MYOEPITHELIAL CARCINOMA: A DIAGNOSTIC CHALLENGE IN CYTOLOGY

Ruchi Agarwal, Kulwant Singh, Anjali Sindhu, Parveen Rana, Swarn Kaur, Deepthi Agarwal

Department of Pathology

Bhagat Phool Singh Government Medical College for Women, Khanpur Kalan, Sonepat (Haryana), India

ABSTRACT

Myoepithelial carcinoma is a very rare neoplasm of salivary gland constituting less than 1% of all salivary gland neoplasm. The most common site of involvement is major salivary gland mainly parotid gland while minor salivary glands are rarely involved. In this case report, we are presenting a case of 29 year old male who presented with a gradually increasing pedunculated painless mass in the oral cavity for 3 months. Cytological diagnosis of cellular pleomorphic adenoma was given. Histopathological examination of excised mass confirmed the diagnosis of myoepithelial carcinoma.

KEYWORDS: Buccal, Myoepithelial carcinoma, Plasmacytoid, Minor salivary gland.

INTRODUCTION

Myoepithelial carcinoma (MC) is a very rare tumor of salivary gland having a reported incidence of less than 1% of all salivary gland tumors (1). Parotid gland is the most commonly involved site (48%-75%), followed by minor salivary glands and the submandibular gland (2-3). The tumour is composed of myoepithelial cells exhibiting both epithelial and smooth muscle differentiation. As myoepithelial differentiation is seen in many benign and malignant neoplasms of salivary glands and MC is a rare neoplasm with diverse cytological findings, it is very difficult to diagnose it alone on aspiration smears. Hence, histopathology and immunohistochemistry are necessary to make a definite diagnosis. With very limited data regarding the cytomorphological features, we present a case of MC of minor salivary gland in a young male along with its cyto-differentials.

CASE REPORT

A 29 old male presented with a slowly progressive swelling, not associated with pain in right oral cavity since 7 months in the surgery outpatient department. The past history and family history were insignificant. There was no history of tobacco chewing and smoking. No history of diabetes, tuberculosis and hypertension was present. The systemic examination was unremarkable. Local examination revealed a soft to firm, grayish pink in colour, non-tender, pedunculated lobulated mass in right buccal mucosa, measuring 1.5x1.5cm in size. On radiological examination, no involvement of the underlying bone was noted. Fine needle aspiration cytology smears revealed cellular smears with large tissue fragments and loosely cohesive groups of neoplastic cells intermixed with metachromatic stromal fragments. Tumour cells have abundant pale cytoplasm with ill defined cytoplasmic borders. The nuclei are eccentrically placed, moderately pleomorphic with inconspicuous nucleoli (Figure 1 a-f).

Figure 1: a: Photomicrograph of FNA from swelling shows cellular smears revealing large tissue fragments and cohesive group of neoplastic cells (Romanowsky stain 40X); b, c, d, e, f: Photomicrographs of FNA smears showing mainly plasmacytoid neoplastic cells arranged in loose cohesive clusters and singly scattered (Romanowsky stain 100X, 200X, 400X)

Based on the above cytological features, the case was diagnosed as cellular pleomorphic adenoma with atypia. Wide local excision of the tumour was done and specimen was sent for histopathological examination (HPE). HPE revealed a tumor mainly composed of nest and lobules of atypical cells focally...
Studies conducted in the past, have reported the varied cytological characteristics of myoepithelial carcinoma. In the study by Di Palma et al (2), out of ten cases of MC, tumour was located in the parotid gland in eight cases. The tumour cells were round epithelioid cells to spindle-shaped and stellate cells and displayed reactivity for high molecular weight keratins and smooth muscle actin. Chhieng and Paulino (7) reported four cases of MC, out of which, three cases were from the parotid gland, and one presenting as recurrent tumor in the minor salivary gland of the hard palate. The aspirates of two cases comprised of predominantly spindle cells, while one case had predominantly epithelioid cells and one case showed mixture of both spindle and epithelioid/plasmacytoid cells. Based on these cytological features, two cases were diagnosed as malignant spindle cell neoplasm, not otherwise specified and one case was interpreted as a pleomorphic adenoma with atypia. They observed that the presence of plasmacytoid/epithelioid cell type with marked nuclear pleomorphism has been found to be reliable diagnostic feature for MC in cases reported in the past.

Grossly, these tumours are unencapsulated soft to firm multinodular masses with infiltrative margins and size ranging from 2-20 cm (9). The cut surface shows areas of haemorrhage, cystic degeneration and necrosis. Histologically, the tumour exhibits mostly nodular or solid sheet like pattern of neoplastic cells with intervening hyaline or myxoid stroma of variable amount. Tumour cells exhibit variety of cytological appearance including spindle, plasmacytoid, epithelioid, and clear cells, with one of these cell types being predominant (1). Infiltration of tumour cells into adjacent normal tissue is the most important histological feature to differentiate malignant from benign myoepithelial neoplasms and hence it should be considered the minimum requirement for diagnosis of MC (6).

Due to its wide cytomorphological features, MC poses a diagnostic challenge for the pathologist. The various differential diagnosis include pleomorphic adenoma,
Myoepithelial carcinoma of the salivary glands have a relatively high recurrence rate and metastasis rate. The current knowledge of myoepithelial carcinomas is limited because these have been underrecognized and underreported in the past. Hence, awareness and knowledge of its characteristic cytoarchitectural, histomorphological patterns and immunohistochemical profile is crucial for accurate identification.

The clinical outcome of myoepithelial carcinoma is variable. Few studies done in the past reported long time patient survival in highly aggressive tumours while other studies reported early metastasis and death in patients with low grade tumours showing mild atypia (6-7,10). Due to rarity of this tumour, very limited treatment experience is available for it. Complete surgical resection is generally the treatment of choice. Large, multicentric studies need to be conducted to assess the role of chemotherapy and radiotherapy as the potential treatment modality for myoepithelial carcinoma.

CONCLUSION

Myoepithelial carcinoma of the salivary glands have a relatively high recurrence rate and metastasis rate. The current knowledge of myoepithelial carcinomas is limited because these have been underrecognized and underreported in the past. Hence, awareness and knowledge of its characteristic cytoarchitectural, histomorphological patterns and immunohistochemical profile is crucial for accurate identification.

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