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# **Surgical treatment of liver inflammatory pseudotumor-like follicular dendritic cell sarcoma: A case report and review of literatures**

liver inflammatory pseudotumor-like follicular dendritic cell sarcoma

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**Abstract**

BACKGROUND

Inflammatory pseudotumor-like follicular dendritic cell sarcoma (IPT-like FDCS) is rare with a low malignant potential. Hepatic IPT-like FDCS has similar clinical features to hepatocellular carcinoma (HCC), making it extremely difficult to distinguish between them in clinical practice. We describe the case of a young female patient diagnosed with HCC before surgery, which was pathologically diagnosed as IPT-like FDCS after the left half of the liver was resected. During 6 mo of follow-up, the patient recovered well with no signs of recurrence or metastasis.

CASE SUMMARY

A 23-year-old female patient with a 2-year history of hepatitis B presented to the Affiliated Hospital of Guizhou Medical University. She was asymptomatic at presentation, and the findings from routine laboratory examinations were normal except for slightly elevated alpha-fetoprotein levels. However, ultrasonography revealed a 3-cm diameter mass in the left hepatic lobe, and abdominal contrast-enhanced computed tomography revealed that the tumor had asymmetrical enhancement during the arterial phase, which declined during the portal venous phase, and had a pseudo-capsule appearance. Based on the findings from clinical assessments and imaging, the patient was diagnosed with HCC, for which she was hospitalized and had undergone laparoscopic left hepatectomy. However, the tumor specimens submitted for pathological analyses revealed IPT-like FDCS. After surgical removal of the tumor, the patient recovered. In addition, the patient continued to recover well during 6 mo of follow-up.

CONCLUSION

Hepatic IPT-like FDCS is difficult to distinguish from HCC. Hepatectomy may provide beneficial outcomes in non-metastatic hepatic IPT-like FDCS.

**Key Words:** Hepatocellular carcinoma; Liver; Pseudotumor-like follicular dendritic cell sarcoma; Surgery; Tumor; Case report

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**Core Tip:** Inflammatory pseudotumor-like follicular dendritic cell sarcoma (IPT-like FDCS) is a type of FDCS with low malignancy potential. We investigated the clinical and pathological characteristics, diagnosis, and treatment in a 23-year-old woman diagnosed with hepatic IPT-like FDCS. She underwent laparoscopic left hepatectomy, with an uneventful postoperative course. It is difficult to distinguish hepatic IPT-like FDCS from hepatocellular carcinoma based on clinical features. Therefore, most patients with hepatic IPT-like FDCS are found after surgery. However, surgery may be the best treatment option for patients with hepatic IPT-like FDCS. At present, no abnormality has been found on the patient during the six-month follow-up.

## INTRODUCTION

Follicular dendritic cell sarcoma (FDCS) is a rare tumor with nonspecific clinical features. FDCS cannot be diagnosed based on clinical findings alone; therefore, its diagnosis depends on pathological examinations of surgically resected tumor specimens. Pathologically, FDCSs express at least two FDC markers in most cases, including CD21, CD35, and CNA-42. There are two morphologic variants of this tumor: conventional and inflammatory pseudotumor (IPT)-like[1]. IPT-like FDCS cells are irregularly arranged and IPT-like FDCS presents with lymphocytic infiltrate, with a positive *in situ* hybridization test for Epstein-Barr virus (EBV)-encoded RNA[2]. Most IPT-like FDCSs affect the liver and spleen. Herein, we report a rare case of a female patient with liver IPT-like FDCS in the context of hepatitis B virus infection. We

investigated the clinical and pathological characteristics, diagnosis, and treatment of IPT-like FDCS.

## CASE PRESENTATION

### *Chief complaints*

A 23-year-old female patient with an underlying hepatitis B virus infection presented, with no symptoms, at the Affiliated Hospital of Guizhou Medical University.

### *History of present illness*

She was asymptomatic.

### *History of past illness*

She had received entecavir as treatment for her hepatitis B infection 2 years previously.

### *Personal and family history*

The patient had an unremarkable personal and family history.

### *Physical examination*

There were no significant findings on initial physical examination.

### *Laboratory examinations*

Laboratory analyses provided the following findings: hemoglobin, 134 g/L; white blood cell count, 5.23 × $10^9$ cells/L; platelet count, 149 × $10^9$ cells/L; red blood cell count, 4.50 × $10^9$ cells/L; anti-HB test results, positive; serum levels of albumin, 50.40 g/L; aspartate aminotransferase level, 39.50 U/L; alanine aminotransferase level, 23.80 U/L; alkaline phosphatase level, 80 U/L; total bilirubin level, 15.40 μmol/L; direct bilirubin level, 5.20 μmol/L; total bilirubin level, 10.20 μmol/L; hepatitis B viral DNA, 2.35e+01 IU/mL (normal ≤10 UI/mL); and alpha-fetoprotein (AFP) level, 12.31 ng/mL (normal = 0.00–7.00 ng/mL).

*Imaging examinations*

Ultrasonography revealed a 3-cm diameter mass in the left hepatic lobe, necessitating the performance of abdominal contrast-enhanced computed tomography (CT). CT showed asymmetrical tumor enhancement during the arterial phase; however, the enhancement declined during the portal venous phase, and the tumor had a pseudo-capsule appearance.

## FINAL DIAGNOSIS

We initially diagnosed the disease as hepatocellular carcinoma (HCC) based on the findings from clinical, laboratory, and imaging assessments.

## TREATMENT

Needle biopsy was recommended to the patient before surgery, but she refused. The patient underwent laparoscopic left hepatectomy.

## OUTCOME AND FOLLOW-UP

Postoperatively, pathological analysis showed that a large number of medium to small lymphocytes were distributed in the liver space-occupying lesion area, and an unequal number of spindle or epithelial cells, and histiocyte-like cells, were distributed alternately. Further, hepatic lobule structure was found in the surrounding liver tissue, some small lymphocytes had infiltrated the portal area, and a few small cells were found in the hepatic sinuses.

Immunohistochemical staining showed that the tumor specimen was positive for CD2, CD3, CD5, CD7, CD8, and TIA-1 in all lymphocytes. A portion of the tissue specimen was CD4 positive. Furthermore, positive expressions of CD21, CD35, Ki-67 (30%), and SMA were observed in a portion of the specimen with spindle-epithelioid tumor cells. *In situ* hybridization test was only positive for EBV-encoded RNA in spindle-epithelioid tumor cells. Based on these pathological findings, the patient was diagnosed with IPT-

like FDCS. During the 6-month postoperative follow-up period, the patient did not have any signs of recurrence and metastasis.

## DISCUSSION

Although IPT-like FDCS is a special type of FDCS, it exhibits characteristic features comparable to that of conventional FDCS[3]. More than 60 cases of IPT-like FDCS have been reported in English literature, mainly located in the liver[4-11] and spleen [12-15], and to a lesser extent in the colon[16-18], lungs[19], and pancreas[20]. More than 20 cases have been reported in the liver, more than 30 cases in the spleen, 1 case in the pancreas, 6 cases in the colon, and 1 case in the lung. The tumor mainly occurs in middle-aged and elderly people, with a female-to-male ratio of 2.2:1, and a median age of 56.5 years[21]. The etiology and pathogenesis of IPT-like FDCS are not clear. Its occurrence may coincide with EBV infection, because in cases with confirmed IPT-like FDCS, the positive rate of Epstein-Barr encoding region (EBER) through *in situ* hybridization was as high as 92.1%[22]. For the diagnosis of IPT-like FDCS, fine-needle biopsy is a feasible preoperative diagnostic method, but many false negative cases have been encountered due to less amount of obtained puncture tissue. Its definitive diagnosis mainly relies on immunohistochemical and *in situ* hybridization analyses of surgically obtained tumor specimens.

To date, all reported patients with IPT-like FDCS in the liver presented with fever, jaundice, abdominal pain, and/or anemia as the initial clinical manifestations. In addition, other case reports highlighted paraneoplastic arthritis as an initial clinical manifestation[23,24]. However, the patient in the present case did not experience any clinical manifestations prior to the hospital visit, which shows the peculiarity of the disease in clinical practice. In our case, contrast-enhanced CT showed an asymmetrical tumor enhancement and decline during the arterial and portal venous phases, respectively, with a pseudo-capsule appearance of the tumor; these findings corroborated with those of previous studies[25].

Nevertheless, we could not distinguish hepatic inflammatory pseudotumor (HIPT)-like FDCS from HCC based on imaging findings alone. Because the imaging manifestations of IPT-like FDCS of the liver are nonspecific, CT usually shows low-density nodule enhancement, which is characterized by non-uniform enhancement in the arterial phase and resolution in the delayed phase. Some highly differentiated HCCs can also show resolution in the delayed phase; therefore, HIPT-like FDCS should also be differentiated from HCC with internal necrosis *via* imaging findings. The latter often has peripheral structural invasion, accompanied by cirrhosis and portal hypertension, and may have tumor thrombus formation. Generally, it can be differentiated by a history of hepatitis B infection and tumor markers. This patient had a 2-year history of hepatitis B infection as well as a slightly elevated serum AFP level but was asymptomatic. She refused liver puncture before the operation. Finally, after a detailed correlation between clinical and imaging findings, she was preliminarily diagnosed with HCC, which warranted her to undergo laparoscopic left hepatectomy.

Postoperatively, pathological analyses revealed that the tumor was IPT-like FDCS. At the same time, AFP level did not decrease significantly after the operation, which was considered to be related to chronic hepatitis B infection. HCC is the most common malignant tumor of the liver. The patient had chronic viral hepatitis, AFP elevation, and imaging findings suggestive of HCC. These signs are consistent with the general clinical manifestations of HCC. The final diagnosis requires puncture biopsy or pathological analysis after surgical resection.

Apart from HCC, pathological analyses should be performed to distinguish HIPT-like FDCS from other diseases such as hepatic inflammatory pseudotumor (HIPT) and primary liver lymphoma. The symptoms and imaging findings of HIPT are nonspecific, and pathological analyses are required to confirm the diagnosis. The histopathological characteristics of HIPT include the presence of inflammatory lesions consisting of diffused and dense hyalinized collagenosis with inflammatory cells, compact foamy histiocyte proliferation, as well as lymphocyte and plasma cell infiltration[26]. In primary liver lymphoma, clinical features are nonspecific as well, and histological analyses

demonstrate infiltrations limited to the liver. Among the types of lymphoma, diffuse large B-cell lymphoma is the most common type noted[27,28]. HIPT-like FDCS shows a contrasting lymphocyte infiltration pattern compared with that of FDCS, immunohistochemical analyses show positive expression of one of the FDCS markers (CD21, CD35, CD23, or CNA42) in tumor cells, and *in situ* hybridization testing is positive for EBV-encoded RNA in spindle-epithelioid tumor cells. Clinicians can better diagnose HIPT-like FDCS *via* pathological examinations of surgically resected tissue specimens.

HIPT-like FDCS displays combined characteristics of chronic inflammation and malignant tumors on imaging. The final diagnosis is dependent on pathology, which shows that the tumor cells express CD21, CD23, CD35, SMA, and other markers, or do not express these antigens, but are EBER positive. HCC, HIPT, and primary liver lymphoma should be considered in the differential diagnosis. In the treatment of HIPT-like FDCS, complete resection of tumor is the best treatment. Chemotherapy and/or radiotherapy can be used for patients with recurrence or surgery that cannot be cured[29-31]. In terms of prognosis, the recurrence and metastasis rate (15.8%) and mortality rate (3.5%) of previously reported liver cases were very low. In the present case, HIPT-like FDCS was found to be an indolent malignant tumor with no sign of relapse or metastasis noted during the follow-up.

## CONCLUSION

HIPT-like FDCS is extremely difficult to distinguish from HCC due to their similar clinical features. In addition, surgical resection may provide better long-term outcomes in patients with indolent malignant HIPT-like FDCSs.

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