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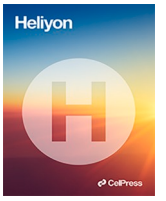

Case report

Epstein-barr virus (EBV)-positive inflammatory pseudotumor-like follicular dendritic cell sarcoma (IPT-like FDCS) presenting as thrombocytopenia: A case report and literature review

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ABSTRACT

Background: Follicular dendritic cell sarcoma (FDCS) represents an exceedingly rare malignant neoplasm. Inflammatory pseudotumor-like follicular dendritic cell sarcoma (IPT-like FDCS) is recognized as a variant manifestation of FDCS. The clinical incidence of this particular disease is remarkably low, resulting in the absence of established standardized clinical protocols for its management and treatment.

Methods: Presented here is a case of primary Epstein-Barr virus (EBV)-positive splenic IPT-like FDCS, noteworthy for manifesting thrombocytopenia as its initial symptom. Our study analyzed the clinicopathologic characteristics of this case and 29 previously reported cases identified in the literature. Also, we conducted a comprehensive review of pertinent literature.

Results: We administered splenectomy to this patient and verified the diagnosis of EBV-positive IPT-like FDCS through immunohistochemical examination. Postoperatively, the patient underwent a one-year follow-up period, demonstrating no signs of recurrence. Analyzing a total of 30 cases revealed that this disease is more prevalent in female patients (F:M = 1.14:1), with a median age of 62 years. Fifteen patients were asymptomatic, and nine patients presented with abdominal discomfort or pain. All patients underwent surgical treatment. Among the cases, histopathological and immunohistochemical information was unavailable for five; however, in the remaining 25 cases, histopathology revealed a distinct inflammatory cell infiltration and spindle tumor cells arranged in sheets or fascicles. These tumor cells had vesicular chromatin and distinct nucleoli and they expressed conventional FDC markers. In situ hybridization analysis of Epstein-Barr virus-encoded small RNA (EBER) showed that all 30 cases were EBV-positive. Follow-up information showed that no patients relapsed and one (3.8 %) patient died.

Conclusion: The clinical diagnosis of EBV-positive IPT-like FDCS poses considerable challenges, necessitating a conclusive diagnosis through pathological immunohistochemical examination. EBER in situ hybridization holds significance for the definitive diagnosis of the disease. We advocate for splenectomy as the treatment of choice for limited splenic IPT-like FDCS.

1. Introduction

The occurrence of follicular dendritic cell tumors was first pointed to by Lennert in 1978 [1]. In 1986, Monda et al. [2] detailed FDCS as a rare malignant tumor arising from follicular dendritic cells. Most of FDCS occurs in the lymph nodes of neck, mediastinum,

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and axillary cavity, with about one-third of cases occurr in extranodal sites [3]. However, a study [4] that reviewed 809 cases of FDCS indicated that the disease predominantly occurs in extranodal sites (79.4 %), such as the liver, spleen, and gastrointestinal tract. Histologically FDCS is mainly classified into conventional and inflammatory pseudotumour (IPT-like) type. IPT-like FDCS predominantly originates in the hepatic and splenic parenchyma, are generally considered associated with Epstein-Barr virus (EBV) [5]. The study noted that the conventional type features spindle cells amidst small lymphocytes [2]. Conversely, the IPT-like variant exhibits sparse spindle cells with conspicuous lymphocytes, plasma cell infiltration, and other inflammatory cells [6]. Due to these morphologic features, IPT-like FDC tumors are commonly misdiagnosed as reactive lesions. This disease is difficult to diagnose preoperatively due to the lack of clinical features and the low specificity of laboratory tests and imaging.

Herein, we present a case of EBV-positive splenic IPT-like FDCS with thrombocytopenia as the initial symptom. In addition to one already reported case [7], the case detailed in this article represents only the second instance of splenic IPT-like FDCS characterized by thrombocytopenia. We have reviewed the existing literature and discussed the disease in light of our findings. To date, there have been 29 published cases [7–19] of splenic IPT-like FDCS, all of which have shown an association with EBV proliferation. Yet, the clinical presentation and pathological characteristics of this condition continue to be a subject of debate.

2. Case presentation

The 67-year-old female patient was admitted with a one-month history of "bleeding from the mouth, nose, and skin" and a two-week history of splenic space-occupying disease. Previously healthy, the spleen was non-palpable during physical examination. No similar disease history in the patient's family. Laboratory findings revealed thrombocytopenia (PLT 1*10^9/L). The patient was treated with recombinant human thrombopoietin (rhTPO), immunoglobulin (IG), methylprednisolone (MP) on the advice of a haematologist. Medication did not work well for this patient (repeat PLT 7*10^9). Computed tomography (CT) scan indicated splenomegaly with a patchy low-density shadow measuring approximately 57*58 mm (Fig. 1). There was moderate peripheral enhancement in the arterial phase, low enhancement in the center, and slightly reduced enhancement in the venous phase. The patient has been preliminarily diagnosed with splenic hamartoma. Refer to the timeline annotations for details on the diagnosis and treatment process of the patient (Fig. 2).

The patient underwent laparoscopic total splenectomy. Gross examination revealed a spleen measuring 11*8*6 cm with a smooth surface and intact peritoneum. Partial dissection exposed a white-gray-red mass measuring 6*5*4.5 cm with a medium texture (Fig. 3). The patient is recovering well post-surgery. No adverse events or complications occurred postoperatively. Cytohistological examination (Fig. 4A) indicates the presence of spindle tumor cells arranged in fascicles amidst proliferating lymphocytes and plasma cells. Some tumor cells exhibit nucleoli, with fibrin-like and histiocyte deposits containing necrosis observed in the blood vessel walls. Immunohistochemical analysis showed partial expression of CD21(Fig. 4B), CD23, CD35. In addition, CD79a, CD56, TIA-1, GB, Bcl-2, S100 were positive. CD30, CD10, Kappa, Lambda, Perforin, Bcl-6 and MUM1 were consistently negative. The plasma cell population in the scant fibrotic areas resulted IgG4-positive. The Ki-67 index 5 % of tumor cells. EBER (EBV-encoded small RNAs) in situ hybridization showed a positive nuclear result in the spindle cells (Fig. 4C). Molecular pathology: B Rearrangement (sequencing) (−).

Given these findings, the conclusive diagnosis is EBV-positive inflammatory pseudotumor-like follicular dendritic cell sarcoma. Post-surgery, the patient did not undergo radiotherapy or chemotherapy. Follow-up assessments at 3 months, six months, and 1 year post-operation revealed normal platelet counts with no evidence of tumor recurrence or metastasis.

3. Literature review

A total of 29 cases of splenic IPT-like FDCS were retrieved from the previous literature. The clinical information of these 29 cases, as well as the new case (NO.30 case) in this study, is detailed in Table 1. In our statistics, there were 16 female and 14 male patients (female to male ratio, 1.14:1). The age range of the 30 patients was 29–79 years (median age 62 years). In all cases, 2 patients had IPT-like FDCS not only in the spleen, but also in the liver in one case and in the pancreas in the other.

The most common symptom was abdominal discomfort or pain (9 cases), although 15 cases had no clinical symptom. Other common clinical features included weight loss (3 cases), malaise (2 cases), back pain (2 cases), skin and oral bleeding (2 cases), anemia (1 case), and fever (1 case).

Of the 30 cases, the two patients with concomitant pancreatic or hepatic IPT-like FDCS underwent distal pancreatectomy with

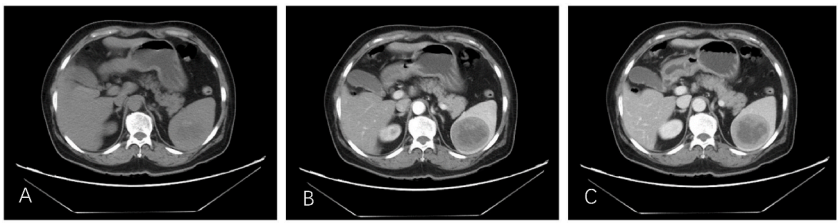

Fig. 1. A. Plain CT scan showed an enlarged spleen with a patchy low-density shadow measuring about 57*58mm. B. Contrast-enhanced CT showed moderate peripheral enhancement and central hypoenhancement of the tumor in the arterial phase. C. The tumor in venous phase has slightly reduced enhancement.

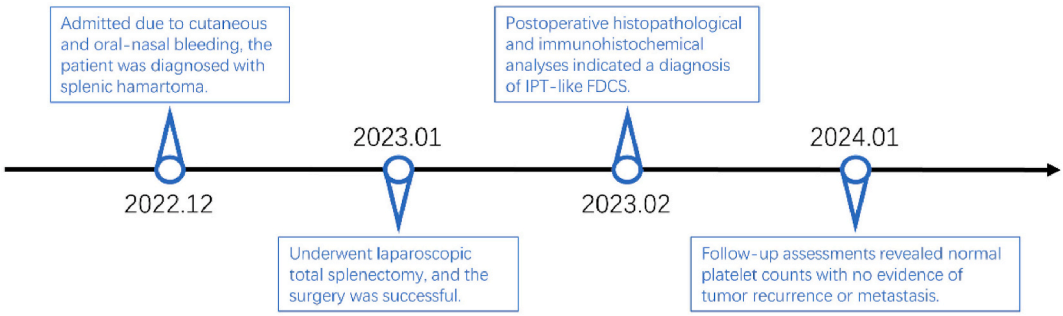

Fig. 2. Timeline of the case report.

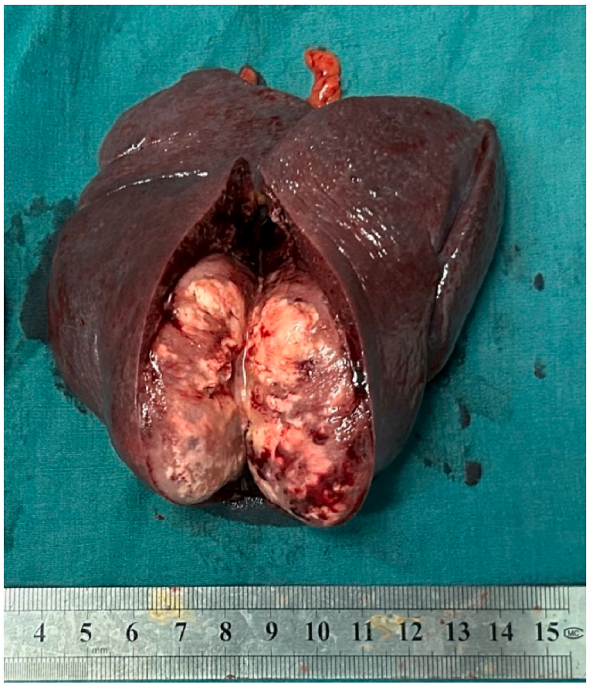

Fig. 3. Gross appearance of the splenic lesion on sectioning.

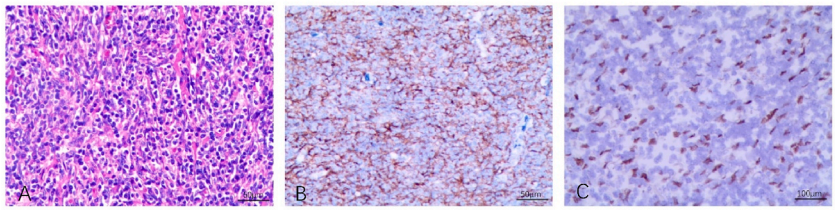

Fig. 4. A. Cytohistological examination indicates the presence of mild and elongated bundles of tumor cells amidst proliferating lymphocytes and plasma cells. B. The immunohistochemical analysis revealed partial expression of CD21. C. Positive EBER expression was seen in tumor cells.

splenectomy as well as splenectomy and radical removal of retroperitoneal lymph nodes, respectively. Of the remaining 28 cases, 26 cases underwent splenectomy and 2 cases underwent splenectomy with removal of splenic hilar lymph nodes.

The maximum diameter of splenic tumors in all 30 cases ranged from 2.3 cm to 22.3 cm (mean, 6.75 cm; median, 6 cm). Among the cases, histopathological and immunohistochemical information was unavailable for five; however, in the remaining 25 cases, histopathology revealed a distinct inflammatory cell infiltration and spindle tumor cells arranged in sheets or fascicles. These tumor cells had vesicular chromatin and distinct nucleoli and they expressed conventional FDC markers. CD21, CD23 and CD35 were usually used,

Table 1
Summary of reported cases of IPT-like FDCS of spleen.

Case	Sex	Age	Location	Maximum diameter (cm)	Presentation	Treatment	Last follow-up (months)	Outcome
1 [1]	F	63	Spleen	15	Rapid and unexplained weight loss	Splenectomy with removal of splenic hilar lymph nodes	NA	NA
2 [2]	F	70	Spleen, Pancreas	Spleen: 3.0; Pancreas: 7.0	Asymptomatic	Distal pancreatectomy and splenectomy	NA	NA
3 [4]	F	54	Spleen	3.5	Intermittent vague pain in the left upper quadrant	Splenectomy with removal of splenic hilar lymph nodes	10	AW
4 [4]	M	79	Spleen	6	Persistent epigastralgia	Splenectomy	18	AW
5 [6]	M	29	Spleen	11	Asymptomatic	Splenectomy	12	AW
6 [7]	M	59	Spleen	9.5	Bleeding gums and epistaxis	Splenectomy	17	AW
7 [8]	F	64	Spleen	7.2	Upper abdominal pain	Splenectomy	8	AW
8 [8]	M	61	Spleen	6.2	Asymptomatic	Splenectomy	16	AW
9 [8]	F	42	Spleen	4	Left-sided flank pain	Splenectomy	9	AW
10 [8]	F	57	Spleen	13.3	Upper abdominal pain	Splenectomy	4	AD: Pulmonary metastasis on admission
11 [8]	M	52	Spleen	3.7	Back pain	Splenectomy	5	AD: Multiple bone metastasis on admission
12 [9]	F	57	Spleen	5	Asymptomatic	Splenectomy	12	AW
Table 1: Summary of reported cases of IPT-like FDCS of spleen								
Case	Sex	Age	Location	Maximum diameter (cm)	Presentation	Treatment	Last follow-up (months)	Outcome
13 [10]	F	59	Spleen	4.5	Occasional left low back pain	Splenectomy	NA	NA
14 [11]	M	57	Spleen	2.7	Asymptomatic	Splenectomy	9	AW
15 [12]	F	64	Spleen	5.5	Asymptomatic	Splenectomy	78	AW
16 [12]	F	72	Spleen	7.2	Asymptomatic	Splenectomy	18	AW
17 [12]	F	53	Spleen	3.2	Asymptomatic	Splenectomy	13	AW
18 [12]	M	76	Spleen	3.2	Asymptomatic	Splenectomy	8	AW
19 [12]	M	72	Spleen	6	Asymptomatic	Splenectomy	18	AW
20 [12]	M	75	Spleen	3.5	Abdominal pain	Splenectomy	30	AW
21 [18]	F	69	Spleen	6	Asymptomatic	Splenectomy	NA	NA
22 [19]	F	66	Spleen	5	Left upper quadrant pain	Splenectomy	6	AW
23 [21]	M	39	Spleen	7.4	Asymptomatic	Splenectomy	40	AW
24 [21]	M	65	Spleen, Liver	Spleen: 22.3; Liver: 5.8	Abdominal epigastric pain, fever, fatigue, anorexia, mild anaemia, weight loss	Splenectomy and radical dissection of retroperitoneal lymph nodes	2	Died at 2 months
25 [21]	M	51	Spleen	8.5	Malaise, weight loss	Splenectomy	19	AW
26 [21]	M	68	Spleen	2.3	Asymptomatic	Splenectomy	6	AW
27 [21]	F	51	Spleen	5.3	Intermittent epigastric discomfort	Splenectomy	5	AW
28 [21]	M	67	Spleen	7.5	Asymptomatic	Splenectomy	5	AW
29 [21]	F	52	Spleen	9	Asymptomatic	Splenectomy	12	AW
30	F	67	Spleen	6	Bleeding from the mouth, nose, and skin	Splenectomy	12	AW

AW, alive and well; AD, Alive with disease; NA, not available.

and all had a high positive rate (68 % for CD21, 68 % for CD23 and 84 % for CD35). Immunohistochemical results for smooth muscle actin (SMA) were 82 % (18/22) positive. The Ki-67 index of tumour cells ranged from 5 % to 30 % in 5 cases. IgG4-positive plasma cells were found in the tumors of 10 cases. Positive immunostaining for LMP-1 was detected in 92 % (12/13) of cases.

With the exception of 4 cases (case1.2.13.21), a total of 26 cases provided follow-up information for 2–78 months (mean, 15.1 months; median, 12 months). At the final follow-up, 1 (3.8 %) patient died of the disease, 2 (7.7 %) patients survived with the disease, and 23 (88.5 %) patients survived without evidence of disease. All patients had no recurrence. The overall mortality rate was 3.8 %.

4. Discussion

In 2001, IPT-like follicular dendritic cell sarcoma (FDCS) was initially characterized as a variant of FDCS [5]. IPT-like FDCS typically exhibits selective involvement of the liver and spleen, and is strongly linked to Epstein-Barr virus (EBV). In situ hybridization in a comprehensive study identified EBER-positive IPT-like FDCS cells in all cases [19], reinforcing the association between EBV and IPT-like FDCS. However, our review of the literature identified two cases [20,21] of EBV-negative IPT-like FDCS that were reported. Additionally, there are documented instances of rare occurrences in atypical locations, such as the colon [22,23], pancreas [24] and lung [25]. IPT-like FDCS originating from the spleen is very rare in clinical practice, with few cases described in the literature. Typically, IPT-like FDCS manifests without specific clinical symptoms, frequently presenting as a painless, gradually enlarging abdominal mass. In our study, 50 % of splenic IPT-like FDCS were asymptomatic, with the most common symptom being abdominal discomfort or pain (30 %, 9/30). Diagnostic challenges arise as imaging studies offer limited assistance, complicating accurate clinical diagnoses. Some studies [11,12] have shown that CT examination of splenic IPT-like FDCS revealed a well-circumscribed, hypodense mass with weak delayed heterogeneous enhancement after contrast-enhancement in a normalized spleen. Kitamura et al. [26] reported that the presence of the capsular like rim on delayed phase on CECT and dynamic contrast-enhanced MR corresponding to a fibrous structure demarcated the tumor from the splenic parenchyma might aid the diagnosis of splenic IPT-like FDCS. However, all of the above studies concluded that it is difficult to make an accurate diagnosis by imaging, and a definitive diagnosis still requires further confirmation by needle aspiration biopsy or surgery. In our patient, thrombocytopenia emerged as the initial symptom, with unremarkable findings on physical examination and other laboratory tests. Conservative internal medicine interventions yielded suboptimal results. Initial CT scan suggested splenic hemangioma or splenic hamartoma. However, definitive diagnosis of splenic IPT-like FDCS was achieved through surgical intervention, coupled with immunohistochemical analysis. In the current case, the tumor displayed no signs of aggressive biological behavior, reinforcing the notion that the IPT-like FDCS is relatively inert and/or has a slow growth rate [16].

IPT-like FDCS predominantly affects females, spanning a broad age spectrum, with a prevalence among adults. In our study, splenic IPT-like FDCS occurred predominantly in older female patients, which is generally consistent with previous reports. Research indicates a notable occurrence of IPT-like FDCS cases in the Asian population, suggesting a potential racial predisposition [6]. The pathogenesis of FDCS remains elusive, lacking definitive insights into genetic contributions to its development. Nonetheless, genetic scrutiny of splenic FDCS has revealed a potential association with unbalanced translocation involving chromosomes X, 3, 5, 7, 8, 9, and 10 [27]. A study by Go et al. [28] identified BRAF gene mutations in 28.5 % (5/27) of IPT-like FDCS cases. However, many other studies have not identified the BRAF mutations previously discussed, and instead have discovered mutations in genes such as RICTOR [29], STAT3 [30], MYC and TP53 [31], CDKN2A and NF1 [32]. Bruehl FK et al. [29] found that comprehensive next-generation sequencing analysis on two cases of IPT-like FDCSs failed to demonstrate any pathogenic variant of potential or strong clinical significance within the targeted regions of the evaluated genes.

IPT-like FDCS predominantly originate from dendritic cells and exhibit clinical, histological, and pathological features similar to inflammatory pseudotumors [33]. Due to the occasional presence of Reed-Sternberg-like cells in a chronic inflammatory environment, these tumors are sometimes confused with Hodgkin's lymphoma, but are uncommon [5]. Notably, EBV-positive IPT-like FDCS is characterized by the neoplastic proliferation of follicular dendritic cells, accompanied by significant lymphoplasmacytic infiltration and persistent EBV presence. The key distinguishing factor between this disease and the other conditions is the atypical spindle/ovoid cells expressing some dendritic markers and the absence of expression of other cell type lineage [34]. Because reactive fibroblasts can morphologically resemble follicular dendritic cells, detecting atypical spindle cells within an inflammatory background poses a clinical challenge [6]. This complexity adds to the difficulty in clinically diagnosing this disease. At present, immunohistochemical analysis has become crucial in facilitating the diagnosis, with significant immunomarkers such as CD21, CD23, and CD35 playing pivotal roles. In our case, immunohistochemistry suggested partial expression of CD21, CD23, and CD35. The plasma cell population in the scant fibrotic areas was IgG4-positive. In a Korean study [16], a large number of IgG4-positive plasma cells were found in six EBV-positive patients with IPT-like FDCS, suggesting that EBV has a key role in IPT-like FDCS. Three cases of IPT-like FDCS, characterized by IgG4 positivity, EBV positivity, and splenic involvement, were also reported in various studies [8,14,17]. Additional markers for FDCS encompass Ki-M4 and CD14 [35], CXCL13 and clusterin [36], gamma-synuclein and D2-40 [8,16], podoplanin and the low affinity NGFR [37,38]. Furthermore, this tumor exhibits positivity for smooth muscle actin, vimentin protein, and myofibrillar protein [39]. Positive expression of Epstein-Barr virus-encoded small RNA (EBER) serves as a crucial indicator for diagnosing IPT-like FDCS. Additionally, the identification of fibrin-like deposits within blood vessels can serve as a diagnostic clue for the disease [34]. Clinicians must exercise vigilance in the differential diagnosis of IPT-like FDCS, distinguishing it from tumors such as classic Hodgkin's lymphoma, interdigitating dendritic cell sarcoma (IDCS), splenic metastases, hamartoma, hemangioma and Inflammatory myofibroblastic tumor.

The available clinical data suggest that surgical resection remains the primary approach in the clinical management of follicular dendritic cell sarcomas (FDCS). Complete surgical resection is typically the preferred treatment for patients with either primary or

recurrent disease. The role of adjuvant radiotherapy and chemotherapy remains a topic of controversy [40]. Chemotherapy is considered for patients who are not suitable candidates for surgical removal or those with involvement of multiple organs. In clinical practice, lymphoma chemotherapy regimens or sarcoma chemotherapy regimens, such as CHOP, DHAP, and AVBD, are often employed for treating FDCS. However, there is no consensus on the optimal clinical use of chemotherapy. Li J et al. [41] observed that the combination of PD-1 inhibitors and chemotherapy as a first-line treatment demonstrates promise in managing metastatic FDCS. Another study indicates that bendamustine could be considered among the chemotherapeutic options for FDCS with multiple organ involvement, although its efficacy requires further investigation [42]. Some scholars propose the feasibility of targeted therapy in patients with these diseases [43]. However, data on targeted therapies, such as complex kinase inhibitors, or immunosuppressive agents are limited, and their response varies. There is insufficient evidence to substantiate the effectiveness of either of these treatment modalities. A more profound comprehension of the genetic mechanisms and molecular drivers of FDCS may pave the way for potential therapeutic strategies. All the 30 cases we collated were treated surgically and all of them were free from postoperative recurrence and metastasis. We present a distinct case of IPT-like follicular dendritic cell sarcoma (FDCS) with thrombocytopenia as the predominant symptom, which is clinically uncommon. Given the nature of being a single case report and literature review, certain limitations should be acknowledged. Our aim is to reduce misdiagnosis and help identify undetected patients. The accumulation of case reports is pivotal for unraveling the pathophysiology of FDCS and establishing systematic treatment approaches.

IPT-like FDCS typically follows a painless course and exhibits a more indolent nature compared to classic FDCS. It was noted that IPT-like FDCS exhibits an inert biological behaviour that allows patients to achieve long-term survival even in the event of relapse [14]. Our findings corroborate this conclusion. Nonetheless, local recurrence is more prevalent in these tumors, with a recurrence rate of approximately 30–40 %, and they may progress to a more aggressive clinical course [5]. We posit that IPT-like FDCS should be characterized as moderately malignant tumors, warranting early-stage complete surgical resection.

5. Conclusion

In conclusion, splenic IPT-like FDCS is a rare entity. The clinical diagnosis of this disease poses considerable challenges, necessitating a conclusive diagnosis through pathological immunohistochemical examination. Epstein-Barr virus-encoded small RNA (EBER) in situ hybridization holds significance for the definitive diagnosis of the disease. We advocate for splenectomy as the treatment of choice for limited splenic IPT-like FDCS.

Ethics statement

Written informed consent was secured from patients for the dissemination of all images and data pertinent to this study. The study involving humans was approved by the First Affiliated Hospital of Soochow University, China. The study was conducted in accordance with the local legislation and institutional requirements.

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Data availability statement

Data included in article/supplementary material/referenced in article.

CRediT authorship contribution statement

Jiawei Jin: Writing – original draft, Methodology, Investigation, Conceptualization. **Xiaolong Zhu:** Writing – review & editing, Data curation. **Yi Wan:** Writing – review & editing, Resources. **Yang Shi:** Writing – review & editing, Validation, Supervision.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

References

[1] K. Lennert, Malignant Lymphomas Other than Hodgkin's Disease, Springer, Berlin, 1978, pp. 51–68.
[2] L. Monda, R. Warnke, J. Rosai, A primary lymph node malignancy with features suggestive of dendritic reticulum cell differentiation. A report of 4 cases, Am. J. Pathol. 122 (3) (1986 Mar) 562–572. PMID: 2420185; PMCID: PMC1888214.
[3] T. De Pas, G. Spitaleri, G. Pruneri, G. Curigliano, C. Noberasco, A. Luini, B. Andreoni, A. Testori, F. de Braud, Dendritic cell sarcoma: an analytic overview of the literature and presentation of original five cases, Crit. Rev. Oncol. Hematol. 65 (1) (2008 Jan) 1–7, https://doi.org/10.1016/j.critrevonc.2007.06.003. Epub 2007 Jul 19. PMID: 17658269.
[4] F. Facchetti, M. Simbeni, L. Lorenzi, Follicular dendritic cell sarcoma, Pathologica 113 (5) (2021 Oct) 316–329, https://doi.org/10.32074/1591-951X-331. PMID: 34837090; PMCID: PMC8720404.

[5] W. Cheuk, J.K. Chan, T.W. Shek, J.H. Chang, M.H. Tsou, N.W. Yuen, W.F. Ng, A.C. Chan, J. Prat, Inflammatory pseudotumor-like follicular dendritic cell tumor: a distinctive low-grade malignant intra-abdominal neoplasm with consistent Epstein-Barr virus association, Am. J. Surg. Pathol. 25 (6) (2001 Jun) 721–731, https://doi.org/10.1097/00000478-200106000-00003. PMID: 11395549.

[6] Y. Li, X. Yang, L. Tao, W. Zeng, M. Zuo, S. Li, L. Wu, Y. Lin, Z. Zhang, J. Yun, Y. Huang, Challenges in the diagnosis of epstein-barr virus-positive inflammatory follicular dendritic cell sarcoma: extremely wide morphologic spectrum and immunophenotype, Am. J. Surg. Pathol. 47 (4) (2023 Apr 1) 476–489, https://doi.org/10.1097/PAS.0000000000002011. Epub 2022 Dec 28. PMID: 36574358.

[7] D.N. Leng, K.J. Yu, J. Wang, Inflammatory pseudotumor-like follicular dendritic cell sarcoma with first clinical manifestation of thrombocytopenia: a case report, Medicine (Baltim.) 101 (52) (2022 Dec 30) e32528, https://doi.org/10.1097/MD.0000000000032528. PMID: 36596072; PMCID: PMC9803453.

[8] F. Pagliuca, A. Ronchi, A. Auricchio, E. Lieto, R. Franco, Inflammatory pseudotumor-like follicular/fibroblastic dendritic cell sarcoma: focus on immunohistochemical profile and association with Epstein-Barr virus, Infect. Agents Cancer 17 (1) (2022 Dec 24) 63, https://doi.org/10.1186/s13027-022-00474-8. PMID: 36566201; PMCID: PMC9789662.

[9] M. Mograbi, M.S. Stump, D.T. Luyimbazi, M.H. Shakhatreh, D.J. Grider, Pancreatic inflammatory pseudotumor-like follicular dendritic cell tumor, Case Rep Pathol 2019 (2019 Dec 5) 2648123, https://doi.org/10.1155/2019/2648123. PMID: 31885993; PMCID: PMC6915151.

[10] R. Ge, C. Liu, X. Yin, J. Chen, X. Zhou, C. Huang, W. Yu, X. Shen, Clinicopathologic characteristics of inflammatory pseudotumor-like follicular dendritic cell sarcoma, Int. J. Clin. Exp. Pathol. 7 (5) (2014 Apr 15) 2421–2429. PMID: 24966952; PMCID: PMC4069939.

[11] J. Zhang, L. He, X. Ma, J. Wang, Y. Qiu, L. Jiang, Imaging findings of inflammatory pseudotumor-like follicular dendritic cell sarcoma of the spleen: a case report and literature review, Oncol. Lett. 24 (5) (2022 Sep 22) 399, https://doi.org/10.3892/ol.2022.13519. PMID: 36276498; PMCID: PMC9533655.

[12] X. Li, Z. Shi, R. You, Y. Li, D. Cao, R. Lin, X. Huang, Inflammatory pseudotumor-like follicular dendritic cell sarcoma of the spleen: computed tomography imaging characteristics in 5 patients, J. Comput. Assist. Tomogr. 42 (3) (2018 May/Jun) 399–404, https://doi.org/10.1097/RCT.0000000000000700. PMID: 29287022.

[13] F. Chen, J. Li, P. Xie, Imaging and pathological comparison of inflammatory pseudotumor-like follicular dendritic cell sarcoma of the spleen: a case report and literature review, Front Surg 9 (2022 Sep 5) 973106, https://doi.org/10.3389/fsurg.2022.973106. PMID: 36132202; PMCID: PMC9483013.

[14] C. Nie, X. Xie, H. Li, Y. Li, Z. Chen, Y. Li, Z. Li, Epstein-Barr virus-positive inflammatory follicular dendritic cell sarcoma with significant granuloma: case report and literature review, Diagn. Pathol. 19 (1) (2024 Feb 16) 34, https://doi.org/10.1186/s13000-024-01457-6. PMID: 38365739; PMCID: PMC10870656.

[15] J.F. Hang, L.C. Wang, C.R. Lai, Cytological features of inflammatory pseudotumor-like follicular dendritic cell sarcoma of spleen: a case report, Diagn. Cytopathol. 45 (3) (2017 Mar) 230–234, https://doi.org/10.1002/dc.23626. Epub 2016 Oct 24. PMID: 27775241.

[16] J.Y. Choe, H. Go, Y.K. Jeon, J.Y. Yun, Y.A. Kim, H.J. Kim, J. Huh, H. Lee, D.H. Shin, J.E. Kim, Inflammatory pseudotumor-like follicular dendritic cell sarcoma of the spleen: a report of six cases with increased IgG4-positive plasma cells, Pathol. Int. 63 (5) (2013 May) 245–251, https://doi.org/10.1111/pin.12057. PMID: 23714251.

[17] I.A. Ungureanu, R.M. Lupinacci, M. Parrens, J.F. Emile, Granulomatous splenic mass with necrosis revealing an EBV-positive inflammatory follicular dendritic cell sarcoma, J. Surg. Case Rep. 2022 (5) (2022 May 5) rjac034, https://doi.org/10.1093/jscr/rjac034. PMID: 35531436; PMCID: PMC9071999.

[18] B. Morales-Vargas, K. Deeb, D. Peker, Clinicopathologic and molecular analysis of inflammatory pseudotumor-like follicular/fibroblastic dendritic cell sarcoma: a case report and review of literature, Turk Patoloji Derg 37 (3) (2021) 266–272, https://doi.org/10.5146/tjpath.2021.01523. PMID: 34514557; PMCID: PMC10510619.

[19] Y. Chen, H. Shi, H. Li, T. Zhen, A. Han, Clinicopathological features of inflammatory pseudotumour-like follicular dendritic cell tumour of the abdomen, Histopathology 68 (6) (2016 May) 858–865, https://doi.org/10.1111/his.12851. Epub 2015 Oct 28. PMID: 26332157.

[20] X. Liu, L. Cao, W. Chin, J. Yu, Y. Liu, S. Zheng, Epstein-Barr virus-negative inflammatory pseudotumor-like variant of follicular dendritic cell sarcoma of the liver: a case report and literature review, Clin Res Hepatol Gastroenterol 45 (1) (2021 Jan) 101457, https://doi.org/10.1016/j.clinre.2020.05.007. Epub 2020 Jun 13. PMID: 32540141.

[21] R. Kazemimood, Hamedani F. Saei, A. Sharif, S. Gaitonde, E. Wiley, P.C. Giulianotti, J.V. Groth, A rare case of epstein-barr virus negative inflammatory pseudotumor-like follicular dendritic cell sarcoma presenting as a solitary colonic mass in a 53-year-old woman; case report and review of literature, Appl. Immunohistochem. Mol. Morphol. 25 (5) (2017 May/Jun) e30–e33, https://doi.org/10.1097/PAI.0000000000000405. PMID: 27299190.

[22] S.T. Pan, C.Y. Cheng, N.S. Lee, P.I. Liang, S.S. Chuang, Follicular dendritic cell sarcoma of the inflammatory pseudotumor-like variant presenting as a colonic polyp, Korean J Pathol 48 (2) (2014 Apr) 140–145, https://doi.org/10.4132/KoreanJPathol.2014.48.2.140. Epub 2014 Apr 28. PMID: 24868227; PMCID: PMC4026805.

[23] J. Hu, D. Huang, C. Xu, Y. Chen, H. Ma, Z. Shen, Epstein-barr virus-positive inflammatory follicular dendritic cell sarcoma presenting as a colonic polyp: report of a case with a literature review, Medicina (Kaunas) 59 (7) (2023 Jul 21) 1341, https://doi.org/10.3390/medicina59071341. PMID: 37512154; PMCID: PMC10385426.

[24] M. Mograbi, M.S. Stump, D.T. Luyimbazi, M.H. Shakhatreh, D.J. Grider, Pancreatic inflammatory pseudotumor-like follicular dendritic cell tumor, Case Rep Pathol 2019 (2019 Dec 5) 2648123, https://doi.org/10.1155/2019/2648123. PMID: 31885993; PMCID: PMC6915151.

[25] H. He, Q. Xue, F. Tan, L. Yang, X. Wang, Y. Gao, Y. Mao, J. Mu, D. Wang, J. Zhao, L. Zhao, S. Gao, A rare case of primary pulmonary inflammatory pseudotumor-like follicular dendritic cell sarcoma successfully treated by lobectomy, Ann. Transl. Med. 9 (1) (2021 Jan) 77, https://doi.org/10.21037/atm-20-4965. PMID: 33553370; PMCID: PMC7859798.

[26] Y. Kitamura, Y. Takayama, A. Nishie, Y. Asayama, Y. Ushijima, N. Fujita, K. Morita, S. Baba, Y. Kubo, K. Shirabe, H. Honda, Inflammatory pseudotumor-like follicular dendritic cell tumor of the spleen: case report and review of the literature, Magn. Reson. Med. Sci. 14 (4) (2015) 347–354, https://doi.org/10.2463/mrms.2014-0052. Epub 2015 Feb 12. PMID: 25740238.

[27] B. Sander, P. Middel, B. Gunawan, H.J. Schulten, F. Baum, M.M. Golas, F. Schulze, E. Grabbe, R. Parwaresch, L. Füzesi, Follicular dendritic cell sarcoma of the spleen, Hum. Pathol. 38 (4) (2007 Apr) 668–672, https://doi.org/10.1016/j.humpath.2006.08.030. PMID: 17367608.

[28] H. Go, Y.K. Jeon, J. Huh, S.J. Choi, Y.D. Choi, H.J. Cha, H.J. Kim, G. Park, S. Min, J.E. Kim, Frequent detection of BRAF(V600E) mutations in histiocytic and dendritic cell neoplasms, Histopathology 65 (2) (2014 Aug) 261–272, https://doi.org/10.1111/his.12416. Epub 2014 May 12. PMID: 24720374.

[29] F.K. Bruehl, E. Azzato, L. Durkin, D.H. Farkas, E.D. Hsi, S.L. Ondrejka, Inflammatory pseudotumor-like follicular/fibroblastic dendritic cell sarcomas of the spleen are EBV-associated and lack other commonly identifiable molecular alterations, Int. J. Surg. Pathol. 29 (4) (2021 Jun) 443–446, https://doi.org/10.1177/1066896920949675. Epub 2020 Aug 12. PMID: 32787485.

[30] M.C. Ramsey, P.J.B. Sabatini, G. Watson, T. Chawla, M. Ko, A. Sakhdari, Case Report: identification of a novel STAT3 mutation in EBV-positive inflammatory follicular dendritic cell sarcoma, Front. Oncol. 13 (2023 Oct 26) 1266897, https://doi.org/10.3389/fonc.2023.1266897. PMID: 37965457; PMCID: PMC10640977.

[31] G. Frigola, M. Bühler, M. Marginet, A. Enjuanes, F. Nadeu, N. Papaleo, M. Salido, E. Haralambieva, J. Alamo, F. Garcia-Bragado, R. Álvarez, R. Ramos, I. Aldecoa, E. Campo, L. Colomo, O. Balagué, MYC and TP53 alterations but not MAPK pathway mutations are common oncogenic mechanisms in follicular dendritic cell sarcomas, Arch. Pathol. Lab Med. 147 (8) (2023 Aug 1) 896–906, https://doi.org/10.5858/arpa.2021-0517-OA. PMID: 36355424.

[32] A. Baber, P. Legendre, P. Palmic, A. Lupo-Mansuet, B. Burroni, C. Azoulay, T.A. Szwebel, N. Costedoat-Chalumeau, K. Leroy, H. Blons, J.Y. Blay, P. Boudou-Rouquette, B. Terrier, EBV-positive inflammatory follicular dendritic cell sarcoma of the spleen: report of an aggressive form with molecular characterization, Int. J. Surg. Pathol. 32 (1) (2024 Feb) 150–154, https://doi.org/10.1177/10668969231168345. Epub 2023 May 8. PMID: 37157817.

[33] P.N. Martins, S. Reddy, A.B. Martins, M. Facciuto, Follicular dendritic cell sarcoma of the liver: unusual presentation of a rare tumor and literature review, Hepatobiliary Pancreat. Dis. Int. 10 (4) (2011 Aug) 443–445, https://doi.org/10.1016/s1499-3872(11)60076-3. PMID: 21813397.

[34] X.Q. Li, W. Cheuk, P.W. Lam, Z. Wang, F. Loong, M.L. Yeong, P. Browett, J. McCall, J.K. Chan, Inflammatory pseudotumor-like follicular dendritic cell tumor of liver and spleen: granulomatous and eosinophil-rich variants mimicking inflammatory or infective lesions, Am. J. Surg. Pathol. 38 (5) (2014 May) 646–653, https://doi.org/10.1097/PAS.0000000000000170. PMID: 24503752.

[35] R. Fonseca, M. Yamakawa, S. Nakamura, P. van Heerde, M. Miettinen, T.W. Shek, O. Myhre Jensen, M.C. Rousselet, A. Tefferi, Follicular dendritic cell sarcoma and interdigitating reticulum cell sarcoma: a review, Am. J. Hematol. 59 (2) (1998 Oct) 161–167, https://doi.org/10.1002/(sici)1096-8652(199810)59:2<161::aid-ajh10>3.0.co;2-c. PMID: 9766802.
[36] L. Lorenzi, C. Döring, T. Rausch, V. Benes, S. Lonardi, M. Bugatti, E. Campo, J. Cabeçadas, I. Simonitsch-Klupp, A. Borges, J. Mehta, C. Agostinelli, S.A. Pileri, F. Facchetti, M.L. Hansmann, S. Hartmann, Identification of novel follicular dendritic cell sarcoma markers, FDCSP and SRGN, by whole transcriptome sequencing, Oncotarget 8 (10) (2017 Mar 7) 16463–16472, https://doi.org/10.18632/oncotarget.14864. PMID: 28145886; PMCID: PMC5369977.
[37] D.K. Marsee, G.S. Pinkus, J.L. Hornick, Podoplanin (D2-40) is a highly effective marker of follicular dendritic cells, Appl. Immunohistochem. Mol. Morphol. 17 (2) (2009 Mar) 102–107, https://doi.org/10.1097/PAI.0b013e318183a8e2. PMID: 18838918.
[38] N. Kasajima-Akatsuka, K. Maeda, Development, maturation and subsequent activation of follicular dendritic cells (FDC): immunohistochemical observation of human fetal and adult lymph nodes, Histochem. Cell Biol. 126 (2) (2006 Aug) 261–273, https://doi.org/10.1007/s00418-006-0157-6. Epub 2006 Feb 10. PMID: 16470387.
[39] M. Martel, D. Sarli, M. Colecchia, J. Coppa, R. Romito, M. Schiavo, V. Mazzaferro, J. Rosai, Fibroblastic reticular cell tumor of the spleen: report of a case and review of the entity, Hum. Pathol. 34 (9) (2003 Sep) 954–957, https://doi.org/10.1016/s0046-8177(03)00399-x. PMID: 14562295.
[40] C. Saygin, D. Uzunaslan, M. Ozguroglu, M. Senocak, N. Tuzuner, Dendritic cell sarcoma: a pooled analysis including 462 cases with presentation of our case series, Crit. Rev. Oncol. Hematol. 88 (2) (2013 Nov) 253–271, https://doi.org/10.1016/j.critrevonc.2013.05.006. Epub 2013 Jun 5. PMID: 23755890.
[41] J. Li, M. Ren, F. Bi, Y. Chen, Z. Li, Favorable response to PD-1 inhibitor plus chemotherapy as first-line treatment for metastatic follicular dendritic cell sarcoma of the spleen: a case report, Front. Immunol. 14 (2023 Aug 25) 1228653, https://doi.org/10.3389/fimmu.2023.1228653. PMID: 37691960; PMCID: PMC10485249.
[42] M. Sasaki, H. Izumi, T. Yokoyama, M. Kojima, A. Hosono, Follicular dendritic cell sarcoma treated with a variety of chemotherapy, Hematol. Oncol. 35 (4) (2017 Dec) 905–908, https://doi.org/10.1002/hon.2364. Epub 2016 Oct 13. PMID: 27734516; PMCID: PMC5763378.
[43] A. Wu, S. Pullarkat, Follicular dendritic cell sarcoma, Arch. Pathol. Lab Med. 140 (2) (2016 Feb) 186–190, https://doi.org/10.5858/arpa.2014-0374-RS. PMID: 26910224.