Clinical Case Report

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Small intestine follicular dendritic cell sarcoma with liver metastasis

A case report

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Abstract

Rationale: Follicular dendritic cell sarcoma (FDCS) is a rare neoplasia composed of spindle or oval cells with follicular dendritic cell differentiation, usually occurring in lymphoid tissue. In this report, we present a case of FDCS of the small intestine with liver metastasis.

Patient concerns: A 19-year-old female presented with recent onset of left upper abdominal pain. Abdominal computed tomography scan showed a large tumor mass in the liver lateral segment with compression to the pancreas upper part, and a smaller mass in the terminal ileum, respectively. High serum levels of amylase and lipase were noted. Resection of the tumors was performed. Microscopically, both tumors consisted of ovoid to spindle-shaped nuclei cells admixed with some lymphocytes arranged in fascicles, whorls, storiform arrays. Immunohistochemistry demonstrated that the tumor cells were positive for follicular dendritic cell markers, including CD21, CD23, and CD35. Epstein–Barr virus encoding small RNA (EBER; Inform EBER probe; Ventana Medical Systems, Tucson, AZ) in situ hybridization was negative.

Diagnoses: According to the clinicopathological features, diagnosis of FDCS of intestinal origin was made.

Interventions: Resection of tumors located in the liver and at the small intestine was performed. After the operation, patient received adjuvant vinblastin chemotherapy.

Outcomes: There was no evidence of recurrence at 8-month follow-up.

Lessons: It was unusual for FDCS of intestinal origin with liver metastasis and expressing with high serum levels of pancreatic enzymes.

Abbreviations: CT = computed tomography scan, FDCS = follicular dendritic cell sarcoma.

Keywords: follicular dendritic cell sarcoma, liver metastasis, small intestine tumor

1. Introduction

Follicular dendritic cell sarcoma (FDCS) is a rare neoplasm, located primarily in the germinal centers of primary and secondary lymphoid follicles of nodal and extranodal sites.[1] It affected mainly young adults with median age of 40 years without gender predilection.[2] Intra-abdomen as the only site of involvement of FDCS was a very uncommon condition and was associated with a particularly aggressive clinical course.[2,3]

2. Case presentation

A 19-year-old female visited the hospital emergency department for recent onset of upper abdominal pain. Physical examination indicated a firm mass in the upper part of the left side abdomen. The serum levels of amylase and lipase were elevated to 195 U/mL (normal range, 25–125 U/mL) and 1410 U/mL (normal range, 8–78 U/mL), respectively. Abdominal computed tomography (CT) scan showed a large tumor located in the left lobe lateral segment of the liver with compression to the pancreas and the stomach, and a smaller tumor at the small intestine (Fig. 1). Resection of both tumors was performed. Macroscopically, a mass sized 4.0 x 4.0 cm was seen in the mesentery of the ileum, 40 cm proximal to the ileocecal valve, and another tumor sized 12.6 x 8.3 cm in the liver lateral segment (Figs. 2 and 3). There was no tumor involvement over regional lymph nodes of the small intestine and liver or other parts of the abdomen. After operation, the serum levels of amylase and lipase returned to normal range (56 and 77 U/mL, respectively). Microscopical examination of the liver and mesentery tumors both showed FDCS, characterized by ovoid to spindle cells arranged in fascicles and whorls admixed with lymphocytes. Mitotic figures were frequently seen [>10/10 high-power field (HPF); Fig. 4]. Necrosis was also noted. The tumor cells showed immunoreactivity for follicular dendritic cell

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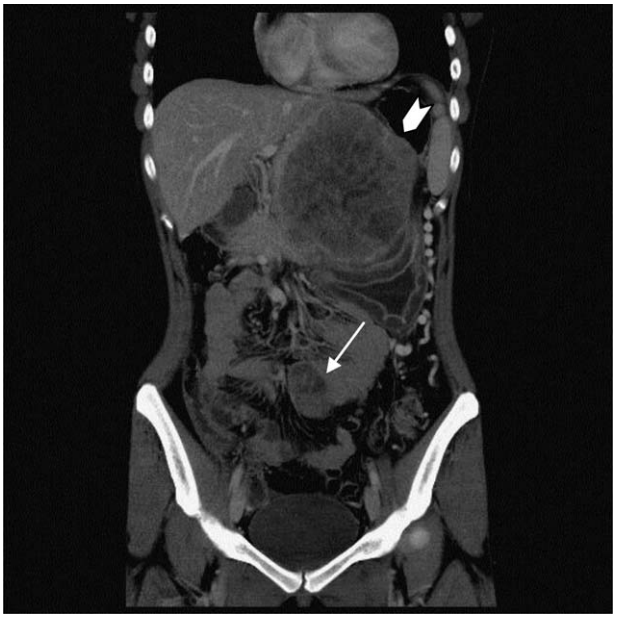

Figure 1. The CT imaging showed a well-defined heterogeneous mass located at the small intestine (arrow) and a larger tumor located at liver lateral segment (arrow head).

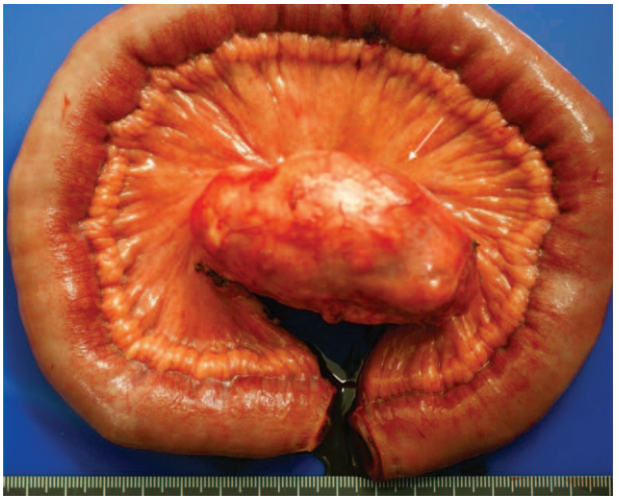

Figure 2. Macroscopic image of the resected tumor arising from the mesentery of ileum as a round to ovoid fleshy mass (arrow).

markers, including CD21 (Clone 2G9, Dilution 1:10; Leica, Germany), CD23 (Clone 1B12, Dilution 1:15; Leica, Germany), CD35 (Clone RLB25, Dilution 1:40; Sigma-Aldrich, Canada). In situ hybridization for Epstein–Barr virus (EBV) encoding small RNA (EBER; Inform EBER probe; Ventana Medical Systems, Tucson, AZ) failed to show positive cells (Fig. 4). A final diagnosis of small intestine FDCS with liver metastasis was made on these findings. Further staging workup showed no evidence of tumor involvement over other sites, including regional lymph nodes or bone marrow, and with no residual tumor over surgical sites. After the operation, patient received adjuvant chemotherapy for 3 months and there was no evidence of recurrence at 8-month follow-up.

3. Discussion

FDCS was first presented by Monda et al in 1986.[4] FDCS mainly affects peripheral lymph nodes, with the cervical nodes being the most common site. Other sites, including the mediastinal, retroperitoneal, and mesenteric areas lymph nodes, were extensively affected.[5,6] However, a wide variety of extranodal sites have been reported, including the tonsils, liver, spleen, oral cavity, gastrointestinal tract, bones, soft tissues, skin, and breasts.[5] Clinical presentation of extranodal FDCS included abdominal pain, intestinal obstruction, rectal bleeding, and dyspepsia, when abdominal involvement was noted. Dyspnea, cough, dysphagia, and dizziness were reported when it comes to extra-abdominal involvement. Image study can be the initial tool for diagnosis of FDCS, and provides delineation of the extent of the mass and staging. Ultrasound and CT are usually the initial imaging modalities of choice used. CT can show smaller, relatively homogeneous masses; however, heterogeneity as a result of necrosis or hemorrhagic areas has been reported in more than 80% cases.[7]

Univariate and multivariate analyses of FDCS prognostic factors was done by Saygin et al[5] in 2013. Fifty cases of FDCS were collected and analyzed. Similar to other soft tissue sarcomas, large tumor size (≥6 cm), presence of coagulative necrosis, high mitotic count (≥5 per 10 HPFs), and significant cytologic atypia were shown to be associated with poor prognosis.[5]

The current case demonstrated characteristic histopathological features of FDCS, including intimate admixture of oval to spindle-shaped tumor cells with mature lymphocytes and plasma cells. There are some unusual clinicopathological features as compared with those described in the literature. First, we present a case of intra-abdominal FDCS with mesentery and hepatic lesions present at the moment of diagnosis, with similar pathological result at both

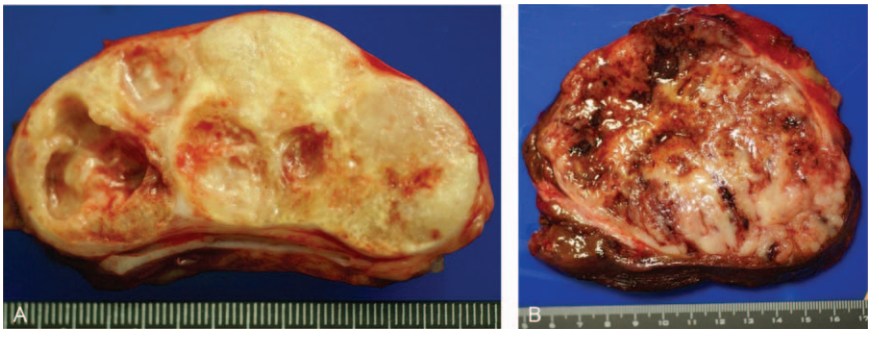

Figure 3. On cut sections, the tumor located at the small intestine (A) demonstrating well-defined border, whereas the tumor located at liver (B) had less defined margin. Both tumors were yellowish with necrotic area.

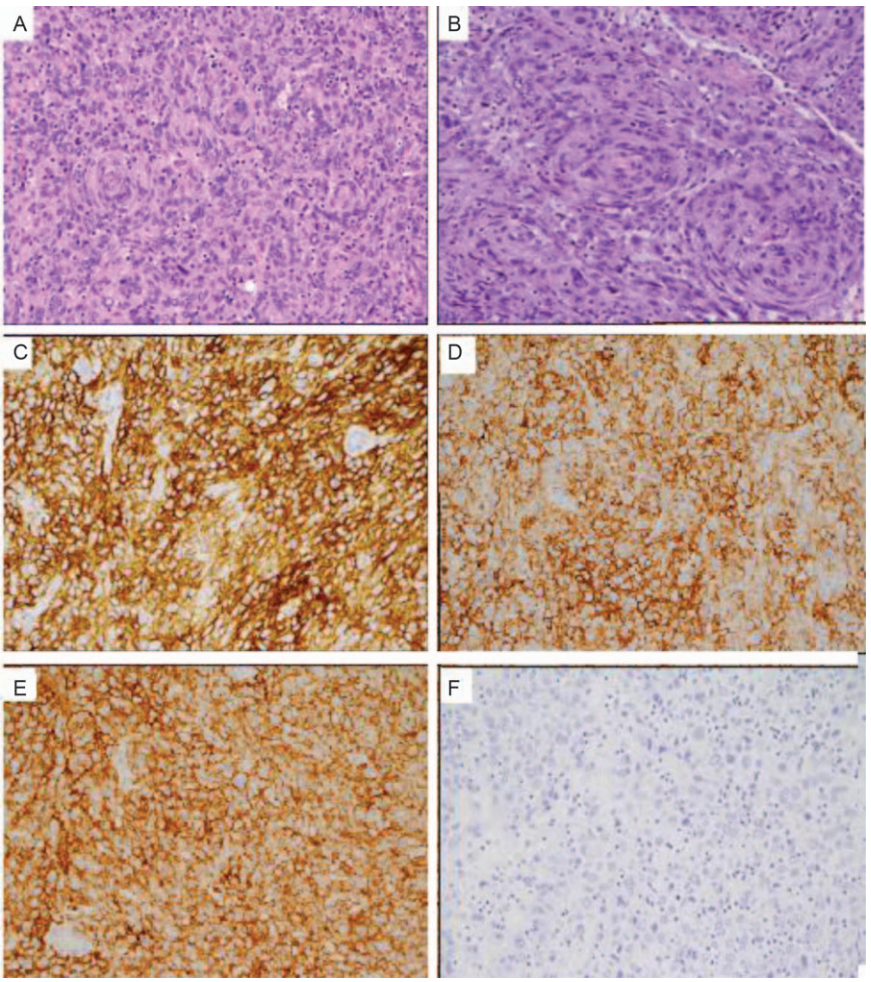

Figure 4. Histological examinations of the small intestine (A) and liver (B) tumors show ovoid to spindle cells arranged in fascicles and whorls admixed with lymphocytes (H&E staining, original magnification ×200). The tumor cells are diffusely positive for CD21 (C), CD23 (D), and CD35 (E). EBER in situ hybridization is negative in the tumor (F) (original magnification ×200).

sites. Second, the tumors presented as acute abdominal pain with elevated serum amylase and lipase level, possibly due to the tumor mass located at the liver left lobe was large and with compression to the pancreas and stomach. Third, in our case, the tumor located at the ileum mesentery and liver both showed aggressive histopathological features, including marked nuclear pleomorphism, brisk mitotic rate that included occasionally atypical forms, nucleolar prominence, and coagulative necrosis.

With positive results of CD21, CD23, CD35, which are specific for FDCS, and classical features of morphology, the diagnosis of FDCS was made. According to the reports, almost all primary hepatic FDCSs were EBV-positive,[8,9] while our case was negative. In addition, it is very unusual for a large FDCS of liver origin with single site involvement of the mesentery and with sparing of other parts of the abdomen. Thus, we inferred that the primary site of the FDCS in our case is located at the intestine mesentery, and the liver lesion was considered consistent with blood-borne metastatic FDCS.

According to the study to date, the first-line treatment of localized FDCS is surgical resection, and there is still no consensus on performing optimal adjuvant chemotherapy and radiotherapy. Some treated FDCS with an aggressive lymphoma regimen, which often includes chemotherapy, whereas others treated FDS as a soft tissue sarcoma, with wide resection and adjuvant radiotherapy.[10] Disseminated type of FDCS is suggested to use lymphoma-type chemotherapy, and the CHOP regimen including cyclophosphamide, adriamycin, vincristine, and prednisone is the most widely used.[10–12] In our presented case, adjuvant vinblastin chemotherapy is used and there is no evidence of recurrence for 8 months to date.

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