

Interdigitating dendritic cell sarcoma presenting in the sigmoid colon mesentery

A case report and literature review

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Abstract

Rationale: Interdigitating dendritic cell sarcoma (IDCS) is an extremely rare disease. It commonly occurs in middle-aged males and mainly involves the lymph nodes. Pathological examination plays an important role in differentiating from other tumors, but far less published literature focuses on the imaging characteristics of IDCS.

Patient concerns: Here, we reported a case of IDCS in a 52-year-old male involving the pelvis with medical imaging and pathologic findings.

Diagnoses: Preoperative unenhanced CT scan revealed a 6.0 × 6.3 × 8.0 cm mass with density equal to that of adjacent muscle, located in the pelvis. On contrast-enhanced CT images, the tumor presented apparent homogeneous enhancement. CT angiography showed that the tumor was supplied by the branches of inferior mesenteric artery. Pelvic magnetic resonance imaging manifested a lobulated solid mass with low signal on T1-weighted and intermediate to high signal on T2-weighted images. Simultaneously, significantly high signal intensity was exhibited on the diffusion-weighted images. This patient underwent operative resection of the tumor. The pathologic diagnosis was IDCS.

Interventions: This patient underwent operative resection of the tumor. The resection margins were negative for the neoplastic proliferation and no distant metastases were found. The patient did not receive advanced radiotherapy or chemotherapy.

Outcomes: Three months after surgery, the follow-up CT scan did not reveal any recurrence or metastases.

Lessons: This case adds to the experience with IDCS by summarizing its characteristics as well as reviewing the literature.

Abbreviations: CA125 = carbohydrate antigen 125, CA724 = carbohydrate antigen 724, CEA = carcinoembryonic antigen, CK19 = cytokeratin 19, CT = computed tomography, FDCS = follicular dendritic cell sarcoma, HU = Hounsfield unit, IDCS = interdigitating dendritic cell sarcoma, IMF = inflammatory myofibroblastic, MFH = malignant fibrous histiocytoma, NSE = neuron-specific enolase, PSA = prostate-specific antigen.

Keywords: interdigitating dendritic cell sarcoma, medical imaging, pathology, sigmoid colon mesentery

1. Introduction

Interdigitating dendritic cell sarcoma (IDCS) is a rare malignancy originating from dendritic cells, which participate in immune response activity as antigen presenting cells by stimulating T lymphocytes.^[1] Most IDCS occurs in the lymph nodes (particularly in the neck, mediastinum, and axilla)^[2]; however, about 1/3 are located in the extranodal organs such as the lung, skin, breast, bone, liver, spleen, and small intestine.^[3–5] Histological findings of IDCS show the tumor cells are medium to large spindle shaped with indistinct cell borders that form a whorled growth pattern.^[6,7] Until now, only about 100 cases of IDCS have been reported in the

English literature and surgical resection remains the mainstay of treatment for early-stage, localized disease.^[6] In this study, we report the first case of IDCS of the sigmoid colon mesentery occurring in a 52-year-old man and describe the medical imaging features. The aim of current study was to better our understanding with IDCS by summarizing its characteristics as well as reviewing the literature. This study was approved by the institutional review board at the Second Affiliated Hospital of Nanjing Medical University in China, with informed patient consent.

2. Case report

In July 2016, a 52-year-old male was admitted to our hospital with a 2-year history of abdominal distension. One week before admission, his symptoms worsened with increased stool frequency. On the physical examination, the patient had a regular pulse of 70 beats/min, a respiratory rate of 20 breaths/min, and a temperature of 36.6°C. His cardiopulmonary and neurologic examinations were normal. No swollen lymph nodes were detected in the neck, axillary, or inguinal regions. His laboratory data such as complete blood cell count and creatinine showed no significant abnormalities. The serum carbohydrate antigen 125 (CA125) was significantly increased at 258 U/mL (normal range, 0–35 U/mL). Other tumor markers [serum carbohydrate antigen 724 (CA724), cytokeratin 19 (CK19), carcinoembryonic antigen (CEA), serum prostate specific antigen (PSA), and neuron-specific enolase (NSE)] were within normal

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Figure 1. (A) Unenhanced CT scan shows a mass in the pelvis. (B, C) On contrast-enhanced CT images, the tumor presents apparent, homogeneous enhancement. CT = computed tomography.

limits. Preoperative unenhanced CT scan of abdomen revealed a $6.0 \times 6.3 \times 8.0$ cm lobulated mass with homogeneous density (mean CT value, 40HU), located in the pelvis (Fig. 1A). On contrast-enhanced CT images, the tumor presented apparent enhancement. The mean CT values were 93 HU in the arterial phase and 87 HU in the venous phase (Fig. 1B and C). The tumor had an unclear interface with the adjacent tissue causing pressure on the sigmoid colon. CT angiography revealed the tumor was supplied by branches of inferior mesenteric artery (Fig. 2). Pelvic MRI showed a lobulated solid mass of low signal intensity on unenhanced T1-weighted images and of intermediate to high signal intensity on T2-weighted images (Fig. 3A and B). The lesion exhibited significantly high signal intensity on the diffusion-weighted images (Fig. 3C). Contrast-enhanced T1-weighted images demonstrated a homogeneously enhancing mass in the pelvis (Fig. 3D). This patient underwent operative resection of the tumor. During surgery, the pelvic mass was found to be located at the sigmoid colon mesentery, which compressed the sigmoid colon causing intestinal edema and partial intestinal obstruction. Postoperative pathology showed the tumor had large, fusiform spindle cells with indistinct cell borders. The shape of the nucleoli was oval with finely dispersed chromatin and the nucleoli were small but prominent (Fig. 4). With immune stains, the tumor cells tested positive for S-100, CD34 and negative for B-cell and T-cell markers, CD21, CD35. The proliferation index, expressed as a percentage of Ki-67 antigen-positive nuclei, was

around 20%. Fifteen mesenteric lymph nodes (with diameters of up to 5 mm) were dissected after surgical removal of the tumor and identified to be negative. Based on the pathologic and immunohistochemical findings, the pelvic mass was diagnosed as interdigitating dendritic cell sarcoma.

3. Discussion

Clinically, IDCS presents as a painless mass occurring between the ages of 6 to 87 years (mean age 51 years), and more often in the male gender (M: F=1.5:1).^[2,5] To our knowledge, the vast majority of the IDCS are presented as case reports without any specific symptoms. In most patients, IDCS arises in a peripheral lymph node and its symptoms are similar to lymphadenitis.^[3,8] The next most commonly affected organ is the liver.^[3,7] In these patients with extranodal IDCS, systemic symptoms such as weight loss and fever are more common.^[1,4,9,10] In the current report, the tumor of $6.0 \times 6.3 \times 8.0$ cm in size was located in the pelvis. It mechanically compressed the sigmoid colon, causing intestinal edema and partial intestinal obstruction. The patient presented with abdominal distention and increased stool frequency. To our knowledge, similar cases of IDCS originating from the sigmoid colon mesentery have not been reported in the previously published literature.

Sarcoma of the dendritic cells is divided into several subtypes including Langerhans cell sarcoma, IDCS, follicular dendritic cell sarcoma (FDSC), dermal dendrocyte sarcoma, interstitial dendritic cell sarcoma, and veiled cell sarcoma.^[11] Among the above neoplasms, FDSC is the most frequently reported with 343 cases in the literature, and only about 100 cases of IDCS have been described.^[6] The etiology of IDCS is unknown. Helbig et al^[2] suggested Epstein–Barr virus and human herpesvirus 8 may be involved in the development of the disease. Fraser et al^[8] presented a case of IDCS associated with chronic lymphocytic leukemia/small lymphocytic lymphoma and demonstrated that these 2 tumors were clonally related. A clonal relationship between IDCS and low-grade B-cell lymphomas had been reported in several studies.^[8,12,13] However, the current patient did not have any significant past medical history of viral infection or leukemia/lymphoma.

Histological and immunohistochemical findings act perform key roles in the diagnosis of IDCS. Histologically, IDCS is characterized by the proliferation of spindle cells with whorled growth pattern,^[14] which is usually absent in melanoma, malignant peripheral nerve sheath tumor, atypical fibroxanthoma, malignant fibrous histiocytoma, rhabdomyosarcoma, and leiomyosarcoma.^[9] Immunologically, IDCS is almost

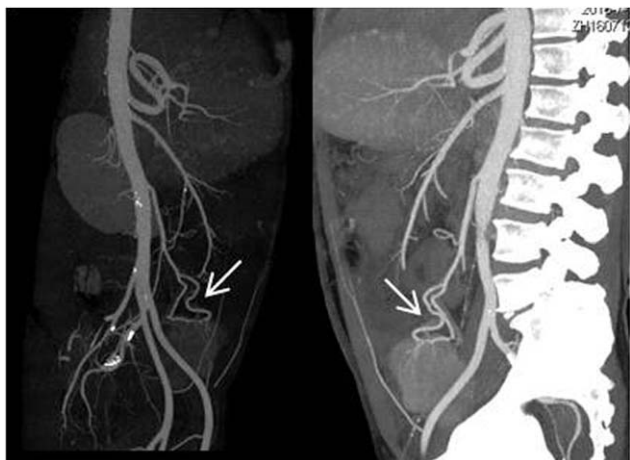


Figure 2. CT angiography reveals that the tumor is supplied by branches of inferior mesenteric artery (white arrow). CT = computed tomography.

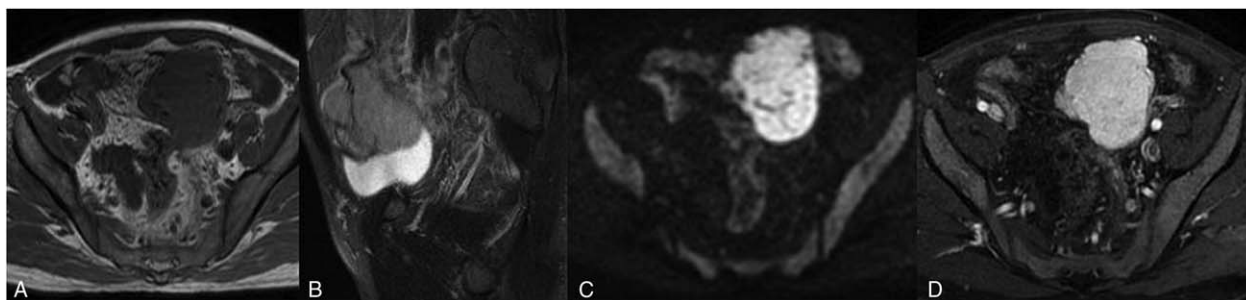


Figure 3. (A–C) The tumor presents as low signal on T1, slightly high signal on T2, and high signal on diffusion-weighted images. (D) On contrast-enhanced MRI images, the tumor shows homogeneous enhancement. MRI = magnetic resonance imaging.

uniformly positive for S-100 protein expression. On contrast, the makers for T cells (CD3, CD5 and CD8), B cells (CD20), FDCCS (CD21, CD35), and Langerhans cells (CD-1 α , CD56) are negative.^[15] In the present case, tumor cells were positive for S-100 and CD34, but there was no immunoreactivity for CD-1 α , CD21, CD35, CD3, and CD20.

According to several published reports, the imaging features of IDCS have been summarized. On CT imaging, IDCS presents as an iso-intense soft tissue mass with apparent enhancement effect.^[6,9,16,17] Positron emission tomography/computed tomography imaging shows increased 18-fluoro-deoxyglucose uptake in the lesion.^[4,15,16] In our current report, CT demonstrated a well circumscribed homogeneous soft tissue mass located in the pelvis. Because of tumor-feeding arteries, the mass was clearly enhanced on contrast-enhanced imaging. We are the first to report the tumor manifesting high signal intensity on diffusion-weighted images. This specific manifestation is possibly related to the density of tumor cells but that is open to future study.

Melanoma, malignant fibrous histiocytoma (MFH), and inflammatory myofibroblastic (IMF) tumor are considered in the differential diagnosis of IDCS in the mesentery. The MRI features of melanoma are described as high signal on T1 and very low signal on T2-weighted images.^[18] According to Park's report,^[19] the CT findings of the MFH in the mesentery were an irregularly margined, heterogeneously delayed enhancing mass with irregular peritumoral strands in the mesentery. MFH frequently expressed vimentin, actin, alpha 1-antichymotrypsin

and CD 68. IMF is a solid tumor that often affects children and shows typically heterogenous attenuating enhancement. Immunohistochemistry for ALK is relatively specific for IMF tumor.^[20]

The results of therapy are unsatisfactory; the median survival for IDCS is 9 months.^[14] Surgical resection remains the primary treatment for localized and resectable IDCS; adjuvant radiotherapy does not significantly improve overall survival or lower the recurrence rate.^[2] Systemic chemotherapy is the mainstay of treatment for the patients with distant metastases.^[6] In the current case, the resection margins were negative for the neoplastic proliferation and no distant metastases were found. The patient did not receive advanced radiotherapy or chemotherapy. Three months after surgery, the follow-up CT scan did not reveal any recurrence or metastases.

In conclusion, our case adds to the experience with IDCS. The above presented case is interesting for 3 reasons: First, IDCS is a very uncommon tumor. Second, a more detailed description of medical images features has been provided. Last, according to our knowledge, this is the first case of primary localization of IDCS in the sigmoid colon mesentery.

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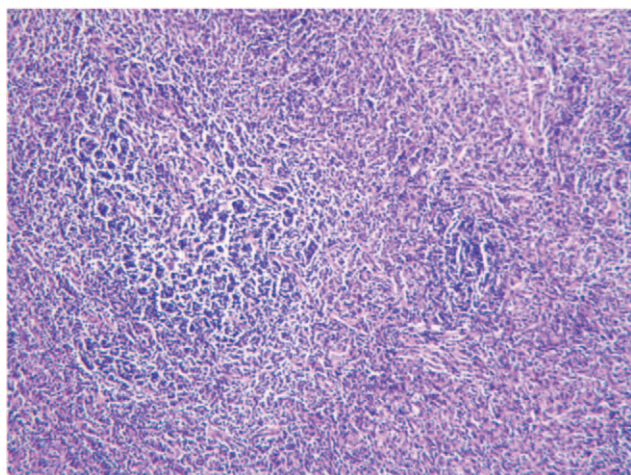


Figure 4. Spindle-shaped tumor cells form a whorled growth pattern (H&E, $\times 100$). H&E = hematoxylin and eosin stain.

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