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Follicular Dendritic Cell Sarcoma of Colon Presenting as Colo-colic Intussusception: a Rare Entity in Gastrointestinal Site With Extremely Rare Presentation

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ABSTRACT

Follicular dendritic cell sarcoma is a rare mesenchymal tumor of follicular dendritic cells arising from nodal and extranodal site. FDCS generally involves the lymph nodes while many extranodal sites are also affected. We report a case of follicular dendritic cell sarcoma primarily arising in colon and presenting as colo-colic intussusception.

Keywords: Follicular dendritic cell sarcoma, Colo-colic intussusception, Extranodal.

Abbreviation: EBV- Epstein-barr virus, FDCS- Follicular dendritic cell sarcoma, VEGF- Vascular endothelial growth factor.

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INTRODUCTION

Follicular dendritic cells are accessory cells of immune system, also known as dendritic reticulum cells, are required for humoral response by providing antigen presentation and B-cell response. They are present in the germinal centers of primary and secondary lymphoid follicles of the nodal and extranodal sites.¹

Follicular dendritic cell sarcoma is a malignant neoplasm composed of spindled to ovoid cells showing morphological and immunophenotypic features of FDCs. Extranodal FDSC has been described in patients of age group ranging from 9-82 years of age with average age group in fourth decade.² There is no sex predilection. However, there is a female predominance present in EBV(+) inflammatory FDSC, a separate entity described by recent WHO GIT classification, which was previously called as inflammatory pseudotumor like FDSC.³ Gastrointestinal tract is an uncommon site for this tumor and a total of 18 cases in gastrointestinal tract have been reported in literature so far³, out of which only one case has presented with colonic intussusception.⁴ We report a case of follicular dendritic cell sarcoma arising in gastrointestinal tract in a 19 year old female and presenting as colo-colic intussusception.

CASE REPORT

A 19 year girl presented to the emergency with complaints of pain abdomen and chronic constipation for 3 months. She had also complaints of loose motion and bleeding per rectum for last 10 days.

There was no history of trauma. No history of malena, hematemesis, jaundice, urinary disturbance was present.

Ultrasound abdomen was done and showed circumferential thickening of rectum. Due to gaseous abdomen, further assessment could not be done properly. X-ray abdomen was advised to rule out large bowel obstruction and CECT was advised for any rectal growth.

CECT was performed and showed a heterogenous mass extending from left colonic flexure till the rectum, proximal to it the colonic flexure was infiltrated with mesenteric fat and vascular structures that lead towards the mass. The findings were consistent with colo-colic intussusception.

Exploratory laparotomy was done and an intraluminal colonic growth was found. Reduction of transverse colo-colic intussusception with resection of colonic growth with transverse colo-colic resection and loop diversion was done.

We received a specimen of large intestine measuring 7x7x5.5 cm. A well-circumscribed polypoidal solid fleshy growth was identified protruding through the mucosa measuring 6x5x3.5 cm. On cut the growth was grey-white and homogenous. There were no areas of hemorrhage or necrosis seen.

Microscopically a tumor was identified predominantly involving the submucosa and in few areas reaching upto the mucosa. The tumor cells were arranged in sheets, whorls and fascicles with admixed lymphocytes. Individual tumor cells were oval to spindle to plump having eosinophilic cytoplasm. They were showing moderate pleomorphism. Binucleated and multinucleated forms were also seen. Mitosis including atypical mitosis were seen. Mitotic count was 8/10 high power field. No areas of hemorrhage or necrosis was seen. Based on histomorphology, the differential diagnosis that were considered.

Follicular dendritic cell sarcoma, Gastrointestinal stromal tumor, Undifferentiated carcinoma, Anaplastic large cell lymphoma, Interdigitating dendritic cell sarcoma, Inflammatory myofibroblastic tumor, Smooth muscle tumor, Histiocytic sarcoma, Metastatic melanoma, Langerhans cell sarcoma. Immunohistochemistry was performed and the tumor cells were positive for Vimentin, CD-21 & CD-23. They were negative for CD-117, CD-34, DOG-1, CK, LCA, S-100, SMA, Desmin, CD-68, HMB-45 & CD1a. Ki-67 labelling index was >50%.

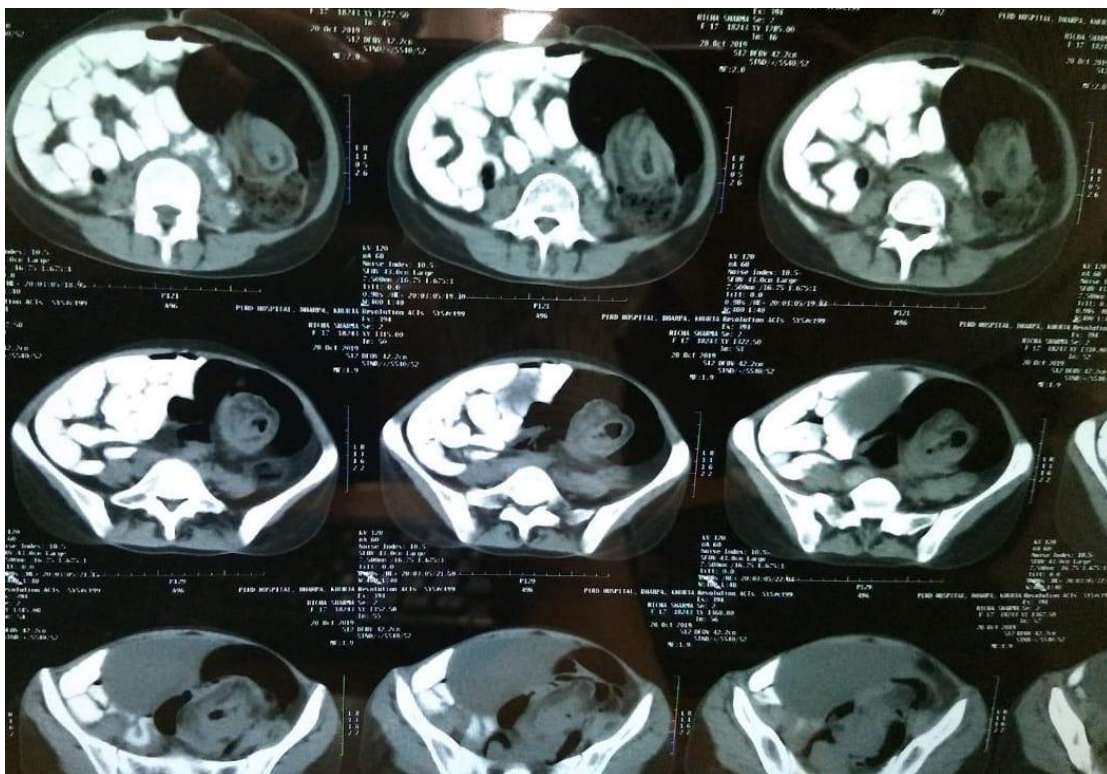
Based on morphology and immunohistochemistry other differentials were ruled out.

Thus based on characteristic morphology of oval to spindle cells arranged in fascicles and whorls

with sprinkling of lymphocytes and immunohistochemical staining of these cells for CD-21 & CD-23, a final diagnosis of follicular dendritic cell sarcoma was made



Xray showing proximal bowel loop dilatation



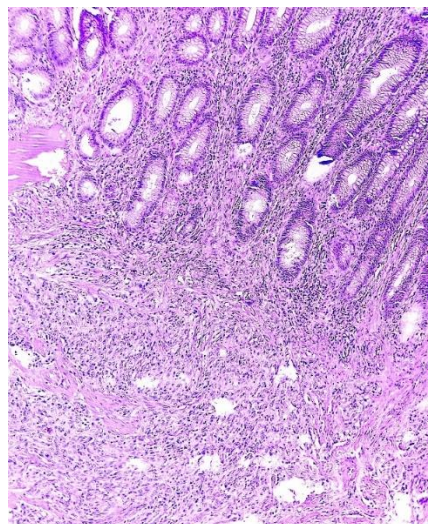
CECT showing features of colo-colic intussusception



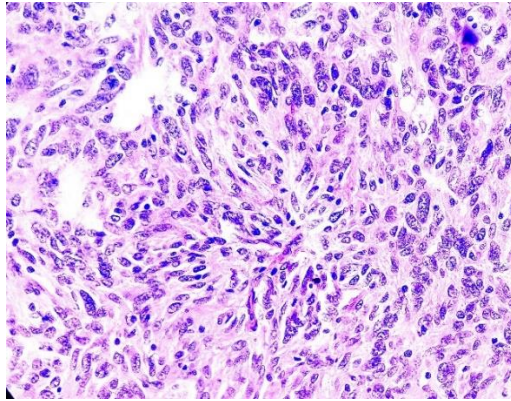
Polypoidal mass protruding through mucosa



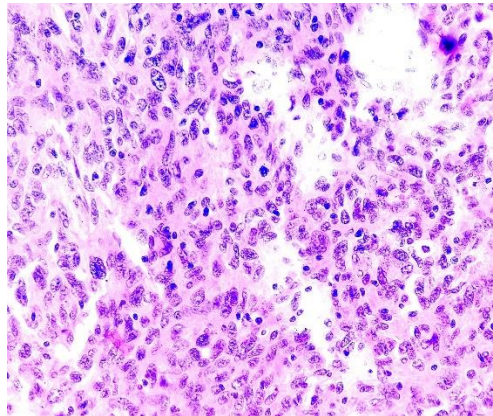
Cut section was grey white and homogenous



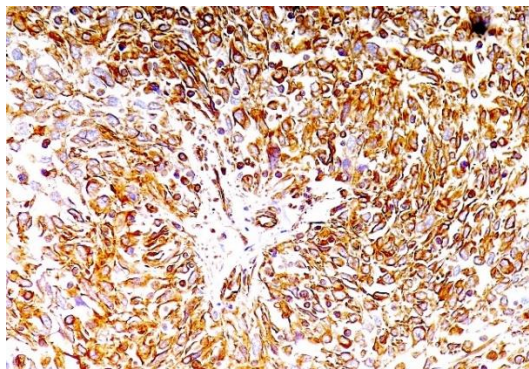
H&E 10X : Tumor cells mostly in the submucosa



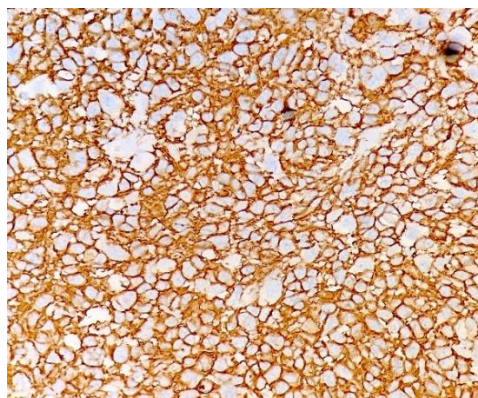
H&E 40X : Tumor cells forming whorls with admixed lymphocytes



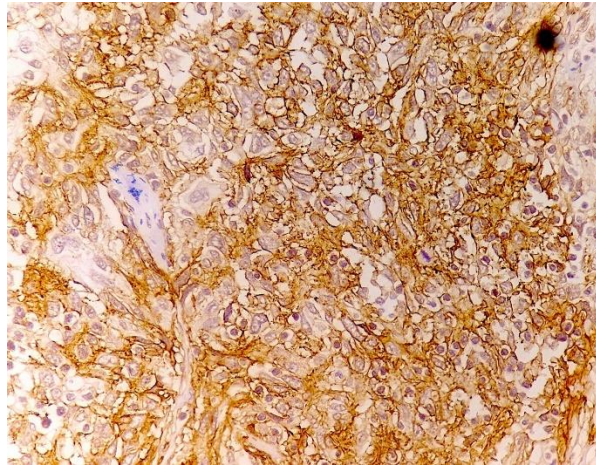
H&E 40X :Tumor cells showing moderate pleomorphism and atypical mitosis



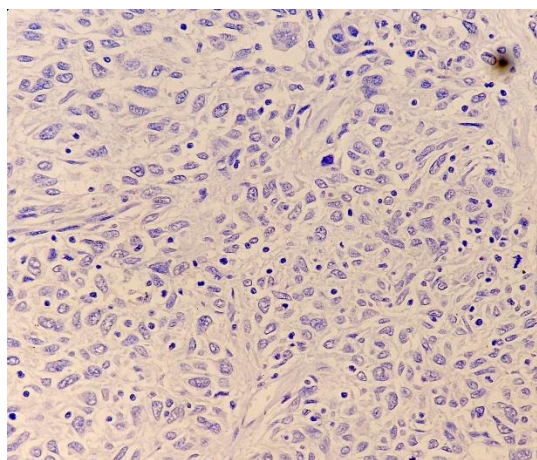
Tumor cells showing cytoplasmic staining for vimentin



Tumor cells showing membranous staining for CD-21



Tumor cells showing membranous staining for CD-23



Tumor cells negative for DOG-1

DISCUSSION

Follicular dendritic cell sarcoma is a low to intermediate grade malignancy³ arising from follicular dendritic cells. FDCS was first described by Monda et al in 1986 and first extranodal site was reported by Chan et al in 1994.^{5,6} Cervical and axillary lymph node groups are most commonly affected. Among the extranodal sites⁷ includes stomach⁸, small bowel⁹, large bowel¹⁰, omentum¹¹, mesentery¹², liver¹³, nasopharynx¹⁴, oral cavity¹⁵, tonsil¹⁶, soft tissues of head and neck region¹⁷, mediastinum¹⁸, spleen¹⁹, lung²⁰ and breast²¹.

Dendritic cells are important cells in immune system. They help in antigen processing and antigen presentation. Follicular dendritic cells which are found in primary and secondary lymphoid follicles²², are mesenchymal in origin²³⁻²⁵. How follicular dendritic cells become

neoplastic is still debatable. There is no driver mutation or translocation found. But FDCS is associated with chromosomal instability, dysregulation of cell cycle progression, nuclear factor kappa beta activation, mitogen-activated protein kinase activation and immune evasion.²⁶⁻²⁸

Histologically the tumor is composed of spindle to plump shaped cells with moderate cytoplasm, oval to plump nuclei, moderate pleomorphism. Whorl, storiform pattern are seen with sprinkling of lymphocytes. Important differential diagnosis include gastrointestinal stromal tumor (GIST), undifferentiated carcinoma, anaplastic large cell lymphoma, interdigitating dendritic cell sarcoma, inflammatory myofibroblastic tumor (IMT), smooth muscle tumors, histiocytic sarcoma, metastatic melanoma and Langerhans cell sarcoma. These entities need to be ruled out by

a panel of long list of immunohistochemical markers. But a careful look at the morphology (oval to spindle cells forming fascicles or whorls with sprinkling of lymphocytes) will clinch the probable diagnosis and immunohistochemistry will aid in the final diagnosis.

A pooled analysis of literature shows local recurrence and distant metastasis rates of 28% and 27% respectively¹. Large tumor size (≥ 6 cm), coagulative necrosis, high mitotic count and substantial cytological atypia are associated with a worse prognosis.^{7,29,30} One analysis of 60 cases of FDCS of extranodal site found that size ≥ 5 cm, high grade histology (mitotic count $\geq 5/10$ hpf or Ki-67 labelling index $\geq 10\%$, coagulative necrosis and cellular/nuclear atypia) were significantly associated with tumor recurrence.² The authors used these features to develop a model for recurrent risk assessment :

Low risk : Tumor size < 5 cm, low or high grade histology; local recurrence 16%

Intermediate risk : Tumor size ≥ 5 cm, low grade histology; local recurrence 46%

High risk : Tumor size ≥ 5 cm, high grade histology; local recurrence 73%.

Our case falls into high risk category with tumor size being > 5 cm and high histology grade (cellular/nuclear atypia, high mitotic count and high Ki-67 labelling index).

Treatment is usually resection of the tumor. Role of adjuvant therapy (radiotherapy or chemotherapy) is not yet proved. FDCS generally has an indolent course but 50% cases develop local recurrence and aggressive clinical course has been reported with metastasis to lung, lymph nodes or liver.^{2,29,31,32} Radiotherapy, chemotherapy or targeted therapy may be useful in cases of refractory or recurrent disease. Advanced disease means unresectable due to size/location or metastatic to other sites. A number of patients of locally advanced disease may be treated with initial neoadjuvant chemotherapy and then they may become surgically resectable candidate.³³ The efficacy of systemic treatment with chemotherapy and

targeted agents for the management of disseminated FDCS is not well defined. The available evidence suggest that the same regimens that are used in advanced soft tissue sarcoma with gemcitabine plus docetaxel are effective in FDCS.^{34,35} Few case reports of multitargeted kinase inhibitors such as pazopanib and sorafenib (target VEGF) are described.³⁶ A case of follicular dendritic cell sarcoma of lung and pleura was treated by surgery, RT and CT but the patient developed disease progression. Mutation analysis showed BRCA-2 mutation gene and further treatment with carboplatin and veliparib achieved disease stabilization.³⁷ BRCA-2 testing may become promising for therapy in future.

To the best of our knowledge, there is only one case report of follicular dendritic cell sarcoma presenting as colonic intussusception². This case has been presented due to its unusual location and unusual presentation as colo-colic intussusception.

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