

# Cytological diagnosis of myoepithelial carcinoma of minor salivary gland: A rare entity

Vandana U. Grampurohit, Hephzibah Rani, S. S. Parinitha, U. S. Dinesh

Department of Pathology, SDM College of Medical Sciences and Hospital, Sattur, Dharwad, Karnataka, India

## ABSTRACT

Neoplastic cells with myoepithelial differentiation are often present in both benign and malignant salivary gland neoplasms. Potential diagnostic problems may arise due to morphologic heterogeneity of myoepithelial cell-rich lesions in fine-needle aspiration cytology (FNAC). Myoepithelial carcinoma is a malignant salivary gland tumor composed exclusively of cells with myoepithelial differentiation. Histopathologically, it is a well-established entity but its cytological features have rarely been reported. We report a case of myoepithelial carcinoma of minor salivary gland in a 62-year-old female diagnosed preoperatively by FNAC. Awareness of diverse cytoarchitectural patterns and immunohistochemical profile is crucial for accurate identification.

**Key words:** Fine-needle aspiration cytology, minor salivary gland, myoepithelial carcinoma

## INTRODUCTION

Neoplastic cells with myoepithelial differentiation are often present in both benign and malignant salivary gland neoplasms.<sup>[1]</sup> Lesions that contain abundant myoepithelial cells may present as a diagnostic challenge in fine-needle aspiration cytology (FNAC). Potential diagnostic problems may arise due to morphologic heterogeneity of myoepithelial cell-rich lesions and difficulty in predicting malignancy in FNAC.<sup>[2]</sup>

Myoepithelial carcinoma (MEC) or malignant myoepithelioma is a malignant salivary gland tumor composed exclusively of cells with myoepithelial differentiation. The tumor represents the malignant counterpart of myoepithelioma and is characterized by infiltrative growth and potential for metastasis.<sup>[3]</sup> These are rare, unusual and intriguing tumors. Histopathologically,

MEC is a well-established entity but its cytological features have rarely been reported. Our knowledge with respect to its FNAC features is deficient, possibly due to its rare occurrence and lack of comprehensive case reports in the literature. The presence of pleomorphism, coarse chromatin, prominent nucleoli, mitotic figures and necrosis should raise the possibility of MEC in FNA specimens from myoepithelial cell-rich lesions.<sup>[2]</sup> This case is reported in view of rarity of preoperative FNAC diagnosis of this entity with cytological features of MEC and to highlight importance of FNAC to plan a proper surgical management.

## CASE REPORT

A 62-year-old female complained of a painless swelling in right cheek for 6 months duration. On examination, there was a hard mass present in right maxillary region involving right buccal space with palpable right submandibular cervical lymph node. Ultrasonography revealed right submandibular cervical lymphadenopathy and a soft tissue mass in right buccal space with increased vascularity, focal infiltration into buccinators and necrotic areas.

Fine-needle aspiration cytology of mass showed high cellularity composed of pleomorphic discohesive plasmacytoid cells in sheets, clusters and singles. The cells showed abundant cytoplasm, irregular hyperchromatic

### Access this article online

#### Quick Response Code:



#### Website:

www.cci-journal.org

#### DOI:

10.4103/2278-0513.138087

**Address for correspondence:** Dr. Vandana U. Grampurohit, Department of Pathology, SDM College of Medical Sciences and Hospital, Sattur, Dharwad, Karnataka, India. E-mail: vandanoday@yahoo.com

eccentric nucleus with prominent nucleoli. Binucleate cells, tumor giant cells, cells with cytoplasmic hyaline globules and vacuolations, many atypical mitoses were seen. No ductal differentiation was seen. Background was hemorrhagic with foci of necrosis [Figure 1]. The lymph node aspirate showed metastatic cells with similar features. With these cytological features the diagnosis of “myoepithelial carcinoma of minor salivary gland with metastasis in right submandibular lymph node” was offered.

Radical neck dissection specimen showed a globular tumor mass measuring 6 × 5 × 5 cm. Cut surface was variegated, grey white with foci of necrosis and hemorrhage. 18 lymph nodes were isolated, largest measuring 2.5 × 1.5 × 1 cm [Figure 2a].

Histopathologically, mass showed tumor composed of markedly pleomorphic discohesive cells arranged in solid sheets, trabeculae and reticular pattern [Figure 2b]. Cells showed granular, glassy and at places vacuolated cytoplasm with hyperchromatic eccentric nucleus showing prominent nucleoli. Atypical mitoses ranging from 1 to 10/high power field were noted. Stroma showed myxoid and necrotic areas. Four out of 18 lymph nodes showed metastatic deposits. Immunohistochemistry showed reactivity to S-100 protein, epithelial membrane antigen (EMA), vimentin, but was negative for actin leading to the diagnosis of MEC [Figure 3].

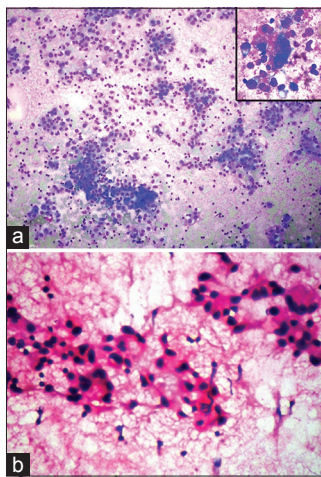
## DISCUSSION

Myoepithelial carcinomas are very rare and constitute only 0.2% of epithelial salivary gland neoplasms.<sup>[4]</sup> According to Dean and Sierra *et al.*, the term myoepithelioma was first

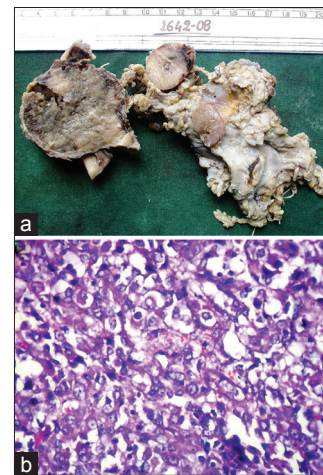
used by Sheldon in 1943, and MEC was first described by Stromeyer *et al.* in 1975.<sup>[5]</sup> MEC was included in the updated histological classification of salivary gland tumors by World Health Organization in 1991.<sup>[3]</sup>

Myoepithelial carcinoma is commonly located in parotid followed by submandibular gland and is rarely seen in minor salivary gland. Except for palate, intraoral sites are only rarely affected.<sup>[6-8]</sup> It is a tumor of adults with wide age range of 14-86 years. Males and females are affected equally.<sup>[3,4]</sup> About 60-70% of MEC develop in pleomorphic adenomas and the remainder arise *de novo*.<sup>[1]</sup> The present case developed “*de novo*” in normal salivary gland. Criteria to establish a diagnosis of myoepithelial carcinoma are – the neoplastic cells must be characterized as myoepithelial and the tumor must be morphologically and biologically malignant.<sup>[4,6,8]</sup>

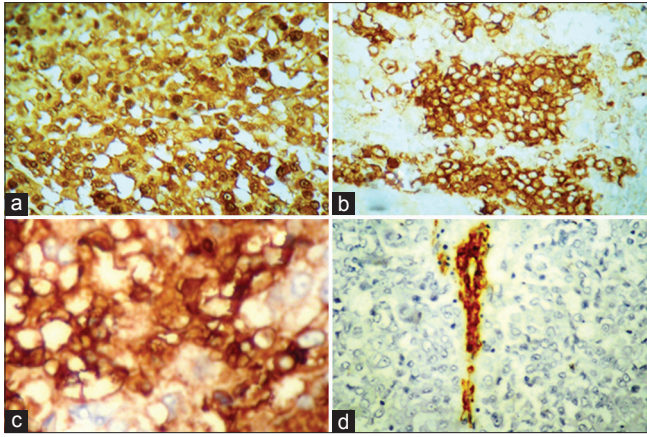
Grossly these tumors are soft to firm, un-encapsulated and have infiltrative borders and destructive tumor extensions into adjacent salivary gland.<sup>[6]</sup> The tumor cells show a wide variety of morphology comprising spindle, plasmacytoid (hyaline), epithelioid and clear cell subtypes. Combinations of these cell types may be present within the same tumor. Stroma may be myxoid or hyalinized.<sup>[6,9]</sup> Same features are depicted even in FNAC. Immunohistochemical studies show that these neoplasms consistently express S-100 protein, cytokeratin, glial fibrillary acidic protein, EMA and vimentin.<sup>[1,6,9]</sup> Smooth muscle actin is seen in spindle cell type of myoepithelial cells but not necessarily in plasmacytoid or hyaline cells. It is now well-established that neoplastic epithelium does not always retain actin expression.<sup>[6]</sup> Our case hence met both criteria: (1) Increased mitotic activity, cellular pleomorphism, necrosis in cytology and



**Figure 1:** Fine-needle aspiration cytology showing. (a) Hypercellular smears composed of pleomorphic discohesive cells in small sheets, clusters and singles. The inset shows plasmacytoid cells, binucleate cells and tumour giant cells (Leishman's, ×10). (b) Pleomorphic cells with abundant cytoplasm, irregular hyperchromatic eccentric nucleus, prominent nucleoli and intracytoplasmic hyaline globules (H and E, ×40)



**Figure 2:** (a) Gross photograph shows a globular tumour mass with lymph nodes and submandibular gland. The cut section of the tumor shows variegated appearance having grey white areas with foci of necrosis and haemorrhage. (b) Microphotograph shows pleomorphic cells in sheets with abundant eosinophilic cytoplasm, hyperchromatic nucleus and prominent nucleoli (H and E, ×40)



**Figure 3:** Immunohistochemical stains ( $\times 40$ ) showing. (a) Reactivity for S-100 protein. (b) Reactivity for epithelial membrane antigen. (c) Reactivity for vimentin. (d) Negative staining for smooth muscle actin

more importantly an infiltrative and destructive growth pattern with metastasis in histopathology proved that the tumor was morphologically and biologically malignant. (2) Immunohistochemistry showed reactivity to EMA, S-100 protein and vimentin, however being plasmacytoid variant showed negative reaction to actin. In addition, the lack of ductal and acinar differentiation also supported the diagnosis of myoepithelial tumor.

Due to its diverse cytologic presentations, MEC raises a variety of neoplasms in the differential diagnosis and depends on the prominent cell type. The differential diagnosis includes amelanotic melanoma, adenocarcinoma, plasmacytoma, epithelial-myoepithelial carcinoma. Immunohistochemical profile of each tumor helps to separate them from myoepithelial tumor.<sup>[1,4,6]</sup>

The clinical behavior of this tumor is variable.<sup>[1]</sup> Rarely metastases are seen in lungs, liver and lymph nodes but local recurrence rate is very high and accounts for poor prognosis.<sup>[4,8]</sup>

Wide surgical resection is the treatment of choice for MEC.<sup>[6]</sup> Early and radical surgery with close follow-up are essential achieving favorable outcome. Therapeutic neck dissection is indicated when there are metastases in cervical lymph nodes. As documented by Dean and Sierra *et al.*, Stromeyer found that the tumor is not sensitive to radiation while Takeda reported a good clinical response to radiation.<sup>[5]</sup>

## CONCLUSION

Myoepithelial carcinoma may pose a diagnostic challenge in FNAC. FNAC plays an important and easy diagnostic modality in these unusual rare cases. Cytological features have rarely been reported for this tumor. A diagnostic dilemma may arise due to rarity of lesion. Awareness of diverse cyto-architectural patterns and immunohistochemical profile is crucial for accurate identification.

## ACKNOWLEDGMENT

We thank Dr. Laxmi Rao, Consultant Pathologist for providing us immunohistochemistry slides and guiding us for the case.

## REFERENCES

1. Chhieng DC, Paulino AF. Cytology of myoepithelial carcinoma of the salivary gland. *Cancer* 2002;96:32-6.
2. Darvishian F, Lin O. Myoepithelial cell-rich neoplasms: Cytologic features of benign and malignant lesions. *Cancer* 2004;102:355-61.
3. Skalova A, Jakel KT. Salivary glands, myoepithelial carcinoma. In: Barnes L, Eveson JW, Reichart P, Sidransky D, editors. *World Health Organization Classification of Tumours, Pathology and Genetics of Head and Neck tumours*. Lyon: IARC Press; 2005. p. 240-1.
4. Luna MA. Salivary glands. In: Pilch BZ, editor. *Head and Neck Surgical Pathology*. 1<sup>st</sup> ed. Philadelphia: Lippincott Williams and Wilkins; 2001. p. 305.
5. Dean A, Sierra R, Alamillos FJ, Lopez-Beltran A, Morillo A, Arévalo R, *et al.* Malignant myoepithelioma of the salivary glands: Clinicopathological and immunohistochemical features. *Br J Oral Maxillofac Surg* 1999;37:64-6.
6. Saveria AT, Sloman A, Huvos AG, Klimstra DS. Myoepithelial carcinoma of the salivary glands: A clinicopathologic study of 25 patients. *Am J Surg Pathol* 2000;24:761-74.
7. El-Mofty SK, O'Leary TR, Swanson PE. Malignant myoepithelioma of salivary glands: Clinicopathologic and immunophenotypic features. *Int J Surg Pathol* 1994;2:133-40.
8. Cosentino TB, Brazão-Silva MT, Souza KC, de Faria PR, de Moraes S, Loyola AM, *et al.* Myoepithelial carcinoma of the submandibular gland: Report of a case with multiple cutaneous metastases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2008;106:e26-9.
9. Nagao T, Sugano I, Ishida Y, Tajima Y, Matsuzaki O, Konno A, *et al.* Salivary gland malignant myoepithelioma: A clinicopathologic and immunohistochemical study of ten cases. *Cancer* 1998;83:1292-9.

**Cite this article as:** Grampurohit VU, Rani H, Parinitha SS, Dinesh US. Cytological diagnosis of myoepithelial carcinoma of minor salivary gland: A rare entity. *Clin Cancer Investig J* 2014;3:438-40.

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