

A Rare Case Report of Gastric Follicular Dendritic Cell Sarcoma: An Unconsidered Differential of Gastrointestinal Stromal Tumour

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Abstract: *Follicular Dendritic Cell Sarcoma (FDSC) is a rare well - defined malignancy presenting with homogenous nodal mass. The rare occurrence of such masses at extranodal locations like the abdomen or mediastinum presents large, heterogeneous masses with regional lymphadenopathy. However, Gastrointestinal stromal tumours are relatively common tumours, but due to their overlapping imaging features with gastric follicular dendritic cell sarcoma, they should be kept as a differential, too. Stomach FDSC are very rare, and a diagnostic suspicion of the radiologist will result in an increased chance of detection and guide the patient in further management.*

Keywords: Gastrointestinal stromal tumor, Gastric Follicular Dendritic Cell Sarcoma, stomach

1. Introduction

Follicular dendritic cell sarcomas (FDSC) usually involves the lymph nodes, especially the cervical and axillary lymph nodes. On the other hand, almost one - third of cases occur in extranodal sites such as the tonsils, soft and hard palate, nasopharynx, parapharyngeal space, liver, gastrointestinal tract, mesentery, and retroperitoneum.^{1,2} Involvement of the gastrointestinal tract is rare, and the stomach is even rarer, with only four cases described to date³. It often presents as a diagnostic challenge for both the pathologist and the clinician and accounts for only 0.4% of soft - tissue sarcomas, and its underlying causes are largely unknown. Its correct diagnosis cannot be over emphasised as the treatment and prognosis of follicular dendritic cell sarcoma (FDSC) are very much different from tumors which come in its differential diagnosis.⁴ Given the rarity of this tumour in gastrointestinal sites and the lack of consensus on treatment, a proper evaluation of this disease must be done.

2. Case Report

A 48 - year - old female presented with diffuse, dull, aching upper abdominal pain with no associated symptoms. A history of loss of appetite and nausea was present.

No palpable lump or organomegaly was found on abdominal

examination, and no other significant findings were present on systemic examination.

Investigations: Biochemical and hematological tests are unremarkable. Abdominal ultrasound revealed diffuse thickening near the cardia, lesser curvature of the stomach, and Cholelithiasis.

UGI endoscopy revealed growth at the cardiac end of the stomach.

CECT Findings: Soft tissue attenuation lesion in the gastro - hepatic area inseparable from gastric lesser curvature measuring approx.5x4cm without significant luminal obstruction in close apposition to the left lobe of the liver with maintained fat planes and minimal post - contrast enhancement possibly suggestive of Gastrointestinal stromal tumor.

MRI Findings: A fairly defined T2 hyperintense with internal hypointensity and T1 hypointensity arising exophytically from the stomach wall in the fundus along lesser curvature in close apposition and compression of fat planes with hepatic and pancreatic body without frank invasion, patchy diffusion restriction and minimal post - contrast enhancement. . . possibly GIST. T2 hypointense calculus - cholelithiasis.



Figure 1: Axial, Coronal, and Sagittal CECT images showing hypodense lesion along the cardiac end extending along the lesser curvature

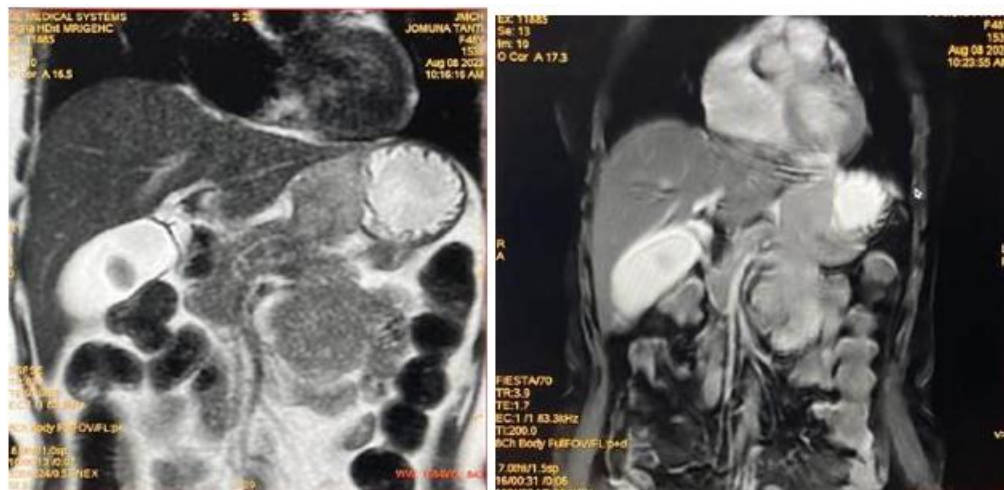


Figure 2: T2WI sequences showing the mass lesion and the hypointense calculus in the gallbladder.

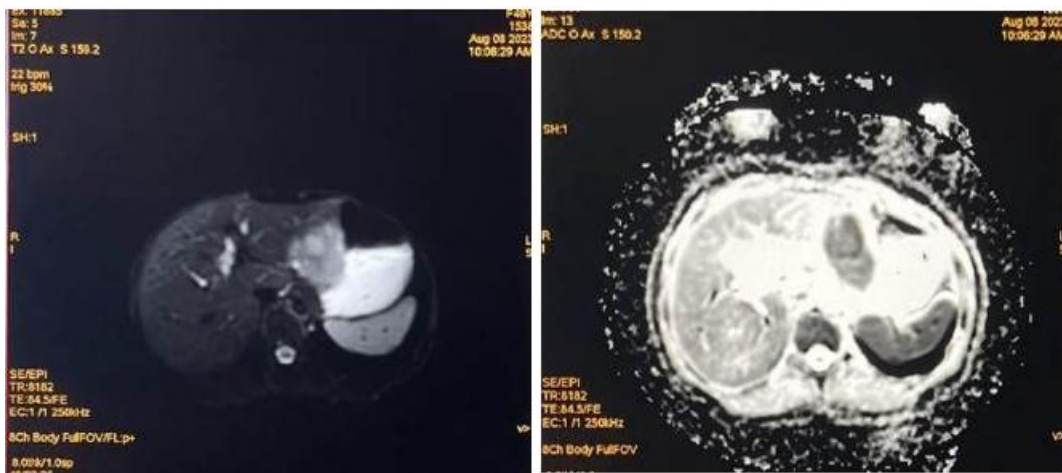


Figure 3: DWI and corresponding ADC, showing patchy diffusion restriction of the lesion.

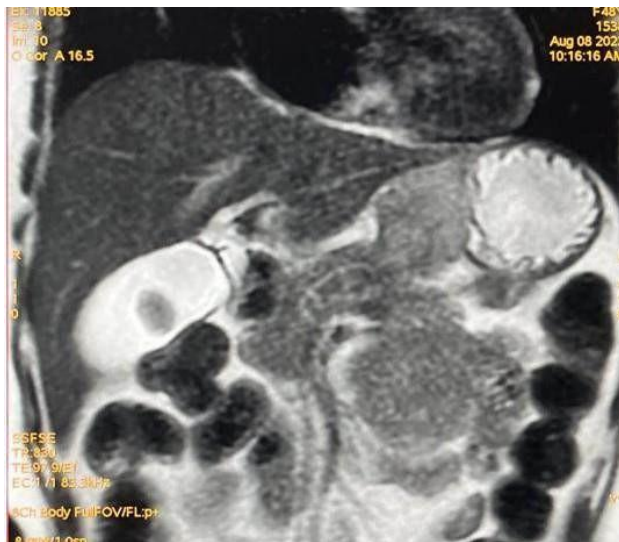


Figure 4: CEMRI showing minimal post - contrast enhancement

Operative Procedure: Open Sleeve Gastrectomy with Cholecystectomy.

Intra - Operative Findings: A firm mass was noted along the lesser curvature of the stomach, showing adhesion with the pancreas.



Figure 5: Before tumor resection and after tumor resection



Figure 6: Post Operative Specimen

suggestive of an **FDCS**

IHC: CD21, CD35 patchy positive, CD 45 positive, Ki67 is 10%vimentin strongly positive and WT1 patchy positive, CD20 andCD3 positive.

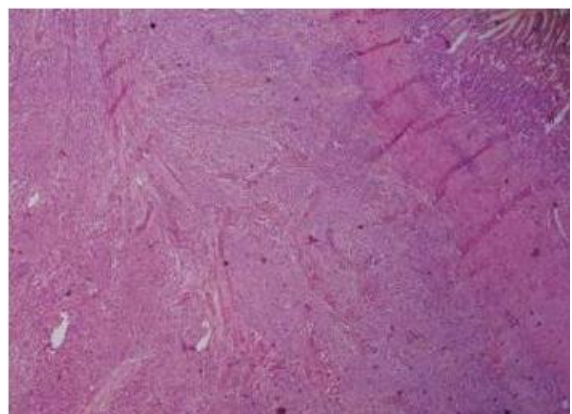


Figure 7: HPE image suggestive of the diagnosis of Follicular dendritic cell sarcoma

Histopathology and Immunohistochemistry

Microscopic examination shows gastric mucosa with a submucosal tumor composed of spindle cells arranged in a haphazard to vaguely storiform pattern with elongated nuclei and open nuclear chromatin and occasional mitotic figures

3. Discussion

Follicular dendritic cell sarcoma is an uncommon tumor, and the extra-nodal disease accounts for only one-third of FDCCS cases⁵. Gastrointestinal FDCCS tumors often present as slow-growing and painless masses. However, their location is variable, and the abdominal pain can be non-specific. For the abdominal component of this disease, common imaging findings include a well-defined mass with regional lymphadenopathy and homogenous enhancement with internal necrosis and often with internal calcifications^{6, 7}. FDCCS may be suspected when the tumor exhibits distinct microscopic features, such as a storiform arrangement of spindle-shaped cells, indistinct cell borders with vesicular nuclei and distinct nucleoli and a background of lymphocytes scattered throughout the neoplastic cells.⁸ The primary differential diagnosis includes gastrointestinal stromal tumor and primary gastrointestinal lymphomas. The inflammatory pseudotumor-like FDCCS variant must also be considered.

4. Conclusion

The complex entity FDCCS and its gastrointestinal variant are poorly described in the literature. Lack of clinical and radiological suspicion and challenges in pathological diagnosis are the reasons that this uncommon entity is poorly understood. FDCCS is a rare tumor having distinct morphology and phenotype which if known can be correctly diagnosed. Therefore, knowledge of its varied location, morphology, and phenotype is very important to correctly diagnose this tumor and to prevent misdiagnosis and mistreatment⁴. This necessitates the need for researchers to bring in clinical suspicion and consider this uncommon entity that is commonly missed as a differential of GIST.

References

- [1] Shia J, Chen W, Tang LH, et al. Extranodal follicular dendritic cell sarcoma: clinical, pathologic, and histogenetic characteristics of an underrecognized disease entity. *Virchows Arch* 2006; 449: 148–158
- [2] Jain P, Milgrom SA, Patel KP, et al. Characteristics, management, and outcomes of patients with follicular dendritic cell sarcoma. *Br J Haematol* 2017; 178: 403–412
- [3] Sassi F, Sahraoui G, Charfi L, Achouri L, Doghri R, Mrad K. An incidental discovery of a gastric follicular dendritic cell sarcoma: A rare case report and a literature review. *Rare Tumors*.2023 Apr26; 15: 20363613231172077.
- [4] Hassan U, Rana IA, Mushtaq S, Azam M, Akhter N. Follicular Dendritic Cell Sarcoma of Gastrointestinal Tract: an Uncommon Lesion, Commonly Missed. *J Gastrointest Cancer*.2019 Dec; 50 (4): 913 - 918.
- [5] Shaw D, Cuisson R, Ito H. Follicular dendritic cell sarcoma of the stomach: case report and review of the literature. *Curr Oncol*.2014 Dec; 21 (6): e775 - 8.
- [6] Youens KE, Waugh MS. Extranodal follicular dendritic cell sarcoma. *Arch Pathol Lab Med* 2008; 132: 1683–7.
- [7] Kang TW, Lee SJ, Song HJ. Follicular dendritic cell sarcoma of the abdomen: the imaging findings. *Korean*

J Radiol 2010; 11: 239–43.

- [8] Shia J, Chen W, Tang LH, Carlson DL, Qin J, Guillem JG, Nobrega J, Wong WD, Klimstra DS: Extranodal follicular dendritic cell sarcoma: clinical, pathologic, and histogenetic characteristics of an underrecognized disease entity. *VirchowsArchiv* 2006, 449 (2): 148 - 158.