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Gastric Follicular Dendritic Cell Sarcoma: A Case Report of a Rare Entity

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Abstract

Follicular Dendritic cell sarcoma arises from the follicular dendritic cells present in the lymphnode. Though is commonly seen in the head and neck area but is extremely rare in the abdomen. Less than eighty cases are reported in the indexed literature. We herein describe a case of follicular dendritic cell sarcoma arising from the stomach wall with infiltration into the pancreas in an 85 year old patient.

Key words: Follicular dendritic cell sarcoma; CD 21, CD 23, CD 35; Gastrointestinal spindle cell tumour

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INTRODUCTION

Follicular Dendritic Cell Sarcoma is a rare neoplasm, arises from the follicular dendritic cell of the germinal center. Follicular dendritic cells are mesenchymal in origin and are involved in Humoral Immunity. These cells express CD 21, CD23, CD 35. These can arise from both nodal and extranodal site[1]. FDCS cases reported in the english literature are extreemly rare and less than eighty in number[2-5]. The involvement of GIT is extremely rare. To our knowledge, only few cases have been described to date and these were located mostly in the colon and retroperitoneum[6-7]. Upon review of literature, we identified only three additional cases of FDCS presenting as primary stomach tumour[8]. To add to the existing literature, we present a case of follicular dendritic cell sarcoma of stomach infiltrating into pancreas.

CASE REPORT

An 85-year-old male with no co morbidities presented with loss of appetite, and early satiety for three months. He had intermittent abdominal pain for two months. Vomiting, altered bowel habits and melaena were absent. A well-defined 14 cm ×10 cm mass involving the left hypochondriac, lumbar, and umbilical areas was found. There were no ascites or palpable neck nodes.

Ultrasonography revealed a solid mass of 15×13 cm with cystic component in the centre with increased vascularity. CT scan (Figure 1) showed mass lesion with large necrotic area, appearing to arise from the posterior wall of stomach with retroperitoneal extension. The lesion was PET avid. EUS guided FNAC was suggestive of either low grade neuroendocrine carcinoma or epitheloid Gastro-intestinal stromal tumour (GIST). Serum chromogranin was 372ng/ml. Routine blood investigations were within normal range.

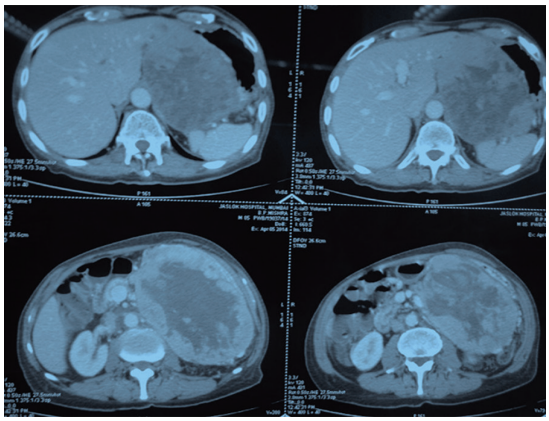

Figure 1
CECT Abdomen

In view of the possibility of GIST being real, he was put on Imatinib Mesylate trial for two weeks. Patient had generalised serositis including pericardial effusion. Reassessment with PET-CT showed no response.

The patient underwent surgical exploration (Figure 2a, 2b) with en-bloc resection of the tumour with wedge gastrectomy with distal pancreatico-splenectomy.

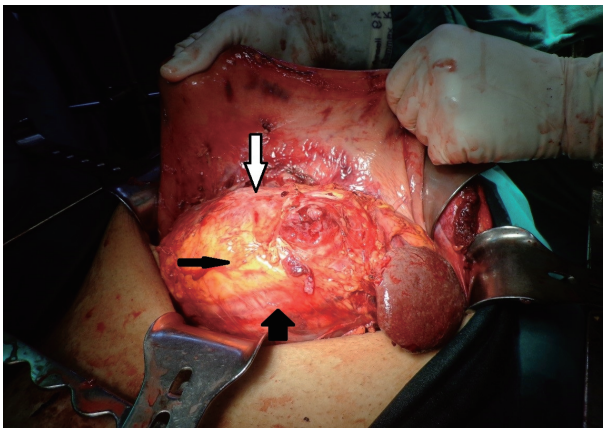

Figure 2a
Tumour Insitu Arising From Stomach Posteriorly Along the Lesser Curvature (White Arrow) With Infiltration Into The Body and Tail Of Pancreas (Black Arrow-Thin). Splenic Vein Was Pushed Inferiorly (Black Arrow- Thick)

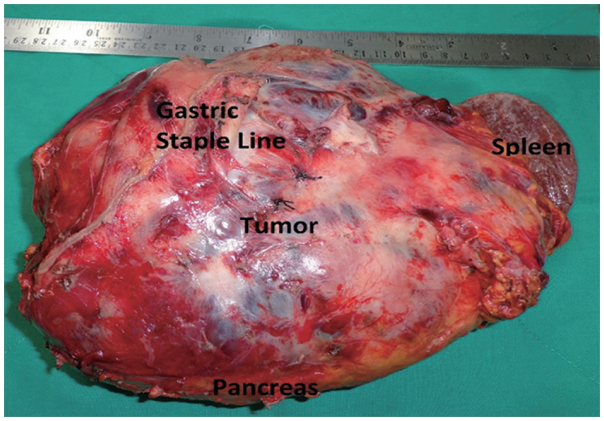

Figure 2b
Enbloc Resected Specimen

On gross examination it was an 18×14 cm solid mass arising from the posterior wall of stomach with infiltration into body and tail of pancreas weighing 2.2 kgs. Histopathological examination (Figure 3) showed spindle shaped cells with a few scattered severed small lymphocytes. There were three mitoses /10 hpf. Tumour cells showed marked pleomorphism. Histopathological examination was suspicious of follicular dendritic cell sarcoma.

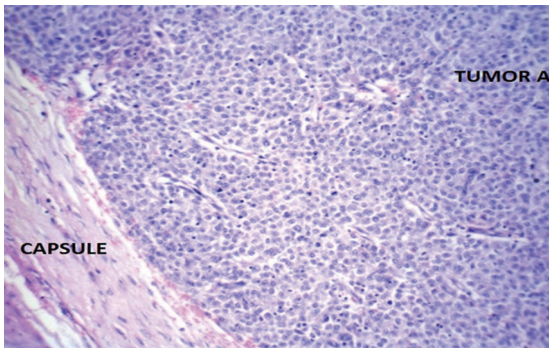

Figure 3
Haematoxilin and Eosin Stained (10X) photomicrograph Showing a Well Formed Capsule and Tumour Cells

On Immunohistochemistry, the tumour cells expressed CD21, CD23, and CD35 (Figures 4, 5 and 6). It was immunonegative for CD117, CD34, DOG1, Desmin, SMA. Thus the diagnosis of follicular dendritic cell sarcoma was confirmed. No adjuvant therapy was recommended. Follow up CECT abdomen at six weeks and 6 months revealed no recurrence.

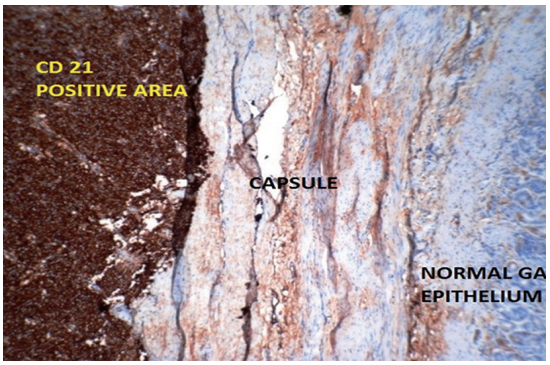

Figure 4
Photomicrograph (4X) Showing a Positive CD 21 Immunoreaction With Capsule and Normal Stomach Interface

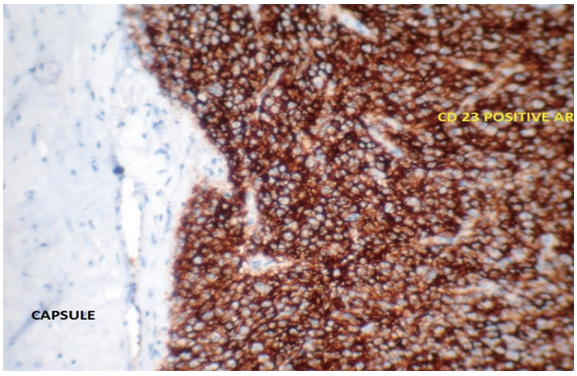

Figure 5
Photomicrograph (4X) Showing a Positive CD23 Immunoreactivity by Tumour Cells

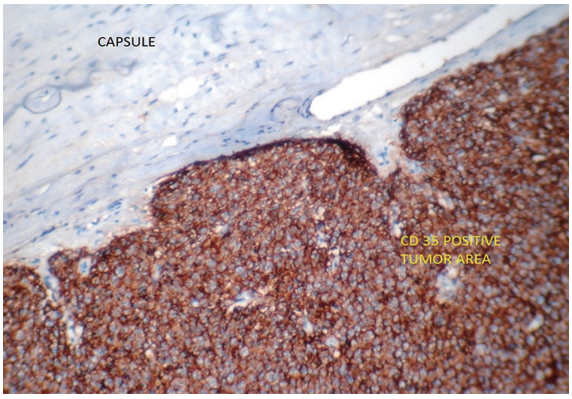

Figure 6
Photomicrograph (4X) Showing a Positive CD 35 Immunoreactivity by Tumour Cells

DISCUSSION

The available indexed English literature search was made in pubmed, directory of open access journals (DOAJ) and google search till date. Mesh terms "Follicular dendritic cell sarcoma", "extranodal fdcs" were used. The relevant literature is reviewed.

FDCS is a rare tumor of dendritic-histiocytic cell origin, less than 80 cases have been reported in the English literature[2-5]. Monda et al. first described FDCS in 1986 and Chan et al reported the first extranodal tumor in 1994[9-11]. Since then the tumor has been reported in various sites like palate, tonsils, stomach, mesocolon, pancreas, breast, small intestine [1-2,6]. Wang et al found that more than half of the reported cases fall in extranodal group. This neoplasm affects both sexes equally, and the disease occurs over a wide age range from 14 to 77 years (mean age: 47 years). The disease typically presents as a painless, slow growing, well-defined mass lesion at the involved site[12]. The causes of FDCS are largely unknown. Associations with Castleman disease[13] and Ebstein-Barr virus[14] have been reported. In addition, relationships have been found with autoimmune disease such as paraneoplastic pemphigus[8] and myasthenia gravis[8]. The diagnosis was challenging as the neoplasm appeared consistent to be gastrointestinal stromal tumour based on morphology and histopathological examination. To our surprise, finally it turned out to be a FDCS, after the detailed immunohistochemistry markers were studied. Immunohistochemical study in our case was helpful as FDCS is consistently negative for CD34, CD117 (positive in GIST), S-100 (positive in Melanoma and interdigitating reticulum cell tumors), cytokeratin (positive in sarcomatoid carcinoma), sma (positive in smooth muscle tumors). CD21, CD23, CD 35 markers were positive.

The tumor is considered to be a low grade neoplasm with a tendency to recur. More aggressive behavior has been noted by some, when the tumor is found in the intra-abdominal location [15]. Podoplanin, clusterin, fascin are the recently empoyed markers apart from CD21, CD23, CD35[16-17]. The behavior of these tumors is more akin to that of a low-grade soft tissue sarcoma than a malignant lymphoma, and is characterized by local recurrence in 36% of cases and metastases in 28%[18]

Complete surgical resection is the treatment of choice whenever feasible.

There are no consensus on optimal chemotherapy and adjuvant therapy[19]. The CHOP (cyclophosphamide–hydroxydaunorubicin–vincristine–prednisolone) chemotherapy regimen is perhaps the most widely used regimen[20]. Polyethylene glycol liposomal doxorubicin has also been used, with a fauvorable response[21]. Adjuvant radiotherapy or chemotherapy appears to be indicated in cases having adverse pathological features and in recurrent or incompletely resected lesions. Cases with microscopic features such as significant cytological atypia, extensive coagulative necrosis, high proliferative index (mitotic count of >5/10 hpf), or tumor size greater than 6 cm have poor prognosis, whereas lesions arising in the lymph nodes behave as low-grade sarcoma with a relatively good prognosis[16]

Naturally FDCS did not respond to imatinib trial since they don't express CD 117, as is evident in our case. CD 117 and CD 34 are negative in less than 5% of cases diagnosed as GIST [22]. A differential of FDCS seems logical in such scenario.

CONCLUSION

Follicular dendritic cell sarcoma should be considered as a differential diagnosis in cases of CD34 and CD117 negativity in a spindle cell tumour of gastrointestinal tract. Surgical resection is preferred choice of treatment. Further research into adjuvant therapy may improve the outcomes and survival.

Conflict of interest: The authors declare that they have no conflicting interests.

Author contributions: All the authors have contributed in the surgical management of the case, literature search and preparation of the case report.

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