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MASC UNMASKED- A CASE REPORT AND WHAT WE KNOW TILL DATE

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ABSTRACT We report a case of MASC (mammary analogue of secretory carcinoma) in a 24-year-old male who presented with swelling in the parotid region and provisional diagnosed as pleomorphic adenoma and superficial parotidectomy was done. On histopathological examination and immunohistochemistry, it was diagnosed as MASC which is a less common variant of salivary gland tumor and completely a pathological diagnosis, not possible to diagnose preoperatively. It shows resemblance to pleomorphic adenoma both clinically and histopathologically. The treatment varies from simple excisions to radical surgery with or without neck dissections, adjuvant radiotherapy and or adjuvant systemic chemotherapy. Post-operative radiotherapy is reserved for patients with close surgical margins (5mm), incomplete resection, perineural invasion and / T3 or T4 tumours which are not present in our case.

KEYWORDS : MASC ,Mammary analogue of secretory carcinoma, pleomorphic adenoma, parotidectomy.

INTRODUCTION

MASC or Mammary Analogue Secretory Carcinoma is a newly recognized variant of salivary gland malignancy. It shares histologic, immunohistochemical and genetic features with secretory carcinoma of the breast. The clinical behavior of MASC ranges from slowly growing tumors that infrequently recur after surgical resection to aggressive tumors that cause widespread metastasis and death. Many cases of MASC were discovered in archived cases previously classified as acinic cell carcinoma, mucoepidermoid carcinoma and adenocarcinoma not otherwise specified.

CASE REPORT

A 24 year old male attended the surgical clinic with chief complaint of swelling on left side of his face for 2 years, which was gradually increasing in size and not associated with pain, fever, or change in size during eating or swallowing. No history of loss of weight or appetite, difficulty in swallowing, tubercular infection or mumps noted. He was a known tobacco chewer and alcoholic for ten years.

On physical examination, he was afebrile with average body built and good nutritional status. His BMI was 28.2. Examination of other systems did not reveal any significant abnormality. On local examination, a single, firm, non-tender, globular swelling of size 4x4 cm occupying the hollow between mandible and mastoid on left side and elevating the left ear lobule, deep to the parotid fascia and superficial but non-fixed to the masseter with normal overlying skin was found. Examination of deep lobe and Stensons duct was within normal limits. The facial nerve was clinically not involved. No ipsilateral or contralateral lymph nodes were palpable. Other salivary glands were clinically normal.

His routine biochemical and haematological investigations were within normal limits. ELISA test for HIV was negative. Markers for viral hepatitis were negative. Ultrasound of neck was suggestive of a well-defined heteroechoic lesion of size 27.8 x 23.3mm in superficial lobe of parotid gland with mobile internal echoes and central hyperechoic floating contents, differentially diagnosed as necrotic parotid abscess or a neoplastic swelling. No cervical lymphadenopathy was noted. Opposite side parotid gland was normal. USG guided FNAC from the swelling was inconclusive.

A provisional diagnosis of pleomorphic adenoma of parotid was made and patient was taken for excisional biopsy. Intraoperative findings of a 4x 3 cm, well defined mass, limited to the superficial lobe of left parotid gland was noted and hence superficial parotidectomy with facial nerve preservation with sternomastoid muscle flap was done. Postoperative period was uneventful and patient was discharged on POD-4. The specimen was sent for histopathological examination which revealed features of MASC. It was staged as T2N0M0, since it was no more than 4 cm in dimension and without any extraparenchymal extension. The extraparenchymal extension includes clinical or macroscopic evidence of invasion of soft tissues or nerve. Microscopic evidence of invasion alone does not constitute extraparenchymal extension[1].

Grossly, the specimen was a single, well circumscribed and encapsulated tissue measuring 3x3x4 cm. Cystic and solid areas were identified. The cyst was filled with brown gelatinous substance. The solid areas were yellow in color and friable. No capsule breach was noted.

On microscopy, the well encapsulated tumor had cells arranged in the form of tubules, papillae and solid sheets (Fig. 1a). Cystic areas show intraluminal eosinophilic secretions (Fig 1b). Cells were small to intermediate in size with amphophilic and, vacuolated and granular cytoplasm. The nuclei showed mild pleomorphism with granular chromatin and prominent nucleoli. At places, the cells show hobnailing appearance (Fig 2b). Interspersed congested blood vessels along with foci of haemorrhage were also present. Mitotic activity was low. The adjacent tissue showed unremarkable parotid gland parenchyma separated from the tumor by thick fibrous band. Four lymph nodes included in specimen had features of dermopathic lymphadenitis.

Immunohistochemical staining was done further to characterize the tumor. S-100 showed dense positivity (Fig. 2a) while EMA showed moderate positivity in tumor cells (Fig. 2c). Ki67 proliferating index was 0-1 %.

The case was then discussed in multidisciplinary tumor board for consensus regarding further management in this case and it was decided to follow up the patient regularly on outpatient basis with clinical and if required, radiological examination. Post 11 months of discharge, patient is doing fine, without any symptoms or signs of recurrence.

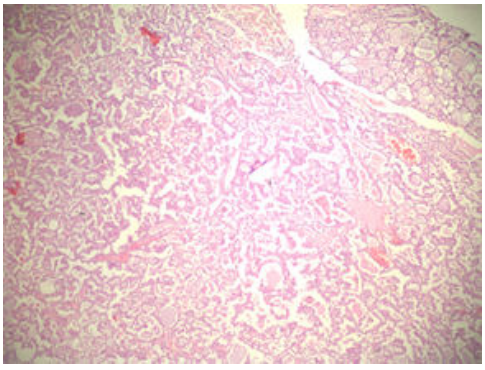

Fig. 1a. Cells arranged in the form of tubules, papillae and solid sheets

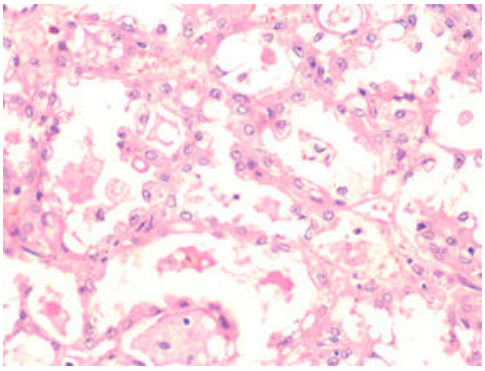

Fig. 1b. Cells with amphophilic, vacuolated and granular cytoplasm with cystic spaces with intraluminal eosinophilic secretion.

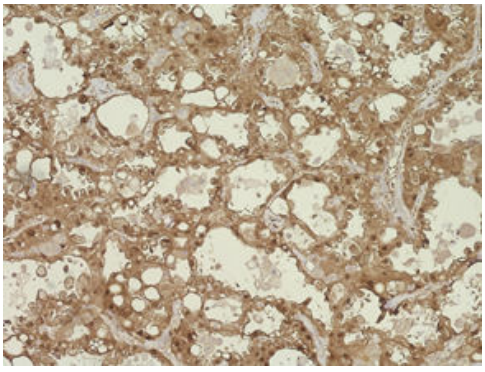

Fig 2a. Dense S 100 positivity of cells.

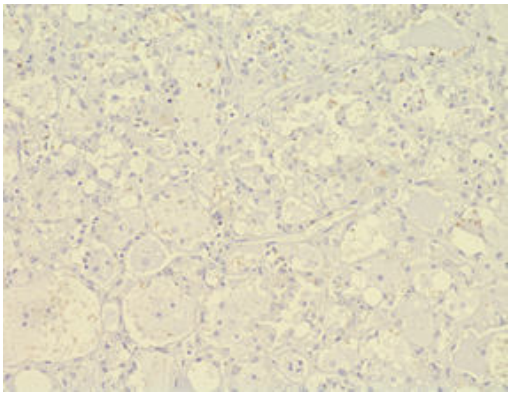

Fig 2b. Hobnail appearance of cells

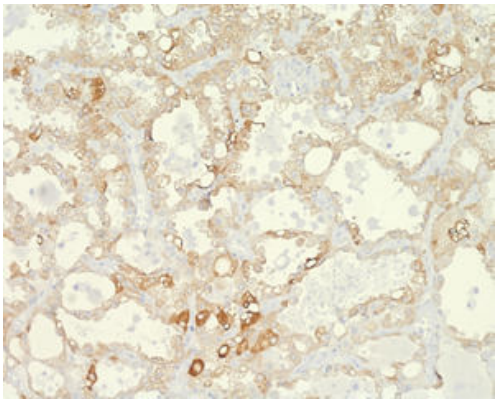

Fig 2c. Moderate EMA positivity of cells

DISCUSSION

Since its first case report, hundreds of cases of MASC have been reported, really well, with some controversies, some dilemmas, some yet to establish protocols. It's no more rare, shouldn't be called so, as it's now a well-defined, a well-known entity, being diagnosed more frequently than ever before. This discussion means to summarise all that we know about it till date.

Secretory breast carcinoma is a very rare subtype of invasive breast cancer and accounts for <0.1% of all invasive breast cancers .(Lee et al., 2014).MASC shares its morphological and immunohistochemical features with this carcinoma, and hence is equally rare.

It commonly affects parotid gland, 70% of all the cases while 7% of them have been reported in submandibular gland. They may also occur in minor salivary glands of lips, soft palate, base of tongue and buccal mucosa.(Shukla et al., 2018)

MASC or mammary analogue secretory carcinoma is completely a pathological diagnosis, not possible to differentiate it clinically, preoperatively from other salivary gland tumors. It is not clear as to how preoperative diagnosis of MASC will alter the management since it will be managed like any other low-grade malignancy. They are also staged by TNM staging similar to any other major salivary gland tumor including parotid, submandibular and sublingual glands, since minor salivary gland tumors are staged according to their anatomic site of origin.(*NCCN Guidleines*, n.d.)

MRI is considered the most appropriate modality for evaluation of parotid gland tumors since it has high contrast resolution for soft tissues and capability of visualizing deep lobe tumors while also providing excellent morphological and volumetric assessment, relationship with surrounding structures and assessment of cervical lymph nodes as well. MRI features of MASC, though not done in our case, have been reported to include hypointense lesion in T1 weighted imaging while isointensity is seen in T2 image, usually with well-defined borders.

They are similar to other salivary gland neoplasms especially pleomorphic adenoma, both clinically and histopathologically. Both are painless, well defined slow growing and will have diffuse S- 100 positivity but adenomas are negative for mammaglobin and/ muc-4. MASC can be focally positive for CK5/6 to resemble mucoepidermoid carcinoma but typical mucocytes will be absent. Acinus cell carcinoma is said to be the most common misdiagnosis for MASC due to histopathological resemblance but is negative for mammaglobin and S100.(Bissinger et al., 2017)

The treatment of MASC has not been standardized yet and is still being treated like any other salivary gland tumor of low

malignant potential. This varies from simple excisions to radical surgery with or without neck dissections, adjuvant radiotherapy and or adjuvant systemic chemotherapy. Post operative radiotherapy is reserved for patients with close surgical margins (5mm), incomplete resection, perineural invasion and / T3 or T4 tumors, none of which were present in our case, hence patient was only followed up in OPD. (Wang et al., 2016)Doses between 60-66 Gy are delivered in fractions of 1.8 to 2 Gy for between five to six treatments per week. But the value of this PORT is still unclear.

MASC has been known to show a widely varied clinical course ranging from being very indolent to very aggressive. Several studies have tried to estimate the risk of progression and death but patients enrolled in the studies have undergone varying degree of surgical resections and some received PORT while others don't. Some have received chemotherapy as well with unspecified agents. Hence, the results of the available data need to be interpreted carefully , since there has been no standard protocol yet. However, Chiosa et al followed 14 patients from the authors' institutional cohort, and 14 additional patients culled from the literature. The mean disease-free survival, using death or recurrence as the end-point, was 92 months (95% CI, 71-115).(Sethi et al., 2014)

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