IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 20, Issue 12 Ser.8 (December. 2021), PP 29-34
www.iosrjournals.org

Rare presentation of Follicular Dendritic Cell Sarcoma arising from transverse colon: a Case Report

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Abstract
Follicular dendritic cell sarcoma (FDCS) is a rare low-grade sarcoma that most commonly presents as a slowly growing, painless mass with the histologic appearance of spindle-shaped cells in a whorled pattern. Occasional cases have an inflammatory presentation associated with fever and weight loss and high-grade histologic features with cellular atypia. This is a case report that FDCS occurs in the transverse colon. To our knowledge, abdominal FDCS is an infrequent entity, and there is a paucity of cases in the literature. A 60-year-old female patient was found to have an intraabdominal mass during a physical examination. Computed tomography revealed a sizeable lobulated oval enhancing solid space-occupying lesion behind the peritoneum. Intraoperative findings discovered a 10x10cm mass in the proximal transverse colon, which was adhered to the adjacent greater curvature of the stomach. The tumour was diagnosed as FDCs by histopathology and immunohistochemical testing. The patient had an unfavourable recovery due to the aggressive clinical course of the tumour.

Keywords: *Transverse colon, Follicular dendritic cell sarcoma, CD23, CD21, CD68,*

Date of Submission: 08-12-2021 Date of Acceptance: 23-12-2021

I. Introduction

A rare tumor with lymph nodes and extranodal region involvement is characteristic of Follicular dendritic cell sarcoma (FDCS). It appears in adult individuals without gender susceptibility. On histopathologically, FDCS contains mainly ovoid and spindle cells budding in follicles, layers, spirals, or a storiform design with slight lymphocytes scattering throughout the neoplasm [1]. The neoplasm cells usually clear follicular dendritic cell indicators comprising CD23, CD21, and CD35. Furthermore, CXCL13, D2-40 and clusterin are frequently positive however not precise. These diverse pathological descriptions identify nodal FDCS quite upfront, assuming that not many kinds of spindle cell neoplasm happen in lymph nodes. Neoplasm of the follicular dendritic cells sarcoma denotes an unusual disease with merely around 200 cases stated globally [2]. Most of the cases appear predominantly in axillary, cervical or abdominal lymph nodes. Though, extranodal source from ancillary lymphatic tissue, same as Waldeyer's ring and tonsils, is also standard [3]. Association of the GI tract is occasional. A study conducted in 2014 in which 106 cases of FDCS had been described with the tumour in 46 cases arising within the abdomen [4]. It is classified alongside with neoplasm of dendritic cells and histiocytes in the WHO taxonomy of neoplasms [5]. Mainly FDCS is diagnosed by histopathological investigation supplemented by immunohistochemistry. Surgical treatment remains the basis of management for FDCS of any site with or without ancillary radiotherapy and chemotherapy [6 7]. Here, we present a patient with FDCS rising from the transverse colon, she was managed with surgery, but the prognosis was poor due to an aggressive clinical course.

II. Case Report

A 60-year-old female came with the complaint of abdominal pain with discomfort. She was also experiencing heaviness, anorexia, weight loss, generalized weakness, loose stools for the past four to six months. Apart from a medical history of vitiligo since 20yrs, she had no other significant comorbidities or history of hospitalization in the past. During the physical examination, she was deeply pallor with moderate dehydration. There was no fever or complaints of dysuria, and her vitals were within normal ranges. Her cardiopulmonary system was unremarkable on systemic examination, but her abdominal examination showed

abdominal distention with hepatosplenomegaly. There was also palpable non-mobile solid mass noted over the umbilical & paraumbilical region.

Complete laboratory workup was done as shown in Table 1. There was inflammation and infection as her total leukocyte count (TLC) was very high. In addition, due to obstruction of bile ducts related to the space-occupying lesion, her alkaline phosphatase was highly elevated. The radiological investigation showed a normal chest X-ray and ECG. Ultrasound abdomen was ordered and showed a lobulated hypoechoic mass in the umbilical and paraumbilical region, which could be lymph node mass. CT abdomen was done in June 2020 due to high suspicion of mass in the abdomen, which showed a sizeable lobulated oval enhancing solid SOL behind the peritoneum. It was located in a right intrahepatic region with focal low attenuation areas of calcifications with hepatosplenomegaly (Figure 1).

Initially, the patient was started on IV antibiotics, PPIs, antiemetic, blood transfusion and other supportive care for four days. After a thorough evaluation and pre-op fitness, the patient was planned for surgical exploration as a treatment option. The exploratory laparotomy with adhesiolysis was done, extended to right hemicolectomy with block excision of greater curvature covering ileostomy.

Intraoperative findings revealed a 10x10cm mass in the proximal transverse colon, which was adhered to the adjacent greater curvature of the stomach (Figure 2). Multiple pericolic lymph nodes present. No signs of liver, peritoneal or omental metastasis was present (Figure 3). The entire resected specimen was sent for histopathology in 10% formalin.

Histopathology findings revealed a brownish tubular structure attached to an irregular mass. The intestinal segment measured 38.0 cm in length, and the diameter varied from 1.5 to 11.0cm. Mass measured 15.0 x 12.0 x 10.0 cm. The outer surface shows perforation and hemorrhagic areas with mesenteric nodules (Figure 4). Cut section of the lumen showed a mass invading the colon and impinging on the serosa, obstructing the lumen. There was no mucosal growth. Its cut section showed variegated areas with necrosis and multiple hemorrhagic areas. Numerous lymph nodes were isolated. On microscopy, sections showed severe atypia with a round to oval cells, high N: C ratio, hyperchromatic nuclei, mild to moderate anisokaryosis, rare mitotic figures. Areas of coagulative necrosis were seen without any lymphovascular invasion. There were seven lymph nodes found to be involved (Figure 4). A diagnosis of poorly differentiated carcinoma was given, and an immunohistochemistry panel was ordered. An extensive immunohistochemistry panel was performed (Table 2). The tumour was positive for HLA-DR, CD 23, CD68, EBV, EBER (focal and weak), vimentin, EMA (focal and weak), Lysozyme, ki 67- low index. It was negative for panCK, PAX5, ALK1, SMA, MUM1, CD43, S100, Desmin, CD34, CD1a. Thereby a confirmatory diagnosis of follicular dendritic cell sarcoma was established.

Postoperatively the patient was under strict observation in SICU for two days. On the 2nd day, the patient was shifted to the regular ward as she was hemodynamically stable with normal vitals. On post-op day 4, she started tolerating liquid diet orally and further was given a soft diet successfully. The postoperative phase was uneventful, and she was managed on I/V antibiotics, antiemetic, PPIs, blood products (PRBCs) and dietary supplements with parenteral feed. Consequently, the patient was discharged on a postoperative day 9 with a colostomy bag and drain in situ. Unfortunately, our patient had an aggressive clinical course and died after few months of surgical intervention.

III. Discussion

The WHO taxonomy of neoplasms of lymphoid tissues and hematopoietic, issued in 2017, outlined FDCS as a cancerous growth of spindle-shaped to oval cells presenting immunophenotypic and morphologic qualities of follicular dendritic cells [8]. Initially, an author in 1978 called it a tumour with many facets and unique etiologic links in his detailed report. The blend of ultrastructural & straightforward microscopic and immunohistochemical evidence is constant with the lymphoid fascicules of follicular dendritic cells [9]. Monda et al. in 1985 described four cases concerning lymph nodes with description indicative of dendritic cell distinction [10]. After that, there were cases regarding FDCs in the literature described by various authors. A considerable study by Saygin et al., available in 2013, indicated that few cases comprise the lymph nodes in the head & neck and the mediastinum, whereas most cases had extranodal involvement and the rest presented with the shared nodal and extranodal disorder [11]. FDCS arising from the extranodal region usually includes the gastrointestinal, nasopharynx, tonsils, soft tissue, Omentum, retroperitoneum, lung and mediastinum [11 12]. Our case showed FDCS from the transverse colon involving lymph nodes, a rare site for dendritic cell tumour.

Histology of FDCS is comprised of fascicles, spirals (like meningiomas) and storiform design of spindle-shaped, elongated and ovoid cells with modest eosinophilic cytoplasm oval nuclei with finely dispersed or vesicular chromatin and lesser diverse nucleoli and unclear cell margins. Cytological atypia is not expected but is seen in few cases. However, some cases may display much complex mitotic function, prominent nuclear atypia and centers of necrosis [13]. Histopathology in our case displayed severe atypia with a round to oval cells, high N: C ratio, hyperchromatic nuclei, mild to moderate anisokaryosis, rare mitotic figures. Similar to our case, Soriano et al. issued a series of fourteen patients with FDCS with 5 cases exhibiting high-grade qualities

histopathologically. Additional three cases revealed crucial high-grade descriptions [14]. Another study from South Asia [15] presented a case series of FDCs in 18 cases. In nine out of eighteen cases, colon mainly was involved. In four cases, they reported positive lymph nodes for metastasis in gross specimens. Total cases were analyzed grounded on appearance sustained by positivity for immunohistochemical dyes CD21 and CD23. Histologic aspects linked with hostile behaviour were appreciated in a majority of the cases. This is significant as these cases were from the same region like ours, and there could be a link in similar clinical course occurring in our part of the world. Li et al. issued an article on FDCS about the abdomen concentrating on the computed tomography (CT) results of these neoplasms. They suggested that FDCS must be integrated with diagnosing abdominal masses when these are comparatively huge, solitary or multiple with precise or poorly-defined edges, complicated inside arrangement with noticeable necrosis and central calcification and assorted prominence outline in the solid portion [16]. CT abdomen in our case was done due to high suspicion of mass in the abdomen, which showed a sizeable lobulated oval enhancing solid SOL behind the peritoneum. It was precisely located in a right intrahepatic region with focal low attenuation areas of calcifications with hepatosplenomegaly.

FDCS grow gradually with an average age of 45 to 50 years, and there is a nearly equal division in females and males [11]. Choi et al. reported a 79-year-old female who showed widespread involvement of bilateral lymph nodes of the head, neck, chest and abdomen [17]. The age of our patient was 60 years, and she had extensive involvement of colon. FDCs are usually significant, as reported by Saygin et al., mean size of 7 cm. However, size ranges from 1 to 15 cm subjected to the position. The biggest FDCs have been described in the abdomen and mediastinum, whereas the lowest was stated in the head and neck area. In our case, the gross findings showed a massive mass around 10x10 cm arising from the transverse colon attaching towards the greater curvature of the stomach.

Few common tumours can look like FDCS morphologically. However, they can be diagnosed separately from FDCS by relating appropriate immunohistochemistry (IHC) panels which can assist in distinguishing these tumours. On IHC, FDCS explicit CD21, CD23 and CD35 and clusterin are generally negative in other dendritic cell neoplasms. Additional markers usually stated comprise fascin, desmoplakin, EGFR, vimentin, podoplanin, and CXL13. Inconsistently stated markers are S100 protein, EMA, CD68. Thus, CD21, CD23 and CD35 are indicators of FDC distinction and are extremely specific and sensitive for identifying FDCS. Table 2 shows the immunohistochemical results in our case, and CD35 was negative in our patient. Nevertheless, other markers associated with the IHC panel for FDCS defined in the literature.

Thorough surgical removal is the utmost extensively used modality and stays the central pillar of management. Due to the paucity of these neoplasms, there is still no agreement on management strategies. Radiotherapy and chemotherapy have frequently been used for partly operated or extensive tumours with variable success levels. The precise role of radiotherapy and chemotherapy in FDCS remains ambiguous. The surgical option was the preliminary management in most of the cases with contained neoplasms [18]. According to findings by Saygin et al. [11] and Chan et al. [8], unfavourable prognostic aspects comprise above five mitoses per high power fields, size bigger than 6 cm, and marked nuclear atypia. Limited neoplasms have an improved prognosis. In our case, the patient underwent surgical treatment, i.e., right hemicolectomy with block excision of greater curvature with covering ileostomy. The mass measured 10 x 10 cm rising from the transverse colon, which was adhered to the adjacent greater curvature of the stomach and significant nuclear atypia on histology, leading to her poor prognosis.

IV. Conclusion

In our case report, the transverse colon was involved. Lymph nodes were involved but negative for metastasis. Unlike a male predominance in most cases, our patient was female with a large abdominal mass measuring 10x10 cm. she was mainly diagnosed based on histopathological descriptions and immunohistochemical markers positivity for CD23 and CD68. Histopathological features associated with aggressive clinical behaviour and our patient died of illness 24 months subsequent surgical removal. Follicular dendritic cell sarcoma is an unusual neoplasm, and treatment is quite possible with an expert surgical intervention and appropriate course in the hospital. More studies to elucidate the role of definitive therapy, i.e., radiation or chemotherapy for FDCS, are recommended.

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Table 1: Laboratory Investigations of our Patient

Analyte	*Units*	*Normal range*	*Results*
Haemoglobin	g/dl	12-15.5	**5.5**
TLC	10^9 /L	4-11	**38.1**
Platelets	10^9 /L	150-400	**366**
Sodium	mEq/L	135-145	**134**
Potassium	mEq/L	3.5-5.0	**3.8**
Urea	mg/dl	7-30	**8**
Creatinine	mg/dl	0.7-1.2	**0.6**
SGOT	U/L	5-34	**15**
SGPT	U/L	< 55	**10**
Alkaline phosphate	U/L	40-150	**873**
Albumin	g/dl	3.5-5.0	**3**

Red: High; Blue: Low

Table 2: Immunohistochemistry report confirming FDCS diagnosis

Markers	*Results*
HLADR	+++
CD 23	+++
CD68	+++
EBV	+++
EBER	***+ (focal and weak)***
Vimentin	+++
EMA	***+ (focal and weak)***
Lyzozyme	+++
ki 67	***+ (low index)***
PanCK	-
PAX5	-
ALK1	-
SMA	-
MUM1	-
CD43	-
S100	-
Desmin	-
CD34	-
CD1a	-

- , negative; +, partially positive; +++, positive.

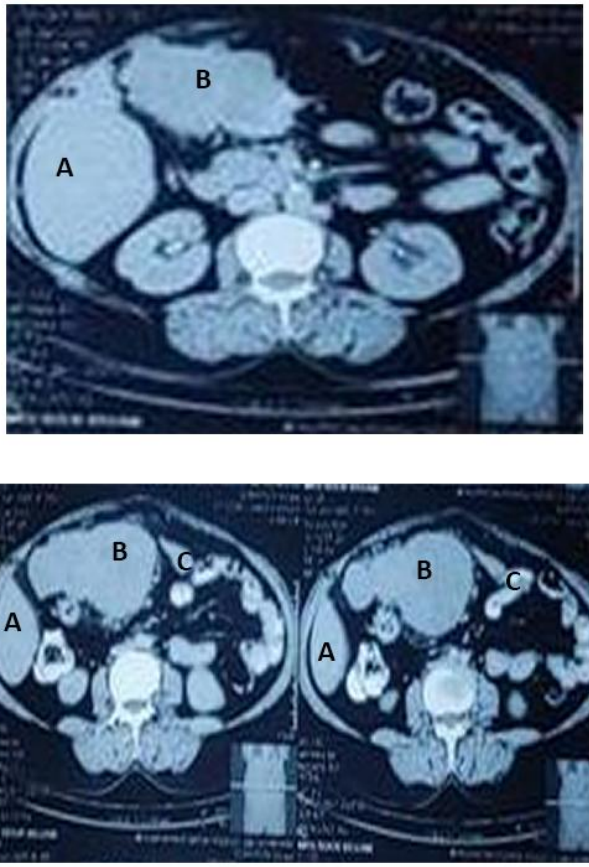

Figure 1: CT Scan Abdomen
A: Liver, B: Large Lobulated Mass; C: Transverse Colon

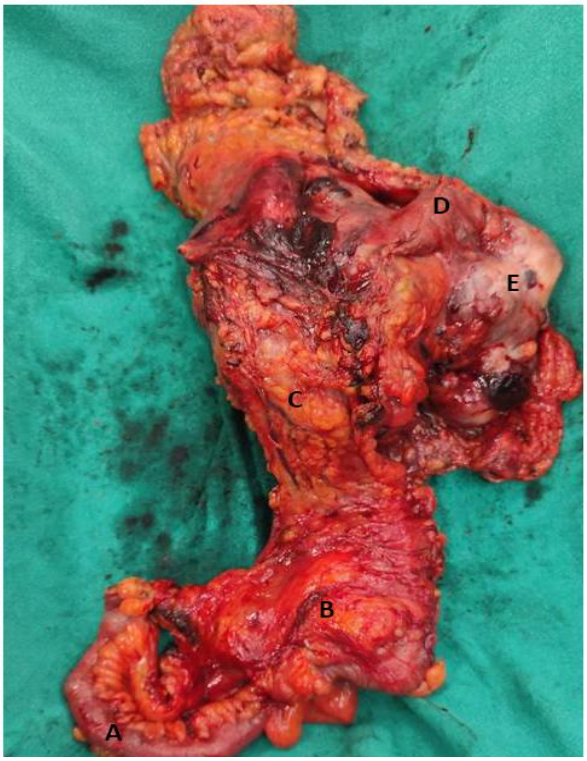

Figure 2: Tumor arising from transverse colon and densely adhere to posterior wall of stomach
A: Ileum; B: Ascending colon; C: Transverse colon; D: Stomach; E: Tumor (Mass)

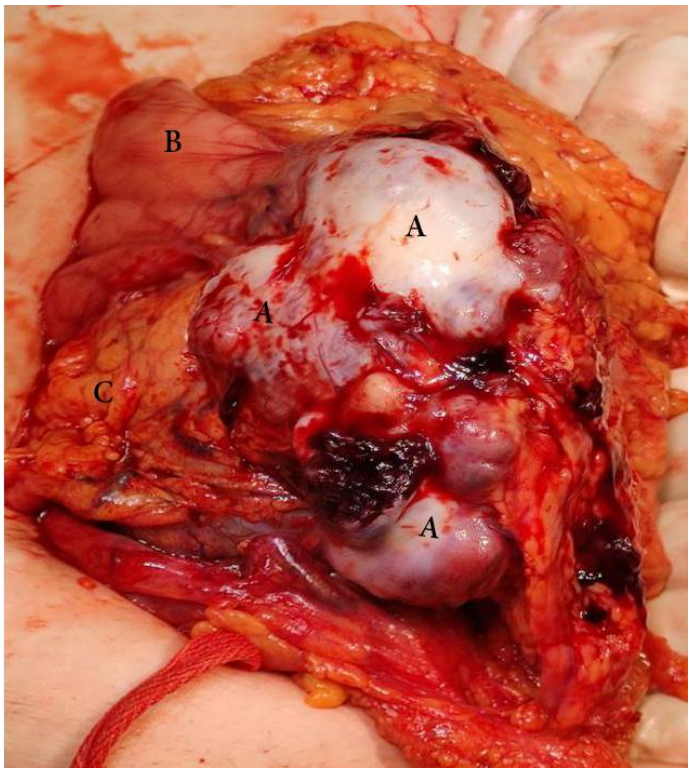

Figure 3: Surgical Specimen of FDCS
A: Tumor depicting FDCS; B: Stomach; C: Omentum

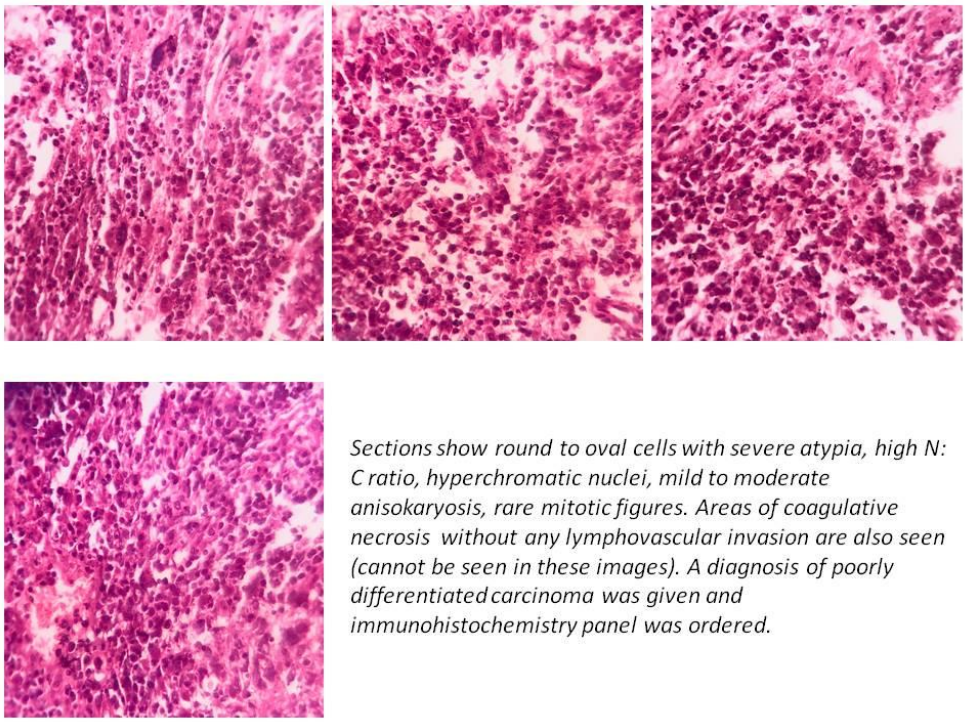

Figure 4: H&E staining of tumor cells

Dr Dipen Patel, et. al. "Rare presentation of Follicular Dendritic Cell Sarcoma arising from transverse colon: a Case Report." *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, 20(12), 2021, pp. 29-34.