

Subcutaneous Myoepithelial Carcinoma of Extremity: A Case Report

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Abstract: *Myoepithelial carcinoma of soft tissue is an uncommon malignant tumour. Development of this tumour is considered to be of uncertain differentiation and histogenesis (1). Myoepithelial carcinoma arising from salivary gland is a well known entity, but that in the soft tissue is less reported. We report a case of myoepithelial carcinoma in a 50 year old male, presented with a subcutaneous swelling in left forearm. Microscopically, diagnosed by the lobular growth pattern with cells having eosinophilic and clear cytoplasm with vesicular nuclei, mitotic figures and areas of necrosis. Stroma was hyalinized and myxoid. IHC panel aided in the final diagnosis.*

Keywords: Myoepithelial tumours, Myoepithelial carcinoma of soft tissue

1. Introduction

Myoepithelial tumors comprise clinicopathologically heterogeneous group of tumors, that ranges from benign to malignant lesions. Myoepithelial carcinomas of soft tissue share the morphological, immunophenotypic and genetic features with their counterparts in salivary gland (1). Recent molecular pathogenesis includes EWSR1 gene rearrangement in soft tissue myoepithelial carcinoma (2).

2. Case Report

50 year old male patient presented with swelling on medial aspect of left forearm for 2 months duration. Clinical examination revealed single, painless, firm, swelling with restricted mobility. MRI showed well defined heterogeneously hyperintense encapsulated lesion involving deep subcutaneous plane in medial aspect of forearm showing heterogenous enhancement.

Patient underwent wide local excision. We received a grey white nodular soft tissue with attached fibrofatty tissue measuring 3x2.8x2.5cm. Cut section shows circumscribed lesion with glistening grey white areas. Foci of hemorrhage and necrosis was present. Microscopy showed a focally invasive neoplasm composed of cells arranged in sheets, nests and trabeculae. Neoplastic cells are round to polygonal having eosinophilic and clear cytoplasm with vesicular nuclei. Mild to moderate anisonucleosis of tumour cells, low mitotic count perineural invasion and areas of necrosis were present. Stroma showed hyalinisation and myxoid change (Figure 1, 2, 3, 4). Immunohistochemistry revealed strong positivity for panCK and p63. SMA showed weak positivity and S100 was negative. Ki67 increased - 20%. (Figure 5) Final diagnosis was given as Myoepithelial carcinoma.

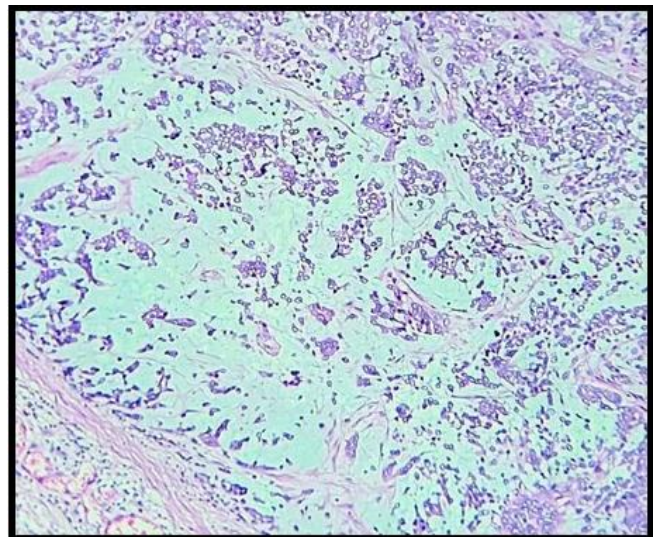


Figure 1

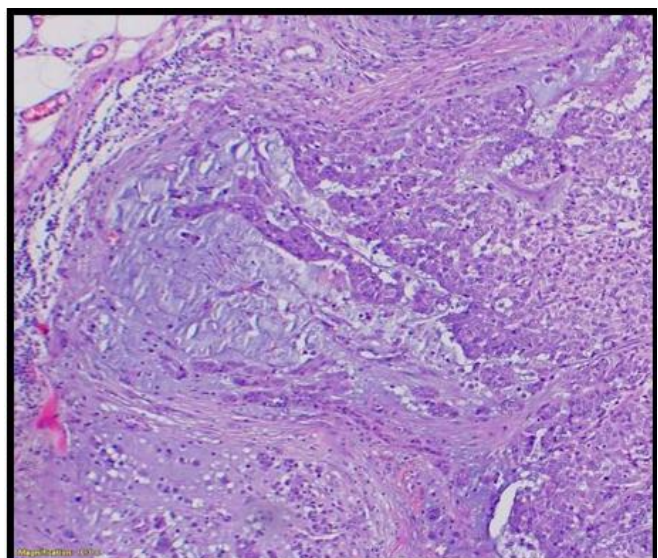


Figure 2

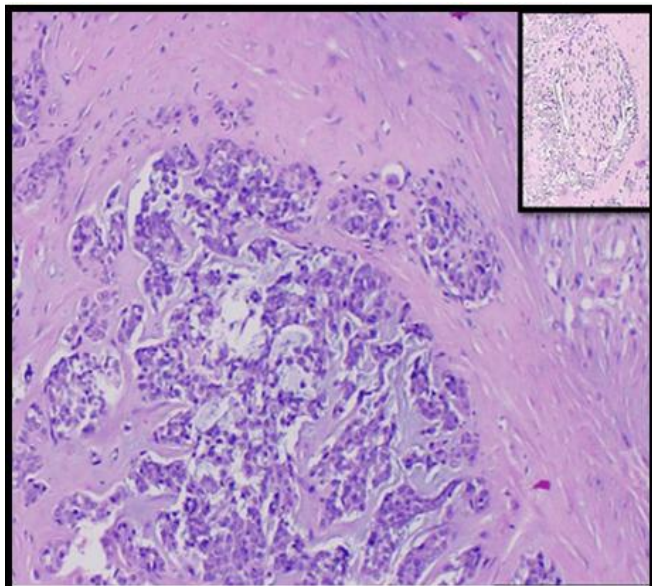


Figure 3

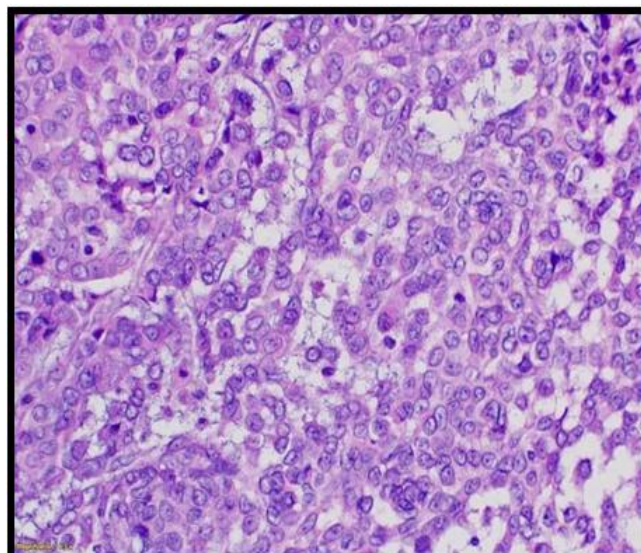


Figure 4

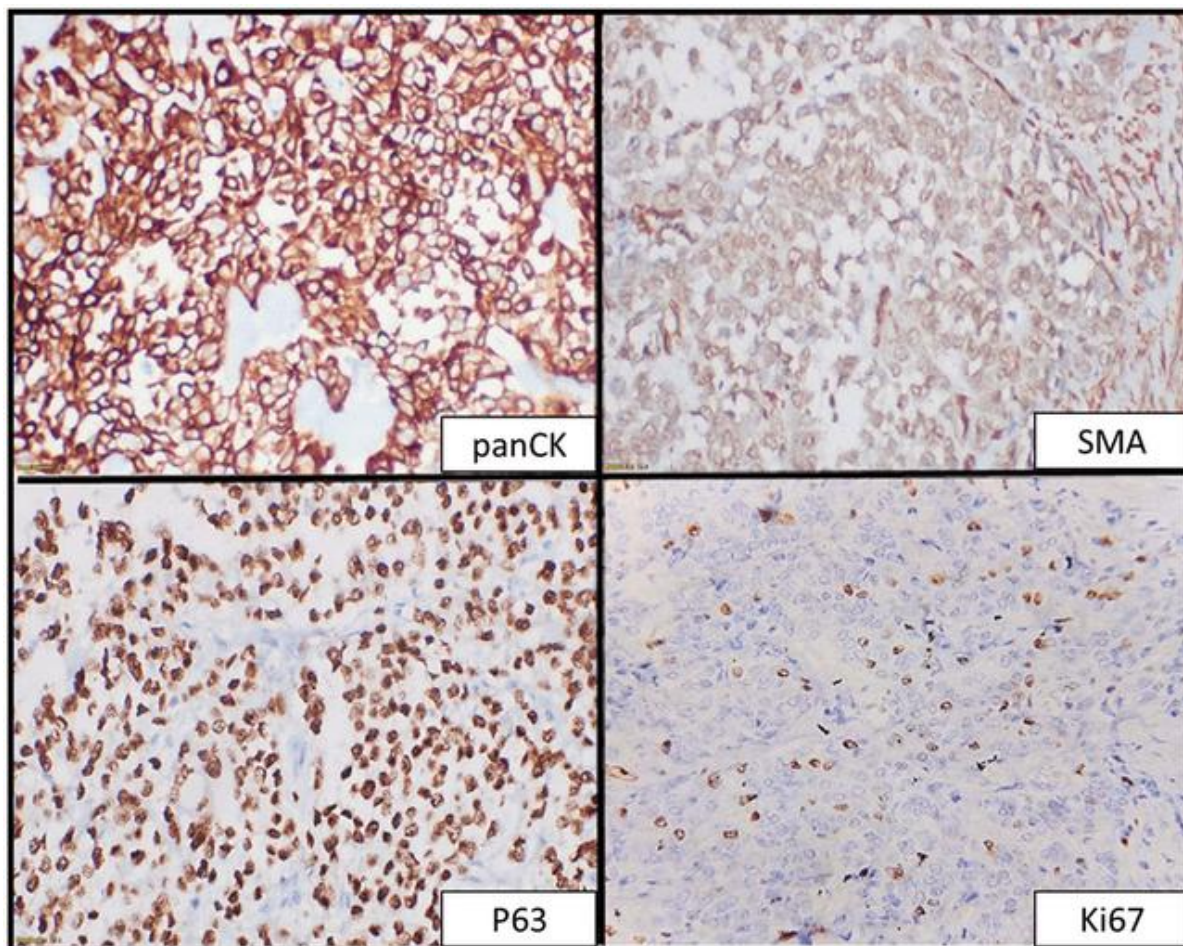


Figure 5

3. Discussion

Soft tissue myoepithelial carcinomas are subgroup of myoepithelial tumours, most commonly arising in the extremities (3). Few cases of intramuscular and subfascial location were reported. Incidence is common in young and middle-aged adults with equal sex predilection. Molecular pathogenesis includes EWSR1 gene rearrangements with

fusion partners like POU5F1 and PBX1 and minority of cases with FUS1 or PLAG1 rearrangement (4). Grossly these tumors have gelatinous, glistening and fleshy appearance. Microscopy exhibit wide morphological spectrum ranges from trabecular, lobular or reticular growth pattern with prominent myxoid stroma. Neoplastic cells are epithelioid, ovoid or spindle having eosinophilic to clear cytoplasm with nuclear atypia, vesicular nuclei, high mitotic count and

necrosis. Osseous, cartilaginous, squamous, adipocytic differentiation were noted in few cases (1). Immunohistochemistry reveals positivity with panCK, S100, EMA, p63, SOX10, SMA. There are no standard criteria for grading of these tumors. Treatment includes complete surgical resection. But tumour show 40-45% recurrence and 20-30% show metastasis to lung, lymph node and bone (2).

4. Conclusion

Soft tissue tumors presenting in the extremities requires multidisciplinary evaluation and histopathologic diagnosis. Myoepithelial carcinomas are rare, aggressive tumors with high chance of local recurrence and metastasis. Effective treatment and follow up required for better prognosis. Newer systemic therapies are under research and clinical trails (5).

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