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CYTOLOGICAL POINTERS TO THE DIAGNOSIS OF A RARE PAROTID GLAND TUMOR - EPITHELIAL-MYOEPITHELIAL CARCINOMA

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Abstract: *Epithelial-Myoepithelial carcinoma (EMC) of the salivary gland is a rare tumor representing 1% of all salivary gland neoplasm. Fine needle aspiration cytology (FNAC) is the best tool for preoperative diagnosis of salivary gland neoplasms. However there are only few documented case reports of preoperatively diagnosed cases of EMC in English literature. We present a third case of cytologically diagnosed EMC and highlight the fact that three dimensional clusters, acellular basement membrane material, prominent central nucleoli and attempted tubule formation are important pointers to the cytological diagnosis of EMC at cytology.*

Key words: *Epithelial-Myoepithelial carcinoma, salivary gland neoplasm, Cytology*

Introduction

Epithelial-Myoepithelial carcinoma (EMC) of the salivary gland is a rare tumor first described by Donath et al in 1972 and recognized as a distinct pathologic entity in the 1991 World Health Organization Classification. [1] This rare carcinoma represents 1% of all salivary gland neoplasm. [2]

Case Report

A 61year old male presented to surgery OPD with left parotid mass which increased in size within 10-15 days. There was no history of fever, pain or weight loss. Clinically a diagnosis of pleomorphic adenoma was made.

On examination a mass was palpable in the left parotid region, measuring 4 x 3 cm, non-tender and firm in consistency. Fine needle aspiration cytology (FNAC) was requested to assess the necessity for surgery. The mass was aspirated with 25 gauge needle attached to a 10 ml syringe. All the smears were stained with Giemsa, Hematoxylin and Eosin and Papanicolaou stain.

Cytological Findings

The smears revealed abundant cellularity of large ductal cells arranged in clusters, sheets, pseudo papillae and vague tubule formation. (Figure 1A, inset shows pseudo papillae) Clusters showed overlapping and overcrowding of tumor cells. The ductal epithelial cells were round to polygonal to elongated with moderate amount of cytoplasm, round nucleus and fine chromatin. Columnarization of cells was seen. (Figure 2 A inset) Many of them had single prominent nucleoli. (Figure 1 B inset) Amidst these cells were myoepithelial cells which were spindle shaped cells to round with oval dark nuclei. (Figure 1 B) Basement membrane material was seen in the background (Figure 2 A) Bare nuclei were seen in the background. FNAC diagnosis of epithelial myoepithelial carcinoma was made. Total parotidectomy was performed.

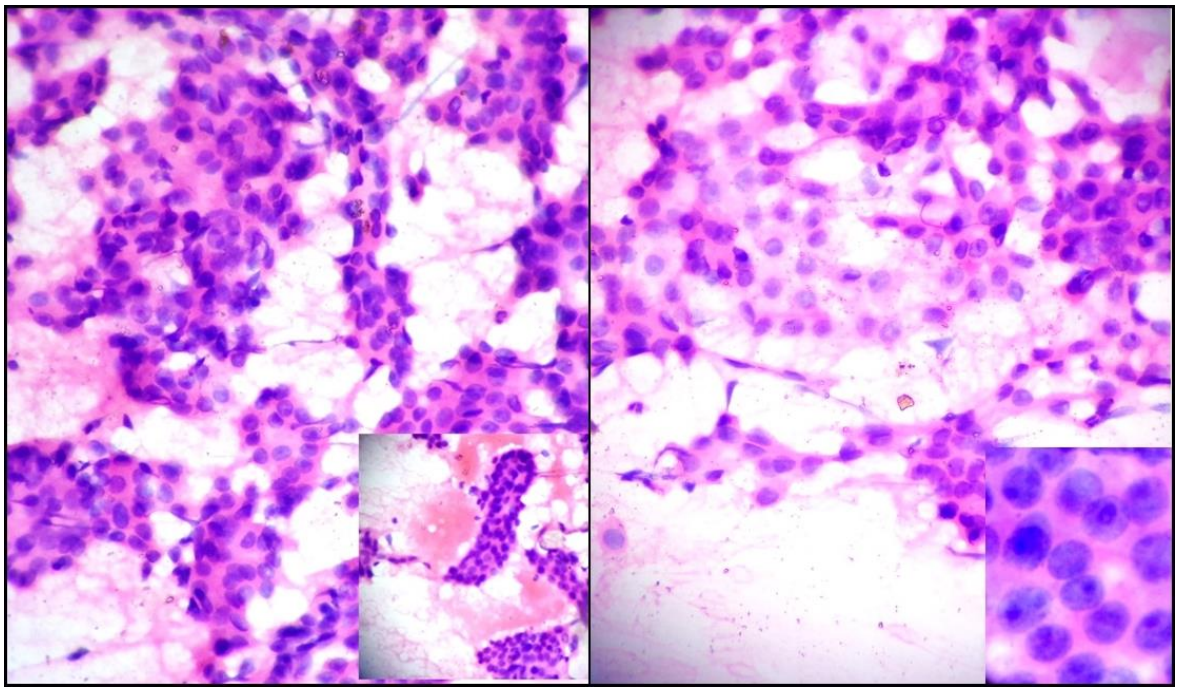

Figure 1:
A. Cytology smears showing tumor cells in clusters, sheets and tubule formation.(H & E, × 100) Inset shows pseudo papillae formation.(H & E, × 100)

B. Cytology smears showing dual population of epithelial and spindle shaped myoepithelial cells.(H & E, × 100) Inset shows epithelial cells with single prominent nucleoli.(H & E, × 400)

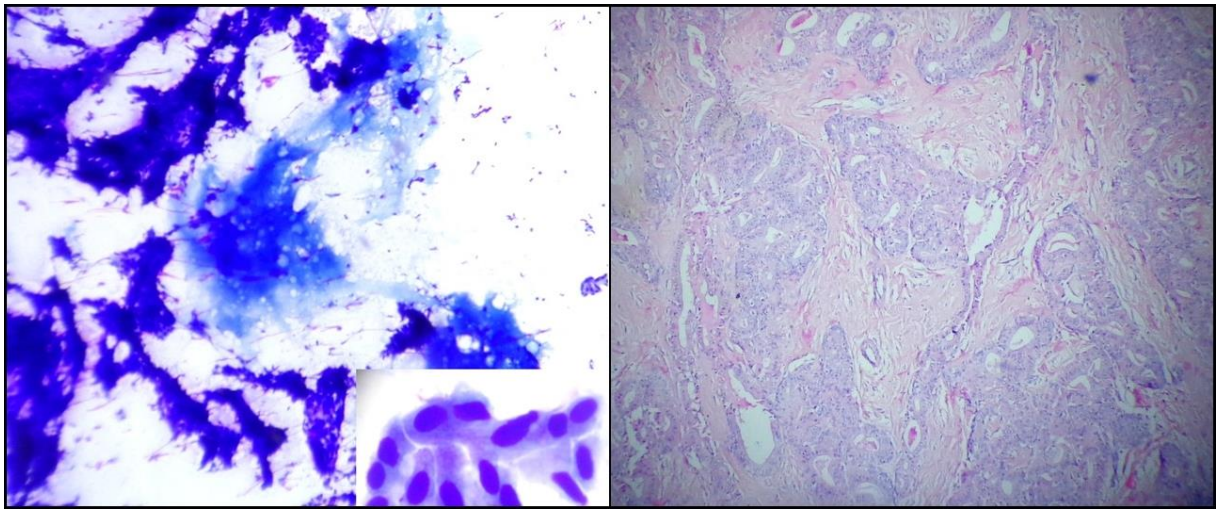

Figure 2:
A. Cytology smears showing basement membrane material adjacent to the tumor cell clusters. (MGG,× 40) Inset shows columnarization of cells. (MGG,× 400)

B. Section showing tubules lined by columnar epithelium with eosinophilic secretions in the lumen along with surrounding single layer of moepithelial cells. Stroma shows basement membrane material (H & E,× 100)

Histopathological Findings

Gross: Specimen consisted of a mass measuring 4.5 x 4.5 cm. Cut surface was solid grey white.

Microscopy: Histopathologically, the hematoxylin – eosin stained sections showed multinodular tumor comprising predominantly of glandular pattern along with clusters. The glands were lined by columnar epithelium which at places showed stratification. The individual cells had abundant cytoplasm, round to oval nuclei, vesicular chromatin and single large prominent nucleoli which was the replica of the FNAC findings. The lumens of the glands were filled with pink eosinophilic secretions. Also seen were spindle shaped cells with dark nucleus and eosinophilic cytoplasm. Stroma showed extensive hyalinization and deposits of basement membrane material. (Figure 2 B)

Periodic acid Schiff stain highlighted the basement membrane material by magenta colour. Hence a final diagnosis of epithelial myoepithelial carcinoma was made.

Discussion

Epithelial-Myoepithelial carcinoma (EMC) is a rare, low grade, malignant neoplasm with a distinctive histomorphology, accounting for 1% of salivary gland neoplasms.[2] EMC occurs more frequently in the seventh decade of life. It affects females more frequently than males. EMC is defined as a malignant tumor composed of duct like structures lined by single layer of ductal cells, surrounded by single layer or multiple layer of myoepithelial cells. The relative proportion of these cells varies from case to case as well as within the same lesion.

Clinically EMC presents as a slowly enlarging painless mass of long duration. Though commonly involving parotid gland, EMC has also been reported in minor salivary glands, and rarely in the para nasal sinuses, pharynx, bronchus and palate. [3]

Preoperative diagnosis of EMC is of utmost importance since it is a low grade malignancy and is prone for recurrence and distant metastasis to the lungs. [4]

Cytological features of EMC have been described rarely in the literature with only two case reports of EMC diagnosed preoperatively at cytology. [3] Most of the time bimodal pattern is not always discernable at cytology. In addition due to fragile cytoplasm the myoepithelial cells present as naked nuclei in the background adding to the diagnostic dilemma. [5, 6]

Adenoid cystic carcinoma, polymorphous low-grade adenocarcinoma, and cellular pleomorphic adenoma closely mimic EMC at cytology because all these contain epithelial and myoepithelial cells along with hyaline globules in common. [5]

Pleomorphic adenoma being the most common tumour of the parotid gland is the closest differential diagnosis of EMC. However the characteristic chondrohyaline myxoid stromal fragments are typical of pleomorphic adenoma. Similarly angulated, hyperchromatic nucleus with nuclear molding clinches the diagnosis of an adenoid cystic carcinoma. Polymorphous low grade carcinoma is mostly limited to the minor salivary glands. And also pseudopapillary trabeculae with bland nuclei helps pathologist to make an accurate diagnosis. [6]

According to Carillo et. al., findings of three dimensional clusters, accompanied by acellular hyaline material in the background can suggest diagnosis of EMC at cytology. [7]

In our case, in addition to the three dimensional clusters, presence of attempted tubule formation and prominent nucleoli are good pointers to the diagnosis of EMC.

At histopathology, classically the tumour is bimodal with infiltrative margins, perineural and vascular invasion. Inner layer of ductal epithelial cells show positivity for cytokeratin and outer myoepithelial cells, which can be either clear, spindle or plasmacytoid, are positive for S100 at immunohistochemistry. [3].These ducts are further enveloped by PAS positive basement membrane material. The attempted tubule

formation seen at cytology recapitulates the same at histopathology

Treatment strategy includes wide surgical resection. Local spread to lymph nodes has been reported, which mandates lymph node resection. Adjuvant therapy may be adopted to prevent local recurrence. [8, 9]

Conclusion

We present a rare a preoperative, cytologically diagnosed case of EMC of parotid gland. with emphasis on the fact that in addition to the three dimensional clusters and acellular basement membrane material, prominent central nucleoli and attempted tubule formation are the two important pointers to the cytological diagnosis of EMC. Hence cytology of salivary holds implication in planning appropriated treatment strategy.

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