

## Inflammatory Pseudotumor-Like Follicular/Fibroblastic Dendritic Cell Sarcoma: Focus on Surgical Treatment Approach

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### ABSTRACT

*Inflammatory pseudotumor-like follicular/fibroblastic dendritic cell sarcoma (IPT-like FDC sarcomas, also known as FDC/FRC sarcoma) is a subgroup of follicular dendritic cell sarcoma (FDC), a rare neoplasm that often occurs in the spleen [1]. FDC/FRC sarcomas can affect other organs including the liver, colon, and the peripancreatic region [2-4]. The tumor has a predilection for young to middle-aged adults and may present with subset of symptoms including fever, abdominal pain, sweats, fatigue, and commonly a splenic solitary mass formation [5]. FDC/FRC sarcoma comprises of neoplastic spindle cells, typically with a follicular dendritic cell differentiation and over 90% of the cases are associated with EBV infection [6]. Here we present the a case of a IPT-like FDC sarcoma in a 49-year old female treated with splenectomy and a review of the literature.*

### Case Report

#### Clinical Findings

The patient is a 49-year-old female, who started to experience headaches and visual changes approximately one year prior to her initial presentation. She was diagnosed with uveitis involving one eye, and was started on steroid eyedrops. A limited work-up including autoimmune testing was negative for antinuclear antibodies; however, she had an elevated HSV type I titer and was started on antiviral medications. More recently, her serum protein electrophoresis studies showed a polyclonal gammopathy and immunoglobulin G subclass, which resulted in a presumptive diagnosis of IgG4 related disease, and a CT scan of the body was performed, which showed a 6.5 cm heterogeneous splenic mass. Due to this finding a biopsy of the splenic mass was performed which was consulted with the National institute of health and was most consistent with an EBV-positive inflammatory pseudotumor-like follicular/fibroblastic dendritic cell sarcoma positive for CD79a, MUM-1, CD30 (weak), IgG4 (4%), SMA in the myofibroblasts, and EBER ISH throughout the splenic cores. The patient underwent a splenectomy 2 months after, and the pathology report was consistent with the prior core biopsy of EBV-

positive inflammatory pseudotumor-like follicular/fibroblastic dendritic cell sarcoma. On the most recent follow at 1year, she has persistent uveitis with no other symptomatology.

#### Surgical Procedure

The patient was positioned in 10 degrees reverse Trendelenburg with a right lateral decubitus position. A Hassan trocar was placed in the supraumbilical area, an 8mm robotic port in the right upper quadrant was used for retraction of both the liver, stomach, and eventual pancreas during dissection, and a tip-up bowel grasper was used. Two robotic ports were placed on the patient's left side in mid-clavicular and lateral clavicular lines at level 3-4cm inferior to the umbilicus. A laparoscopic port was placed on the left lateral robotic port and the robot was docked over the patient's right upper side. Using a bowel grasper, a bipolar, and a vessel sealer, the lesser sac was opened. Larger vessels were additionally clipped with a 5mm clip applicator. The mass was seen abutting the capsule of the spleen but away from the hilum. The stomach was retracted superiorly with the right lateral arm and careful dissection along the superior aspect of the pancreas allowed isolation of the splenic artery. This was isolated with a

vessel loop and used to retract superiorly. The splenic vein was seen and dissected free from the tail of the pancreas with bipolar long-tip Maryland and scissors along with clips as needed. The splenic vein was ligated with an endo-GIA following ligation of the splenic artery. The tail of the pancreas was completely free from the spleen, the splenic attachments were incised, and the large spleen was free. The splenic bed along with the lateral stomach was inspected and found to be hemostatic. Due to the size of the spleen and concerns about the tumor inside and to prevent rupture, a 5cm right subcostal incision was made to deliver the spleen and send fresh to pathology. Again, the splenic artery and vein and their ligated branches were inspected along with the tail of the pancreas and found to be free of bleeding and

no injury. A 19Fr Blake drain in the LUQ. All bowel and port sites were inspected and found to be injury free. The fascia was closed as well as the skin. The patient was awakened, extubated, and no post-op complications were reported.

### Literature Review

A literature review was performed with PUBMED and MEDLINE search using the terms “follicular dendritic cell tumor” combined with “inflammatory pseudotumor of the spleen” and “minimally invasive splenectomy”. The articles that included the cases of the inflammatory pseudotumor of the spleen were selected as well as minimally invasive splenectomy and the references from these articles were reviewed to identify other relevant publications.

### Pathological Findings

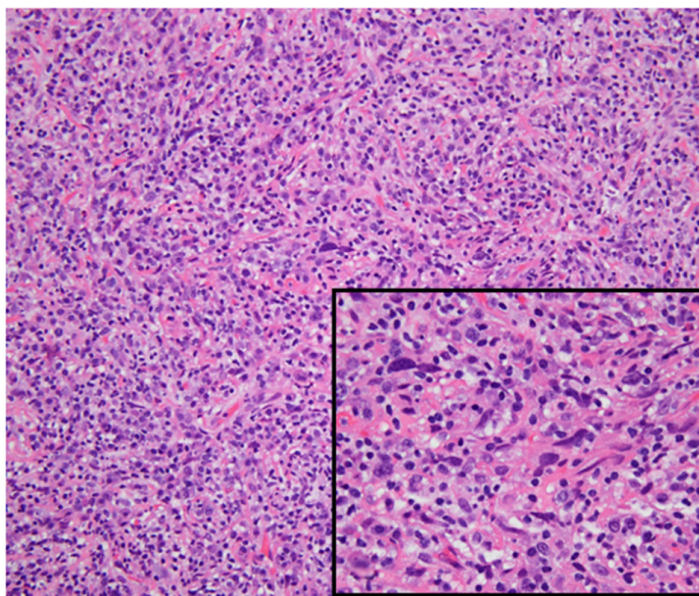


Image 1. HE section @ 20X of splenic core biopsy (inset @ 40X)  
Sections show absence of normal splenic architecture a with diffuse polymorphous infiltrate with a fasciculate architecture. Small to medium sized lymphocytes with presence of plasma cells, histiocytes, and rare eosinophils.

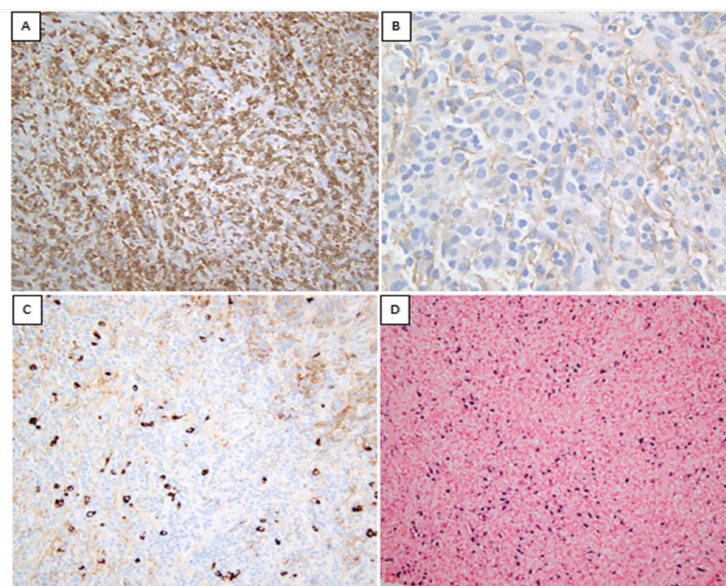


Image2. A-D Immunostains from the splenic core biopsy specimen.  
A. CD3 showing numerous atypical T cells. B. Smooth muscle acting (SMA) weak staining highlighting spindle cells. C. IgG4 staining highlighting a small subset of plasma cells (less than 50/hpf). D. EBER ISH demonstrating many positive cells diffusely throughout.

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## Discussion

Follicular dendritic cells (FDC) are mesenchymal-derived dendritic cells located in B-follicles where they play a pivotal role in triggering and maintaining B-cell adaptive immune response [7]. Inflammatory pseudotumor-like follicular/fibroblastic dendritic cell sarcoma is a relatively rare neoplasm, with less than 50 reported cases in the literature, including the current case. Although its clinicopathologic features have not been fully studied, it is generally considered a distinctive variant from the conventional FDC sarcoma with several distinguishing features [8]. IPT-like FDC sarcomas consistently occur in the spleen or liver, in contrast, to the conventional follicular dendritic cell FDC sarcomas, which can occur at various anatomical sites including the lymph nodes [9]. Second, IPT-like FDC sarcomas demonstrate a marked female predilection (female to male ratio, 2.2:1), whereas FDC sarcomas do not show a gender-specific tendency [10]. Third, IPT-like FDC sarcomas have intense lymphoplasmacytic infiltrates, which make it difficult to distinguish them from inflammatory pseudotumor; however, majority of conventional FDC show only minimal inflammatory cell aggregates. Fourth, IPT-like FDC sarcomas are strongly associated with EBV positivity, which is rare in conventional FDC sarcomas [5]. Fifth, although both IPT-like FDC sarcomas and conventional FDC sarcomas show generally an indolent clinical behavior, intra-abdominal cases of conventional FDC sarcomas usually behave more aggressively than IPT-like FDC sarcomas, and maybe associates with high recurrence, metastasis, and even mortality [11]. On the contrary, IPT-like FDC sarcomas rarely recur or metastasize, especially in cases with splenic involvement [8].

In all cases reviewed, a splenectomy was performed and no additional therapy was carried out. Follow-up after splenectomy showed no recurrence or metastasis [12]. FDC/FRC is a rare subtype of FDCS with a favorable clinical course, and surgical treatment alone is often the therapeutic approach [13]. Most patients remained asymptomatic afterward and no complications were reported.

Laparoscopic splenectomy was first introduced and reported in 1991 by Delaitre and Maignien, who performed a laparoscopic splenectomy (LS) on 1 patient [14]. Minimally invasive splenectomy has become the standard of care in the management of surgical diseases of the spleen, especially hematologic diseases. Many studies have shown decreased patient discomfort, hospital stay, and recovery time compared to open procedures [14,15]. Laparoscopic splenectomy can be a difficult procedure to master taking into account different factors including the anatomical location of the spleen and its high vascularization, particularly in patients with splenomegaly, hematological malignancies, obesity or previous laparotomies [16]. It has some disadvantages, including two-dimensional (2D) visualization and rigid instrumentation, which can make splenectomy for splenomegaly or partial splenectomy challenging [17].

In order to conquer these limitations, robotic surgery (da Vinci, Intuitive Surgical, and Sunnyvale, CA, USA) has been developed with a 'wrist-like' action of the instruments and with three-dimensional (3D) visualization, resulting in a high-resolution binocular view of the surgical field.

Analyzing the comparison of the laparoscopic approach vs. the robotic in difficult splenectomies, the robotic approach had a longer total operative time which might be associated to the docking time, less blood loss reported, and decreased risk of hemorrhagic complications during surgery [18]. This review suggest that robotic approach in splenectomy could be beneficial in difficult splenectomies such as partial splenectomy, splenic tumors, and splenomegaly. Laparoscopic splenectomy remains the approach of choice for simple splenectomies in the surgical treatment for common indications [15,18].

The literature review demonstrated that robotic and laparoscopic splenectomies have comparable perioperative outcomes with similar rates of conversion to an open procedure, procedure time, and length of hospital stay. The robotic approach may reduce the volume of intraoperative blood loss [16]. It is suggested that the optimal indications for the robotic technique could be all those challenging conditions that would render conventional laparoscopy a high-risk procedure [19].

EBV plays a major role in the development of many lymphoproliferative disorders including IPT-like FDC sarcomas [8,20]. It is postulated that EBV induces the activation of cytidine deaminase in B-cells, which mediates somatic hypermutation and may be responsible for mutations and translocations in B-cell lymphomas via the expression of viral proteins [21]. Two very closely related diseases, both expressing EBV gene uptake by insitu hybridization include EBV-positive IPT-expressing FDC markers and IPT-like FDC tumors, which are likely of the same disease entity, although more discussion is required to clarify their relationship. In both situations, EBV is thought to play a significant role in the pathogenesis of the disease, although one is considered a benign condition (IPT) versus its malignant component IPT-like FDC [22].

## Conclusion

Inflammatory pseudotumor-like follicular/fibroblastic dendritic cell sarcoma (IPT-like FDC sarcomas, also known as FDC/FRC sarcoma) is a subgroup of follicular dendritic cell sarcoma (FDC), a rare neoplasm that often occurs in the spleen and has been managed with splenectomy as a definitive therapy. Most cases have not shown recurrence of disease and have remained asymptomatic. More recently, splenectomy is performed as a minimally invasive procedure with different surgical approaches available including laparoscopic and robotic-assisted. Review of the literature has demonstrated similar outcomes with both techniques.



**Table 1:** Summary of reported cases of inflammatory pseudotumor-like follicular/fibroblastic dendritic cell sarcoma of the spleen treated with splenectomy.

Sex	Age	Diameter (cm)	Last F/U (Months)	Outcome
F	54	3.5	10	Alive, ND
F	64	5.5	78	Alive, ND
F	72	7.2	18	Alive, ND
F	53	3.2	13	Alive, ND
M	76	3.2	8	Alive, ND
M	72	6	18	Alive, ND
M	75	3.5	30	Alive, ND
M	54	12	48	Alive, ND
F	37	9	5	Alive, ND
M	63	7	18	Alive, ND

\*ND: No disease

The table shows equal distribution for sex, and an age range of 37-76, with a predilection for middle-aged adults for the occurrence of IPT-like FDC sarcomas. Splenectomy was performed in all cases and all patients remained alive with no recurrence of disease at follow-up, suggesting splenectomy as the therapeutic and definitive management for this condition. Some of the cases showed a single, well-demarcated splenic mass in preoperative imaging studies and underwent splenectomy under the clinical suspicion of metastatic tumor or other diseases and other presented with a variety of symptoms. Scarce information regarding the surgical approach in these specific cases was found in the literature review. Based on the current revision minimally invasive splenectomy with either laparoscopic or robotic assisted splenectomy must have been performed [13-15,17-19,23].

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