J Surg Med. 2022;6(11):929-931.
DOI: 10.28982/josam.1096683

Journal of Surgery and Medicine
e-ISSN: 2602-2079

Follicular dendritic cell sarcoma of the spleen: A case report

Mehmet Sait Özsoy [1], Nuray Colapkulu Akgul [2], Aman Gapbarov [1], Ozgur Ekinci [1], Salih Tosun [1], Orhan Alimoglu [1]

[1] İstanbul Medeniyet University, Goztepe Prof. Dr. Suleyman Yalcin City Hospital, General Surgery Clinic, Istanbul, Turkey
[2] Department of General Surgery, Gebze Fatih Public Hospital, Kocaeli, Turkey

ORCID ID of the author(s)

MSO: 0000-0003-2935-8463
NCA: 0000-0002-3033-8702
AG: 0000-0001-5892-1169
OE: 0000-0002-2020-1913
ST: 0000-0002-5033-4477
OA: 0000-0003-2130-2529

Corresponding Author
Mehmet Sait Ozsoy
Istanbul Medeniyet University, Goztepe Prof. Dr. Suleyman Yalcin City Hospital, Dr. Erkin Street No:1 Kadikoy, Istanbul, Turkey
E-mail: saitozsoy@gmail.com

Informed Consent
The authors stated that the written consent was obtained from the patient presented with images in the study.

Conflict of Interest
No conflict of interest was declared by the authors.

Financial Disclosure
The authors declared that this study has received no financial support.

Previous Presentation
This study was presented as a poster at the 21st National Surgery Congress in Antalya, Turkey on April 11 – 15, 2018.

Published
2022 November 26

Copyright © 2022 The Author(s)
Published by JOSAM
This is an open access article distributed under the terms of the Creative Commons Attribution-Non Commercial-NoDerivatives License 4.0 (CC BY-NC-ND 4.0) where it is permissible to download, share, remix, transform, and buildup the work provided it is properly cited. The work cannot be used commercially without permission from the journal.

Abstract

Follicular dendritic cell sarcomas (FDCS) are spindle cell lesions that are large, slow-growing masses in either nodal or extranodal regions or both and originate from B-cell follicles of the lymph nodes. Most tumors originate from the cervical lymph nodes, but retroperitoneal and mediastinal origins have also been reported. Extranodal areas include soft tissues, skin, tonsils, gastrointestinal tract, liver, and spleen. The spleen is an uncommon location for an FDCS and for this reason, the tumor may be underdiagnosed or overlooked because of confusion with other solid tumors. In this study, we present a patient with a splenic FDCS who presented clinically with abdominal pain and diarrhea. The patient underwent a splenectomy and had an uneventful remission.

Keywords: Follicular dendritic cells, Sarcoma, Spleen

Introduction

Follicular dendritic cells may demonstrate as antigen presenting cells in B-lymphocyte mediated humoral immunity. A follicular dendritic cell sarcoma (FDCS) is an uncommon type of tumor with a low incident rate. Although these tumors are primarily found in cervical lymph nodes, they can be localized in extranodal regions. Extranodal areas include soft tissue, skin, tonsil, gastrointestinal tract, liver, and spleen. FDCS have a serpiginous growth pattern of the lymph nodes that may progress by leaving residual areas between the nodules [1]. FDSCs may clinically present with regional growth of lymph nodes or intraabdominal masses. Due to their aggressive biological patterns, they are reported to have high local recurrence and metastasis rates and can cause a decrease in patient survival [2]. This article presents a case of a FDCS that was localized in spleen.

How to cite: Özsoy MS, Akgul NC, Gapbarov A, Ekinci O, Tosun S, Alimoglu O. Follicular dendritic cell sarcoma of the spleen: A case report. J Surg Med. 2022;6(11):929-931.

Case presentation

We received written consent from the patient after which we designed the study. A 58-year-old male with no history of chronic disease presented with ongoing abdominal pain and diarrhea over the course of one month. Physical examination revealed a closed Traube's space and splenomegaly. Laboratory investigations showed elevated levels of white blood cells (17.3 x 10^3/L), C-Reactive Protein (CRP) of 8.28 mg/L (reference range: 0-5 mg/L), cancer antigen-15-3 (CA-15-3) of 30.1 U/mL (reference: <30 U/mL), and CA-125 of 45 U/mL (reference: <35 U/mL). No other laboratory abnormalities were found. On a computed tomography (CT) scan, the long axis of the spleen was found to be remarkably increased by 168 mm. A bilobular, heterogeneously enhanced, solid lesion measuring 14 cm extending from the hilus of the spleen to the lower pole was detected, and the left kidney was compressed by the lesion inferomedially (Figure 1). Endoscopic evaluation showed no pathological findings in the upper or lower gastrointestinal tract. The patient underwent surgery for explorative laparotomy and was diagnosed with a primary splenic tumor. The tumor had not invaded the surrounding tissues and was limited to the splenic capsule. The tumor was excised with en-bloc splenectomy (Figure 2). Abdominal exploration revealed no additional macroscopic pathology. Histopathological examination confirmed that the lesion was an FDCS with spindle cell fascicles that formed a storiform pattern (Figure 3). The splenic capsule was intact, and the surgical margins were found to be negative in agreement with a macroscopic examination. Significant cytological atypia and extensive coagulative necrosis were detected in the lesion. Based on immunohistochemistry, examination showed positive cluster of differentiation (CD)-23 and -21 whereas CD-3, -20, -117, and -138 were found to be negative. The percentage of Ki-67-stained tumor in the hotspots was found to be 60%. The patient was discharged without any post-operative complications and has completely recovered. The patient was referred to the oncology clinic for follow-ups.

Figure 1: Computed tomography (CT) view of the heterogeneously enhanced, bilobular solid splenic lesion (arrow)

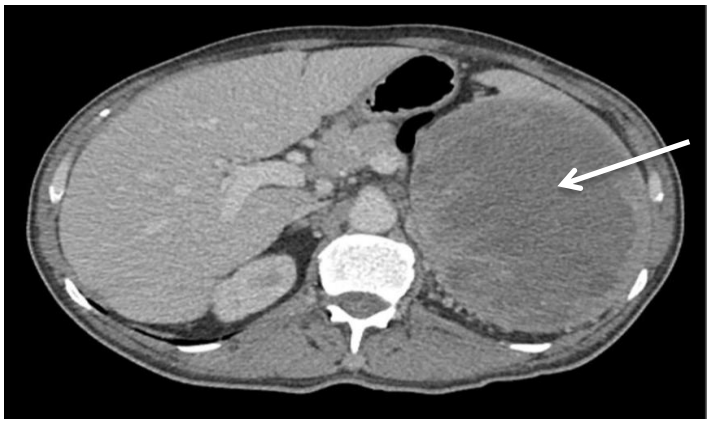


Figure 2: Macroscopic view of the excised tumor with en-bloc splenectomy.

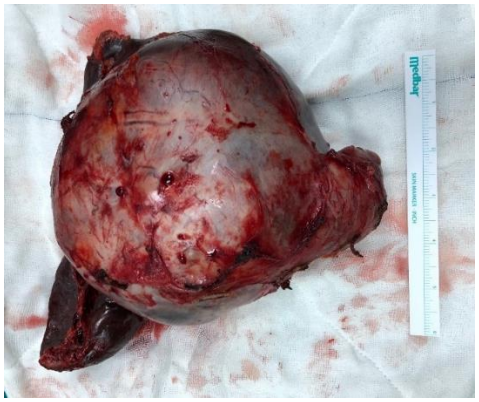
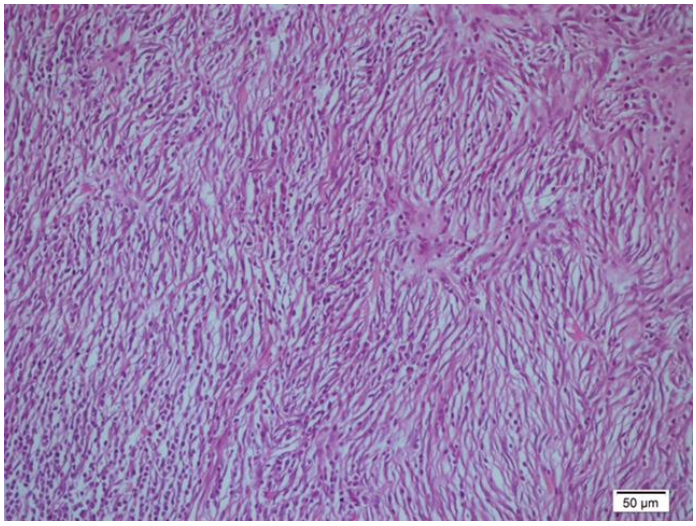


Figure 3: Histopathological view of the spindle cell fascicles that formed a storiform pattern in the resected spleen material. Oval cells and lymphocytes are found between the spindle cells.



Discussion

FDCSs are recently frequently described in the literature [3]. Most of these tumors originate from the cervical lymph nodes, whereas mediastinal and retroperitoneal origins have also been reported. Retroperitoneal tumors have only been reported as sporadically developed tumors. Due to their aggressive histopathological nature, high incidence of local recurrence and metastasis and poor survival rates have been reported [2]. Although patients often present with regional enlarged lymph nodes, spleen sarcomas mostly present as a palpable mass in the abdomen [4]. FDCS are rare malignancies with heterogeneous outcomes. Patients with bulky, extranodal, and/or intra-abdominal disease at presentation usually have poor outcomes [5].

Physical examination revealed a closed Traube's space and splenomegaly. Elevated WBCs, CRP, and tumor markers were reported. Radiological evaluation revealed a bilobular, well-bordered splenic mass that had not invaded the surrounding tissues.

FDCSs have a serpiginous growth pattern of the lymph nodes that may progress by leaving residual areas between the nodules. Immunohistochemically, the tumor has a unique phenotypic profile, including follicular dendritic cell markers: (10 CD21 and (2) CD35. They also express CD23, CD68, fascin, and clusterin, whereas CD1a, S100, desmin, actin, and cytokeratin expression is not found [1].

CT imaging with focal, course, single, or multiple masses occurring intra-abdominally and/or retroperitoneally, which are relatively larger sized and have significant internal necrosis, should lead to consideration of a possible FDCS [6]. Magnetic resonance imaging is helpful in determining the extent of the lesion, its relationship with the surrounding tissues, and the tumor stage. T1-weighted sections indicated an isointense expansive mass, whereas T2-weighted sections indicated a homogeneous, slightly hyperintense mass after a gadolinium infusion was administered [6].

FDCS is a rare type of tumor with low incidence rates. No specific treatment guidelines for FDCSs are available. Resection of the tumor with a wide surgical margin is usually recommended much the same as is done for other sarcomas [7]. Currently, surgery is the primary choice of treatment, and in the case of resectable FDCS, adjuvant chemotherapy or radiotherapy can be used to improve survival rates [8].

Conclusion

In conclusion, FDCS is an uncommon tumor that can remain underdiagnosed or overlooked since it has imaging features similar to other solid tumors. The current report describes the main clinical features and discusses the diagnostic challenges and treatment options. Immunohistochemistry and specific FDCS markers are important for determining the tumor differentiation grade. Currently, complete surgical resection is warranted. After proper surgical resection, FDCS generally has a favorable prognosis.

References

1. Hsi ED. Hematopathology E-book (3rd ed.). 2017, Elsevier Health Sciences. Retrieved from https://www.perlego.com/book/2938601/hematopathology-ebook-pdf (Original work published 2017).
2. Carboni F, Covello R, Bertini L, Valle M. Uncommon retroperitoneal tumour: follicular dendritic cell sarcoma. Acta Chir Belg. 2021;121:219–21. doi: 10.1080/00015458.2019.1689646
3. Nakashima T, Kuratomi Y, Shiratsuchi H, Yamamoto H, Yasumatsu R, Yamamoto T, et al. Follicular dendritic cell sarcoma of the neck; a case report and literature review. Auris nasus larynx. 2002;29:401–3. doi: 10.1016/s0385-8146(02)00056-1
4. Hu T, Wang X, Yu C, Yan J, Zhang X, Li L, et al. Follicular dendritic cell sarcoma of the pharyngeal region. Oncol Lett. 2013;5:1467–76. doi: 10.3892/ol.2013.1224
5. Jain P, Milgrom SA, Patel KP, Nastoupil L, Fayad L, Wang M, et al. Characteristics, management, and outcomes of patients with follicular dendritic cell sarcoma. Br J Haematol. 2017;178:403–12. doi: 10.1111/bjh.14672
6. Clement P, Saint-Blancard P, Minvielle F, Le Page P, Kossowski M. Follicular dendritic cell sarcoma of the tonsil: a case report. Am J Otolaryngol. 2006;27:207–10. doi: 10.1016/j.amjoto.2005.09.003
7. Wang L, Xu D, Qiao Z, Shen L, Dai H, Ji Y. Follicular dendritic cell sarcoma of the spleen: A case report and review of the literature. Oncol Lett. 2016;12:2062–4. doi: 10.3892/ol.2016.4826
8. Chen HM, Shen YL, Liu M. Primary hepatic follicular dendritic cell sarcoma: A case report. World J Clin Cases. 2019;7:785–91. doi: 10.12998/wjcc.v7.i6.785.

The National Library of Medicine (NLM) citation style guide has been used in this paper.