

## Case Report

# Laparoscopic Splenectomy for Follicular Dendritic Cell Sarcoma with Pseudotumor-Like Feature is Viable Alternative Option

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**Background:** Follicular dendritic cell (FDC) sarcoma is an extremely rare type of tumor. It is located in the lymphoid follicle that originates in the antigen presenting cell of B-cell follicles in the germinal center. Most FDC sarcomas are found in lymph nodes. Some are reported to be found in the liver or spleen.

**Objective:** The present study was reported the rare case of splenic sarcoma and the alternative option of treatment by laparoscopic splenectomy.

**Material and Method:** A case report study in patient who presented with splenic mass and performed laparoscopic splenectomy. The pathological results reported FDC sarcoma with pseudotumor-like feature of spleen.

**Results:** In the present study, we report a case of a 69-year old woman who visited our hospital for scheduled annual examination. She had no symptoms. During ultrasound, we discovered abdominal mass, so we made further investigations by CT scan, which confirmed our suspicions. Although this operation is normally performed by standard open surgery, we performed a successful laparoscopic splenectomy to remove the mass. Our experienced surgeons and excellent facilities enabled this to be possible. There were no post-operative complications. The pathological and immunohistochemical results reported FDC sarcoma with pseudotumor-like feature of spleen. At the six-month follow-up, we found no evidence of tumor recurrence.

**Conclusion:** In summary, we believe that in certain selected cases of splenic tumors, laparoscopic splenectomy is a viable alternative option, provided that the performing surgeon is experienced and has access to suitable facilities.

**Keywords:** Follicular dendritic cell, Pseudotumor-like feature, Sarcoma, Laparoscopic, Splenectomy

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Follicular dendritic cell (FDC) sarcomas are classified into histiocytic and dendritic cell neoplasms, according to the classification of hematopoietic and lymphoid tumors of The World Health Organization (WHO) in 2016<sup>(1)</sup>. FDC is located in the lymphoid follicle that originates in the antigen presenting cell of B-cell follicles in the germinal center. FDC tumor is a rare condition. The majority of FDC tumors occur in the lymph nodes. A small number of cases are found in the spleen or liver<sup>(2)</sup>. Most patients do not present with clinical symptoms. In 1986, Monda et al described the SDC sarcoma as extremely rare<sup>(3)</sup>. Now, many reports of extranodal FDC sarcoma are increasing. In this paper, we report the case of FDC sarcoma with pseudotumor-like features of the spleen. The patient had not received

systemic chemotherapy or radiotherapy after complete surgical resection. After surgery, the patient had good remission and was healthy.

### Case Report

A 69-year-old woman visited our out-patient Department for scheduled annual medical examination. During abdominal ultrasonography, the radiologist discovered a splenic mass. The patient did not have gastrointestinal symptoms, compressive symptoms, or urinary symptoms. The patient was sent for further investigation by abdominal Computer Tomography (CT) (Fig. 1, 2). The axial helical CT scan of her entire abdomen showed a lobulated hypervascular mass measuring 5.4x5x5.9 cm in the spleen. The spleen appeared normal in size and shape. Other findings showed a small wedge-shape hypodense lesion in the subcapsular region of the hepatic segment IVb, which was focal fat infiltration, or benign lesion. Neither intraabdominal lymphadenopathy nor ascites were found. Differential diagnosis included atypical

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**Fig. 1** Splenic mass in horizontal view of abdominal CT scan.



**Fig. 2** Splenic mass in coronal view of abdominal CT scan.

Hemangioma, Angiosarcoma, or metastasis. The patient underwent laparoscopic splenectomy. Before surgery, the patient had vaccinations to reduce the risk of sepsis that included pneumococcal vaccine, H. influenza vaccine, and meningococcal vaccine. During the operation, we found the splenic mass size to be 5x7x6 cm. The mass did not invade other organs. The inside of the patient's abdomen appeared normal, including size and appearance of the liver, and no evidence of lymphadenopathy or ascites. A successful laparoscopic splenectomy was performed, without any ruptures or



**Fig. 3** Complete laparoscopic splenectomy.



**Fig. 4** Tumor inside spleen.

spillage (Fig. 3, 4). The operative time was 90 minutes. Estimated blood loss was 100 cc. No complications were experienced during the operation. The patient was admitted and discharged five days after surgery. The

tumor was sent for pathological examination. The pathology reported that the specimen measured 9x6.5x5 cm and weighed 164.6 grams. The whole tumor was limited to the spleen. The surgical margin was free of the tumor. The pathologic diagnosis was spindle cell tumor. Immunohistochemistry techniques were used to differentiate the type of tumor, including CD3, CD20, CD68, CD30, CD31, CD34, CD8, CD21, CD23, Anaplastic Lymphoma Kinase (ALK) 1, Desmin, S-100, Smooth Muscle Actin (SMA), Kappa, Lambda, Ki-67, Epstein-Barr virus-encoded small RNAs (EBER), Leukocyte Common Antigen (LCA), CD1a and Epithelial Membrane Antigen (EMA). The neoplastic cells showed positive immunoactivity for CD21, CD23 (focal), SMA, Ki-67 in 5% and EBER. The neoplastic cells showed negative immunoactivity for CD3, CD20, CD30, kappa, lambda, AKL1, CD31, CD34, CD8, desmin, S-100, CD68, LCA, CD1a and EMA. Thus, the definite diagnosis was follicular dendritic cell sarcoma with inflammatory pseudotumor-like features. The patient had not received systemic chemotherapy or radiotherapy after complete surgical resection. The follow-up time was six months after surgery. The CT chest and abdomen scans were performed for evaluation and showed no evidence of local recurrence or metastasis (Fig. 5, 6). The patient's health condition was normal.

### Discussion

There are four different types of FDC existing in lymph nodes, which are histiocytic, fibroblastic, interdigitating, and follicular<sup>(4)</sup>. FDC is localized in the germinal center of lymphoid follicle and stimulates B-cell growth and proliferation<sup>(5)</sup>. FDC sarcoma is a rare neoplasm. The disease occurs at the nodal and extra-nodal sites. For the nodal site, symptoms are lymphadenopathy, especially in the axilla and cervical areas. For the extra-nodal sites, the tumors are uncommon and occur in many locations, such as the liver, spleen, pancreas, mesentery, oral cavity, and tonsils<sup>(6)</sup>. Most patients with extra-nodal FDC sarcoma do not show obvious signs or symptoms. Body imaging is a utility for diagnosis, especially intra-abdominal FDC sarcoma. Abdominal CT scans with contrast show the combination characteristics of delay, thus enhancing the tumor, together with a capsular-like rim that differentiates FDC sarcoma from other splenic tumors<sup>(2)</sup>. Inflammatory Pseudo-Tumor (IPT) was represented by the benign reactive condition. The histology of IPT was presented with proliferation of the spindle cell tumor with background of inflammation and small vessels in the lymph node capsule without an

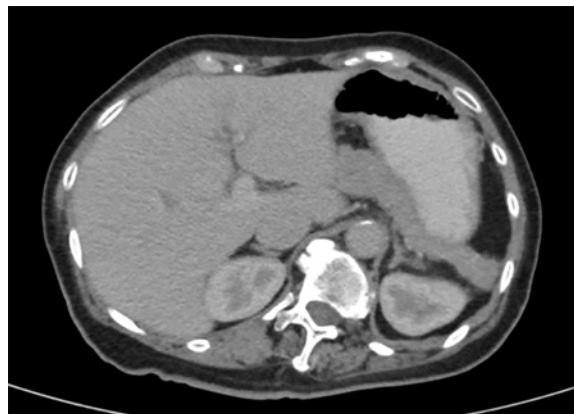


Fig. 5 Horizontal view of abdominal CT scan at 6 months after operation.

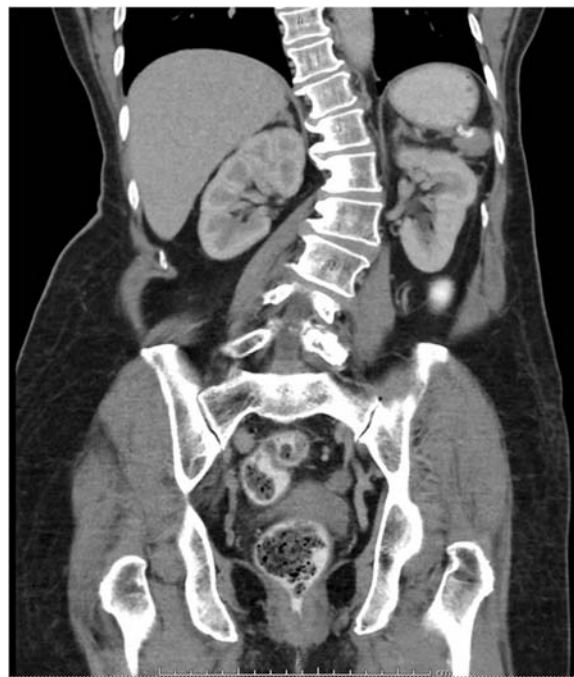


Fig. 6 Coronal view of abdominal CT scan at 6 months after operation.

aggressive growth pattern. The histological results of IPT were divided into many subtypes, such as inflammatory myofibroblastic pseudo-tumor and follicular dendritic reticulum cell tumor, which are distinguished by immunohistochemistry<sup>(7)</sup>. In 2014, Rong Ge et al reported the clinic-pathologic characteristics of inflammatory pseudotumor-like follicular dendritic cell sarcoma, and described the distinctive variants of IPT-like FDC sarcoma compared

to conventional FDC sarcoma. For example, that the majority of IPT-like FDC sarcoma patients are female, IPT-like FDC sarcoma was mostly found at the spleen and liver, whereas conventional FDC sarcoma involved lymph nodes. In addition, conventional FDC sarcoma was more aggressive than IPT-like FDC sarcoma and IPT-like FDC sarcoma was strongly associated with Epstein-Barr virus (EBV)<sup>(8,9)</sup>. The immunohistochemistry played an important role in diagnosing IPT-like FDC sarcoma. CD21 and CD35 were the most widely-used diagnostic markers. CD23 was limited because of a low positive rate and some focal stain. Immunohistochemistry positive results in SMA, S-100, Ki67, EMA and CD68 can be a diagnosis of IPT-like FDC sarcoma<sup>(8)</sup>. Approximately 12% of IPT-like FDC sarcomas are associated with EBV<sup>(10)</sup>. Most IPT-like FDC sarcomas are positive to EBER marker because EBV has an important role for genesis of IPT-like FDC sarcoma, but the mechanism is still unclear<sup>(11)</sup>. CD1a and CD45 lack expression in FDC sarcoma<sup>(9)</sup>.

Like any soft tissue sarcoma, the curative treatment in FDC sarcoma was curative surgical resection with a free margin from the tumor. Conventional solutions are open standard abdominal surgery to enable the surgeon to fully control tumor handling and any ruptures or spillage. Nowadays, with new facilities, experienced surgeons can consider a laparoscopic approach to fully and successfully remove cancerous tumors with a free margin and without disadvantages when compared to open surgery, proven by this patient's case. In previous studies, systemic chemotherapy was used for treatment, including a cyclophosphamide, hydroxydaunorubicin, oncovin, carboplatin or Adriamycin<sup>(4)</sup>. The indication for systemic therapy included incomplete surgical resection, bulky tumor, or initial diagnosis of non-Hodgkin's lymphoma. At present, the role of systemic therapy or radiotherapy is not clearly defined.

### Conclusion

IPT-like FDC sarcoma is a rare condition. Most patients present without obvious signs and symptoms. Imaging and immunohistochemistry play an important role for useful diagnosis. Although chemotherapy and radiotherapy are useful treatments, the best and only full cure is surgical resection. In this case, the patient received curative surgical laparoscopic resection that completely removed the tumor, without systemic chemotherapy or radiotherapy treatment. At the follow-up time, the patient was healthy and showed no signs of recurrence or metastasis. Experienced surgeons

with adequate facilities can consider a laparoscopic approach as well as the traditional open standard abdominal surgery.

### What is already known on this topic?

Follicular dendritic cell (FDC) sarcoma is an extremely rare type of tumor. It is located in the lymphoid follicle that originates in the antigen presenting cell of B-cell follicles in the germinal center. The majority of FDC tumors occur in the lymph nodes. A small number of cases are found in the spleen or liver. The best and only full cure is surgical resection.

### What this study adds?

In this case, the patient received curative surgical laparoscopic resection that completely removed the tumor. At the follow-up time, the patient was healthy and showed no signs of recurrence or metastasis. The laparoscopic approach as well as the optional treatment for this condition.

### Potential conflicts of interest

None.

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### การผ่าตัดม้ามผ่านกล้องในโรค *Follicular Dendritic Cell Sarcoma with Pseudotumor-like feature*

ธีรวิศุทธิ์ รักชอบ, ไชยรัตน์ ทรัพย์สมุทรชัย, ปัตตาเวีย ช้อยเครือ

**ภูมิหลัง:** *Follicular dendritic cell (FDC) sarcoma* เป็นโรคเนื้องอกที่พบน้อยเกิดใน *lymphoid follicle* ซึ่งกำเนิดจาก *antigen presenting cell of B-cell follicles* ใน *germinal center*. *FDC sarcomas* ส่วนใหญ่พบบริเวณต่อมท่อน้ำเหลืองมีบางรายงานพบที่ตับและม้าม

**วัตถุประสงค์:** เพื่อรายงานภาวะโรค *FDC Sarcoma with Pseudotumor-like feature* ของม้ามซึ่งเป็นภาวะที่พบน้อยรวมถึงการเสนอวิธีการรักษาด้วยการผ่าตัดม้ามผ่านทางกล้อง

**วัสดุและวิธีการ:** เคสผู้ป่วยที่ตรวจพบก้อนที่ม้ามและได้รับการรักษาด้วยการผ่าตัดม้ามผ่านทางกล้องและวินิจฉัยว่าเป็น *FDC Sarcoma with Pseudotumor-like feature*

**ผลการศึกษา:** ในการศึกษานี้ได้รายงานผู้ป่วยหญิงอายุ 69 ปี ตรวจพบก้อนที่ท้องจากการตรวจร่างกายประจำปีโดยวิธีการทำอัลตราซาวด์ โดยผู้ป่วยไม่มีอาการผิดปกติ และได้ส่งตรวจเอกซเรย์คอมพิวเตอร์เพื่อยืนยันการวินิจฉัยในภาวะนี้ โดยทั่วไปรักษาด้วยการผ่าตัดปิดช่องท้อง แต่ในผู้ป่วยรายนี้ใช้วิธีการผ่าตัดรักษาโดยวิธีการผ่าตัดผ่านกล้องเพื่อนำม้ามออก (*laparoscopic splenectomy*) ซึ่งสามารถทำได้ในทีมที่มีความพร้อมทั้งเครื่องมือและแพทย์ผู้ผ่าตัดที่มีประสบการณ์และความชำนาญในผู้ป่วยรายนี้ไม่พบภาวะแทรกซ้อนหลังการผ่าตัด ผลขึ้นเนื้อรายงานพบ *FDC sarcoma with pseudotumor-like feature* ของม้ามระหว่างการติดตามการรักษาที่ 6 เดือน ไม่พบรอยโรคกลับเป็นซ้ำ

**สรุป:** การผ่าตัดรักษาก้อนเนื้องอกที่ม้ามโดยวิธี *laparoscopic splenectomy* เป็นอีกทางเลือกหนึ่งที่ได้ผลดีหากอยู่ในสถานที่ที่มีความพร้อมทั้งเครื่องมือและแพทย์ผู้ผ่าตัดที่มีประสบการณ์และความชำนาญ