The Image Presentation of Inflammatory Follicular Dendritic Cell Sarcoma Mimic Primary Cholangiocarcinoma in Epstein-Barr Virus Prevalent Country: A Case Report and Literature Review

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Case report

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Abstract

Background: Follicular dendritic cell sarcoma (FDCS) is a very rare malignant soft tissue tumor. About half of the cases occurred in lymph node, mostly around neck. The uncommon inflammatory variant of FDCS is an Epstein-Barr virus (EBV)-associated neoplasm. It was first introduced as Inflammatory pseudotumor-like FDC in 2001. Few cases had been found in liver and spleen as primary tumor. It mimicked the presentation of cholangiocarcinoma in this case and few had been reported as primary liver tumor yet.

Case report: We represent a 53-year-old man who was referred to our department with a liver tumor. The tumor was accidently found during health exam at lateral segment, which was suspected to be cholangiocarcinoma on Magnetic Resonance Imaging(MRI).

The liver segment was complete resected via laparoscopic hepatectomy. The tumor turned out to be inflammatory FDCS. The tumor specimen was margin free. No recurrence was detected at a 12-month follow-up

Clinical Discussion: Inflammatory FDCS often found in liver and spleen. Taiwan was EBV high-prevalence country, but the occurrence seemed not related. Current management as FDCS, with radiation therapy and chemotherapy if total resection was not feasible.

Conclusion: Inflammatory FDCS is a rare malignant tumor in abdomen. Complete resection represents favorable outcome.

Introduction

Follicular dendritic cell sarcoma is a rare low-grade malignant sarcoma that most commonly presents as a slowly growing, painless mass, or lymph node invasion. It was first introduced in 1986 and several cases have been reported since then[1]. Histologically, FDCS is composed of spindle-shaped cells with eosinophilic cytoplasm and bundles in fascicles or trabeculae. Its nuclei features with pseudoinclusions, distinct nucleoli and vesicular chromatin pattern[2]. Only few cased of FDCS are associated with EBV, demonstrating an inflammatory pseudotumor like morphology. It was first introduced as Inflammatory pseudotumor-like FDC and associated with EBV in 2001[3]. The variant was present with high grade morphology, such as atypical spindle cells with indistinct borders between cells. Since than few cases had been reported. These tumors had most been found in liver and spleen as primary tumor if in abdomen, or present as lymphoma mimicking Hodgkin's disease[4]. Immunohistochemically, the FDCS was present with cell marker include: CD21, CD23, and CD35, also expressing dendric cell marker. On the other hand, the inflammatory variant too expressing expresses CD21, and CD35, but is positive for EBV[5]. We present a case was suspect to be cholangiocarcinoma on image study initially, but later proven as Inflammatory follicular dendritic cell sarcoma. This presentation hadn't been reported in literature yet.

Case Report

The 53-year-old male patient was referred to our department with a liver tumor at lateral segment. The tumor was found during health exam at 2019/4. Reviewing his personal history, he had systemic disease of hypertension and under medication control. He had no remarkable family history. He used to be incineration plant worker for 5 years, railway maintenance worker for 4 years, and worker in metal processing factory for another 10 years. No direct exposure to chemicals or toxin was recorded. Serum tumor marker level: CA19-9: 6.4U/mL, CEA: 1.3ng/mL, AFP: 2.6ng/ml. Hepatitis marker level: HBsAg: Nonreactive (0.253), Anti-HCV: Nonreactive (0.032). The MRI study showed: two markedly enhanced tumors identified respectively in the posterior and in the lateral(20.4mm) segment. The lesion located at posterior segment is markedly enhanced in arterial phase, and presented centripetal enhancement in the portal and equilibrium phases, favor hemangioma. Whereas the lesion in lateral segment, left side at the left hepatic vein, was brighten in diffuse weight image(Fig. 1A,B), dark in ADC mapping(Fig. 1C,D), with well-defined margin, favoring malignancy. Also, the lesion was seen to be mildly enhanced in arterial phase(Fig. 1E) and become more and more enhanced in the portal(Fig. 1F) and equilibrium phases(Fig. 1G). Delayed enhancement was considered, highly suspect malignancy. The preoperatively diagnosis was malignancy favor cholangiocarcinoma.

Since the tumor was suspected to be cholangiocarcinoma of liver, the patient received laparoscopic segmental hepatectomy. The operation was conducted successfully. Post-op course was smooth without immediate and late complications. He was then discharged within days. Surprisingly, we didn't found infiltrative irregular glands pattern or fibrodesmoplastic stromal reaction under microscope. Instead, the tumor was well circumscribed and capsuled. Fibroblast bundled in fascicles (Fig. 2A). Back ground was composed of lymphocyte and plasma cell inflitration(Fig. 2B). The tumor neuclei showed vesicular chromatin and almost come in pair(Fig. 2C). The necrosis pattern were also present in the specimen(Fig. 2D). On the other hand, the tumor present with follicular dendritic cell marker: CD21(Fig. 2E) and EBV related pattern: EBER positive(Fig. 2F). The pathologic proved the tumor to be EBV(+) inflammatory follicular dendritic cell sarcoma. PET scan was later arranged and report no definite evidence of residual disease, regional or distant metastasis. He went on receive outpatient department follow up and computed tomography scan for disease follow up.

Discussion

Inflammatory follicular dendritic cell sarcoma of liver is a very rare case as liver primary tumor. Reviewing literature, about 20 cases had been found in liver and about 30 of the cases were found in spleen[6–8]. Almost all patient had different degree of abdominal pain or discomfort in liver cases. Since there was higher prevalence rate of EB virus in Taiwan, almost above 95% of adults were seropositive, most known of association to nasopharyngeal cancer (NPC)[9]. The mechanism of NPC progression was based on the expression of viral proteins, especially latent membrane protein 1 (LMP1). The protein had demonstrated the ability in tumor progression in epithelium cell[10]. But, interestingly, the virus was never found in non-cancerous cells. Some LMP variant was prevalent in south Asia and responsible for cell transforming, resulting higher occurrence of NPC[11]. However, the role of EB virus in liver or infected organ still were still remain unclear. It is important to investigate if population in Taiwan or south Asia was associate with

higher incidence. We review case reports in Taiwan but found on data was available. There were two cases in our record, one is this case, present as hepatic mass. The other one was present with lymphoma like picture with mediastinum mass and multiple distant metastasis. The prevalence seems not to associate with high EBV prevalence. Still, more data or case should be extract before we jump to the conclusion. For local FDCS, which usually presumed as hepatocellular carcinoma, cholangiocarcinoma or splenic mass, resection was indicated if feasible. But for disseminated disease, diagnosed often based on biopsy, then management with radiation therapy and chemotherapy[12]. On the other hand, complete resection seemed to had better long term outcome by reducing recurrence rate.

Conclusion

Inflammatory FDCS were rare disease found as primary tumor in liver. It was found to be related with EB virus. Most cases were treated as primary liver tumor initially and resected if possible. Others disseminated disease were management with radiotherapy and chemotherapy. To date, no guideline had been established for such disease. Complete resection seems to provide better outcome and surveillance. We should look for more cases worldwide to establish standard treatment and keep investigate its incidence in EBV prevalent countries.

Declarations

Ethical Approval

Ethical approval has been given by IRB in Keelung Chang Gung memorial hospital. No.201902149A3C591

Consent of publication

All four author agreed to submit the article to World journal of surgical oncology and publication if accepted.

Availability of data and materials

All data was extract from medical records stored in Keelung Chang Gung memorial hospital.

Competing interests

All of authors have nothing to declare in any competing interest.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

Yi-Chan Chen and Rueyshyang Soong conducted the surgery. Cheng-Han Yang exam the slide and report the pathologic result. Wu-Po Chao wrote the manuscript. Rueyshyang Soong reviewed the manuscript.

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Figures

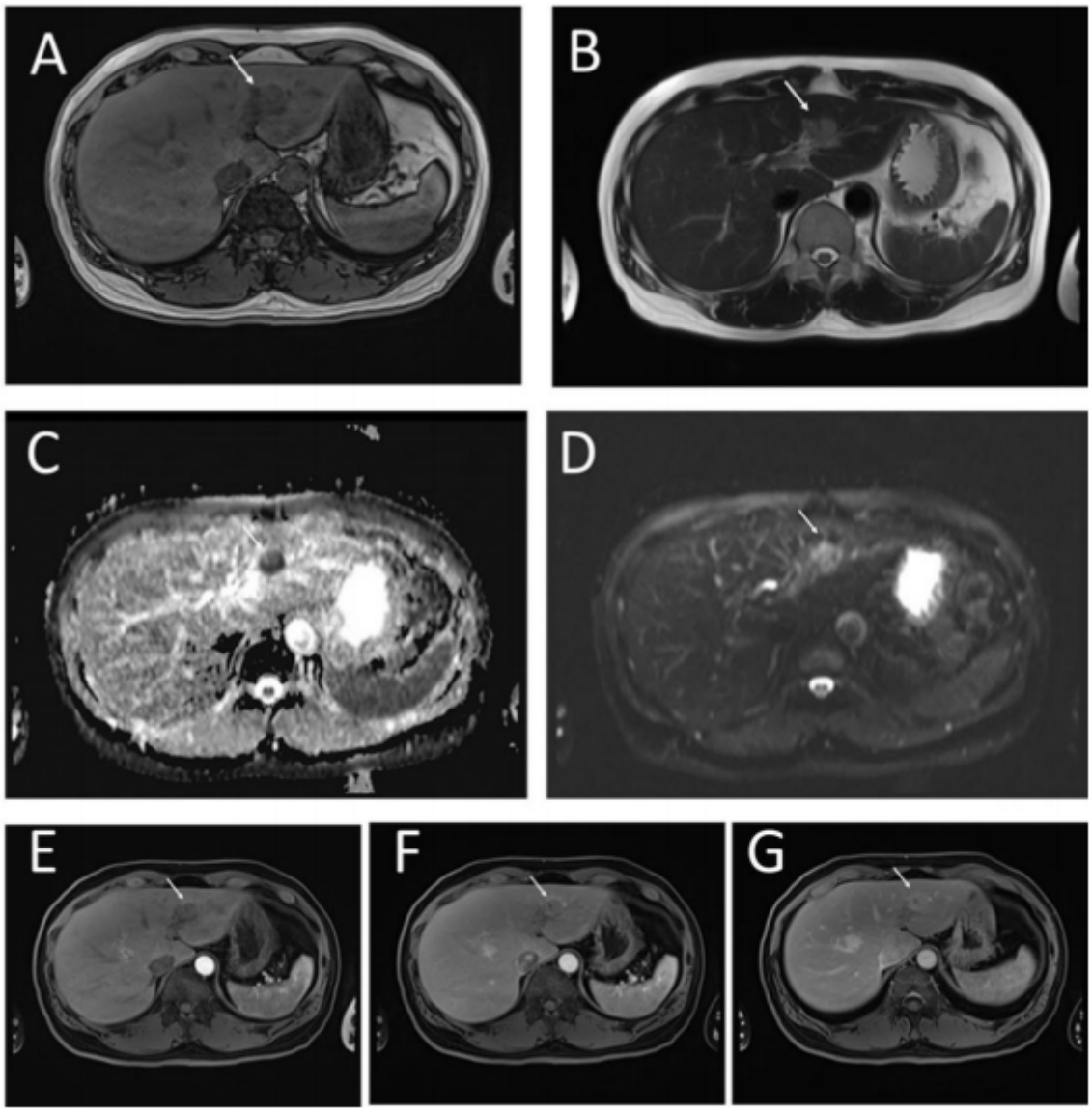

Figure 1

The tumor at lateral segment was dark in T1(A), but brighter in T2 (B), favor tumor. The lesion is brightening in diffuse weight image(D), dark in ADC mapping(C), with well-defined margin, favoring malignancy. The lesion is seen to be mildly enhanced in arterial phase and become more and more enhanced in the portal and equilibrium phases, (E) arterial phase (F)portal phase, (G)equilibrium phase.

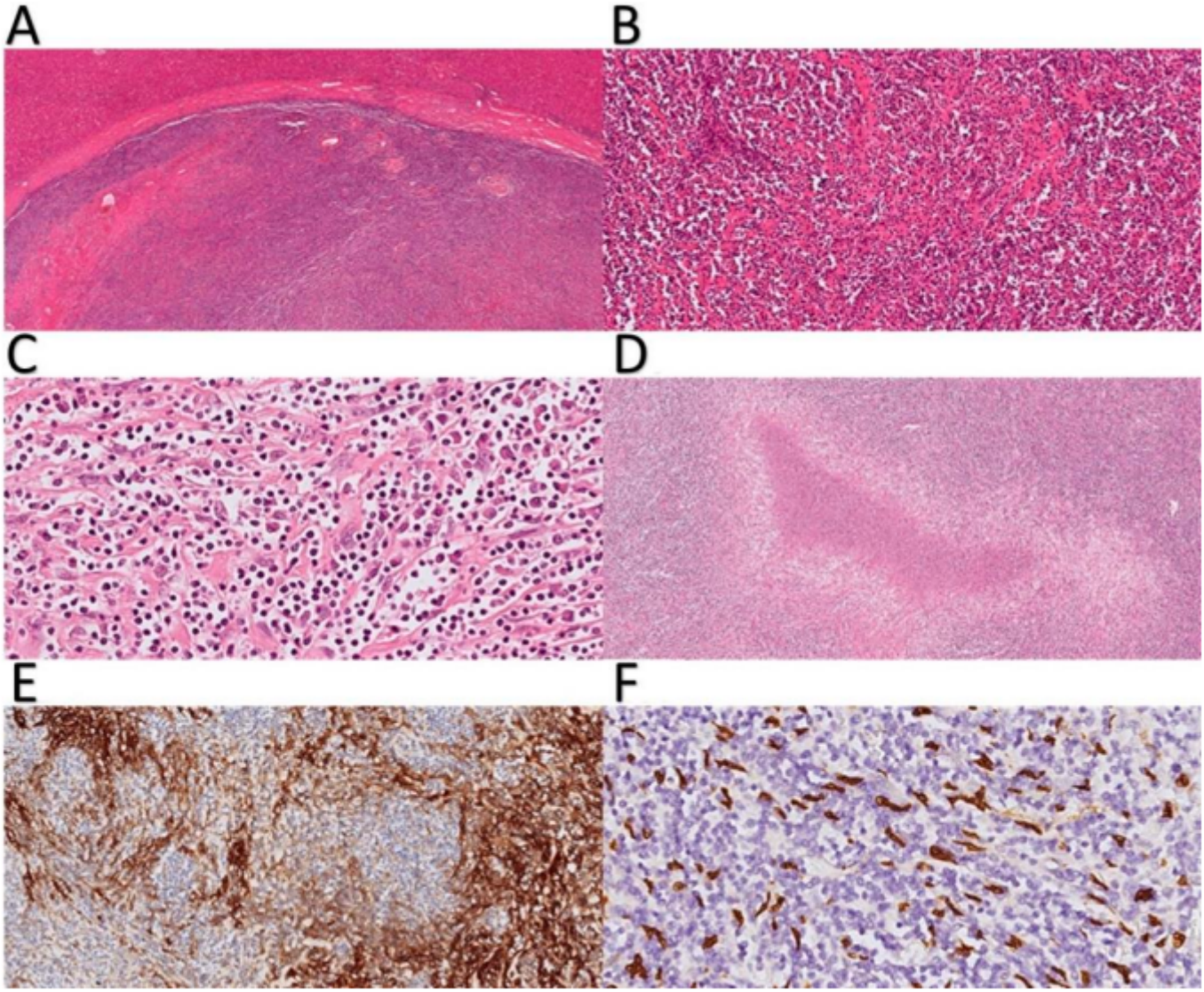

Figure 2

The tumor was well circumscribed and capsuled. Fibroblast bundles in fascicles (A). Back ground compose of lymphocyte and plasma cell(B). The tumor nuclei showed vesicular chromatin and almost come in pair(C). Necrosis were present in the specimen(2D). The tumor present with follicular dendritic cell marker: CD21(E). The tumor also presented with EBV related pattern: EBER positive(F).