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CLINICAL CASE REPORT

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Extranodal Follicular Dendritic Cell Sarcoma Originating in the Pancreas: A Case Report

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Abstract: Follicular dendritic cell (FDC) sarcoma is a type of malignant tumor that originates from immune system-related FDCs. Pancreatic FDC sarcoma is a rare disease, and the specificity of the clinical presentation and laboratory results is unknown. We report the clinical process and imaging features of one case of pancreatic FDC sarcoma.

A 67-year-old woman presented with a hypoechoic mass between the spleen and left kidney during a medical examination. The patient was hospitalized for further diagnosis. Her laboratory results did not present any obvious abnormal changes. Unenhanced and contrast-enhanced pancreatic computed tomography scans indicated a round mass with heterogeneous attenuation in the pancreatic tail, and a 3.5 × 3.6-cm solid mass with a cystic component was noted. Clear-cut, slight contrast enhancement was present in the solid part, whereas contrast enhancement was not observed in the cystic part. In addition, no obvious dilation was observed in the pancreatic duct, and no swollen lymph nodes were noted in the posterior peritoneum. Routine and contrast-enhanced pancreatic magnetic resonance imaging scans showed an abnormal signal indicative of a mass in the pancreatic tail, with a diameter of ~35 mm and a clear boundary. A T2-weighted imaging scan showed a slight hyperintensity coupled with part of a hyperintensity, whereas T1-weighted imaging showed a slight hypointensity coupled with part of a hypointensity, and diffusion-weighted imaging showed a heterogeneous hyperintensity. The solid part of the lesion showed poor contrast enhancement through contrast-enhanced scanning, but contrast enhancement was not observed in the cystic part. Surgical tumor resection was performed, and the pathological diagnosis was pancreatic FDC sarcoma. The tumor did not recur based on short-term CT reexamination.

Pancreatic FDC sarcoma is a rare disease, and the established clinical examinations and laboratory tests lack specificity. Imaging reveals a solid mass with a cystic component and a clear boundary. In addition, the solid part exhibits poor contrast enhancement. Although pancreatic occurrence is rare, a clinical pancreatic solid tumor with a cystic component should be identified by differential diagnosis.

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Abbreviations: CT = computed tomography, DWI = diffusion-weighted imaging, FDC = follicular dendritic cell, MRI = magnetic resonance imaging, T1WI = T1-weighted imaging, T2WI = T2-weighted imaging.

INTRODUCTION

Follicular dendritic cell (FDC) sarcoma is an indolent tumor that originates from FDCs.[1] FDCs might form a tight meshwork in primary and secondary lymphoid follicles while interacting with B or T lymphocytes and thus play a role in the immune system.[1] The pathological presentations of FDC sarcoma are a neoplastic growth of spindle-shaped or oblong cells and cell phenotypes presenting as FDCs. Epstein–Barr virus infection is hypothesized to be related to the occurrence of FDC sarcoma in the liver and spleen.[2,3] FDC sarcomas predominantly occur in the lymph glands, accounting for ~2/3 of cases, whereas in the other cases, the occurrence is in various extranodal sites.[4] The most common extranodal locations for occurrence are the abdominal cavity and pelvic region, followed by the neck and chest; rarely are the soft tissues of the breast, thigh, groin, dura mater encephali, and skin involved.[4] To our knowledge, up till now only three cases of pancreatic FDC sarcoma have been reported.[5–7] Here, we describe the clinical presentation and imaging characteristics of one case of pancreatic FDC sarcoma in detail.

CONSENT

The patient provided informed consent regarding publication of the case information.

CASE REPORT

The patient was a 67-year-old woman who had visited a local hospital for a medical examination 11 days prior. An ultrasound examination had yielded the following notes: ''fatty liver, block mass with hypoecho exists between spleen and kidney. And further examination was suggested.'' At that time, the patient presented with no symptoms, including no fever, chill, nausea, vomiting, cramps, or bloating. The patient was referred to another local hospital for further computed tomography (CT) examination. The results showed a solid mass with a cystic component in the pancreatic tail that was hypothesized to be a solid pseudopapillary tumor. Subsequently, the patient was referred to our hospital for further treatment. The patient had no symptoms, including no fever, chill, nausea, vomiting, cramps, bloating, yellow skin, yellow eyes, or yellow urine. The outpatient record listed ''pancreatic occupying: pancreatic solid pseudopapillary tumor?''; the patient was thus hospitalized. The patient did not present with an obvious recent weight change. In addition, the patient had a 10-year history of hypertension that was treated with antihypertensive drugs, and she reported that her blood pressure was controlled. The patient had a history of penicillin allergy manifesting in symptoms of

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urticaria; had no history of diabetes, heart disease, nephrological disease, or infectious disease; and did not drink or smoke. The patient's skin was not yellow and her abdomen was flat. Peristalsis and a peristaltic bowel wave were not visible and her bowel sounds were 3 times/minute. Shifting dullness was negative and the abdomen was soft. Tenderness and rebound tenderness were not observed and a significant mass was not palpable. Her triglyceride level was 2.13 mmol/L (reference range, 0.30–1.70 mmol/L), and her low-density lipoprotein level was 3.30 mmol/L (reference range, 1.31–3.29 mmol/L). The remaining biochemical indexes and the electrolyte indicators of her liver and kidney function were all within the normal range. Routine blood, urine, and stool levels were also in the normal range. Tumor markers, including carbohydrate antigens (153, 199, and 125), carcinoembryonic antigen, alpha-fetoprotein, and ferritin, were normal, and coagulation function, an electrocardiogram, and chest film had no abnormal signs.

Unenhanced and contrast-enhanced pancreatic CT scans showed an $\sim 3.5 \times 3.6$-cm round solid mass with a cystic component visible in the pancreatic tail, with a smooth border. The internal density was heterogonous (Figure 1). The solid part had slight contrast enhancement after the contrast-enhanced scan, whereas contrast enhancement with a clear boundary was not visible in the cystic part. The pancreatic duct was not obviously expanded, and swollen lymph nodes were not observed in the posterior peritoneum. The imaging indicated a solid mass with a cystic component in the pancreatic tail; it was considered to be a solid pseudopapillary tumor or pancreatic cancer. Routine and contrast-enhanced pancreatic magnetic resonance imaging (MRI) scans showed an abnormal signal with a clear boundary in the pancreatic tail (Figure 2). A T2-weighted imaging (T2WI) scan showed a slight hyperintensity coupled with part of a hyperintensity, whereas T1-weighted imaging (T1WI) showed a slight hypointensity coupled with part of a hypointensity, and diffusion-weighted imaging (DWI) showed a heterogeneous hyperintensity. The solid part of the lesion showed poor contrast enhancement through contrast-enhanced scanning, but contrast enhancement was not observed in the cystic part.

Excision of the pancreatic tail and splenic excision were performed after the examination. An $\sim 4 \times 4 \times 4.5$-cm-sized tumor with medium texture and a clear boundary was visible in the pancreatic tail during the operation. The tumor was near the splenic vein and splenic artery. The profile appeared as a solid tumor with a cystic component. The intraoperative pathological diagnosis was a possible malignant pancreatic tumor. Pancreatic tail and splenic excision specimens were obtained, and a gray neoplasm was observed in the pancreatic sections. The scope included the following (Figure 3): tumor cells presented as a spindle shape and were woven in a vortex arrangement; cell pleomorphism was apparent, and the mitotic count could be determined easily; patchy necrosis was observed in the area; and immunohistochemistry showed CD21 (+), CD23 (+), CD117 (−), S-100 (−), Desmin (−), CD34 (−), SMA (−), DOG-1 (−), Ki-67(40%), CK (−), CgA (−), Syn (−), and EBER (−). The final pathological diagnosis was an FDC sarcoma. The patient recovered well after surgery. No tumor reoccurrence was observed by short-term CT reexamination.

DISCUSSION

In 1986, Monda et al reported the first four cases of FDC sarcoma.[8] Individual cases and group cases have been reported continuously.[4] The age of patients described as having an extranodal FDC sarcoma has ranged from 9 to 82 years of age, with the average age of onset in the fourth decade.[4] Men and women have similar morbidity.[4] A previous study reported pancreatic FDC sarcoma in men with various ages of onset, including 56, 63, and 64.[5–7] The patient in our report was an elderly woman. In two previous reports, the occurrence was in the pancreatic head, whereas one report did not note the specific site in the pancreas.[5–7] In our case, the pancreatic FDC sarcoma occurred in the pancreatic tail. The clinical presentations of pancreatic FDC sarcoma include weight loss, an abdominal mass, nausea, anorexia, and bloating.[5,6] There were no clinical symptoms in the case described here, possibly because of the tumor position and because it was relatively small.

FDC sarcoma has been regarded as a type of low-grade sarcoma.[9] Chan et al reported that this disease is prone to recurrence and should be considered as an intermediate-grade malignancy, particularly in cases of intraperitoneal FDC sarcomas.[10] In 2006, Shia et al compared 47 cases of extranodal FDC sarcoma and found that 43% of the cases had recurrence.[11] In 2010, Li et al reported a recurrence rate of up to 42% for extranodal FDC sarcomas, with the local recurrence rate and remote shift rate being 23% and 21%, respectively.[4] Therefore, extranodal FDC sarcoma presents with relatively high

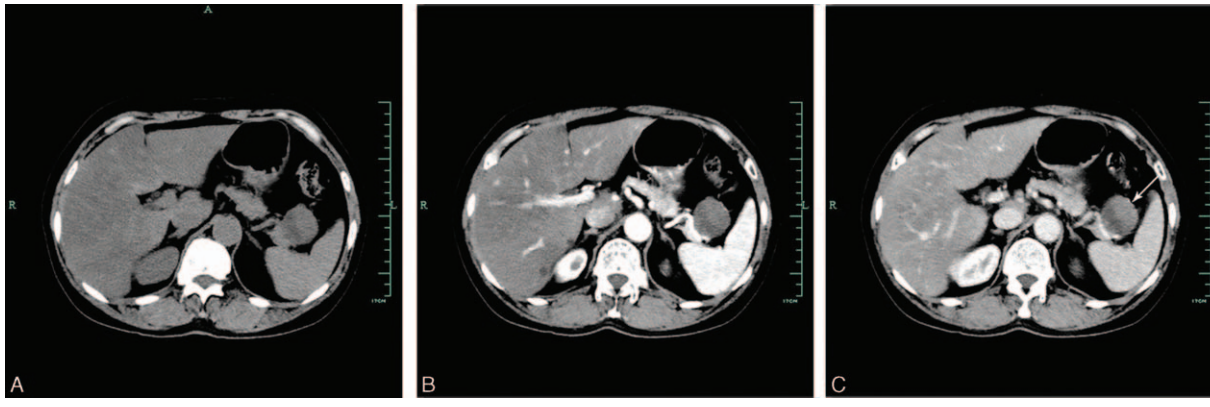

FIGURE 1. Patient pancreatic CT. (A) The unenhanced pancreatic CT scan showed that a round soft tissue block existed in the pancreatic tail (white arrow) and that there was heterogeneous attenuation, striped hypodense visible, and a smooth tumor border with a clear boundary. (B, C) Contrast-enhanced CT showed that the tumor's solid part exhibited poor contrast enhancement and that contrast enhancement was not observable in certain areas (white arrow). CT = computed tomography.

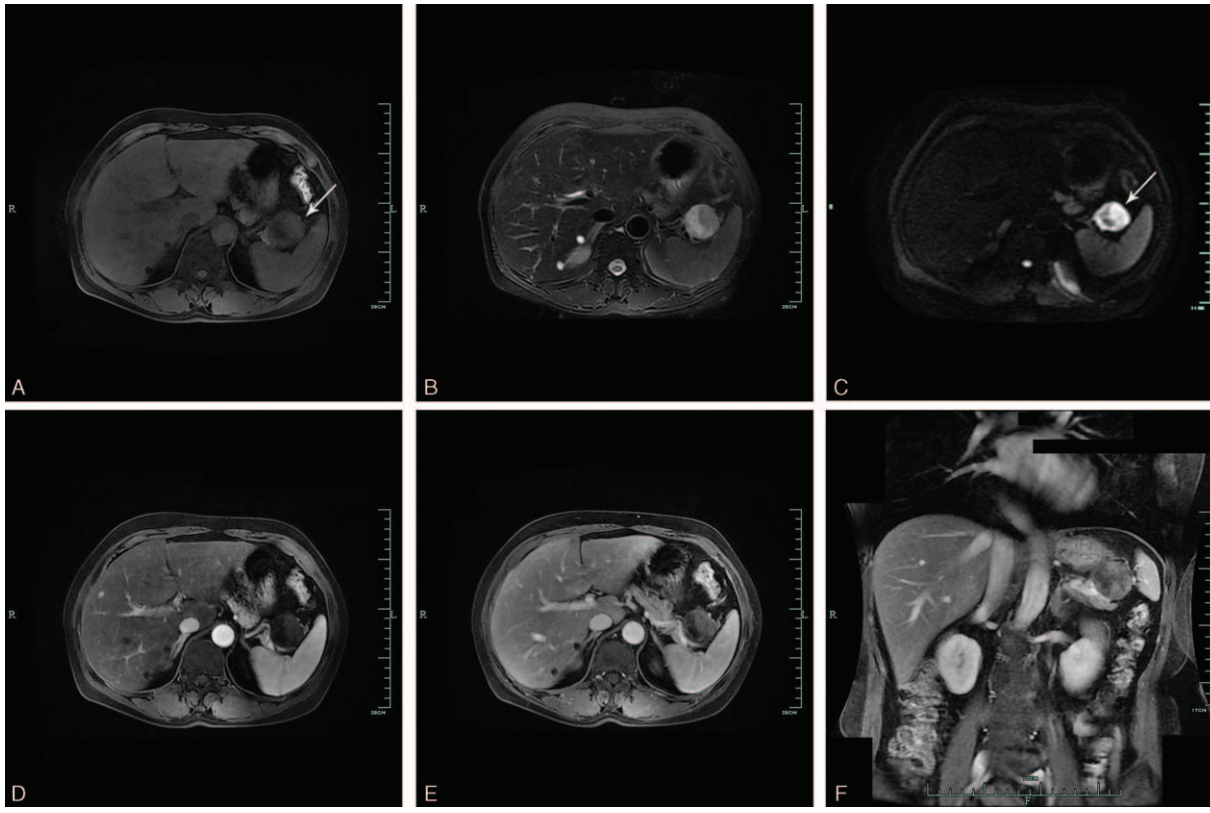

FIGURE 2. MRI of patient's pancreas MRI. (A) Routine pancreatic MRI scan showed a round mass in the pancreatic tail. T1WI took on a slight hypointensity coupled with a part hypointensity. (B) T2WI presented a slight hyperintensity coupled with a part hyperintensity. (C) In terms of the DWI sequence, an obvious hyperintensity was visible for the solid part (white arrow). (D–F) Contrast-enhanced MRI showed that the solid part presented poor contrast enhancement and that no contrast enhancement occurred in the cystic part. DWI = diffusion-weighted imaging, MRI = magnetic resonance imaging, T1WI = T1-weighted imaging, T2WI = T2-weighted imaging.

aggression and is not a low-grade sarcoma. The size of the tumor (5 cm) and the mitotic count (5/10 HPFs) have been confirmed to have an obvious relationship with tumor recurrence.[4]

Past studies have focused on the clinicopathological features of extranodal FDC sarcoma; our focus was on the imaging presentation of the disease. Previously, images of FDC sarcoma cases occurring in the pancreatic head were reported minimally, presenting as a heterogeneous mass accompanied by biliary dilatation.[6] In our case, the tumor occurred in the pancreatic tail, which did not lead to biliary or duct expansion, and our patient had no significant clinical symptoms. Because the tumor was a solid mass with a cystic component, differentiation of lesions, such as solid pseudopapillary neoplasms, neuroendocrine tumors, differentiated carcinomas, acinar cell carcinomas, and metastatic neoplasms, was needed for the differential diagnosis. A solid pseudopapillary tumor of the pancreas is typically observed in young women in the form of a large

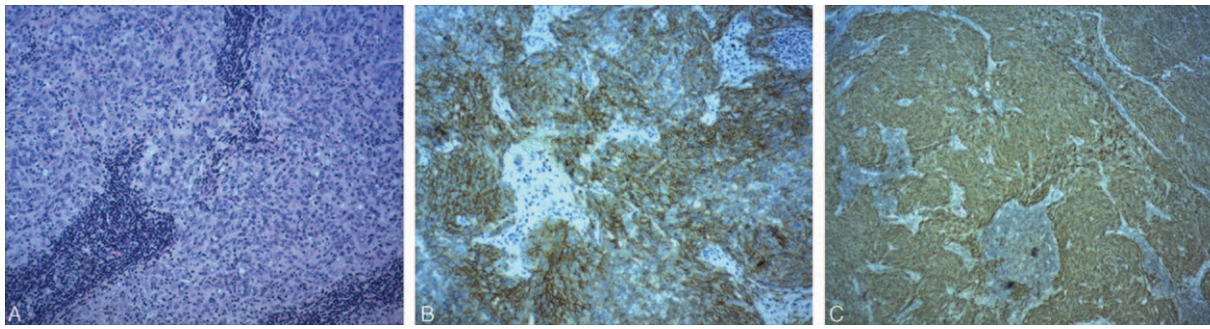

FIGURE 3. (A) Tumor cells presented a spindle shape with a vortex arrangement. Cell pleomorphism was apparent (hematoxylin and eosin staining). (B) CD21 expression was diffusely positive in the tumor cells. (C) CD23 expression was diffusely positive in the tumor cells. (D) CD2 expression was negative in the tumor cells. CD2 = cluster of differentiation 2, CD21 = cluster of differentiation 21, CD23 = cluster of differentiation 23.

lesion.[12] The cystic and solid parts, with various contents caused by tumor hemorrhaging, are shown by CT.[12] A contrast-enhanced solid part and calcification on the lesion border are visible.[12] CT showed that the pancreatic FDC sarcoma in our case had a similar presentation to that of a solid pseudopapillary tumor of the pancreas. However, the T1WI sequence of the MRI, performed to confirm a typical solid pancreatic pseudopapillary tumor, displayed hyperintensity resulting from hemorrhaging, whereas the pancreatic FDC sarcoma appeared as hypointensity because of tumor necrosis. Patients with cystic neuroendocrine tumors typically have no symptoms and frequently show no function.[13] On imaging, neuroendocrine tumors frequently occur in the pancreatic tail, and the solid part shows an abundant blood supply.[13] In contrast, in the pancreatic FDC sarcoma case in the present study, a poor blood supply was observed by contrast-enhanced CT and MRI. Thus, the pancreatic FDC sarcoma representation of contrast enhancement was different from that of a neuroendocrine tumor. The cystic change probability of pancreatic ductal carcinoma is 1% to 8%.[14] The typical cystic change occurs in poorly differentiated pancreatic carcinomas with relatively large diameter.[14] Obvious ductal obstruction and invasion often appear on these tumors and cause metastasis of lymph gland or distant organs. However, there was no ductal obstruction or invasion occurring in our case of pancreatic FDC sarcoma. Pancreatic acinar cell carcinomas usually present as an oval heterogeneous mass with a clear or ill-defined edge, which is likely to be accompanied with necrosis.[15] Its solid part has mild and persistent enhancement in a contrast-enhanced CT or MRI.[15] Indeed, the imaging findings of pancreatic FDC sarcoma in our case are similar to that of pancreatic acinar cell carcinomas. However, pancreatic acinar cell carcinomas are more likely to occur in elderly men, and more than half of the patients present a moderate or intense enhancement;[15] these features may contribute to the differential diagnosis of both. Sometimes, metastatic tumors of pancreas may be a solid mass with cystic change, usually accompanied with disseminated disease.[14] The patients of pancreatic metastatic tumors often have a history of primary cancer, especially a medical history of ovarian cancer or renal carcinoma.[14] In our case, because there was no medical history of other tumors, metastatic tumor of the pancreas could be excluded easily.

The typical treatment method for an FDC sarcoma is excision or excision with chemotherapy.[4] The tumor recurrence rate is 42% with surgery alone, whereas the recurrence rate is up to 48% with surgery plus chemotherapy.[4] Our patient was treated with excision alone, without chemotherapy. Recurrence was not observed in the short term.

CONCLUSION

An extranodal FDC sarcoma is uncommon, and a pancreatic FDC sarcoma is rare. Clinical and laboratory examinations of pancreatic FDC sarcomas lack specificity. The imaging presentations of a pancreatic FDC sarcoma are a solid mass with a cystic component and a clear boundary, accompanied by poor contrast enhancement of soft tissue. Pancreatic FDC sarcomas should be distinguished from other pancreatic solid tumors with a cystic component.

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