Sclerosing mucoepidermoid carcinoma of minor salivary gland

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Abstract

Sclerosing mucoepidermoid carcinoma (SMEC) is extremely rare variant of the mucoepidermoid carcinoma, which is the most common primary malignancy of the salivary glands. As its name suggests, SMEC is characterized by an intense central sclerosis that occupies the entirety of an otherwise typical tumor, frequently with an inflammatory infiltrate of plasma cells, eosinophils, and/or lymphocytes at its peripheral regions, but its uncompanionship with inflammatory cell infiltration might explain its progressive stage of the sclerosis. The sclerosis associated with these tumors may obscure their typical morphologic features and result in diagnostic difficulties. Tumor infarction and extravasation of mucin eventuating in reactive fibrosis are two mechanisms of formation that have been suggested as underlying this morphologic variant. Morphologic evidence in support of the mucin extravasation hypothesis was identified, as small pools of mucin were present throughout the tumor.

Keywords: Inflammatory cell infiltration, mucin extravasation, mucoepidermoid carcinoma, sclerosing variant, tumor infarction

Introduction

Mucoepidermoid carcinoma is the most common malignancy of the salivary gland, accounts for about 34% of the malignant epithelial tumor.[1] More than eight potential variants of mucoepidermoid carcinoma have been introduced,[2] which includes unicystic, sclerosing, oncocytic, clear cell, spindle cell, goblet cell aggressive, psammomatous and sebaceous types. However, sclerosing mucoepidermoid carcinoma (SMEC) is an extremely rare morphologic variant. It has been described almost exclusively in the major salivary glands, rarer in minor salivary glands. It epitomizes the stromal fibrosis and hyalinization accompanied by a dense lymphoplasmacytic infiltrate with or without eosinophilia.[2] Until date, only 20 cases were reported in the literature. This case report documents an unusual case of SMEC without lymphoplasmacytic cells arising from minor salivary glands.

Case Report

The 45-year-old female complained of painless swelling in the upper right posterior region of the jaw since 1-year [Figure 1]. An intraoral clinical examination revealed a single diffuse swelling on right retro molar region extending toward ramus and right posterior part of the hard palate and soft palate up to right lateral pharyngeal wall [Figure 2]. Three months back diffuse extraoral swelling appeared, which gradually increased and measured approximately 4 cm × 4 cm [Figure 3]. Two bilateral submandibular lymphnodes were palpable of size approximately 0.5 cm × 0.5 cm, roughly oval, firm, mobile and nontender on palpation. Orthopantogram revealed a single well defined multicystic irregular corticated lesion involving lower right 6, angle and coronoid process of ramus of mandible of size approximately 4 cm × 5 cm, resulting in the destruction of coronoid process of the mandible [Figure 4]. Computed Tomography Scan revealed an evidence of expansile lytic lesion with multiple thin bony separations suggestive of a destructive lesion [Figures 5,6,7].

Based on the clinicoradiological examination, provisional diagnosis of salivary gland malignancy, odontogenic tumor or the connective tissue malignancy was hypothesized. After an incisional biopsy patient was subjected to surgery with her consent. During the surgery a large cystic lesion was observed, contains large cystic spaces full of mucoid material. Hemimandibulectomy of the right side was performed, and the patient was kept under long-term follow-up [Figure 8]. After 6 and 12 months follow-up no abnormality was detected.

On gross examination, resected specimen retrieved from the right side of the mandible involving half of the posterior 2/3rd of the body of the mandible to the right condyle. The cut surface showed heterogeneity with some well demarcated cystic area filled with mucin and firm areas at places.
Microscopically, examined hematoxyline and eosin stained tissue section was showing areas of sheets of epidermoid cells admixed with some cystic structures [Figures 9 and 10]. Small pools and droplets of mucicarmine, periodic-acid Schiff 1, Alcian blue positive [Figures 11-13] mucin were identified throughout the section.

The tumor nests cells displayed clear to eosinophilic cytoplasm, well-defined cytoplasmic membranes, inconspicuous intercellular bridges, and only minimal nuclear pleomorphism, with prominent nucleoli and few abnormal mitotic figures [Figures 14 and 15]. At places, some mucous cells as well as intermediate cells were visible within the sheets of epidermoid cells which were highlighted by the mucin specific stains [Figures 11-13]. The nests were surrounded by extensive hyalinized stromal sclerosis devoid of lymphocytic infiltration [Figures 9 and 16]. More than 60% stroma of the examined tissue section was hyalinized. No evidence of perineural invasion or necrosis. Thus on the consolidation of these histological features the reported case was classified as a Sclerosing variant of Intermediate grade Mucoepidermoid carcinoma.
The sclerosing variant is an extremely rare entity. Among total 20 reported cases till date, the first case was reported by Chan and Saw, showing a dense infiltrate of lymphocytes, plasma cells and immunoblasts at the periphery of the tumor.\cite{3} Prior to our current case, only two cases were documented in the literature by Sinha et al. and Fadare et al. with no inflammatory infiltrate.\cite{4} Suggestive possibility might be that a temporal evolutionary spectrum is operational with those cases without an inflammatory infiltrate representing tumors at a later stage of the process. Prior to this case, only one case of SMEC of minor salivary glands (parapharyngeal space) of a 65/M patient in 1999 had been reported by Sinha et al. but the present case must be the first case of SMEC of minor salivary gland of the retromolar area devoid of inflammatory infiltrate makes it unique in itself.
There is an interesting debate regarding the histogenesis of SMEC.\textsuperscript{[4,5,6]} Reactive fibrosis in this variant might be the result of two possible mechanisms. (i) Tumor infarction and (ii) host tissue reaction eventuating due to extravasated mucin pools.\textsuperscript{[7-9]} Very often this pronounced desmoplasia
in the salivary gland malignancy may be interpreted as an exaggerated mucous escape reaction. Morphologic evidence in support of the mucin extravasation hypothesis is present in this case, as small pools of mucin were identified throughout the tumor.

Several conditions of the salivary glands may manifest extensive stromal sclerosis or fibrosis, including sclerosing polycystic adenosis, hyalinizing clear cell carcinoma, mixed tumors, sclerosing sialadenitis and polymorphous low-grade adenocarcinoma which might be a host tissue response to the tumor infarct or due to extravasated mucin pools into the connective tissue stroma.

**Conclusion**

This is a rare case of infrequent subtype of SMEC. It may be misdiagnosed as several above mentioned other benign conditions of the salivary gland. A larger study is required to determine whether this pronounced desmoplasia reflects simply secondary defensive response to tumor growth or its sometimes association with lymphoplasmacytic infiltration represents an important background lymphoepithelial lesion. Uncompanion of SMEC with inflammatory infiltrate as in our present case might reflect late progressive stage of the tumor. It is not known if the presence of sclerosis provides any prognostic information outside of that provided by traditional, albeit controversial, histopathologic grading schemes for mucoepidermoid carcinoma. To comment on the prognostic factor of SMEC as a separate entity is of little significance due to inadequacy of the literature and rarity of this variant. Multivariate analyses with a larger number of cases are required truly to evaluate whether the intense sclerosis observed in these cases emerge as an independent prognostic factor.

Complete surgical resection with the tumor-free surgical margin adjunct with the postoperative radiation therapy is of great significance. Patient should follow-up regularly with the serial magnetic resonance imaging studies of the tumor bed altogether with the clinical evaluation for the regional lymphadenopathy and chest to predict the prognostic factor, rate of recurrence and to determine the metastasis.

**References**

1. Kayal L, Jayachandran S, Niranjena PA. A rare case of sclerosing mucoepidermoid carcinoma of minor salivary gland - A diagnostic enigma. Int J Dent Case Rep 2011;1:43-8.
2. Ide F, Horie N, Shimoyama T, Saito I. Sclerosing mucoepidermoid carcinoma: Specific histologic variant or nonspecific morphologic pattern? Oral Med Pathol 2010;15:53-5.
3. Chan JK, Saw D. Sclerosing mucoepidermoid tumour of the parotid gland: Report of a case. Histopathology 1987;11:203-7.
4. Fadare O, Hileeto D, Gruddin YL, Mariappan MR. Sclerosing mucoepidermoid carcinoma of the parotid gland. Arch Pathol Lab Med 2004;128:1046-9.
5. Ide F, Obara K, Enatsu K, Mishima K, Saito I. Sclerosing mucoepidermoid carcinoma of the oral cavity. J Oral Pathol Med 2005;34:187-9.
6. Mardi K, Madan S. Sclerosing mucoepidermoid carcinoma of the submandibular gland: Report of two rare cases. Clin Cancer Invest J 2012;1:36-8.
7. Shinhar SY. Sclerosing mucoepidermoid carcinoma of the parotid gland: case report. Ear Nose Throat J 2009;88:E29-31.
8. Muller S, Barnes L, Goodurn WJ Jr. Sclerosing mucoepidermoid carcinoma of the parotid. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1997;83:685-90.
9. Urano M, Abe M, Horibe Y, Kuroda M, Mizoguchi Y, Sakurai K, et al. Sclerosing mucoepidermoid carcinoma with eosinophilia of the salivary glands. Pathol Pract 2002;198:305-10.

**How to cite this article:** Lohiya PG, Chaudhary MS, Patil S, Agrawal SA. Sclerosing mucoepidermoid carcinoma of minor salivary gland. Contemp Clin Dent 2014;5:564-8.

**Source of Support:** Nil. **Conflict of Interest:** None declared.