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Follicular Dendritic Cell Sarcoma in Colon: A Rare Case Report

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ABSTRACT
Follicular dendritic cells are localized in germinal centers of lymphoid follicles and play an integral role in regulation of the germinal center reaction and present antigens to B cells. A 22 year male patient presented with a well-defined mass around splenic flexure. On resection cut surface was smooth, fleshy and tan. On microscopy large pleomorphic cells with vesicular nuclei and prominent nucleoli intermingled with mature lymphocytes were present. Three mitotic figures /10HPF were noted. IHC was positive for Vimentin, CD21, CD23. Based on histological appearance and immunoprofile of the tumor, the diagnosis of Follicular dendritic cell sarcoma was made.
Key words: *FDCS, IHC, CD21, CD23.*

INTRODUCTION

FDCS is a neoplastic proliferation of spindle to ovoid cells showing morphologic and immunophenotypic features of follicular dendritic cells. It is an extremely rare neoplasm accounting only for 0.4% of all soft tissue sarcomas[1].

Earlier studies showed nodal disease in 31% of cases, extranodal in 58% and in 11% combined nodal and extranodal disease[2]. The scarcity may be partially due to under-recognition of this entity, particularly when they occur in extra-nodal sites.

Most important characteristics of this tumor is its similarity, morphologically and immunophenotypically, with both Hodgkin and Non-Hodgkin lymphomas. FDC is positive for EBV as it expresses CD21 (EBV receptor). Many studies have also found its association with Castleman's disease and it may represent as its precursor lesion. FDC sarcoma has a significant recurrent and metastatic potential [1] and for these reasons it should be viewed as an intermediate grade malignancy [3].

CASE REPORT

A 22 year old male presented with abdominal pain and partial intestinal obstruction. On radiological examination, well defined, round to oval intestinal mass, of approximately 5.5 cm in diameter was seen around splenic flexure. Based on these findings, intestinal resection was advised. On gross, resected specimen of colon measuring 18 cm in length received. Outer surface showed a nodular well circumscribed mass. Cut surface showed intra-luminal non-ulcerated firm, smooth, fleshy and tan mass measuring 6 x 5 x 5 cm in size. No areas of necrosis, perforation and hemorrhage were noted (Figure 1). Surrounding mucosa was normal. On microscopy, large pleomorphic cells with vesicular nuclei and prominent nucleoli were seen intermingled with mature lymphocytes (figure 2, 3). Three MF/10HPF was noted. A large cell neuroendocrine tumor was suspected and IHC was advised that showed positivity for Vimentin, CD21, CD23 (figure 4) and was negative for CK7, CK20, CK8&18, CK5&6, synaptophysin, chromogranin, S100, MelanA, HMB45, CD117, LCA, CD20, CD3. Based on histological appearance and immunoprofile of the tumor, the diagnosis of Follicular dendritic cell sarcoma was rendered.

Figure 1: Gross appearance of the mass.

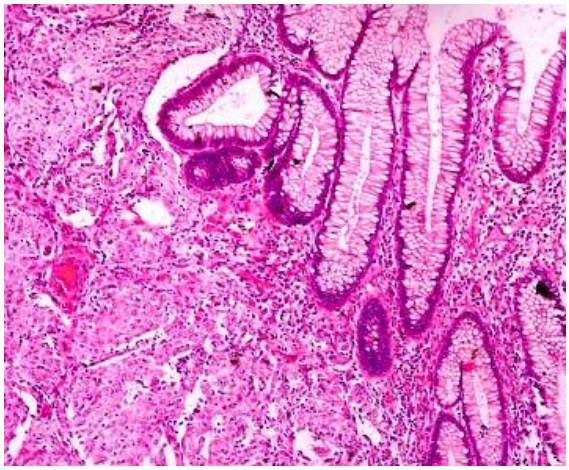

Figure 2: Low power view showing tumor mass adjacent to intestinal mucosa (H&E 100x)

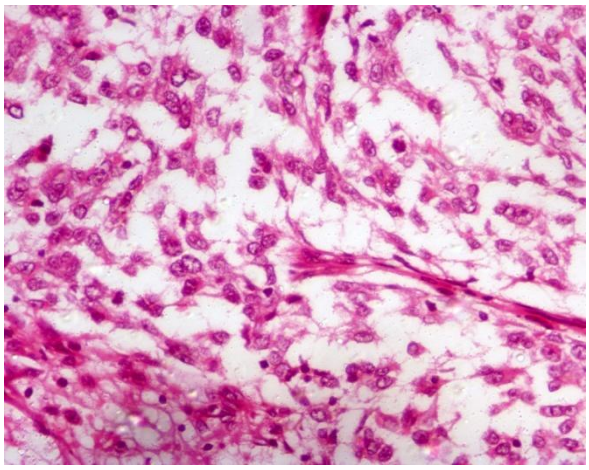

Figure 3: High power view showing large pleomorphic cells intermingled with mature lymphocytes (H&E 400x)

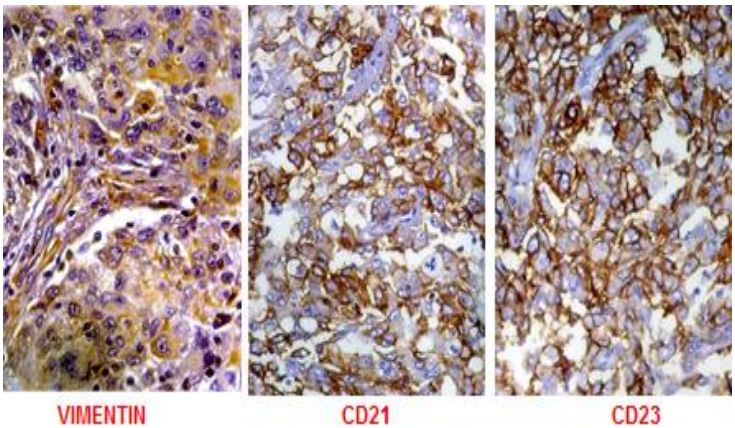

Figure 4: IHC showing Vimentin, CK21 and CK 23 positive (IHC 400x)

DISCUSSION

FDCS is an extremely rare tumor. Moreover its misdiagnosis has added to its rarity. In recent years, better understanding of its immuno profile has helped us make the diagnosis. Its biological behavior is usually indolent, much like a low or intermediate grade soft tissue sarcoma [3]. Most patients are treated by complete surgical resection, with or without adjuvant radiotherapy or chemotherapy, though the local recurrence rate is more than 50% and metastases occur in about 25% of patients [4] .

Based on recent literature, any "undifferentiated carcinoma" that shows syncytial cells containing vesicular nuclei and lymphocytic infiltration, should raise the suspicion for FDCS. Thereafter, a panel of Immunohistochemistry would help to confirm the diagnosis. As the treatment and prongosis for FDCS and undifferentiated carcinoma or lymphoma, are entirely different, therefore the correct diagnosis is of utmost importance.

Proliferation of FDC is characteristic of many neoplastic conditions including follicular hyperplasia, follicular lymphoma, nodular lymphocyte predominant Hodgkin's disease and angioimmunoblastic T-cell lymphoma. Pathological diagnosis is challenging and may require a combination of morphological, immunophenotypical, cytochemical, and electron microscopic analyses. On gross pathology, FDCSs are well-circumscribed tumors with a tan cut surface along with areas of necrosis and cystic changes, especially within larger tumors. On microscopy plump spindled to ovoid cells with eosinophilic cytoplasm and distinct cell borders are seen. These cells are arranged in a fascicular, whorled or storiform pattern, typically infiltrated by scattered small lymphocytes. Necrosis, marked cellular atypia, high mitotic rate and abnormal mitosis may occur and indicate an aggressive behavior [5].

Entities most commonly mimicing FDCS include undifferentiated carcinoma, lymphoma, malignant fibrous histiocytoma, peripheral nerve sheath tumor, inflammatory pseudotumor, granulomatous inflammation, gastrointestinal stromal tumor, and unclassified sarcoma [6].

IHC (immunohistochemistry) is required to confirm the diagnosis. FDCS usually show reactivity for CD21, CD23 and CD35. However, diagnosis may be difficult, mainly because follicular dendritic cell markers are not included in a routine IHC panel.

Etiopathogenesis of FDCS remains unclear. Castleman's disease, which is a benign lymphoproliferative disorder, has been suggested, as a possible precursor lesion for this tumor [7]. As in the hyperplasia-dysplasia-neoplasia sequence proposed for development of some epithelial neoplasms, FDCS may occur in lymph nodes harboring dysplastic FDC, eg. in Castleman's disease. Some studies have reported clonal expansion of FDC in Castleman disease [8]. Epestein-Barr virus is involved in pathogenesis of a small subset of FDCS, but most of them were reported as "inflammatory pseudotumor - like follicular dendritic cell sarcoma". It is a variant of FDCS with different clinical and pathologic features. Due to the rarity of EBV infection associated with classical FDCS [9]. The pathogenesis of this variant may be different. There are also some reports suggesting an association between FDCS and autoimmunity.

Biological mutations of FDCS have been exploited for diagnostic purposes. Characteristically FDCS have mircotubuloreticular structures (MTRS) and increased levels of intracellular clusterin. MTRS contribute to microtubule formation of many structures including the mitotic spindle during cell division. Clusterin is a heterodimeric protein that

aids in the clearance of cellular debris and is involved with apoptosis. Clusterin can be stained to help distinguish FDCS and is involved in the many important cancer hallmarks including resistance to cell death and evading growth suppressors[10, 11].

FDCS is a rare entity that can clinically mimic other tumors. Identification of a different pattern of histopathological features requires further analysis with immunohistochemical stains for follicular dendritic cells. Proper characterization and treatment planning are mandatory due to their recurrent and metastatic potential.

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