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Mammary Analogue Secretory Carcinoma - An Unusual Clinical Entity at an Unusual Site

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Abstract

Mammary analogue secretory carcinoma is a recently described entity mainly occurring in major salivary glands.This is a case report of a non-healing ulcer on the hard palate of a 30-year old female patient with no other systemic diseases. Biopsy of the ulcer resulted in the differential diagnoses of acinic cell carcinoma and mucoepidermoid carcinoma requiring wide excision of the same. Biopsy result of the excised specimen revealed it to be mammary analogue secretory carcinoma. Specimen was positive for S-100 protein and ETV6-NTRK3 fusion gene, thus confirming the diagnosis. Adjuvant radiotherapy was also administered postoperatively.

Keywords

Mammary analogue secretory carcinoma (MASC), carcinoma of salivary glands, hard palate.

Introduction

Mammary analogue secretory carcinoma (MASC) is a recently described salivary gland tumour that shares the same histologic appearance and *ETV6* gene (12p13) rearrangement as secretory carcinoma of the breast [1-3]. MASC usually occurs in adults but afflicts patients across a wide age range (13–

77 years), with a mean age of about 45 years, and is slightly more common in males. Most patients present with a slow-growing, painless mass with a size of about 2 cm and with a known duration varying from 2 months to several years. MASC involves the parotid gland in about 70% of cases, the submandibular gland in about 7% of cases, and, less commonly, other sites, such as the soft palate, buccal mucosa, base of the tongue, and lip [4].

Case Report

A 30 year old lady - without significant previous medical or family history - presented with a mildly painful, non-healing ulcer on the left hard palate for one and a half months (Fig.1). On examination, the ulcer measured 0.5x0.5 cm. It was tender and had inflamed, non-indurated margins. Posterior margin was located at the junction of hard and soft palate. No other positive findings were noted on local and general examination.

As it was a non-healing ulcer, she was advised to undergo biopsy. The histopathologic examination of the biopsy specimen was reported with a differential diagnoses of mucoepidermoid carcinoma (MEC) and acinic cell carcinoma (AcCC). She was advised to undergo wide excision of the lesion. Pre-operatively, a CT scan with contrast of the neck and jaw was done which showed no involvement of the bone or lymph nodes. Pre-operative blood profile was normal. Wide excision of the ulcer with 1 cm of margin was done. The periosteum was not involved by the disease, thus subperiosteal dissection was done without removing the bone. Greater palatine neurovascular bundle was ligated at its exit from the greater palatine foramen. No graft was used to close the defect, leaving it to heal secondarily. Post-operative recovery was uneventful. Speech and deglutition were normal. The excised specimen was sent for histopathological examination. Upon primary examination, mammary analogue secretory carcinoma (MASC) was suspected along with other similar differential diagnoses like AcCC and MEC. The cranial margin of the specimen at the greater palatine nerve bundle was positive. Immunohistochemistry was performed, which was positive for S-100 protein and negative for P 63, DOG-1 and GCDFP-15. For further confirmation, fluorescence in situ hybridization (FISH) was performed for presence of ETV6-NTRK3 fusion gene which was found to be positive. Thus, we arrived at a final diagnosis of MASC. Consequently, adjuvant radiotherapy was administered from third post-operative week.

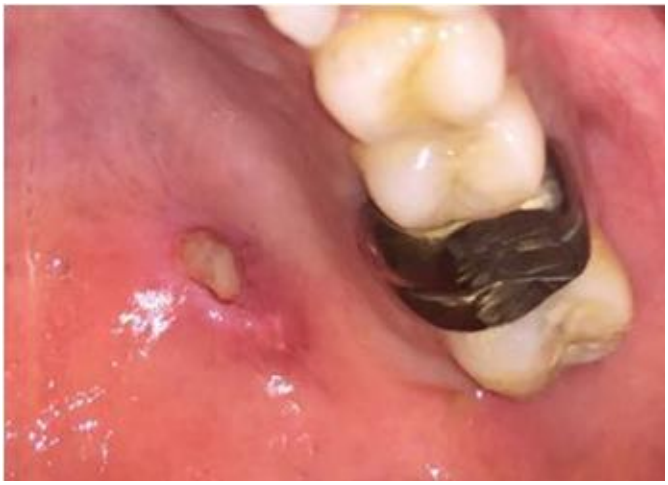
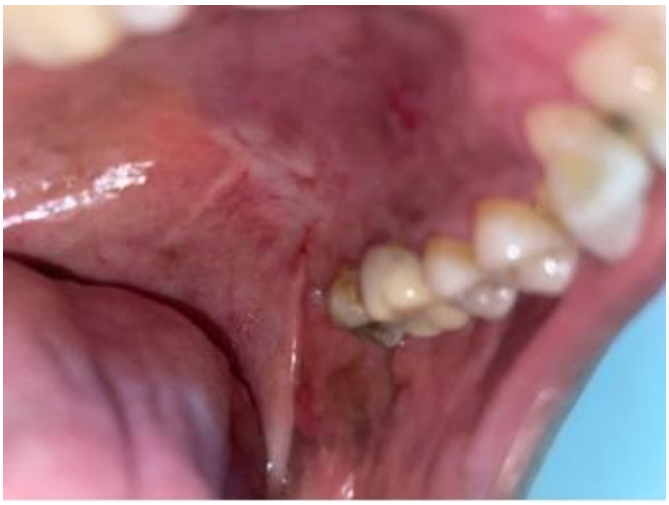


Fig. 1: Clinical picture of the ulcer

Management

Some surgeons have treated this clinical condition with surgical resection followed by radiotherapy. Sethi et al [7] treated all their patients with surgical resection of tumour with or without neck dissection, with a subset of their patients receiving postoperative radiotherapy alone or a combination of radiotherapy and chemotherapy. Skalova et al [5,6, 8-11] have done extensive work on MASC with all the cases requiring surgical resection (radical/non-radical excisions). In a considerable number of those cases there were recurrences which required re-exploration with or without radiotherapy and chemotherapy. Roy et al [12], in their case series, treated their patients with local excision of the lesions without adjuvant therapy.

Our patient underwent wide excision of the lesion with 30 cycles of postoperative radiotherapy. She is currently in remission for the past 3 years (Fig. 2).



Discussion

MASC is a low grade carcinoma of the salivary glands bearing similarities to secretory carcinoma of breast and acinic cell carcinoma of the salivary glands [1]. Pathologic features of MASC include a well-circumscribed and uncapsulated tumour with cystic components on gross examination and cystic architecture (maybe unicystic or multicystic) with at least some infiltration of the margins on histopathologic examination[5,6].Differential diagnoses include AcCC, low grade salivary duct carcinoma (LGSDC), high grade salivary duct carcinoma (HGSDC), MEC and polymorphous low grade adenocarcinoma (PLGA)[Table 1].[1]

Histological features	MASC	AcCC	LGSDC	HGSDC	MEC	PLGA
Architectural features	Varying proportions of macrocystic and microscystic patterns (Fig. 3).	Architecturally similar to MASC but with greater cytologic diversity.	Intraductal proliferation of low grade ductal cells.	Extensive infiltration with abundant cribriform architecture and comedonecrosis.	Variable cystic and solid architecture alongwith inflammation and fibrosis.	Cord-like and whirling growth pattern.
Cytologic features	Eosinophilic cytoplasm abundant with mucicarmine-positive mucin. Absence of zymogen granules.(Fig. 4)	Basophilic cytoplasm with PAS positive zymogen granules.	Cytoplasm consists of yellow lipofuscin-like pigment.	Abundant oncocytoid cytoplasm with well-defined cell borders.	Presence of goblet type mucous cells, intermediate and squamoid/ epidermoid cells.	Lack of bubbly pink cytoplasm unlike MASC.
Immunohistochemistry (IHC)	Mammaglobin and S-100 positive.	Strong cytoplasmic and canalicular expression of DOG-1	Cannot help differentiate from MASC	Cannot help differentiate from MASC	P63 and cytokeratin positive	S-100 and mammaglobin positive
ETV6-NTRK3 gene rearrangement	*Positive*	Negative	Negative	Negative	Negative	Negative

Table 1: Differential diagnosis with IHC

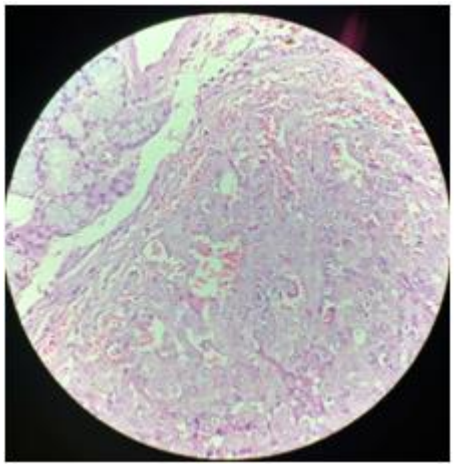


Fig. 3: Tubular and microcystic pattern in 40 X

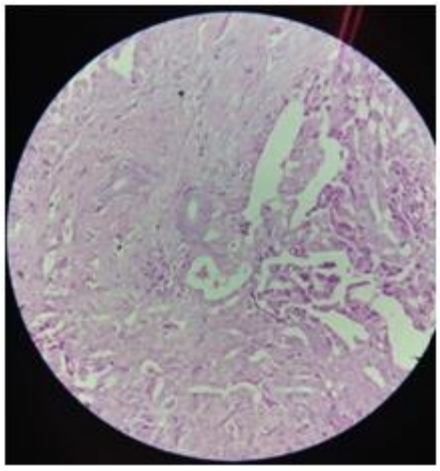


Fig. 4: Cells are having round small nuclei with moderate eosinophilic cytoplasm (40 X).

Conclusion

MASC was first described by Skalova et al in 2010, making it a fairly recent entity, which makes it more likely to be diagnosed as other similar salivary gland tumours. Incidence of MASC in hard palate is very rare. Approximately 300 cases of MASC have been reported in the literature to date [13] and only 18 of these cases have been reported in minor palatal salivary glands [14]. A careful histopathological examination with IHC is the key to identification of this tumour.

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