Original Article

# Imaging features of inflammatory pseudotumor-like follicular dendritic cell sarcoma of the spleen

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**Background:** Inflammatory pseudotumor (IPT)-like follicular dendritic cell sarcoma (FDCS) is an extremely rare malignant neoplasm.

**Methods:** Retrospective analysis of imaging features of splenic IPT-like FDCS, including ultrasonography, computed tomography (CT), and magnetic resonance (MR) and contrast-enhanced imaging were performed.

**Results:** When the masses were small, the ultrasound images showed homogeneous hypoechoic signals, clear boundaries, and complete capsules. Abdominal plain CT scans showed equal density (easy to miss diagnosis), unclear boundaries, and no capsules. Magnetic resonance images (MRI) showed slightly shorter T1, slightly shorter T2, and clear boundaries. When the masses were large, the ultrasound images still showed clear boundaries and complete capsules, but the echoes of the masses were not uniform, and some of the masses showed dendritic hyperechoic centers. Abdominal plain CT scans showed irregular low densities in the center (unclear boundaries) and equal densities in the periphery. MRI showed short T1 and T2, but the central signals were mixed. When the mass was accompanied by extensive necrosis, abdominal plain CT scan showed mostly cystic lesions and slight calcifications in low density lesions. Contrast-enhanced CT showed only moderate enhancement in peripheral and septal areas. MRI showed that T1 and T2 were mainly mixed signals. Contrast-enhanced MR showed moderate enhancement of peripheral areas and septum.

**Conclusions:** This is the first report to describe the IPT manifestations of the spleen (ultrasonography, CT, and MR). The diagnosis of IPT can be made by combining three imaging features.

**Keywords:** Imaging features; inflammatory pseudotumor (IPT); follicular dendritic cell sarcoma (FDCS); spleen; sarcoma

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## Introduction

Inflammatory pseudotumor (IPT)-like follicular dendritic cell sarcoma (FDCS) is an extremely rare malignant neoplasm, which almost exclusively arises from the liver or spleen (1). The pathogenesis and causes are still unknown, but EBV is designated as one of the most important etiologies of this tumor (2). Despite its slow-growing and low-grade malignancy, the recurrence rate is about 10% (3). Due to morphological similarities, this tumor is frequently confused with other inflammatory myofibroblastic proliferations, such as benign reactive IPT and inflammatory myofibroblastic tumor (4-6). The diagnosis of this disease requires auxiliary tests, including

immunohistochemical expression of FDC markers (such as CD21, CD23, or CD35), and *in situ* hybridization probe labeling EBV-encoded early nuclear RNAs (EBER) (1). Preoperative diagnosis of IPT-like FDCS remains difficult due to its unique immunophenotype and typical histopathological features, because it is extremely rare, and lacks specific imaging features. Previously, its pathological characteristics have been reported (1,3,5,6); however, imaging findings are lacking. Previous reports of spleen imaging findings of IPT-like FDCS were limited to case reports, and most of the imaging data were incomplete (only a single CT or MR imaging feature was described), ultrasound imaging features were lacking, and there were no combined imaging features of the three (7). This study summarizes the imaging features of splenic IPT-like FDCS, including ultrasound, CT, MR, and contrast-enhanced imaging. Moreover, we found that different sizes of tumors have different imaging performance. According to our search of literature, we found that this is the first time that the relationship between tumor size and imaging performance has been described. We present the following article in accordance with the STROBE reporting checklist (available at https://dx.doi.org/10.21037/apm-21-2776).

## Methods

### *Patients*

From 2010 to 2018, a total of 7 patients diagnosed with splenic IPT-like FDCS including 2 men and 5 women with a mean age of 59.6 years (range, 40–81 years), respectively, from Ningbo First Hospital, Ningbo Li Huili Hospital, Ningbo Second Hospital, Affiliated Hospital of Medical College of Ningbo University, and Xiangshan People's Hospital were enrolled in this retrospective cohort study. The diagnoses for all were confirmed using pathology and immunohistochemistry. IPT-like FDCS pathological diagnostic criteria: It is composed of oval to spindle-shaped tumor cells, scattered in a mesh pattern against the background of obvious lymphoplasma cell infiltration, and the cells have variable nuclear atypia; at the same time, immunohistochemistry is required to assist in diagnosis (Expression of EBER, CD21, CD23, and CD35). Retrospective analysis of imaging features of splenic IPT-like FDCS included ultrasonography, computed tomography (CT), MR, and contrast-enhanced imaging. All procedures performed in this study involving human participants were in accordance with the Declaration of Helsinki (as revised in 2013). The study was approved by the ethics committee of Ningbo First Hospital (No.: 2021RS057). Individual consent for this retrospective analysis was waived.

### *Imaging analysis*

Ultrasound, CT, and magnetic resonance images (MRI) were retrospectively evaluated by two experienced radiologists who reached consensus. These two reviewers were unaware of the definitive diagnosis and other imaging information at the time of the analysis. The features of ultrasound images consist of size, number, morphology, boundary, echo, Doppler signal, and enhancement patterns of the lesion (SonoVue: Bracco SpA, Milan, Italy). CT images features include size, number, morphology, boundary, density, and enhancement patterns of the lesion (Iohexol, GE Healthcare Co., Ltd., Shanghai, China). MRI describe the following characteristics: size, number, morphology, boundary, T1 and T2 signals, internal enhancement (if intravenous contrast agent is injected), and fluid level.

### *Statistical analysis*

All data were analyzed using SPSS13.0 statistical software (Chicago, IL, United States). The numerical data were expressed as the mean ± SD. A P value of less than 0.05 was considered statistically significant.

## Results

### *Clinical data*

Among 7 patients (2 males and 5 females; age range of 40 to 81years; mean age of 59.6 years) with splenic IPT-like FDCS, 4 patients (57.1%) had no clinical symptoms; 1 of them presented with soreness of the abdomen, and 2 presented with dull pain in the upper left abdomen. Laboratory tests showed that hemoglobin decreased in 4 patients (hemoglobin range 87–114 g/L, average 97.3 g/L), decrease of leukocyte ($2.1\times10^9$ L) and neutrophil ($1.1\times10^9$ L) in 1 patient, increase of neutrophil percentage in 2 patients (71.6%, 78.3%) and slight elevation of carbohydrate antigen 125 in 1 patient (36.6 U/mL). No significant abnormalities were found in other laboratory examinations (*Table 1*).

 

**Table 1** Clinical and imaging features of inflammatory pseudotumor-like follicular dendritic cell sarcoma

| Features | 1 | 2 | 3 | 4 | 5 | 6 | 7 |
|---|---|---|---|---|---|---|---|
| Gender | Female | Female | Male | Female | Female | Female | Male |
| Age (year) | 45 | 40 | 81 | 59 | 54 | 71 | 67 |
| Symptom | Soreness of the waist | No clinical symptoms | No clinical symptoms | Dull pain in the left upper abdomen | Dull pain in the left upper abdomen | No clinical symptoms | No clinical symptoms |
| Laboratory tests | | | | | | | |
| Hemoglobin (g/L) | 89 | 114 | Normal | 99 | 87 | Normal | Normal |
| Carbohydrate antigen 125 (U/mL) | Normal | Normal | 36.6 | Normal | Normal | Normal | Normal |
| Number of lesions | 3 | 1 | 1 | 1 | 1 | 1 | 1 |
| Maximum diameter of lesion (cm) | 2.9, 1.7, 1.5 | 7.3 | 8.1 | 15 | 3.6 | 4.5 | 6 |
| Lesion morphology | Round | Round | Round | Round | Round | Round | Round |
| Ultrasound features | | | | Unexamined | | | |
| Boundary | Well-defined | Well-defined | Well-defined | | Well-defined | Well-defined | Well-defined |
| Echoes | Homogeneous hypoechoic | Peripheral hypoechoic and dendritic hyperechoic centers | Peripheral hypoechoic and dendritic hyperechoic centers. | | Homogeneous hypoechoic | Hypoechoic peripheral and irregular hyperechoic central echoes | Hypoechoic peripheral and irregular hyperechoic central echoes |
| Color doppler flow imaging | No blood flow signal | A little blood flow signal | A little blood flow signal | | No blood flow signal | A little blood flow signal | A little blood flow signal |
| Contrast-enhanced ultrasonography | Short enhancement and then the tumor mass returned to hypoechoic in the arterial phase | Unexamined | Unexamined | Unexamined | Unexamined | Unexamined | Unexamined |
| Computed tomography | | | | | | Unexamined | |
| Boundary | Unclear | Unclear | Unclear | Well-defined | Unclear | | Unclear |
| Density | Isodensity, no mass was found | Irregular low density in the center and equal density in the periphery | Irregular low density in the center and equal density in the periphery | Most cystic lesions and slight calcification in low density lesions | Isodensity, no mass was found | | Irregular low density in the center and equal density in the periphery |

**Table 1** (*continued*)

**Table 1** (*continued*)

| Features | 1 | 2 | 3 | 4 | 5 | 6 | 7 |
|---|---|---|---|---|---|---|---|
| Contrast-enhanced computed tomography | Moderate homogeneous enhancement | Unexamined | Unexamined | Only moderate enhancement in peripheral and septal areas | Moderate homogeneous enhancement | | Uneven moderate enhancement, (low enhancement area in the center) |
| Magnetic resonance | | | Unexamined | | Unexamined | | Unexamined |
| Boundary | Well-defined | Well-defined | | Well-defined | | Well-defined | |
| T1 | Short | Short (mixed central signals) | | Short (mixed central signals) | | Short | |
| T2 | Short | Short (mixed central signals) | | Short (mixed central signals) | | Short | |
| Contrast-enhanced magnetic resonance | No enhancement | Progressive enhancement (lower than that of the spleen) with no enhancement in the center | | Only moderate enhancement in peripheral and septal areas | | Progressive enhancement (lower than that of the spleen) with no enhancement in the center | |

### *Ultrasound features*

Ultrasonography was performed in 6 out of 7 patients. Ultrasound showed that there were 8 lesions in 6 patients, including 3 lesions in 1 patient and 5 single lesions in 5 patients. The maximum diameter of the lesion ranged from 1.5 to 8.1 cm (mean 4.2 cm). All 8 lesions were round and well-defined. Four lesions (2 patients, 1 patient with 3 lesions) were homogeneous hypoechoic (lesion diameters were 1.5, 1.7, 2.9 cm, and 3.6 cm, respectively), and color Doppler flow imaging showed no blood flow signal (*Figure 1*). The echoes of the 4 lesions were heterogeneous (the diameters of the lesions were 4.5, 6, 7.3, and 8.1 cm, respectively), and color Doppler flow imaging showed few blood flow signals. All 4 cases showed hypoechoic peripheral and irregular hyperechoic central echoes, and 2 showed dendritic hyperechoic centers (*Figure 2*). Contrast-enhanced ultrasonography (CEUS) was performed in only 1 patient, who had 3 lesions. Contrast-enhanced ultrasonography of the spleen showed a short enhancement in the hypoechoic area on the non-contrast-enhanced ultrasonography, and then the tumor mass returned to being hypoechoic in the arterial phase (*Figure 1; Table 1*).

### *CT features*

CT examination was performed in 6 out of 7 cases. Ultrasound showed 2 cases of homogeneous hypoechoic lesions (4 lesions), and no lesions were found on a plain CT scan (*Figure 1*). After enhanced CT, the lesions showed moderate homogeneous enhancement (2 larger lesions were slightly lower than that of the spleen, and 2 smaller lesions were equivalent to that of the spleen) (*Figure 1*). There were 4 patients with irregular hyperecho in the peripheral hypoechoic center, where 3 patients received plain CT scans, and 1 patient received an enhanced scan. Plain CT scans showed irregular low densities in the center (unclear boundaries) and homogeneous densities at the periphery (*Figure 2*). One case showed uneven moderate enhancement after an enhanced scan, and a low enhancement area in the center. One case had necrosis (the diameter of the lesion was 15 cm), and ultrasonography was not performed. Plain CT scans showed that cystic lesions were more common, and there was slight calcification in low density lesions (*Figure 3*). Contrast-enhanced CT scan showed only moderate enhancement in peripheral and septal areas, most of which were non-enhanced areas (*Figure 3; Table 1*).

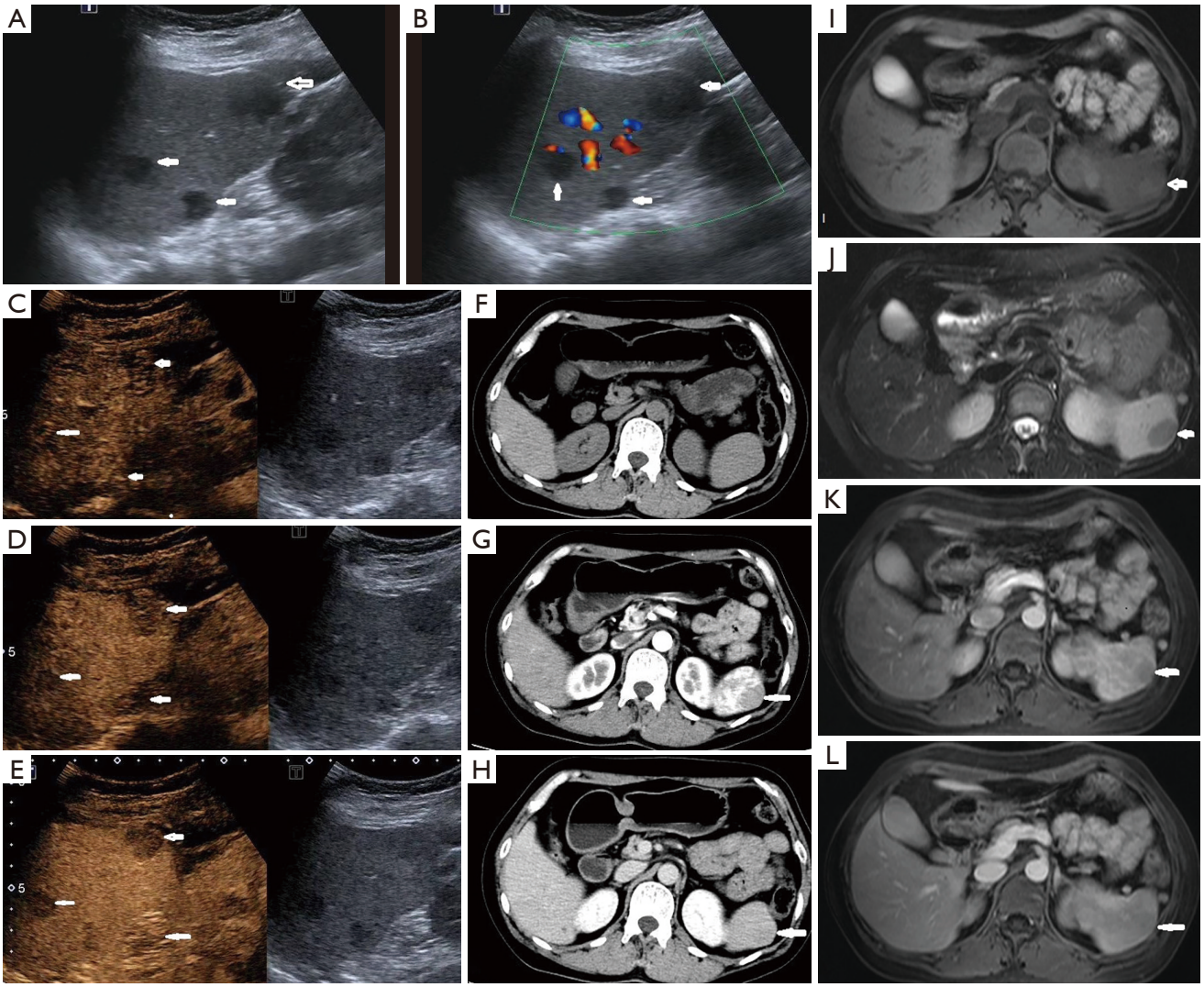


**Figure 1** Inflammatory pseudotumor (IPT)-like follicular dendritic cell sarcoma (FDCS) imaging features of spleen with small mass. (A,B) In this case, 3 lesions (1.5, 1.7, and 2.9 cm in diameter, respectively) with homogeneous hypoechoic were found using two-dimensional ultrasound (A, arrows), and there was no blood flow signal in color Doppler flow imaging (B, arrows). (C-E) Contrast-enhanced ultrasonography showed isoechoicity shortly after contrast administration (15 s) (C, arrows), which returned to hypoenhancement in the arterial phase (26 s) (D, arrows). The tumor mass is still hypoechoic on the delayed-phase ultrasound images (E, arrows). (F) Abdominal plain computed tomography (CT) scan did not find any obvious abnormalities in the spleen. (G,H) Contrast-enhanced CT showed a well-demarcated, hypointense lesion in the lower pole of the spleen (arrows). (I,J) Three well-defined hypointense lesions were detected on both T1- and T2-weighted magnetic resonance imaging (MRI) (arrows). (K,L) Contrast-enhanced T1W MRI showed the lesion without obvious enhancement in contrast to the significantly enhanced spleen parenchyma (arrows).

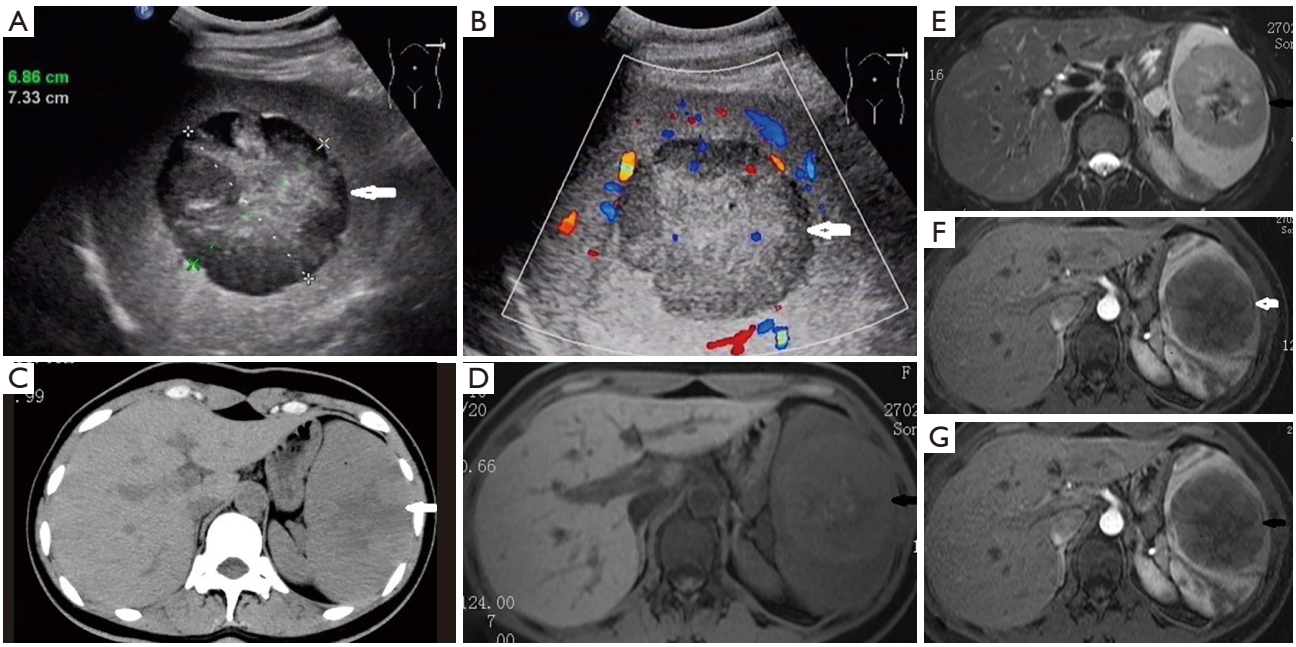


**Figure 2** Inflammatory pseudotumor (IPT)-like follicular dendritic cell sarcoma (FDCS) imaging features of spleen with large mass. (A,B) The echo of this lesion (7.3 cm in diameter) was heterogeneous (dendritic hyperechoic center) (A, arrow). Color Doppler flow imaging showed few blood flow signals (B, arrow). (C) Abdominal plain computed tomography (CT) scan showed irregular low densities in the center (unclear boundary) and homogeneous densities in the periphery (arrow). (D,E) The lesion showed short T1 (mixed central signals) and T2 (mixed central signals) on magnetic resonance imaging (MRI) (arrow). (F,G) Contrast-enhanced MRI showed progressive enhancement (lower than that of the spleen) with no enhancement in the center (arrows).

### *MRI features*

MRI examination was performed in 4 out of 7 cases. It revealed 6 lesions in 4 patients, including 1 case with 3 lesions and 3 cases with single lesions. All 6 lesions showed short T1 and T2 on MRI (*Figure 1*). Three single lesions showed mixed central signals on MRI (*Figure 2*) (One case was accompanied by necrosis, mainly with mixed signals, *Figure 3*). One patient with three lesions showed no enhancement (*Figure 1*). In one case, regarding the lesion with necrosis, enhanced MR showed moderate enhancement of peripheral areas and septum, but no enhancement in most of the central areas (*Figure 3*). The other 2 cases showed progressive enhancement (lower than that of the spleen) with no enhancement in the center (*Figure 2; Table 1*).

## Discussion

Because of the lack of relevant imaging reports, preoperative imaging diagnosis remains difficult. The diagnosis mainly depends on immunohistochemistry. In imaging, splenic fibrohistiosarcoma and hemangioendothelioma are often manifested as malignant tumors with unclear borders and irregular shapes. Because IPT-like FDCS is a low-grade malignant tumor with low invasiveness, it usually appears as a benign tumor with clear borders and regular morphology. All cases in this study showed clear and round borders on ultrasound and MR. As with any other sarcomas, a resection of the tumor with a wide margin is generally recommended where feasible (8). If the tumor recurs or cannot be treated by surgery, chemotherapy or radiotherapy can be given. Therefore, patients can get timely and correct treatment using accurate preoperative imaging diagnosis.

The clinical data of this study showed that the disease was common in women around 60 years old, and most of them had no symptoms or mild symptoms. Except for 57.1% (4/7) of patients with hemoglobin decline, no significant characteristics were found in other laboratory tests. Clinical manifestations and laboratory examinations showed that the disease was easy to miss and misdiagnose. Therefore, it is of great importance for clinicians to

 

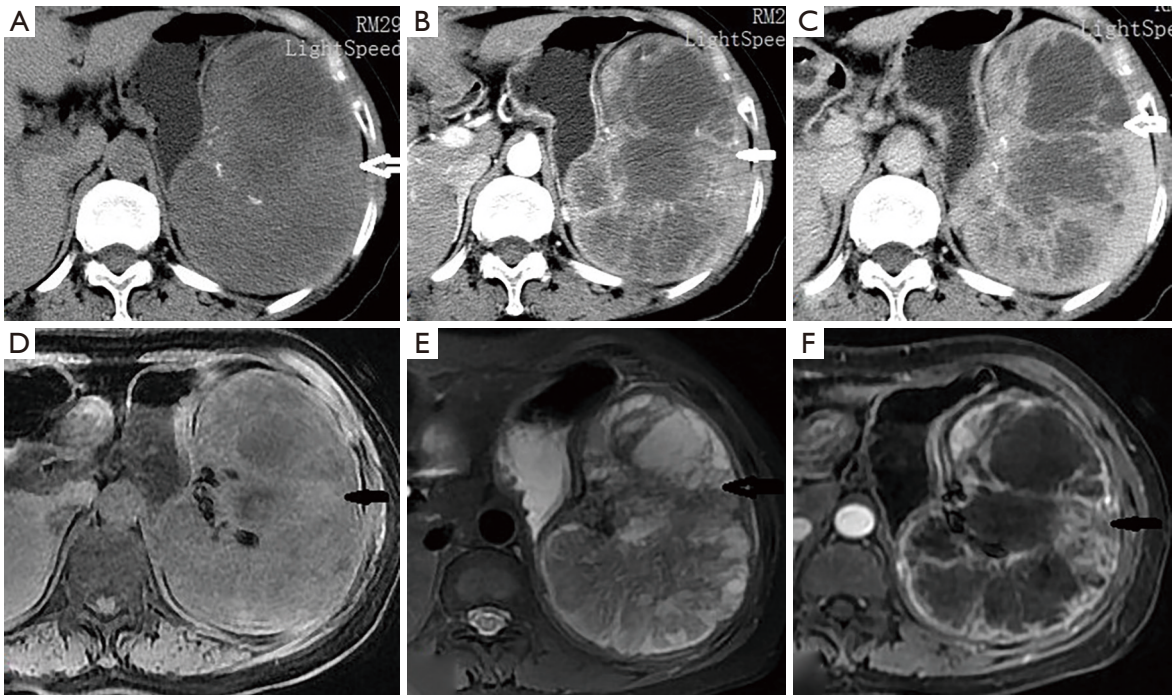


**Figure 3** IPT-like FDCS imaging features of spleen with extensive necrosis. (A) This case was accompanied by necrosis (the diameter of the lesion was 15 cm), the plain computed tomography (CT) scan showed that cystic lesions were more common, and there was slight calcification in low density lesions (arrow). (B,C) Contrast-enhanced CT scan showed only moderate enhancement in peripheral and septal areas, most of which were non-enhanced areas (arrows). (D,E) The lesion showed short T1 (mainly with mixed signals) and T2 (mainly with mixed signals) on magnetic resonance imaging (MRI) (arrows). (F) Contrast-enhanced MRI showed moderate enhancement of peripheral and septum, but no enhancement in most of the central areas (arrow).

understand accurate imaging examinations.

We retrospectively analyzed the imaging features of splenic IPT-like FDCS, which will be helpful for the future diagnosis of splenic IPT-like FDCS. All tumors were round. When the masses were small (1.5–3.6 cm), the ultrasound images showed homogeneous hypoechoic signals, clear boundaries, complete capsules, and no blood flow signals. Contrast-enhanced ultrasound images showed a short enhancement, and then the tumor mass returned to hypoechoic in the arterial phase. Abdominal plain CT scans showed equal density (easy to miss diagnosis), unclear boundaries, and no capsules. Contrast-enhanced CT images showed moderate homogeneous enhancement, which were slightly lower than those of the spleen. MRI showed slightly shorter T1, slightly shorter T2, and clear boundaries. Contrast-enhanced MRI showed lesions without obvious enhancement. Ultrasound and MRI of these tumors are easily misdiagnosed as splenic lymphoma, but lymphoma usually has unclear boundaries and no capsules (9). Because the CT images of these tumors showed equal density and moderate homogeneous enhancement, this easily leads to missed diagnoses. In addition, splenic lymphoma usually showed low density on CT images. Therefore, ultrasound or MR combined with CT imaging can diagnose the disease.

When the masses were large (4.5–8.1 cm), the ultrasound images still showed clear boundaries and complete capsules, but the echoes of the masses were not uniform (hypoechoic peripheral and irregular hyperechoic central echoes), and dendritic hyperechoic centers were seen in some of the masses (pathological results showed fibrous scars). Color Doppler showed few blood flow signal inside the mass. Abdominal plain CT scan showed irregular low densities in the center (unclear boundaries) and uniform densities at the periphery. Contrast-enhanced CT images showed uneven moderate enhancement with low enhancement areas in the center. MRI showed short T1 and T2, but the central signals were mixed. This type of mass was easily diagnosed using CT as primary malignant fibrous histiocytoma of

 

the spleen. However, MR showed equal or long T1, short T2, and long T1 and T2 in the center. Ultrasound showed uneven echo, no blood flow signals and no echoes in the interior. Unlike malignant fibrous histiocytoma of the spleen, necrosis occurs in the center, and fibrosis (partly dendritic) occurs in the center of IPT-like FDCS.

When the mass was accompanied by extensive necrosis (15 cm), abdominal plain CT scan showed that most cystic lesions had slight calcifications in low density lesions. Contrast-enhanced CT showed only moderate enhancement in peripheral and septal areas, most of which were non-enhanced areas. MRI showed that T1 and T2 were mainly mixed signals. Contrast-enhanced MR showed moderate enhancement of peripheral and septum, but no enhancement in most of the central areas. When the mass has extensive necrosis, it is difficult to differentiate it from hemangioma and angiosarcoma with extensive necrosis.

This study has several limitations. First of all, the case samples were limited, and large samples were needed for further research and confirmation. Secondly, some patients in this study did not undergo the three examinations of ultrasound, CT, and MR at the same time. Again, Similarly, this study did not investigate its image formation mechanism.

In summary, this is the first report to describe the IPT manifestations of the spleen (using ultrasonography, CT, and MR). The diagnosis of IPT can be made by combining three imaging features.

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## Footnote

*Reporting Checklist:* The authors have completed the STROBE reporting checklist. Available at https://dx.doi.org/10.21037/apm-21-2776

*Data Sharing Statement:* Available at https://dx.doi.org/10.21037/apm-21-2776

*Conflicts of Interest:* All authors have completed the ICMJE uniform disclosure form (available at https://dx.doi.org/10.21037/apm-21-2776). The authors have no conflicts of interest to declare.

*Ethical Statement:* The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study involving human participants were in accordance with the Declaration of Helsinki (as revised in 2013). The study was approved by the ethics committee of Ningbo First Hospital (No.: 2021RS057). Individual consent for this retrospective analysis was waived.



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