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Case Report

Pleomorphic Liposarcoma of Rectum: A Rare Entity

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ABSTRACT

Pleomorphic liposarcoma (PLS) is the rarest subtype of liposarcoma. It is an aggressive and fast growing tumor and is commonly located in the deep soft tissues of the lower and upper extremities. The typical age of presentation is 50-70 years. The incidence is approximately 1 in 2,000,000 per year and accounts for 5-10% of all liposarcoma cases. Herein we are reporting a case of pleomorphic sarcoma in a 60 years old male who was clinically diagnosed with carcinoma rectum. This case is being reported in view of its rarity and mimicking clinically as carcinoma rectum.

Keywords: Pleomorphic liposarcoma, Rectum, S-100, Vimentin.

INTRODUCTION

Liposarcoma are malignant neoplasms characterized microscopically by presence of fatty differentiation in the form of lipoblasts. Pleomorphic liposarcoma is the rarest subtype of liposarcoma and is differentiated from other high grade sarcomas by the presence of pleomorphic lipoblasts. [1] This tumor mainly involves the lower limb and upper limb. Other anatomic sites that may be involved are trunk, retroperitoneum, head and neck, abdomen/pelvis and spermatic cord. It is a high grade sarcoma which commonly metastasizes to lungs. The etiology is unknown. Pleomorphic liposarcoma is characterized by diverse chromosomal rearrangements and genomic profiles with complex karyotypes. [2-5] In spite of wide range of histologic appearances, no clinical or pathologic feature is predictive of a more aggressive clinical course.

CASE REPORT

A 60 years male presented to the surgery department of our hospital with complaint of a mass coming from the rectum with bleeding per rectum. On per rectum examination a foul smelling and prolapsed fungating mass measuring 3x2 cms was present. Rest of the physical examination of the patient was within normal limits. An incisional biopsy of the mass was taken and received in our department. Grossly the biopsy consisted multiple grey brown colored soft tissue pieces that measured together 3x2x1.5 cms. Histopathological examination of the stained sections prepared showed extensive areas of haemorrhage, necrosis and dense mixed inflammatory infiltrate along with dispersed population of pleomorphic cells and few multinucleated cells. The cells revealed vacuolated cytoplasm and hyperchromatic nuclei [Fig1]. Few cells showed spindle cell configuration and lipoblastic differentiation were also evident in some of the cells. On

immunohistochemistry the tumor cells were strongly positive for S-100 [Fig 2] and vimentin [Fig 3]. These cells were negative for cytokeratin (CK) [Fig 4], low molecular weight cytokeratin (LMWCK), high molecular weight cytokeratin (HMWCK), desmin, CD 68 and CD 117. Epithelial membrane antigen (EMA) showed focal and patchy positivity. CD 34 was positive in vessels. Based on histomorphology and immunohistochemistry diagnosis of malignant mesenchymal tumor favouring pleomorphic sarcoma was made.

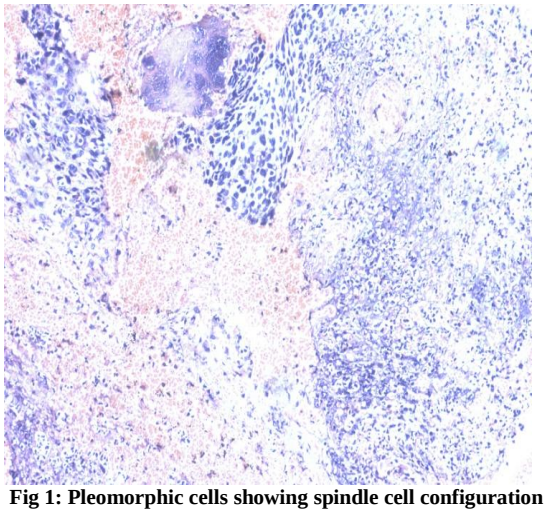

Fig 1: Pleomorphic cells showing spindle cell configuration and lipoblastic differentiation (H&E, 100x)

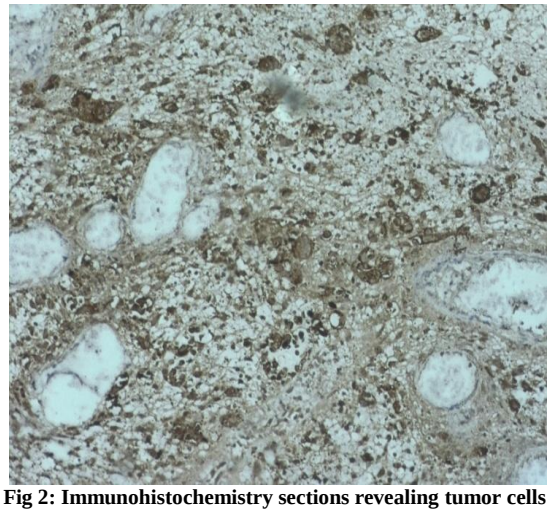

Fig 2: Immunohistochemistry sections revealing tumor cells positive for S-100

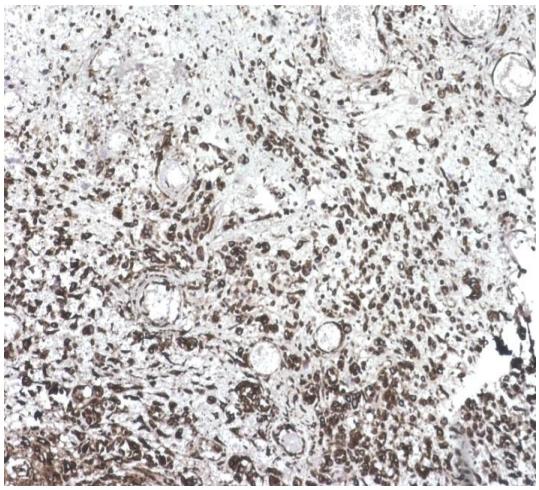

Fig 3: Immunohistochemistry sections revealing tumor Cells positive for vimentin

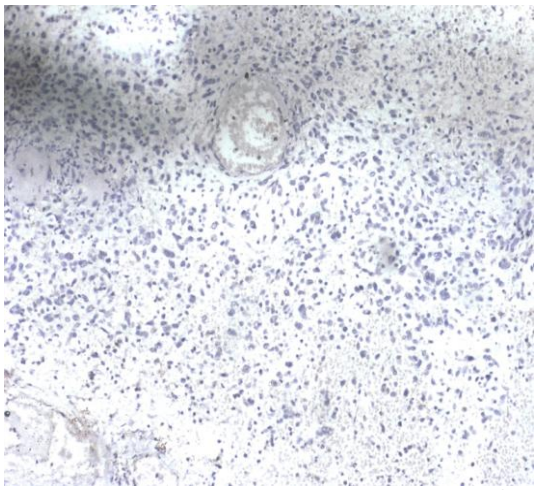

Fig 4: Immunohistochemistry sections revealing tumor for cells positive negative for CK

DISCUSSION

Recognition of pleomorphic liposarcoma requires careful histological and immunohistochemical examination. The differential diagnosis depends upon the predominant pattern which could be epithelioid or Malignant Fibrous Histiocytoma (MFH)-like. The differential diagnosis of epithelioid like variant includes carcinoma, epithelioid sarcoma and epithelioid leiomyosarcoma. The carcinomas which could be difficult to differentiate with epithelioid PLS includes renal cell carcinoma and adrenal cortical carcinoma. Careful histological and immunohistochemical of a panel of markers can help in differentiating and reaching the correct diagnosis. Cells of PLS show large lipid droplets and are negative for EMA but show focal positivity for CK in 50% of cases whereas those of renal cell carcinoma and adrenal cortical carcinoma show fine intracytoplasmic lipid droplets. Also cells in renal cell carcinoma are positive for EMA

while cells in adrenal cortical carcinoma are negative for both CK and EMA, Other differential diagnosis of pleomorphic liposarcoma includes some high grade pleomorphic sarcomas like pleomorphic leiomyosarcoma, pleomorphic rhabdomyosarcoma, pleomorphic malignant peripheral nerve sheath tumor and MFH. Careful light microscopic examination is necessary to differentiate these tumors from pleomorphic liposarcoma. [6]

Pleomorphic lipoma is the only benign condition that can be difficult to differentiate from pleomorphic liposarcoma. It arises as a well circumscribed subcutaneous mass in the posterior neck, shoulders or back in middle aged males. [7] Histologically pleomorphic lipoma consists of adipocytes, atypical hyperchromatic cells and giant cells. However pleomorphic lipoblasts which are characteristic of PLS are absent in pleomorphic lipoma. Also pleomorphic lipomas stain strongly for CD 34 and help in differentiation it from PLS. [8]

The etiology of pleomorphic liposarcoma is unknown. However it is characterized by diverse chromosomal rearrangements and genomic profiles with complex karyotypes. Each category of liposarcoma has a characteristic cytogenetic abnormality. Well differentiated/ dedifferentiated liposarcoma are characterized by ring or giant marker chromosomes derived from 12q13-15. Myxoid/round cell liposarcoma share the characteristic t (12; 16). The cytogenetics of pleomorphic liposarcoma is complex and suggests the probability of multiple molecular aberrations rather than single "oncogenic addictions". Tumor suppressor pathway dysregulations commonly occur in pleomorphic liposarcoma including Rb^{22} and p53. [9] Mutations of p53 contribute to chemoresistance, [10] possibly explaining pleomorphic liposarcoma therapeutic resistance and suggesting that reconstituting p53 function in these tumors may be fruitful. [11]

Treatment involves the surgical excision of the tumor and surrounding normal tissue. In rare cases amputation of the limb is necessary. Tumors that are large or marginally respectable may be treated with preoperative chemotherapy. Adjuvant radiation is recommended if the surgical margin is narrow or positive for sarcoma. Common drug able targets are yet to be discovered. Keeping in mind the molecular complexity, novel therapeutic combinations rather than single target therapies to block multiple pathways and processes may constitute the best anti-PLS approach. Lifelong follow up is necessary in order to monitor for recurrence at the initial site as well as distant metastasis. Pleomorphic liposarcoma has the poorest prognosis of all the subtypes. Five year survival is 59%.

CONCLUSION

Pleomorphic liposarcoma is rare tumor that commonly occurs in adults in the deeper tissues of extremities. Diagnosis requires careful light microscopic and immunohistochemical examination. Early diagnosis is important as it is a fast growing tumor that behaves as high grade sarcoma and often metastasizes especially to lungs.

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