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Case Report

Resection is an effective treatment for recurrent follicular dendritic cell sarcoma from retroperitoneum: unusual presentation of a rare tumor

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Abstract: Retroperitoneum follicular dendritic cell sarcoma (FDCS) is an extremely rare neoplasm. The treatment of this disease is not clear. A 49-year-old Chinese female who had been found a 4.4×4 cm retroperitoneum mass by routine physical examination was received radical resection. Pathology revealed an inflammatory pseudotumor-like follicular dendritic cell tumor. After five years follow-up, a new nodule was noted on the tail of pancreas by routine CT evaluation. Re-resection was performed and pathological examination found a spindle-cell tumor with a great quantity of froth histiocytes. Immunohistochemical stains were positive for CD35 and CD21 which suggested it was a recurrent FDCS. Retroperitoneum FDCS is a very rare tumor. Surgical resection may be the first choice for this disease, even for recurrent tumor, if feasible. A relatively good prognosis often is achieved when compared with other malignancy.

Keywords: FDCS, retroperitoneum, recurrent, resection, pancreas

Introduction

Case presentation

A 49-year-old Chinese woman with a history of 30-years hypertension went to local hospital for annual physical examination. A nodule was noted on the left adrenal gland by ultrasound. Further CT scan demonstrated an occupancy lesion on the left of retroperitoneum with a size of 4.4×4.0 cm (A neuroendocrine tumor was the first consideration).

The patient underwent a radical resection of the left adrenal gland tumor. Upper pole of the left kidney and adrenal gland were showed after open the gerota fascia. The lesion appears on the left adrenal gland adheres tightly to pancreas and spleen. Pathology revealed a 5.0×4.0×4.0 cm spindle cell tumor in a background of froth histiocytes and inflammatory cells infiltrate. Surgical margins of the tumor were negative. The Immunohistochemistry (IHC) showed CK(-), VIM(+), SMA(-), MSA(-), DES(-), CD34(-), CD117(-), S100(-), CD68(+), CD21(++), CD35(+), CD1a(-), ALK-α(-), CD30(-), EBV(-). An inflammatory pseudotumor-like follicular dendritic cell tumor was considered (**Figure 1A**, H&E; **Figure 1B**, VIM; **Figure 1C**, CD21; **Figure 1D**, CD35).

The patient did not receive any adjuvant chemotherapy or radiotherapy after operation but followed up with CT scan every six months. A new lesion was observed on the tail of pancreas five years (58 months) after operation (**Figure 2A**, **2B**, red arrowheads). A recurrent FDCS was considered and distal pancreatectomy plus splenectomy were performed. Pathology revealed a 6.5×5.5×4.0 cm FDCS. Surgical margins of the pancreas and five lymph nodes were negative. The IHC showed CK(-), VIM(+), CD68(+), SMA(-), DES(-), KI67 (30%+), S100(+), CD117(-), CD34(blood vessel +), ALK-α(-), CD1α(+), CD21(+), CD35(++) (**Figure 2C**, H&E; **Figure 2D**, VIM; **Figure 2E**, CD21; **Figure 2F**, CD35).

Discussion

FDCS is a rare neoplasm with less than 400 cases described in the literature [1]. The major-

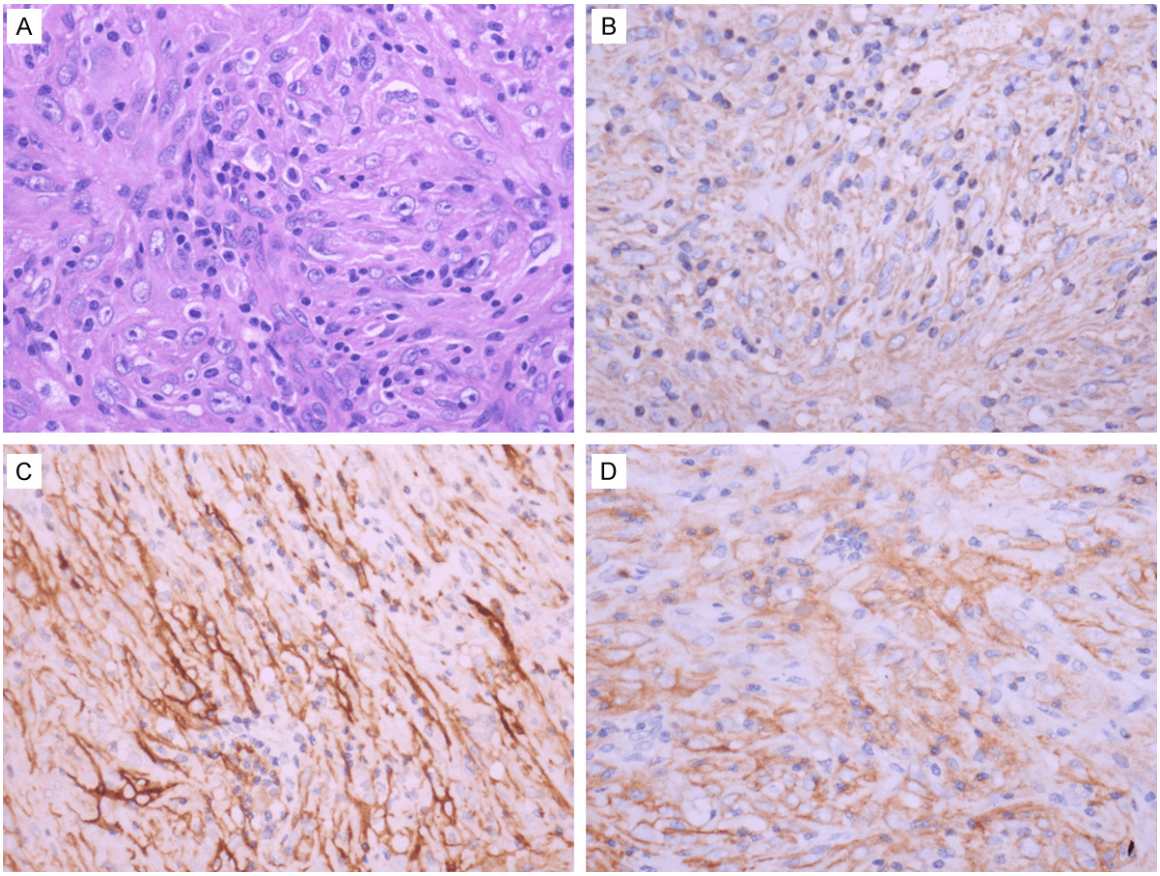

Figure 1. Pathological section for FDCS. After the first resection, an inflammatory pseudotumor-like follicular dendritic cell tumor was considered (A. H&E; B. VIM; C. CD21; D. CD35).

ity of the tumors were found in lymph node. Head and neck area is the most preferred site for nodal FDCS. However, about one-third of the lesions were identified in extranodal sites [2]. Most extranodal FDCS occurred in abdominal cavity (e.g. liver or spleen). Retroperitoneum FDCS is an extremely rare tumor with only 17 cases having been reported in the world [1]. Here we report a case of extranodal FDCS arising within retroperitoneum and relapse in pancreas five years after first resection.

FDCS has been defined by the World Health Organization as a low-grade sarcoma with follicular dendritic cell differentiation. Due to its rarity, FDCS is often initially misdiagnosed. Our patient appeared with an occupancy lesion on adrenal gland and a history of 30 years hypertension, both of which suggested it a neuroendocrine tumor. Biopsy is helpful to make a correct diagnosis. However, it is difficult for pathologist to differentiate FDCS from poorly differentiated carcinomas depend solely on morphology due to limited specimen. In that case, IHC for CD21 (positive in 93% of cases) and CD35 (positive in 89% of cases) is the most commonly used solution to reach the correct diagnosis. Epstein-Barr virus (EBV) infection was demonstrated in majority of the hepatic and splenic lesions and its causative effect has been proposed for the pathogenesis of the tumor [3-6], but the association is poor in most of the tumors from other sites [7, 8]. Our patient showed negative EBV-encoded RNA hybridization in both primary and recurrent tumor.

There is no guideline for treatment of FDCS due to limited experience. Most of the patients with local disease were treated with surgery with or without adjuvant therapy. Caner Saygin [1] analyzed the follow-up data of 282 FDCS patients and found patients who received surgery had a significantly better overall survival (OS) when compared to patients who had other treatment modalities. There is no significant difference on OS between patients who had surgery alone

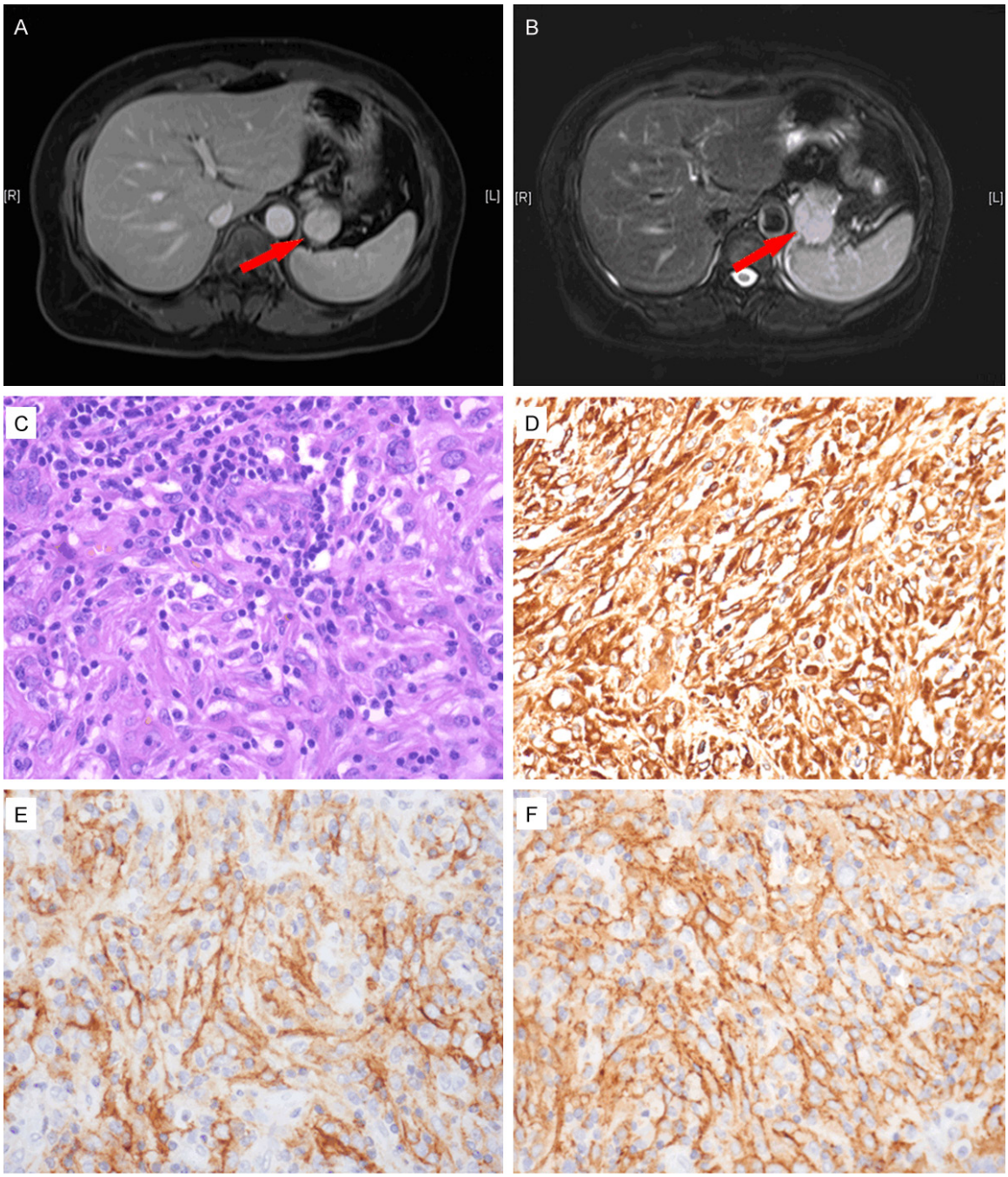

Figure 2. Recurrent FDCS. 58 months after the first operation, a recurrent lesion was observed on the tail of pancreas five years (A, B. Red arrowheads). A recurrent FDCS was considered (C. H&E; D. VIM; E. CD21; F. CD35).

and patients who received adjuvant radiotherapy following surgery.

To our knowledge, this is the first report to describe a case of rare extranodal FDCS arising in retroperitoneum and relapse in pancreas five years after operation. Literature review and experience on this patients suggested that surgical resection may be the first choice for FDCS, even for recurrent tumor. A relatively good prognosis may be achieved when compared with other malignancy. Adjuvant therapy (chemotherapy or radiotherapy) is not necessary for this disease.

Disclosure of conflict of interest

None.

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