therapy, whereas in the group treated by radical methods such as segmental resection with / without treatment of associated neck and / or adjuvant therapy only 4% recurred [10]. Adjuvant radiotherapy is recommended for high-grade tumours [13].

The clinical significance of malignant tumours arising from odontogenic cysts or *de novo* should never be underestimated as illustrated by the present case. This re-emphasises the importance of radical surgery, adjuvant treatment and a careful histopathological evaluation of all excised tissue so that such neoplastic transformation may be identified and treated effectively.

Competing interests

None declared.

Author contributions

DS participated in the preparation of the manuscript and literature search. TS carried out the histopathological studies, participated in the preparation of manuscript and approval of final version; MP carried out the literature search, edited the manuscript for its scientific content and prepared the final version of the manuscript; RD participated in the editing of the manuscript and preparation of final version of manuscript.

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