

Sclerosing mucoepidermoid carcinoma of the submandibular gland: Report of two rare cases

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ABSTRACT

Although mucoepidermoid carcinoma is the most common primary malignancy of the salivary glands, the sclerosing morphologic variant of this tumor is extremely rare, with only 15 reported cases. As its name suggests, sclerosing mucoepidermoid carcinoma 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. The sclerosis associated with these tumors may obscure their typical morphologic features and result in diagnostic difficulties. Two cases of mucoepidermoid carcinoma of the submandibular gland associated with extensive central sclerosis and peripheral lymphoid response are reported. This unusual but distinctive variant of mucoepidermoid carcinoma can be difficult to recognize and may be confused with chronic sialoadenitis or even metastasis to an intra-parotid lymph node.

Key words: Mucoepidermoid carcinoma, sclerosing variant, submandibular gland

INTRODUCTION

Sclerosing variant of mucoepidermoid carcinoma (SMEC) is a rare salivary gland tumor with only a few cases on record so far.^[1,2] Their occurrence in submandibular gland is still rarer with only one case reported so far. We herein report two such rare occurrences and review the pertinent literature.

CASE REPORTS

Case 1

A 58-year-old male presented with swelling below right side of mandible, which he noticed long before and was rapidly increasing in size since 4 months. On examination, there was a firm to hard, nontender swelling, measuring 7 × 4 cm in size situated over right mandible extending in to right submandibular region and posteriorly behind the right angle of the mandible.

The swelling was fixed to the underlying structures. In addition, there were multiple enlarged right cervical lymph nodes. CT scan revealed a heterogenous mass in the right submandibular region measuring 5.6 × 5 × 4.6 cm in size extending medially in to parapharyngeal space, posteriorly in to carotid space, anteromedially in to sublingual space and superiorly in to infratemporal fossa. Decision was taken to excise the mass. Intraoperatively, there was a mass in the right submandibular region, measuring 8 × 6 cm superiorly reaching up to the masseter muscle, inferiorly involving the tendon of digastrics muscle and also involving right hypoglossal, lingual, spinal and accessory nerves. Multiple lymph nodes at levels I to IV levels on right side of neck were also resected. On gross examination, the resected submandibular gland was measuring 9 × 5 × 3 cm. Cut surface revealed a well-demarcated yellowish growth measuring 2.8 × 2.3 cm in size. On microscopic examination, there was central area of extensive sclerosis with keloid-like stroma and an inflammatory infiltrate of lymphocytes and plasma cells along with tumor nests at the peripheral regions [Figures 1 and 2]. The cellular peripheral area showed islands of tumor cells composed of epidermoid, mucin-secreting and intermediate cells with evidence of neural or perineural invasion. Keeping in view these characteristic findings, the tumor was diagnosed as sclerosing variant of mucoepidermoid carcinoma, high grade. All the resected cervical lymph nodes revealed metastatic tumor deposits.

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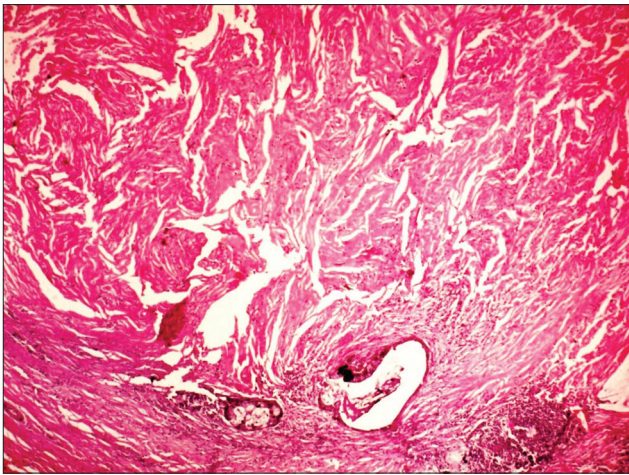


Figure 1: Photomicrograph revealing central keloid-like sclerosis and peripheral tumor nests and lymphoid infiltrates (H and E, 20×)

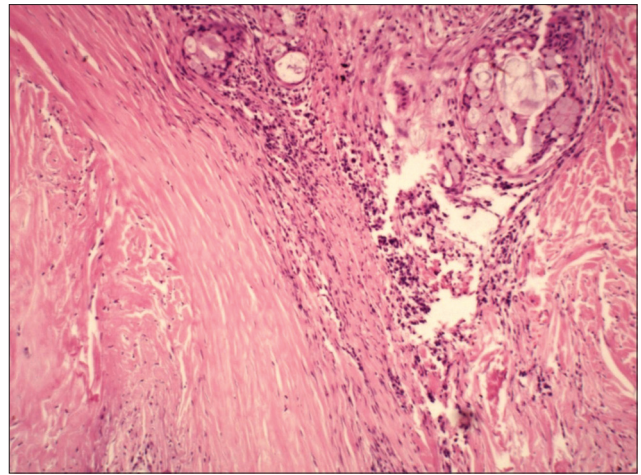


Figure 2: Higher magnification revealing nests of tumor cells surrounded by lymphocytic infiltration at the periphery of tumor (H and E, 40×)

Case 2

A 65-year-old male presented with a left submandibular swelling since 4 years. On examination, there was a hard, nontender mass measuring 3 × 2 cm at the left submandibular region fixed to the mandible. CT scan was suggestive of benign well-defined minimally enhancing lesion in the left submandibular region. The mass was excised under GA. Intraoperatively there was a hard, encapsulated mobile mass, measuring 3 × 3 cm in size and was partially fixed to the underlying structures. On microscopic examination, the tumor predominantly consisted of dense hyalinizing keloid-like sclerosis with lymphoid aggregates, lymphoid follicles with germinal centers and minute cysts predominantly lined by mucous cells with scattered intermediate cells at the periphery. The tumor was diagnosed as sclerosing variant of mucoepidermoid carcinoma, low grade.

DISCUSSION

Mucoepidermoid carcinoma (MEC) is the most common malignant salivary gland neoplasm. Its sclerosing morphologic variant, however, is an extremely rare entity.^[1,2] To the best of our knowledge, only 15 cases of salivary gland SMEC have been previously reported since Chan and Shaw described the first case in 1987.^[3,4] 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. Although MECs sometimes show tumor-associated lymphoid hyperplasia, SMEC shows both marked central keloid-like sclerosis and peripheral lymphoid hyperplasia.

Several conditions of the salivary glands may manifest morphologically with extensive stromal sclerosis or

fibrosis, including sclerosing polycystic adenosis, hyalinizing clear cell carcinoma, mixed tumors, sclerosing sialadenitis, and polymorphous low-grade adenocarcinoma. Mucoepidermoid carcinomas of the salivary glands also frequently show a sclerotic stroma. However, the extensive and obliterative keloidal-type sclerosis is distinctly unusual. Tumor infarction and extravasation of mucin eventuating in reactive fibrosis are two mechanisms of formation that have been suggested as underlying this morphologic variant.^[3] Most investigators prefer the mucin-spillage hypothesis for the origin of the sclerotic stroma.^[3,5,6]

The reported cases have (1) generally occurred across a wide age range (17–65 years), (2) shown a significant female preponderance (5 : 2), (3) shown a predilection for the parotid gland (71%), and (4) been of intermediate sizes (average diameter, 2.9 cm). Their occurrence in submandibular gland is extremely rare with only one reported case so far.^[7]

The sclerosis associated with these tumors may obscure their typical morphologic features and result in diagnostic difficulties. Some major salivary gland lesions show similar sclerotic stroma, and these lesions include sclerosing polycystic adenosis, hyaline clear cell carcinoma, malignant mixed tumor, sclerosing sialadenitis and polymorphous low-grade adenocarcinoma.^[6] Of all the histological features observed in SMEC, a central keloid-like sclerosis rimmed by peripheral lymphoid infiltration is unique enough to distinguish SMEC from the other sclerotic salivary lesions. The percentage of keloid-like areas necessary to characterize SMEC is not defined, because there has been no sizeable study. Consequently, only a brief description of this unique histological change in MEC is given in the current standard text book of salivary gland pathology. On the basis of present findings and previous information, it seems clear that separation of SMEC from

conventional MEC is of little or no merit in view of clinical behavior and prognostic predictability. The real importance lies in its morphologic mimicry of benign sclerosing processes in the salivary glands such as fairly common sialadenitis and extremely rare polycystic adenosis.^[8]

In conclusion, SMEC is a very rare salivary gland tumor with uncertain behavior. Complete surgical excision with tumor-free margins with preservation of facial nerve (in the absence of clinical invasion) is recommended. Postoperative radiation therapy is suggested for positive or close margins given the adherent nature of the tumor and the risk of recurrence and distant disease. Patients should be followed closely with serial MRI studies of the tumor bed with complete clinical evaluation of the regional lymphatics and chest to evaluate for evidence of recurrence or metastasis.^[9]

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