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Sclerosing Central Mucoepidermoid Carcinoma: Rare Case Series and Review

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Mucoepidermoid carcinoma (MEC) comprises around 30% of all salivary gland malignancies, making it a preeminent threatening tumour of the salivary glands. Numerous histologic variants with a great extent of separation have been portrayed. Sclerosing MEC (SMEC) has been portrayed as an uncommon subtype, that can be misdiagnosed as a generous receptive condition or low-grade non-SMEC malignancy. The sclerosing variant of central or intraosseous MEC is extremely rare and no cases are reported to date. We report 2 cases of Sclerosing central MEC, in which histologic examination demonstrated relatively nicely-circumscribed, nonencapsulated tumours composed of significant valuable sclerosis and scattered epithelial islands of low-grade MEC. Within the 2nd case, the tumour confirmed comparable sclerotic stroma; but the epithelial component became of intermediate grade. A Mayer mucicarmine stain and PAS stain were fine in each case and discovered plentiful intracytoplasmic mucin. An analysis of sclerosing crucial mucoepidermoid carcinoma was made. A whole resection of the tumour was executed in both instances and remained sickness free to date.

Keywords: Mucoepidermoid carcinoma; sclerosing variant; histopathology; salivary gland.

1. INTRODUCTION

Mucoepidermoid carcinoma is the most typical exocrine gland malignancy, bills for approximately 34% of the malignant epithelial salivary gland tumors which turned into first defined by Volkmann in 1895 [1]. Of this, the central mucoepidermoid carcinoma incorporates

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3-4% of all MECs and manifests with an unknown pathogenesis. A few subtypes of MEC are stated, which include unicystic, oncocytic, clear cellular, and sclerosing. Among these, the sclerosing version is an extremely rare entity, first identified with the aid of Chan and Saw in 1987 [2,3]. Characteristically, an excessive sclerotic stroma is present in the tumor mass which may also obscure their regular morphologic functions and bring about diagnostic difficulties [1]. To date, no sclerosing version of mucoepidermoid carcinoma has been published in the literature. This case series files rare cases of sclerosing variation of central mucoepidermoid carcinoma.

2. CASE REPORT

2.1 Case I

A 35‑year‑old male patient reported to the outpatient department complaining of a painless swelling in the upper right posterior region of the jaw since one and half months. On intraoral examination, a solitary enlargement on the buccal and palatal alveolus in relation to the right posterior maxilla was identified. The swelling was uneven in shape, reaching from the alveolar area of 14 to the tuberosity of the maxilla, obliterating the buccal vestibule somewhat. A modest swelling was seen on the palatal mucosa that extended to the midline, and the adjacent palatal mucosa was erythematous. The swelling was non-tender, firm, nonvariant, immobile, compressible, and not reducible when palpated. Orthopantamogram revealed a single, well defined, multicystic, irregular corticated lesion involving upper right 16, 17 ,18 and tuberosity of the maxilla of size approximating 6 cm × 5 cm, resulting in the destruction of the right maxillary process involving maxillary sinus. Computed Tomography Scan revealed an evidence of expansile lytic lesion involving alveolar process of maxilla on the right side with soft tissue component filling right maxillary sinus, suggestive of a destructive lesion.

Based on the clinical and radiological examination, a provisional diagnosis of salivary gland malignancy, odontogenic tumour or connective tissue malignancy was considered. After an incisional biopsy, diagnosis of MEC was established and the patient was subjected to surgery with his consent. During the surgery, a large cystic lesion was observed, containing large cystic spaces filled with mucoid material. Complete removal of the lesion was performed, and sent to the Department of Oral Pathology for confirmation of diagnosis.

On gross examination, the lesion was a resected specimen involving the maxillary sinus obtained from the right side of the maxilla. The sliced surface was heterogeneous, with some well-defined cystic spaces filled with mucin and some firm areas (See Fig. 1).

The tissue was subjected to routine tissue processing procedure and stained with haemotoxylin and eosin. The histopathological examination revealed lesional glandular tissue with numerous cystic spaces filled with mucin. The cystic spaces were lined by numerous clear cells and mucous cells against the sclerotic background stroma (Fig. 2). The connective tissue stroma also comprised of a few areas of dispersed nests and groups of intermediate cells and very few epidermoid cells along with irregularly arranged dense bundles of collagen fibres exhibiting areas of hyalinization and the presence of reactive bone formation in many areas. The tumor nests cells displayed clear to eosinophilic cytoplasm and well‑defined cytoplasmic membranes. The nests were surrounded by extensive hyalinized stromal sclerosis without any lymphocytic infiltration.

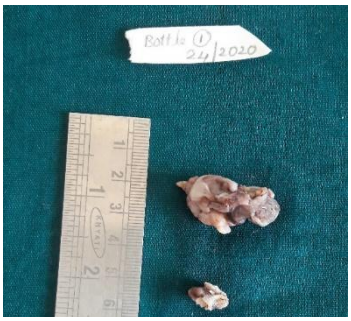

Fig. 1. On gross examination, the lesion was a solid mass which was firm in consistency

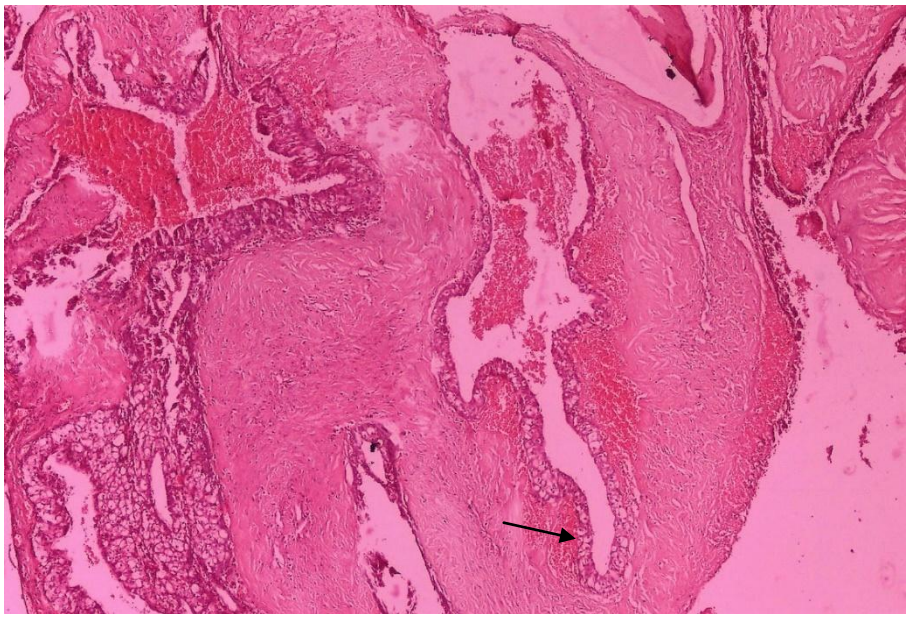

Fig. 2. Hematoxylin and eosin-stained section revealed lesional glandular tissue with numerous cystic spaces filled with mucin. The cystic spaces were lined by numerous clear cells and also mucous cells against the sclerotic background stroma (10x magnification)

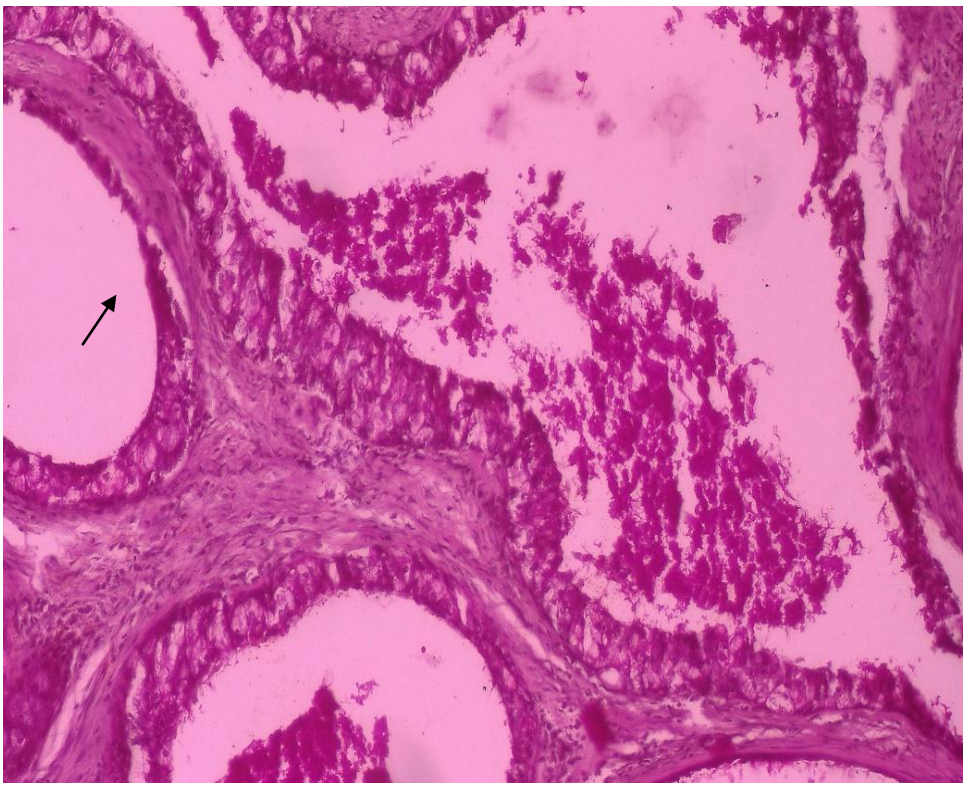

Fig. 3 The mucin filled cystic spaces and mucous cells showed positivity for PAS (40x magnification)

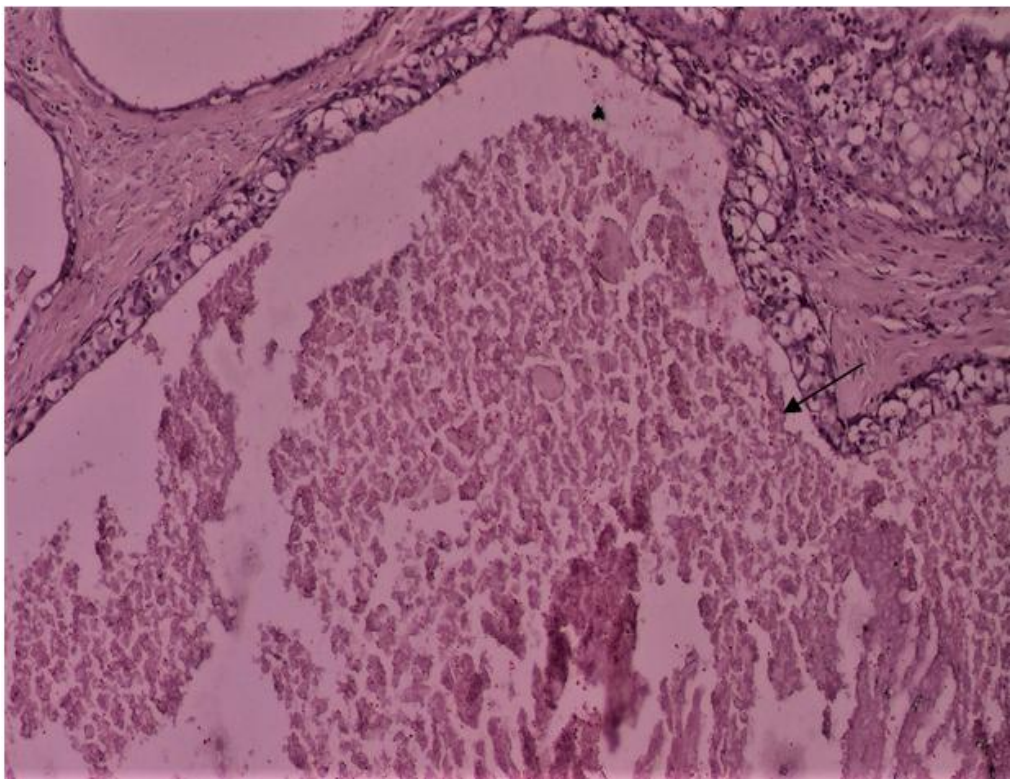

Fig. 4. The mucin filled cystic spaces and mucous cells showed positivity for mucicarmine (20X magnification)

Over 60% stroma of the examined tissue section was hyalinized. No evidence of perineural invasion or necrosis was seen. The mucin-filled cystic spaces and mucous cells showed positivity for PAS and mucicarmine (Fig. 3 & 4). Thus, based on the compilation of these histological features, the reported case was categorized as a Sclerosing variant of low-grade Central Mucoepidermoid Carcinoma.

2.2 Case II

A 49-year-old female patient reported to the outpatient department complaining of a swelling in the lower right posterior region of the jaw for the past 2 years. On examination, intraorally, a solitary swelling was noted on the buccal alveolus with respect to the right posterior mandible and measured about 3 x 3 cm in diameter. The swelling was irregular in shape, extending from 46 to the ascending part of the ramus. On palpation, the swelling was tender. The submandibular lymph nodes in association with the swelling were soft, mobile and palpable. Orthopantomogram revealed a multilocular radiolucency with ill-defined borders, extending anteroposteriorly from 46 to the ascending part of the ramus, and superioinferiorly from the upper border of the mandible till the mandibular canal (Fig. 6). Computed Tomography scan revealed an evidence of buccal and lingual cortical perforation and bone marrow space involvement approximating mandibular canal.

Based on the clinicopathological examination, provisional diagnosis of osteolytic granulomatous lesion or odontogenic cyst was considered. After an incisional biopsy, diagnosis of MEC was established and the patient was subjected to surgery with her consent. During the surgery, a large tumor mass with a few cystic spaces containing mucoid material was observed. Complete removal of the lesion was performed and sent to the Department of Oral Pathology for confirmation of diagnosis.

On gross examination, the lesion was a solid mass which was firm in consistency exhibiting few mucin filled cystic spaces (Fig. 5).

The tissue was subjected to routine tissue processing procedure and stained with haemotoxylin and eosin. Histopathological examination revealed an unencapsulated lesional tissue with few nests of tumour cells and multiple cystic spaces filled with mucin dispersed

sclerotic background stroma. The cystic spaces were lined by numerous clear cells and mucous cells (Fig. 7). Tumor nests comprising of mucous cells, intermediate cells and epidermoid cells with intervening hyalinized area were also observed. Connective tissue was densely collagenous with irregularly arranged bundles of collagen fibers, also exhibiting areas of hyalinization and focal aggregates of chronic inflammatory cells. (Fig. 8) The lesional glandular tissue with a few cystic spaces filled with mucin showed positivity for PAS and mucicarmine (Fig. 9& 10). The described case was classified as a Sclerosing variant of low grade Central Mucoepidermoid Carcinoma due to the presence of certain histological features.

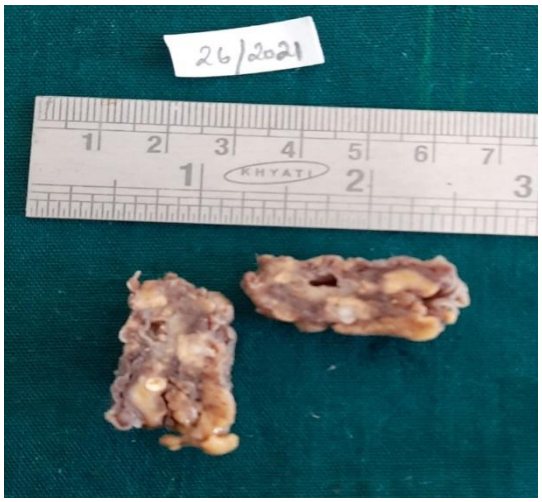

Fig. 5. On gross examination, the lesion was a solid mass which was firm in consistency with cut surface showing cystic spaces

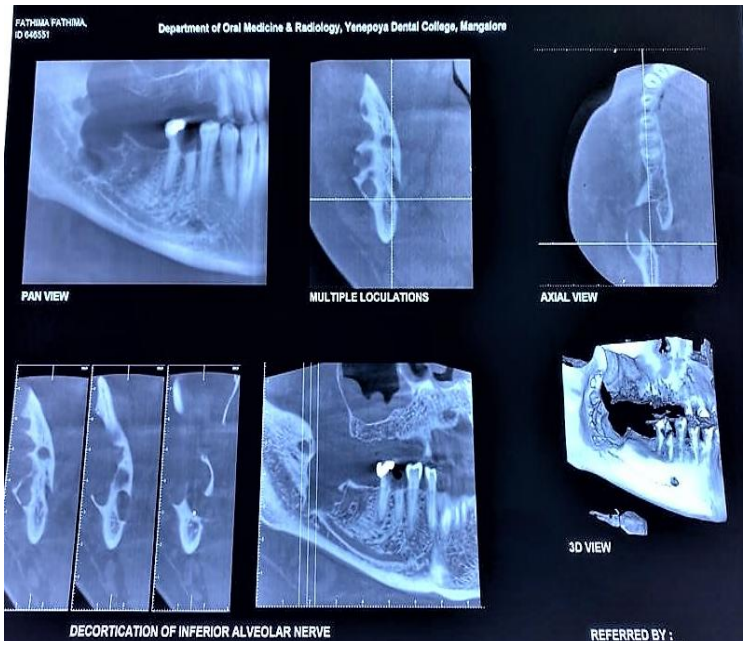

Fig. 6. Orthopantomogram revealed a multilocular radiolucency with ill-defined borders extending anteroposteriorly from 46 to the ascending part of the ramus, and superioinferiorly from the upper border of the mandible till the mandibular canal. Computed Tomography scan revealed an evidence of buccal and lingual cortical perforation and bone marrow space involvement approximating mandibular canal

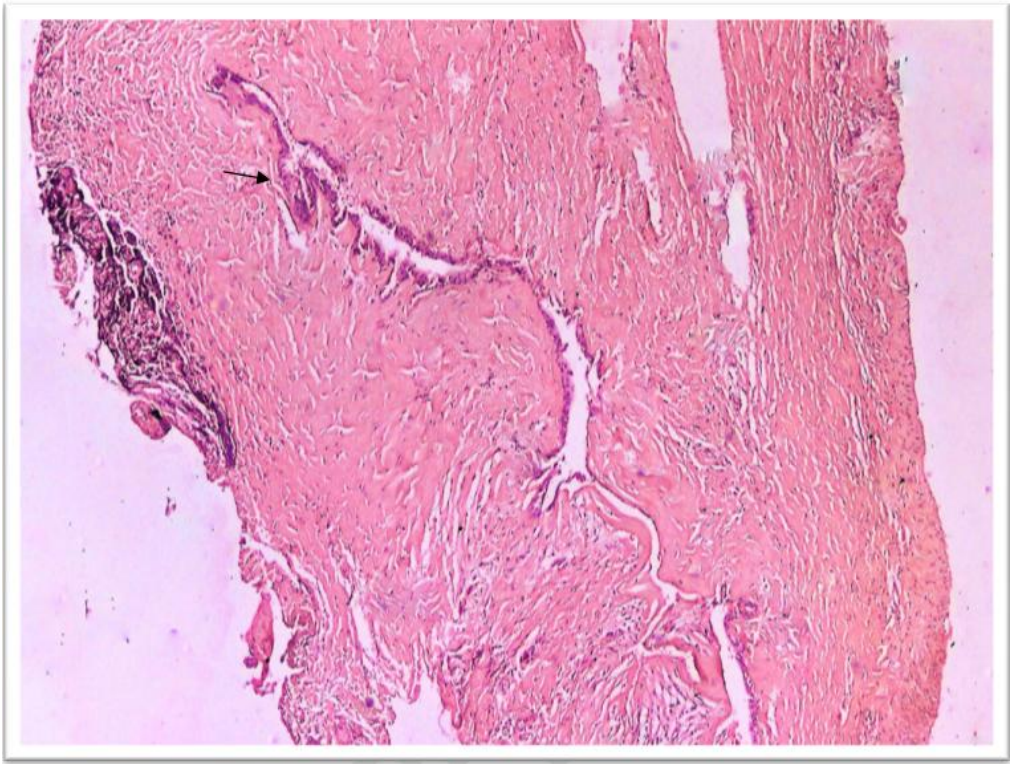

Fig. 7. Hematoxylin and eosin-stained section revealed unencapsulated lesional tissue with few nests of tumour cells and multiple cystic spaces filled with mucin dispersed sclerotic background stroma. (10x magnification)

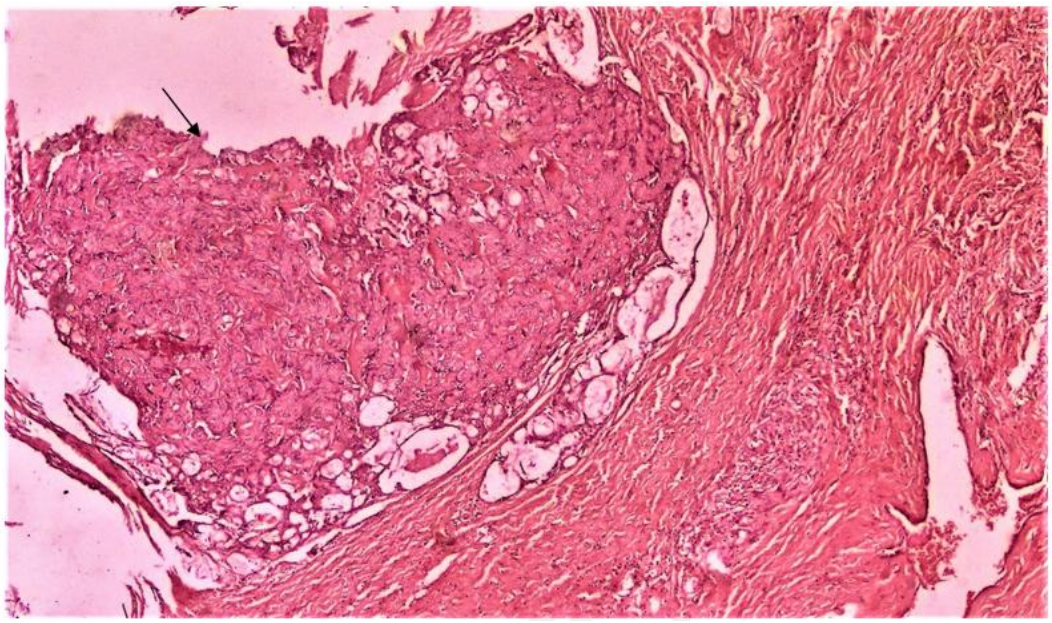

Fig. 8. Hematoxylin and eosin-stained section revealed tumor nests comprising of mucous cells, intermediate cells and epidermoid cells with intervening hyalinized area were also observed. Connective tissue was densely collagenous with irregularly arranged bundles of collagen fibres, also exhibiting areas of hyalinization and focal aggregates of chronic inflammatory cells

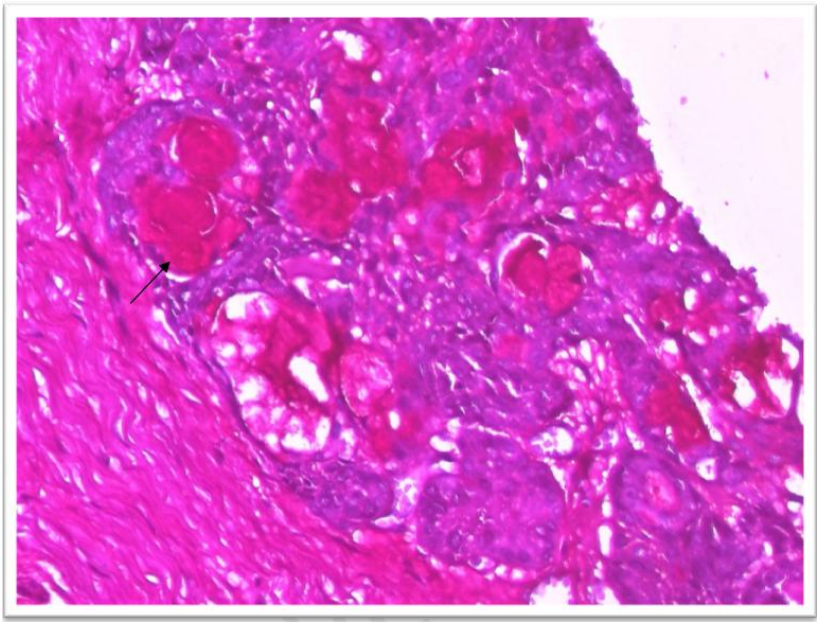

Fig. 9. The mucin filled cystic spaces and mucous cells showed positivity for PAS (40x magnification)

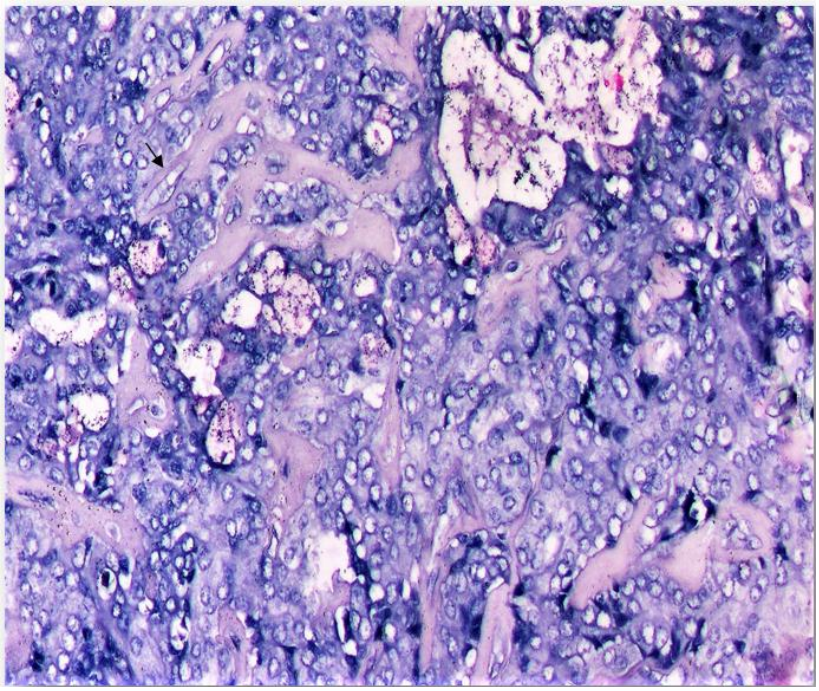

Fig. 10. The mucin filled cystic spaces and mucous cells showed positivity for mucicarmine (40x magnification)

3. DISCUSSION

Mucoepidermoid carcinoma incorporates 16% of all salivary gland tumors and about 30% of all salivary gland malignancies [2]. It is typically visible in women, with the most occurrence in the third and sixth decade of life [4]. The lesion commonly demonstrates particularly variable clinical conduct starting from gradual to indolent to domestically competitive and particularly metastatic tumors. Radiographic appearances in large part, rely upon the grade and manifests as

a radiolucent lesion. Histologically, MEC is characterised through 3 primary cell types: Epidermoid, mucin-generating and intermediate cells originating from the epithelial lining of ducts and is graded into low, intermediate and high grades primarily based on the larger population of a cell type [1,2].

3-5% of Mucoepidermoid Carcinoma comprises of the central variant and is therefore, a rare occurence. Lepp in 1939 reported the first case of Central Mucoepidermoid Carcinoma of the mandible [1]. It's seen generally in the mandible (82%), having a female predominance and unknown pathogenesis. There's no definitive theory about the pathogenesis of central MEC. Several hypotheses are described, including (1) mucous metaplasia and neoplastic metamorphosis of the epithelial lining of an odontogenic cyst; (2) embedding of the submandibular, sublingual, or retromolar mucous glands during embryonic development within the mandible, which latter transforms to neoplastic metamorphosis (3); iatrogenic embedment of minor salivary glands; (4) neoplastic metamorphosis of maxillary sinus epithelium; and (5) remnants of the dental lamella [5].

The criteria for the diagnosis of central MEC include cortical bone devoid of perforation by growth of the mass, radiological substantiation of bone destruction, and histopathological verification. Radiographic expression of central MEC constantly shows bone destruction with a multilocular or cyst-like radiolucent appearance. Also, this mass has the capacity to develop hard tissue and be expressed as a mixed lesion [5,6].

Due to the considerable variation in the type, distribution, and growth pattern of MEC cells, the histopathologic appearance of MEC will differ and show as conventional, sclerotic, unicystic, oncocytic, sebaceous, clear cell, spindle, and psammomatous types [2,7-11]. Amongst these, the sclerosing type of MEC is an extremely rare phenomenon, more so when in combination with the central variant and in this case series, two sclerosing central mucoepidermoid cases is described at maxilla and mandible.

As its name suggests, SMEC is characterized by a strong central sclerosis that occupies the summation of an otherwise typical excrescence, mostly with a seditious insinuate of plasma cells, eosinophils, and/ or lymphocytes at its supplemental regions [12,8]. The possible pathogenic mechanisms causing this type of sclerosis are tumour infarction and mucin extravasation [4]. The mucin acts as a foreign material, ending in fibrosis that forms as an attempt to circumscribe-off the mucin.

Diagnostic challenges may be exacerbated by the sclerosis associated with these excrescences, which may hide their usual morphologic traits. Sclerosing polycystic adenosis, hyaline clear cell carcinoma, malignant mixed tumour, sclerosing sialadenitis, and polymorphous low-grade adenocarcinoma are some of the prominent salivary gland lesions that demonstrate analogous sclerotic stroma [13,9]. Of all the histological features observed in SMEC, a central keloid-like sclerosis rimmed by supplemental lymphoid infiltration is unique enough to distinguish SMEC from the other sclerotic salivary lesions [13]. In our case series, the coupling of such a sclerosing pattern in a central variant was an unusual spectacle that was observed. Neck dissection is recommended in all cases except in those of low-grade small tumours [14]. Prognosis is subject to grade with low grade tumours having 90-98% survival, and low recurrence rate, compared to 30-54% surviving and a veritably high regional recurrence rate for high grade tumours [14,15,8].

4. CONCLUSION

SMEC remains a rare variant of MEC that can mimic benign conditions. Although majority cases are low grade tumours, histologic grading should always be done for prognostic purposes and possible adjuvant remedy since Central Mucoepidermoid Carcinoma of the mandible occurs infrequently and can be misdiagnosed radiographically.

Late recurrences and metastases are common; hence, prolonged follow-up is needed. In our study, both cases haven't reported with any recurrences till date and are under follow up.

CONSENT

Verbal consent was obtained from the patient.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Bhat K, Pandey B, Shetty P, Manohar V, Shruthilaxmi MK, Patidar M. Sclerosing mucoepidermoid carcinoma: a unique case. Sultan Qaboos University Medical Journal. 2014;14(2): e249.
2. Veras EF, Sturgis E, Luna MA. Sclerosing mucoepidermoid carcinoma of the salivary glands. Annals of Diagnostic Pathology. 2007;11(6):407-12.
3. Ide F, Horie N, Shimoyama T, Saito I. Sclerosing mucoepidermoid carcinoma: specific histologic variant or nonspecific morphologic pattern? Oral Medicine & Pathology. 2011;15(2):53-5.
4. Mardi K, Madan S. Sclerosing mucoepidermoid carcinoma of the submandibular gland: report of two rare cases. Clinical Cancer Investigation Journal. 2012;1(2):86.
5. Sepulveda I, Frelinghuysen M, Platin E, Spencer ML, Compan A, Munzenmayer J, Ulloa D. Mandibular central mucoepidermoid carcinoma: A case report and review of the literature. Case reports in oncology. 2014;7(3):732-8.
6. Brandwein MS, Ivanov K, Wallace DI, Hille JJ, Wang B, Fahmy A, Bodian C, Urken ML, Gnepp DR, Huvos A, Lumerman H. Mucoepidermoid carcinoma: a clinicopathologic study of 80 patients with special reference to histological grading. The American journal of surgical pathology. 2001;25(7):835-45.
7. Heavner SB, Shah RB, Moyer JS. Sclerosing mucoepidermoid carcinoma of the parotid gland. European Archives of Oto-Rhino-Laryngology and Head & Neck. 2006;263(10):955-9.
8. Simon D, Somanathan T, Ramdas K, Pandey M. Central Mucoepidermoid carcinoma of mandible–A case report and review of the literature. World Journal of Surgical Oncology. 2003;1(1):1-5.
9. Rasul U, Bradish T, Bashir MT, Shakeel M. Sclerosing variant of mucoepidermoid carcinoma: a diagnostic challenge. BMJ Case Reports CP. 2020;13(10): e236509.
10. Brandwein M, Hille J, Gnepp D, Urken ML, Ivanov K. The many faces of mucoepidermoid carcinoma. AJSP: Reviews & Reports. 2000;5(4):214-20.
11. Batsakis JG, Luna MA. Histopathologic grading of salivary gland neoplasms: I. Mucoepidermoid carcinomas. Annals of Otology, Rhinology & Laryngology. 1990;99(10):835-8.
12. Sinha SK, Keogh IJ, Russell JD, O'Keane JC. Sclerosing mucoepidermoid carcinoma of minor salivary glands: a case report. Histopathology. 1999;35(3) :283-4.
13. Suzuki M, Ichimiya I, Matsushita F, Mogi G. Histological features and prognosis of patients with mucoepidermoid carcinoma of the parotid gland. The Journal of Laryngology & Otology. 1998;112(10): 944-7.
14. Zhou CX, Chen XM, Li TJ. Central mucoepidermoid carcinoma: a clinicopathologic and immunohisto chemical study of 39 Chinese patients. The American Journal of Surgical Pathology. 2012;36(1):18-26.
15. Fadare O, Hileeto D, Gruddin YL, Mariappan MR. Sclerosing mucoepidermoid carcinoma of the parotid gland. Archives of pathology & laboratory medicine. 2004;128(9):1046-9.

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