Case Report

A Case Report of Soft Tissue Myoepithelial Carcinoma in The Neck & Post-Auricular Region: A Diagnostic Challenge

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ABSTRACT

Myoepithelial carcinoma also known as malignant myoepithelioma is a rare aggressive tumor that has been recently described in soft tissue. It is composed exclusively of myoepithelial cells with absence of ductal epithelial structures. We describe a patient with myoepithelial carcinoma of soft tissue occurring in the head and neck. It poses a diagnostic challenge on Fine needle aspiration cytology (FNAC) due to wide spectrum of myoepithelial cell morphology & lack of established criteria for malignancy. The pathologists should be aware of this entity for early and correct diagnosis. To our knowledge, cytological findings of soft tissue myoepithelial carcinoma have not been reported in the literature so far.

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Introduction

The World Health Organisation defines myoepthelioma of soft tissue as a rare tumour composed exclusively of myoepithelial cells in solid, trabecular, myxoid or reticular growth pattern.[1] The tumour cells vary from spindle to plasmacytoid, epithelioid, and clear cells, which have immunohistochemical and electron-microscopic features of myoepithelial differentiation.[1,2] Only 15% of the soft tissue myoepithelial tumors occur in the head and neck region, usually located in the subcutaneous tissue while less than 30% occur in the deep soft tissue.[3] It has been postulated that soft tissue myoepitheliomas arise from deep seated adnexal structures.[4]

Case Report

A 48 yrs female presented with two rapidly enlarging masses in the head & neck region. The midline neck mass, measuring 10 x 8 x 8cm, was present since 5 months. Overlying skin was focally ulcerated. A scar of previous surgery; done one year back for midline neck mass was seen; of which no documents were available. The post-auricular mass, measuring 4 x 3cm was present since 3 months. The ultrasound (USG) neck was suggestive of carcinoma thyroid & FNAC (from outside) was reported as poorly differentiated carcinoma, thyroid.

We performed FNAC from both the masses from multiple sites which revealed similar morphology. The smears were highly cellular predominantly showing spindle shaped cells mixed with some plasmacytoid cells arranged singly, in small groups as well as three-dimensional tissue fragments showing nuclear overlapping and crowding. Abundant pink material (possibly amyloid) and myxoid stroma were also seen intermixed with these neoplastic cells. The spindle cells had scant delicate pale cytoplasm having ill-defined borders, oval to spindle shaped nuclei with moderate anisonucleosis, uniformly distributed fine chromatin & inconspicuous nucleoli [Figure 1(a)].

The plasmacytoid cells were monotonous with moderate amounts of dense non-granular eosinophilic to clear cytoplasm having well-defined borders, round to oval eccentrically located nucleus with moderate anisonucleosis uniformly distributed fine chromatin with few cells showing prominent nucleoli [Figure 1(b)]. Occasional binucleated cells & mitotic figures were identified. No normal thyroid or salivary gland tissue was seen.

In view of the cytological findings & USG neck report, possibility of Medullary Carcinoma, thyroid was considered and the serum calcitonin level was done which came out to be normal. Therefore, CECT neck was advised.

The CECT neck revealed well-defined, heterogenously enhancing, soft tissue density masses in subcutaneous plane in neck & post-auricular region separate from thyroid & salivary glands, along with lytic lesions in cervical vertebrae & upper lobes of bilateral lungs suggestive of malignant lesion.

In view of heterogenous cytomorphology of neoplastic cells and CECT report, various differential diagnoses of soft tissue malignancy having spindle & plasmacytoid cells were considered. Epithelioid MPNST was ruled out by the absence of nuclear palisading, macronucleoli & rhabdoid cells. Epithelioid Sarcoma was ruled out due to lack of pleomorphic nuclei, prominent nucleoli & necrosis. Cutaneous Leiomyosarcoma was excluded by the absence of cigar shaped hyperchromatic nucleus & fibrillary eosinophilic cytoplasm. Moreover, it rarely shows hyalinisation & myxoid change. Amelanotic melanoma was excluded by the absence of hyperchromatic nuclei with prominent nucleoli / intranuclear inclusions. Parachordoma shows spindle to epithelioid cells, with abundant clear, vacuolated cytoplasm. Metastatic sarcomatoid carcinoma generally shows epithelial tumor cells with hyperchromatic nuclei, prominent nucleoli, and a high nuclear to cytoplasmic ratio. Hence, a cytological diagnosis of Myoepithelial carcinoma of soft tissue was suggested and urgent excisison was advised.

Both masses were resected. The midline neck mass measured 10 x 8 x 8cm and the post-auricular mass measured 4 x 3cm. Both the masses were skin covered, solid, well circumscribed, multinodular, grey-white, firm with foci of hemorrhage, necrosis & myxoid change.

Histopathogical examination of both the masses showed well circumscribed, multinodular tumor located in the dermis & subcutaneous tissue, separated by fibrous septa. The tumor was composed predominantly of spindle cells arranged in fascicles & reticular pattern along with nests of plasmacytoid cells. Extensive hyalinisation with foci of necrosis & myxoid change were also seen. The neoplastic cells had moderate amount of eosinophilic to clear cytoplasm, moderate anisonuleosis, vesicular chromatin & 0-1 nucleoli and 2-4 mitosis/HPF [Figure 2].

Immunohistochemically, the tumor cells were positive for pan-cytokeratin, vimentin, smooth muscle actin, calponin, CD10 & S-100 [Figure 3]. While EMA, desmin, myogenin, HMB45, BCL2 & CD34 were negative. Thus, confirming the diagnosis of myoepithelial carcinoma of soft tissue.

Discussion

The myoepithelial tumors are common in salivary gland (1.5%),[5] and rarely occur in extrasalivary locations, such

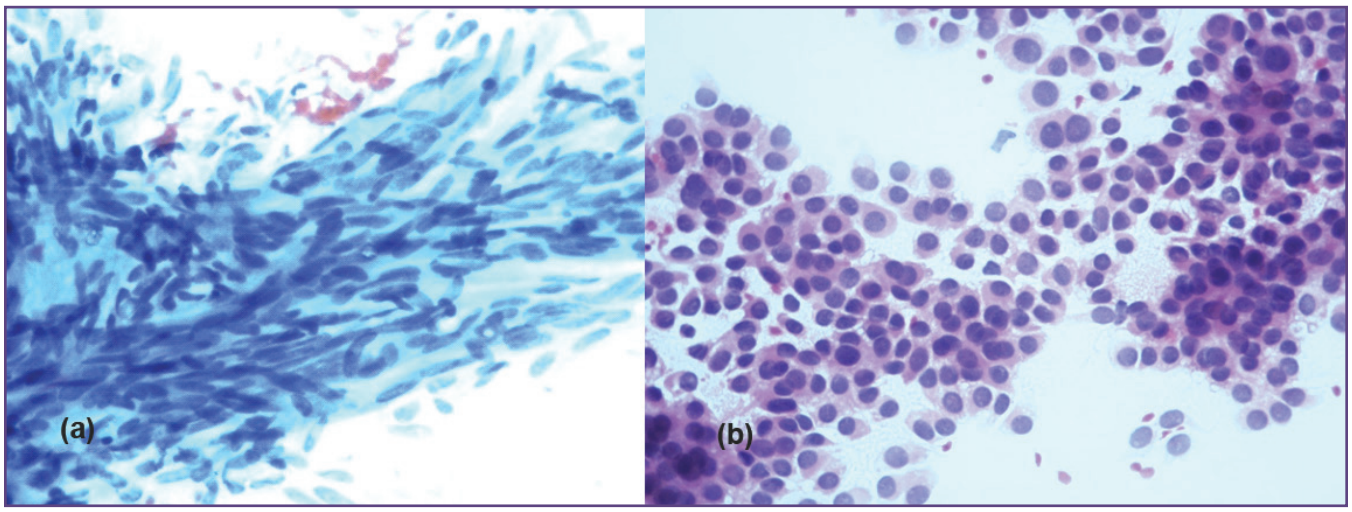

Fig. 1: FNAC smears of neck mass showing (a) spindle shaped cells (Papanicolaou, 400x) & (b) plasmacytoid cells (H&E, 400x)

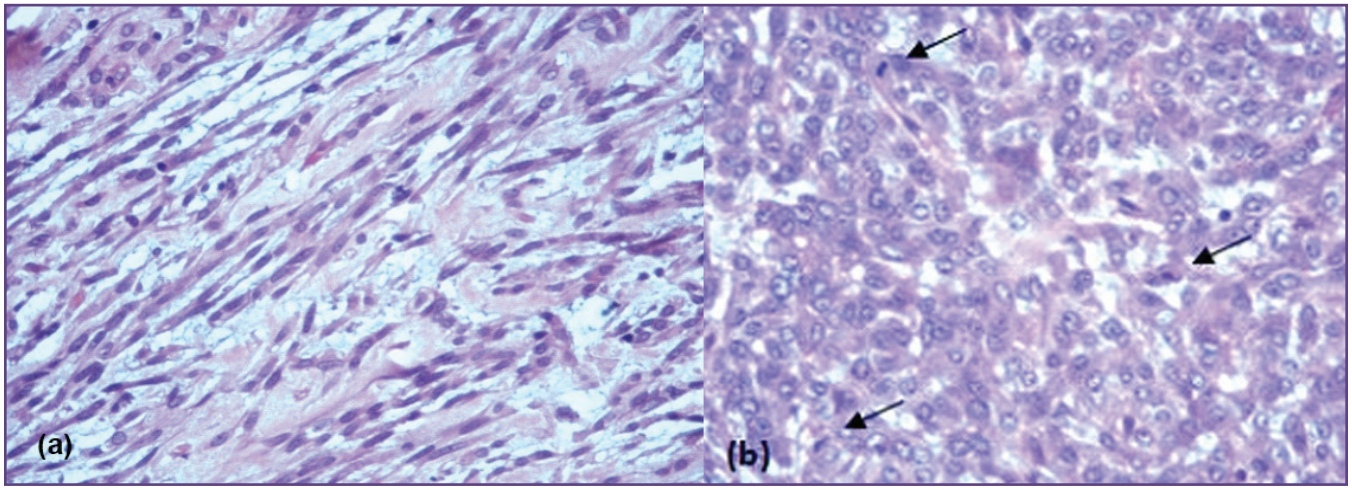

Fig. 2: The tumor revealed (a) fascicles of spindle shaped cells [H&E, 400x] (b) nests of plasmacytoid cells with frequent mitoses (arrow) [H&E, 400x]

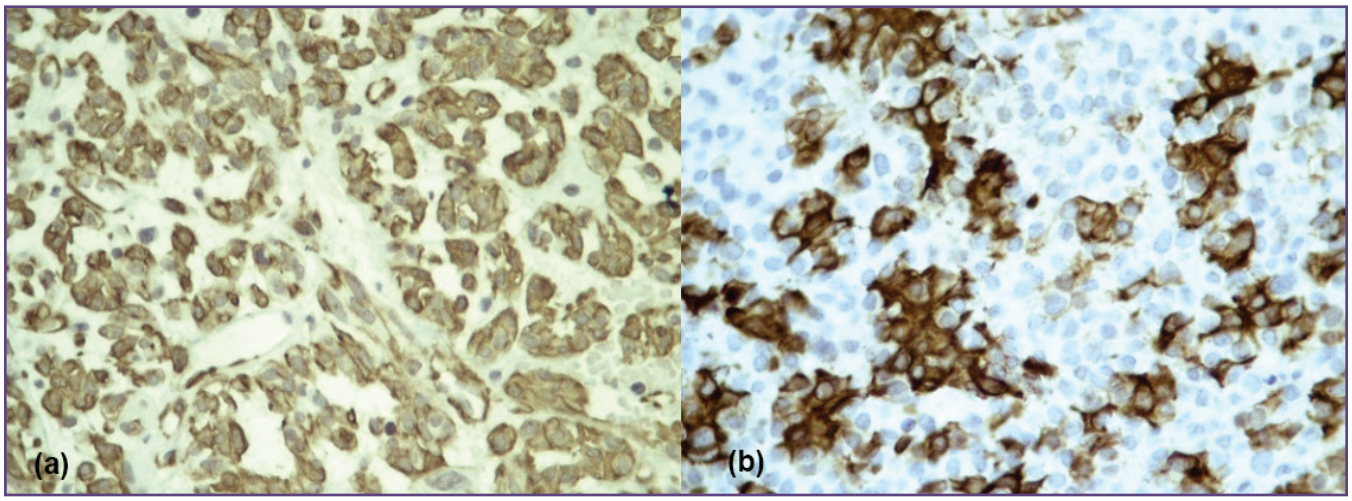

Fig. 3: The tumor cells showing positivity for (a) Calponin [400x] & (b) Pan-cytokeratin (focal) [400x]

as soft tissue.[2-4,6,7] Myoepithelial carcinoma is usually diagnosed in third to fifth decade.[8] Approximately 40% of all soft tissue myoepithelial tumours are malignant.[3]

Preoperative cytological diagnosis of soft tissue myoepithelial carcinoma with diverse cytomorphology is difficult, as it may be mistaken for soft tissue tumor.[6] On cytology, Darvishian F et al and Chhieng DC et al described that nuclear pleomorphism, coarse chromatin, prominent nucleoli, mitosis & necrosis were seen exclusively; however not always, in malignant myoepithelial lesions.[9,10] Hornick and Fletcher et al suggested that even moderate cytological atypia such as prominent nucleoli, vesicular or coarse chromatin & pleomorphism in soft tissue myoepithelioma should be regarded as myoepithelial carcinoma.[3] Histologically, an infiltrative growth pattern is insufficient to diagnose soft tissue myoepithelial carcinoma.[3,7] Moderate to severe nuclear atypia is the only reliable criteria for malignancy.[1] Treatment includes wide surgical excision with lymph node dissection & radiotherapy.[2,8]

Conclusion

Myoepithelial carcinoma should be considered in the differential diagnosis of soft tissue epithelioid and spindle cell neoplasms. Histopathology & immunohistochemistry are essential for unequivocal diagnosis.

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Competing Interests

None declared.

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