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**Inflammatory** **pseudotumor-like** **follicular dendritic cell** **sarcoma: A** **brief report of two cases**

Zhang BX *et al*. Inflammatory pseudotumor-like follicular dendritic cell sarcoma

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**Abstract**

***Background***

Follicular dendritic cell (FDC) sarcoma/tumor is a rare malignant tumor of follicular dendritic cells, which is considered a low-grade sarcoma that can involve lymph nodes or extranodal sites. Conventional FDC sarcomas are negative for Epstein-Barr virus (EBV), whereas the inflammatory pseudotumor-like variant consistently shows EBV in the neoplastic cells.

***CASE SUMMARY***

We report two cases of inflammatory pseudotumor-like FDC sarcoma in the liver that received 3D laparoscopic right hepatectomy and open right hepatectomy separately.

***CONCLUSION***

EBV probe-based *in situ* hybridization and detection of immunohistochemical markers of FDC play an important role in the diagnosis and differential diagnosis of inflammatory pseudotumor-like FDC sarcoma. Complete surgical excision combined with regional lymphadenectomy may be effective in reducing the postoperative recurrence and metastasis and improving long-term survival rates.

**Key words:** Inflammatory pseudotumor-like follicular dendritic cell sarcoma; Epstein-Barr virus; Liver; Spleen; Case report

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**Core tip:** There have been 48 previously reported cases of inflammatory pseudotumor-like follicular dendritic cell (FDC) sarcoma, which occurs almost exclusively in the liver and spleen. Here we report two cases of inflammatory pseudotumor-like FDC sarcoma in the liver that were treated by 3D laparoscopic right hepatectomy and open right hepatectomy separately.

Zhang BX, Chen ZH, Liu Y, Zeng YJ, Li YC. Inflammatory pseudotumor-like follicular dendritic cell sarcoma: A brief report of two cases. *World J Gastrointest Oncol* 2019; In press

**Introduction**

Follicular dendritic cell (FDC) sarcoma/tumor is a rare malignant tumor of follicular dendritic cells, which are mesenchymal cells in the lymphoid follicles with antigen presenting ability. It is considered a low-grade sarcoma that can involve lymph nodes or extranodal sites[1-5]. In 1996, Shek *et al*[6] reported the first case of primary FDC sarcoma in the liver. The histology was similar to an inflammatory pseudotumor and it was related to Epstein-Barr virus (EBV)-related clonal proliferation[6]. Inflammatory pseudotumor-like FDC sarcoma was first described as a distinctive variant of FDC sarcoma and associated with EBV in 2001[7]. There have been 48 previously reported cases of inflammatory pseudotumor-like FDC sarcoma, which occurs almost exclusively in the liver and spleen (Table 1). Ancillary tests, including detection of immunohistochemical markers of FDC such as CD21, CD23, or CD35 and EBV probe-based *in situ* hybridization, are required for this diagnosis. Here we report two cases of inflammatory pseudotumor-like FDC sarcoma in the liver that were treated by 3D laparoscopic right hepatectomy and open right hepatectomy separately.

**CASE PRESENTATION**

***Chief complaints***

**Case 1:** A 31-year-old woman was admitted to hospital for evaluation of a four-week history of anorexia.

**Case 2:** A 48-year-old man stumbled across a liver mass through a routine ultrasound examination.

***History of present illness***

Unremarkable.

***History of past illness***

**Case 1:** Her past medical history was chronic hepatitis B for more than 10 years without antiviral treatment.

**Case 2:** Unremarkable.

***Personal and family history***

Unremarkable.

***Physical examination upon admission***

**Case 1:** Physical examination revealed mild tenderness to palpation in the right upper quadrant.

**Case 2:** Physical examination was unremarkable.

***Laboratory examinations***

**Case 1:** Laboratory tests showed seropositivity for HBsAg, HBeAb, and HBcAb. Furthermore, serum level of hepatitis B virus-DNA was lower than detection limit.

**Case 2:** Laboratory tests were unremarkable.

***Imaging examinations***

**Case 1:** Abdominal magnetic resonance imaging revealed two well-circumscribed masses in the right posterior lobe of the liver (Figure 1).

**Case 2:** An abdominal computed tomography examination revealed an ill-defined 10 cm mass in the right lobe of the liver accompanied with enlargement of hepatic portal lymph nodes (Figure 3).

**FINAL DIAGNOSIS**

***Case 1***

EBV-positive inflammatory pseudotumor-like FDC sarcoma in the liver (Figure 2A-G).

***Case 2***

EBV-positive inflammatory pseudotumor-like FDC sarcoma in the liver with hepatoduodenal ligament lymph node involvement (Figure 4A-H).

**TREATMENT**

***Case 1***

3D laparoscopic right hepatectomy.

***Case 2***

Open right hepatectomy combined with regional lymphadenectomy

**OUTCOME AND FOLLOW-UP**

***Case 1***

Follow-up for 10 mo showed no recurrence or metastasis.

***Case 2***

Follow-up for 2 mo showed no recurrence or metastasis.

**DISCUSSION**

FDC sarcoma is a neoplastic proliferation of spindled to ovoid cells exhibiting morphological and immunophenotypic features of FDCs. Histologically, FDC sarcomas are classified into two types: (A) Conventional FDC sarcoma consisting of spindled to ovoid cells forming fascicles, storiform arrays, whorls, diffuse sheets, or vague nodules with an array of small lymphocytes; and (B) inflammatory pseudotumor-like FDC sarcoma composed of neoplastic spindled cells that are dispersed within a prominent lymphoplasmacytic infiltrate**[3]**. To date, 48 cases of inflammatory pseudotumor-like FDC sarcoma have been reported in the English-language literature, located in the liver (16/48), spleen (32/48), colon (1/48), and peri-pancreas (1/48), respectively. These cases included 19 males and 29 females (male/female ratio of 1: 1.5), with a mean age of 55 years (range, 19-79 years). Clinical manifestations include abdominal pain, abdominal bloating, abdominal mass, weight loss, fever, fatigue, and anorexia, but most cases are asymptomatic (Table 1).

The origin of FDC sarcoma remains controversial. Phenotypic marker studies and *in vitro* experiments with fibroblast-like cell lines have developed FDCs from fibroblast-like cells[8]. The neoplastic cells are often positive for FDC markers, such as CD21, CD23, and CD35, with the staining ranging from extensive to very local. FDCs appear to be closely related to bone marrow stromal progenitors, with several myofibroblast features[9]. Two studies examining the transcriptional profile of FDC sarcoma have revealed: (A) A peculiar immunological microenvironment enriched in follicular helper T cells and Treg populations, with special relevance to the inhibitory immune receptor programmed cell death protein 1 and its ligands, programmed cell death-Ligand 1 and programmed cell death-Ligand 2; and (B) the highly specific expression of the genes encoding for FDC secreted peptide and serglycin[10-11].

Conventional FDC sarcomas are negative for EBV, whereas the inflammatory pseudotumor-like variant consistently shows EBV in the neoplastic cells[7]. EBV-encoded small RNA was detected in both of the present cases by *in situ* hybridization. EBV-encoded latent membrane protein 1, which has been found to have an oncogenic role, has been identified in 74% (26/35) cases of inflammatory pseudotumor-like FDC sarcomas by immunohistochemical staining[7,17,19-21,25]. Recently, Takeuchi *et al*[12] reported increased numbers of EBV-infected cells in IgG4-related lymphadenopathy, compared with other reactive lymphadenopathy or extranodal IgG4-related disease, which suggests that there may be a relationship between IgG4-related disease and EBV[12]. Interestingly, Choe *et al*[17] reported that significant numbers of IgG4-positive plasma cells were found in six cases of EBV-positive inflammatory pseudotumor-like FDC sarcoma of the spleen, suggesting that EBV plays a critical role in inflammatory pseudotumor-like FDC sarcoma and IgG4-related sclerosing disease[17]. Generally, the pathogenic mechanism of EBV in inflammatory pseudotumor-like FDC sarcoma remains unclear and further investigation is required.

FDC sarcoma is usually treated by complete surgical excision, with or without adjuvant radiotherapy or chemotherapy. A pooled analysis of the literature revealed local recurrence and distant metastasis rates of 28% and 27%, respectively. Large tumor size (≥6 cm), coagulative necrosis, high mitotic count (≥5 mitoses per 10 high-power fields), and significant cytological atypia are associated with a worse prognosis[2,5]. Regarding the prognosis of patients with inflammatory pseudotumor-like FDC sarcoma, based on the literature reports of inflammatory pseudotumor-like FDC sarcoma with a median follow-up period of 17 mo, 35 patients had no evidence of disease. Five patients exhibited distant metastasis and two had local recurrence, with traits similar to large tumors and multiple masses. One of the current cases presented with liver and hepatoduodenal ligament lymph node involvement, suggesting that inflammatory pseudotumor-like FDC sarcoma presents an increased risk of lymph node metastasis. Complete surgical excision combined with regional lymphadenectomy may be effective in reducing the postoperative recurrence and metastasis and improving the long-term survival rates.

**CONCLUSION**

In conclusion, there is little specificity in the clinical manifestations of inflammatory pseudotumor-like FDC sarcoma. EBV probe-based *in situ* hybridization and detection of immunohistochemical markers of FDC play important roles in the diagnosis and differential diagnosis of inflammatory pseudotumor-like FDC sarcoma. Radical surgical resection is the main therapeutic intervention for inflammatory pseudotumor-like FDC sarcoma, especially for cases with lymph node involvement, and patients require long-term post-surgical follow-up.

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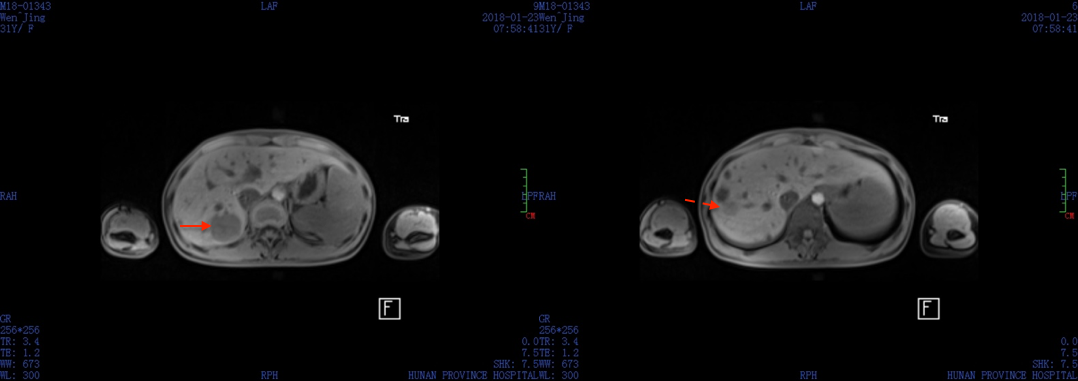
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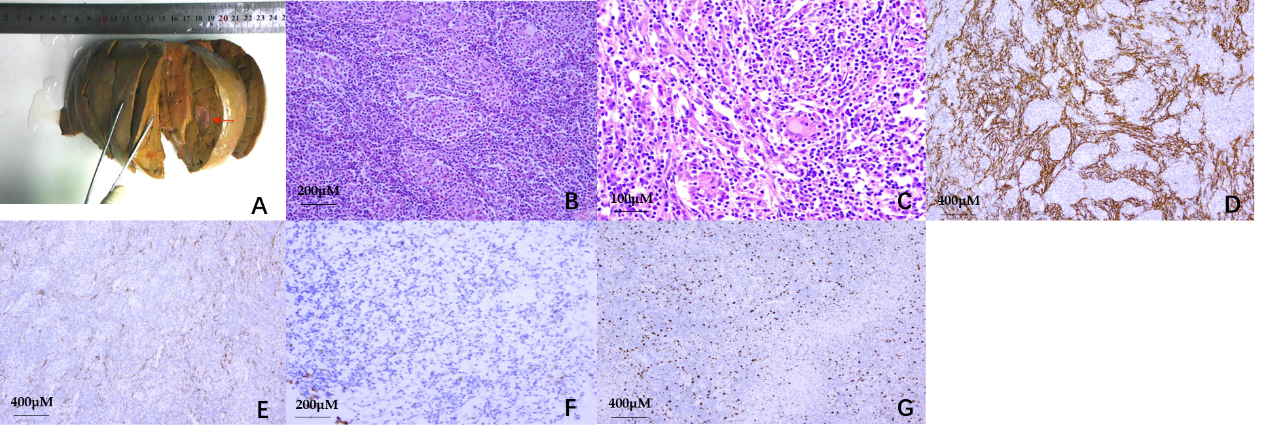
**Table 1 Review of inflammatory pseudotumor-like follicular dendritic cell tumor/sarcoma**

|  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- |
| Ref. | Sex/age | Location | Maximum diameter (cm) | Symptom | Treatment | Follow-up  (mo) | Outcome |
| Li[13] | F/64 | Spleen | 7.2 | Upper abdominal pain | Laparoscopic splenectomy | 8 | NED |
|  | M/61 | Spleen | 6.2 | Asymptomatic | Laparoscopic splenectomy | 16 | NED |
|  | F/42 | Spleen | 4 | Left-sided flank pain | Laparoscopic splenectomy | 9 | NED |
|  | F/57 | Spleen | 13.3 | Upper abdominal pain | Laparoscopic splenectomy | 4 | LWD, pulmonary metastasis |
|  | M/52 | Spleen | 2 masses: 3.7, 2.9 | Back pain | Laparoscopic splenectomy | 5 | LWD, bone metastasis |
| Hang[14] | M/57 | Spleen | 2.7 | Asymptomatic | Laparoscopic partial splenectomy | 9 | NED |
| Ge[15] | F/54 | Spleen | 3.5 | Left-sided flank pain | Splenectomy | 10 | NED |
|  | M/79 | Spleen | 6 | Asymptomatic | Splenectomy | 18 | NED |
| Pan[16] | F/78 | Colon | 3.9 | Abdominal discomfort, bloody stool | Polypectomy | 5 | NED |
| Choe[17] | F/64 | Spleen | 5.5 | Asymptomatic | Splenectomy | 78 | NED |
|  | F/72 | Spleen | 7.2 | Asymptomatic | Splenectomy | 18 | NED |
|  | F/53 | Spleen | 3.2 | Asymptomatic | Splenectomy | 13 | NED |
|  | M/76 | Spleen | 3.2 | Asymptomatic | Splenectomy | 8 | NED |
|  | M/72 | Spleen | 6 | Asymptomatic | Splenectomy | 18 | NED |
|  | M/75 | Spleen | 3.5 | Abdominal pain | Splenectomy | 30 | NED |
| Granados[18] | F/57 | Liver | 13 | Abdominal pain, vomiting | Partial hepatectomy | 24 | NED |
| Cheuk[7] | F/19 | Liver | 12 | Right upper quadrant pain, abdominal mass, weight loss | Partial hepatectomy | 40 | NED |
|  | F/56 | Liver | 15 | Abdominal discomfort | Partial hepatectomy | 56 | LWD, recurrence in liver |
|  | F/40 | Liver | 12.5 | Upper abdominal pain, weight loss | Partial hepatectomy | 108 | LWD, intraabdominal recurrence |
|  | F/49 | Liver | 4.2 | Asymptomatic | Partial hepatectomy | 9 | NED |
|  | M/37 | Liver | 15 | Abdominal mass, weight loss | Partial hepatectomy | 42 | NED |
|  | F/35 | Liver | 20 | Abdominal discomfort, fever, weight loss | Partial hepatectomy | 95 | DOD, disseminated in liver and peritoneum |
|  | F/31 | Liver | 15 | Abdominal distension, weight loss | Partial hepatectomy | 60 | NED |
|  | F/58 | Spleen | 22 | Abdominal mass | Splenectomy | 4 | NED |
|  | F/39 | Spleen | 7.5 | Weight loss, fever | Splenectomy | 2 | LWD, persistent fever |
|  | F/61 | Spleen | 3.5 | Asymptomatic | Splenectomy | NA | NA |
|  | F/49 | Peri-pancreas | 15 | Abdominal distension | Whipple’s operation | NA | NA |
| Li[19] | F/49 | Spleen | 4.7 | Asymptomatic | Splenectomy | NA | NA |
|  | F/56 | Spleen | 8 | Abdominal pain | Splenectomy | 17 | NED |
|  | M/38 | Liver | 8.5 | Anorexia | Partial hepatectomy | 11 | NED |
|  | F/42 | Liver | 2 masses: 2, 1.7 | Abdominal pain | Partial hepatectomy | 36 | NED |
|  | M/50 | Spleen and liver | Spleen: 10 Liver: 3 | Abdominal bloating | Splenectomy and partial hepatectomy | 17 | NED |
|  | F/39 | Liver | 9 | Asymptomatic | Partial hepatectomy | 84 | NED |
| Chen[20] | F/28 | Liver | 6 | Abdominal pain, fatigue, anorexia | Partial hepatectomy | 48 | LWD, recurrence in liver |
|  | M/39 | Spleen | 7.4 | Asymptomatic | Splenectomy | 40 | NED |
|  | M/48 | Liver | 23.3 | Abdominal pain, fever, fatigue | Partial hepatectomy | 23 | NED |
|  | M/65 | Spleen and liver | Spleen: 22.3  Liver: 5.8 (multi masses) | Abdominal pain, fever, fatigue, anorexia, weight loss | Splenectomy | 2 | DOD |
|  | M/51 | Spleen | 8.5 | Weight loss | Splenectomy | 19 | NED |
|  | M/68 | Spleen | 2.3 | Asymptomatic | Splenectomy | 6 | NED |
|  | F/51 | Spleen | 5.3 | Abdominal discomfort | Splenectomy | 5 | NED |
|  | M/67 | Spleen | 7.5 | Asymptomatic | Splenectomy | 5 | NED |
|  | M/60 | Liver | 3 | Asymptomatic | Partial hepatectomy | 3 | NED |
|  | F/52 | Spleen | 0.9 | Asymptomatic | Splenectomy | 12 | NED |
| Kitamura [21] | F/74 | Spleen | 3.6 | Asymptomatic | Splenectomy | 24 | NED |
| Bui [22] | F/50 | Spleen | 6 | Abdominal pain | Splenectomy | NA | NA |
| Vardas[23] | M/61 | Spleen | 10 | Abdominal pain | Splenectomy | 12 | NED |
| Kim[24] | M/76 | Spleen | 3.2 | Asymptomatic | Splenectomy | NA | NA |
| Horiguchi[25] | F/77 | Spleen | 8.5 | Abdominal pain | Splenectomy | 36 | NED |
| Present case | F/31 | Liver | 2 masses: 3.5, 2.5 | Anorexia | 3D laparoscopic right hepatectomy | 10 | NED |
|  | M/48 | Liver and hepatoduodenal ligament lymph node | Liver: 10  Lymph node: 3.5 | Asymptomatic | Open right hepatectomy, lymph node excision | 2 | NED |

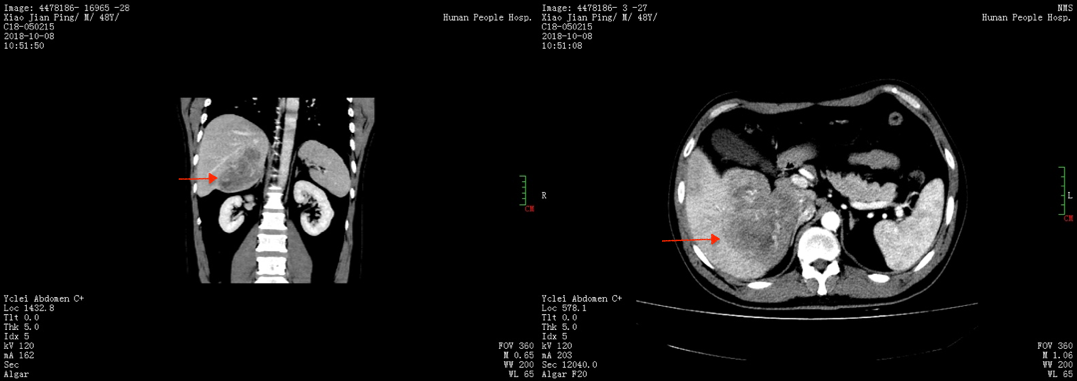
M: Male; F: Female; NED: No evidence of disease; DOD: Dead of disease; LWD: Live with disease; NA: Not available.



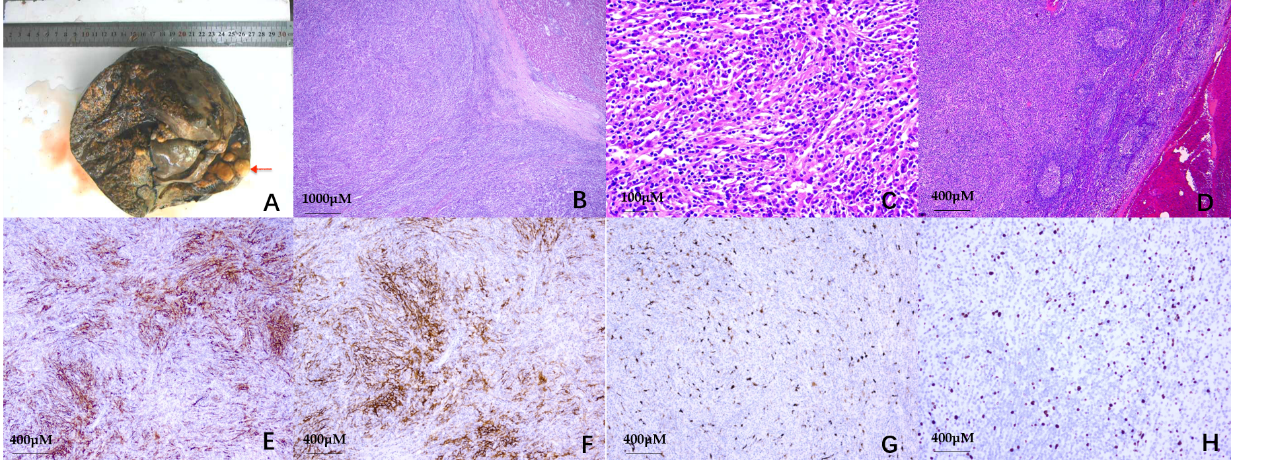
**Figure 1 Magnetic resonance imaging.** Two well-circumscribed lesions with long T1 and long or equal T2 signal (arrows). The multiple lesions with long T1 and long T2 signal are hepatic cysts verified by pathological examination later.



**Figure 2 Epstein-Barr virus-positive inflammatory pseudotumor-like follicular dendritic cell sarcoma in the liver.** a: Gross picture of an inflammatory pseudotumor-like follicular dendritic cell sarcoma of the liver. A well-circumscribed solid nodule was found in the liver. Note the grayish-white colored and soft cut surface with focal hemorrhage (arrow); b: Haematoxylin and eosin stained image showing that the tumor tissue had a meshwork-like architecture (×200); c: On high-power field, the tumor was composed of oval to spindle cells with vesicular chromatin and distinct nucleoli. There was less degree of atypia. The background showed abundant lymphocytes and plasma cells (×400); D: CD21 was detected on the membrane of almost all of tumor cells by immunohistochemistry (×100); E: Smooth muscle actin was detected in the cytoplasm of a part of tumor cells by immunohistochemistry (×100); F: Epstein-Barr virus-encoded small RNA-based *in situ* hybridization demonstrated positive nuclei of the neoplastic dendritic cells (×200); G: Ki-67 was detected in the nuclei of almost all of tumor cells by immunohistochemistry (30%; ×100).



**Figure 3 Abdominal computed tomography examination.** The images show an ill-defined and low-density 10 cm mass (arrows) in the right lobe of the liver, accompanied with enlargement of hepatic portal lymph nodes.



**Figure 4 Epstein-Barr virus positive inflammatory pseudotumor-like follicular dendritic cell sarcoma in the liver with hepatoduodenal ligament lymph node involvement.** a: Gross picture of an inflammatory pseudotumor-like follicular dendritic cell sarcoma of the liver. A large and multinodular confluent tumor was found in the liver (arrow); B: Histologic sections of follicular dendritic cell sarcoma showing an unencapsulated tumor (left) with a sharp margin from the adjacent liver parenchyma (right). The tumor tissue was arranged in whorls (×40); C: On high-power field, the tumor was composed of oval to spindle cells with vesicular chromatin and distinct nucleoli. There was less degree of atypia. The background showed abundant lymphocytes and plasma cells (×400); D: In the hepatoduodenal ligament lymph node, lymphoid follicles were pushed aside by tumor tissue (×100); E: CD21 was detected on the membrane of almost all of tumor cells by immunohistochemistry (×100); F: S100 was detected in the membrance and cytoplasm of almost all of tumor cells by immunohistochemistry (×100); G: Epstein-Barr virus-encoded small RNA *in situ* hybridization demonstrated positive nuclei of the neoplastic dendritic cells (×100); H: Ki-67 was detected in the nuclei of almost all of tumor cells (20%; ×100).