Histiocytic Sarcoma of the Terminal Ileum Presenting As a Large Ulcerating Lesion: CT Diagnosis

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Abbreviations: CT, computed tomography

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Abstract

We report the case of an 84-year-old woman with histiocytic sarcoma arising in the terminal ileum. The lesion was identified on CT of the abdomen as a 10 cm segment of terminal ileum with mild circumferential wall thickening, isoattenuation, and diffuse homogeneous enhancement. The patient was treated with surgery and adjuvant chemotherapy. This case provides an example of when imaging may be useful for the detection of this rare malignancy and its complications.

Introduction

Histiocytic sarcoma is an aggressive malignant hematologic tumor comprising less than 0.5% of all Non-Hodgkin’s lymphomas. We present a case in which the tumor was an ulcerated mass in terminal ileum, involving the muscularis propria and serosa.

Case Report

We present a case of an 84-year-old woman who had a longstanding history of anemia attributed to myelodysplastic syndrome (MDS), chronic renal insufficiency, and chronic iron deficiency. She presented with complaints of dyspnea, left flank discomfort and left lower quadrant abdominal pain. Her past medical history was significant for coronary artery disease, and congestive heart failure. She was status post coronary artery by pass graft, right sided carotid artery endarterectomy, right hip and knee replacement, total abdominal hysterectomy/bilateral oophorectomy and hemorrhoidectomy. She had been receiving multiple blood transfusions. Colonoscopy, an upper gastrointestinal (GI) endoscopy and a capsule endoscopy done in the past did not reveal any ultimate source of blood loss.

Abdominal CT obtained after oral and intravenous contrast agents, was successful in depicting a mild circumferential bowel wall thickening in the terminal ileum (Fig.1). The involved intestinal wall ranged in maximum thickness from 1.0-2.5 cm and the length was 5-10 cm. The lesion showed diffuse homogeneous enhancement with isoattenuation. No definite mural involvement was noted. There was some dilatation of the intestine without perforation. The radiologic findings were interpreted as suspicious for a neoplastic process. Follow up colonoscopy was performed and biopsies
were taken from terminal ileum and the ascending colon. The pathologic diagnosis was ‘small bowel mucosa with acute ulceration and focal changes consistent with ischemia’. There was no evidence of malignancy.

**Figure 1.** 84-year-old woman with histiocytic sarcoma, at initial presentation. Coronal reformatted CT of abdomen and pelvis at initial presentation, showing moderate bowel wall thickening (arrows) with contrast enhancement.

Her clinical course was complicated by progressive increase in requirement for blood transfusions, and recurrent urinary tract infections. She presented a year later with increasing right lower abdominal pain, generalized weakness, dyspnea, and anemia. Her hemoglobin was found to be 8.7 mg/dl and hematocrit of 28.3. Previously, she required transfusion once every five to six months since her diagnosis of MDS. For the past one year, she was transfused eight units of blood.

Contrast enhanced CT scan of the abdomen and pelvis showed an interval increase in the circumferential thickening of the bowel wall, involving a 10-15 cm length of distal ileum. The lesion now ranged in maximum thickness from 2.0-4.5 cm and showed diffuse homogeneous enhancement with isoattenuation. Mucosal ulceration and mural involvement was present without obvious perforation or stranding of mesenteric fat (Fig. 2). Non-bulky scattered lymphadenopathy was also demonstrated. It was localized, involving the small bowel mesenteric and pericolic lymph nodes. The diameter of the short axis of the lymphadenopathy was larger than 10-mm. No peritoneal or omental involvement was noted. Low attenuating lesions were seen in the liver ranging in size from 0.5 to 2 cm, which showed ring enhancement on post contrast images consistent with metastatic disease.
Figure 2A. 84-year-old woman with histiocytic sarcoma, one year after presentation. Coronal reformatted CT of abdomen and pelvis showing marked interval increase in circumferential bowel wall thickening (arrows) with narrowing of the segment of ileum.

Figure 2B. 84-year-old woman with histiocytic sarcoma, one year after presentation. Axial reformatted image of the lower abdomen reveals marked interval increase in the area of thickening (arrows) in the terminal ileum.

Follow up colonoscopy revealed a large ulcerated friable mass in terminal ileum. The initial impression was lymphoma or an adenocarcinoma. Serum Tumor Markers Chorioembryonic Antigen
(CEA) and Cancer Antigen 19-9 (CA19-9) were within normal limits. The preoperative histopathologic diagnosis was consistent with histiocytic sarcoma.

Partial ileocelectomy and appendectomy was performed along with the lymph node dissection. The resected specimen consisted of a 24 cm segment of the terminal ileum and 13 cm of cecum. Findings at gross examination indicated an irregular ulcerated infiltrating mass involving the distal ileum measuring 10.0 cm in maximum dimension (Fig 3). The lesion extended to the ileocecal valve but did not appear to involve the colon and appendix. The cut surface showed a relatively well-circumscribed, solid, variegated gray to yellow tumor extending to the small bowel serosa. Multiple enlarged lymph nodes were identified in mesenteric and pericolic fat measuring 0.5 to 2.5 cm in greatest dimension.

![Figure 3. 84-year-old woman with histiocytic sarcoma, after surgery. Gross surgical specimen obtained after segmental resection of ileum shows an irregular ulcerated mass (arrows).](image)

Microscopic examination showed full thickness infiltration of small bowel wall by malignant tumor composed of many sheets of spindle to epithelioid cells with abundant eosinophilic cytoplasm, oval to irregular nuclei, vesicular chromatin, and large nucleoli (Fig. 4 a). There were interspersed bizarre multinucleated cells, xanthomatous histiocytes and inflammatory cells, such as lymphocytes and plasma cells (Fig. 4 b). Mitoses ranged from 1 to 10 per 10 HPF. Necrosis was not present. The tumor cells were positive for leukocyte common antigen CD45 (LCA), CD45RO (UCHL-1), CD68 (KP1), CD4 (a T-cell antigen present in a subset of histiomonocytic cells), HLA-DR, vimentin, and lysozyme (Fig. 5 a-c). Smooth muscle actin (SMA), desmin, melan A, CAM5.2, CKAЕ1/AE3, EMA, S100 protein, CD1a, CD2, CD5, CD20, CD21, CD30, CD34, CD35 and CD117 (c-kit) were negative. The final diagnosis was ‘histiocytic sarcoma’. All of the regional lymph nodes were involved. The liver biopsy from the right lobe lesion was also similar to the primary ileal tumor.
Figure 4. 84-year-old woman with histiocytic sarcoma, after surgery. A, Photomicrograph shows infiltration of the bowel wall by malignant cells (Hematoxylin-eosin, original magnification x 20). B, Sheets of spindle cells arranged in storiform pattern and scattered bizarre multinucleated cells (Hematoxylin-eosin, original magnification x 40).
Figure 5. Immunohistochemical staining pattern of specimens from 84-year-old woman with histiocytic sarcoma. A, A strong diffuse cytoplasmic CD68 positivity (original magnification x 20). B, The tumor cells are positive for CD4 (original magnification x 20). C, The tumor cells are positive for HLA-DR (original magnification x 20).

The patient was started on adjuvant chemotherapy with ifosfamide, mesna, doxorubicin, and dacarbazine. She received the same therapeutic regimen two times. Each treatment lasted 5 days, with an interval of 21 days. A postoperative CT was performed about one month after the operation, which revealed multiple intra abdominal fluid collections/abscesses. She is currently hospitalized on treatment.

Discussion

Histiocytic sarcoma is a rare, high grade hematopoietic neoplasm, representing <1% of all non-Hodgkin's lymphomas. It is defined in World Heath Organization (WHO) Classification as a malignancy with morphologic and immunophenotypic features that resemble those of mature tissue histiocytes [1]. It commonly occurs in lymph nodes, and in extranodal sites including gastrointestinal tract [2], skin [3] and central nervous system [4]. The site of involvement in gastrointestinal tract includes stomach, ileum, colon, rectum and anus. Regardless of site, this tumor has an aggressive behavior and most often presents with a high stage of disease. In agreement with previous reports, the histiocytic sarcoma case presented here demonstrated nodal spread and liver metastasis at diagnosis. Stage of disease and possibly tumor size are considered significant prognostic indicators.

Histiocytic sarcoma in small bowel commonly manifests as polypoid tumors causing intestinal obstruction [5, 6]. Multiple ulcerations, some with fatal perforation, were found in the esophagus and duodenum of a patient with histiocytic sarcoma [7]. They showed only non-specific inflammatory changes without tumor cell involvement. The ulcers were probably derived from ischemic condition through an embolic process caused by tumor cell infiltration elsewhere in the blood vessels at the periphery of the ulcers. Our case presented as an ulcerated lesion infiltrating full thickness of the bowel wall without perforation. Although, the initial biopsy revealed acute ulceration and focal changes consistent with ischemia'. Like lymphomas of the small bowel, distal ileum seems to be a
preferential site for histiocytic sarcoma. Information on histiocytic sarcoma presenting in terminal ileum is mainly limited to scattered case reports in the clinical and pathology literature and is inconsistently reported in larger series [2, 5, and 6]. To the best of our knowledge, the detailed CT imaging features of histiocytic sarcoma involving the terminal ileum have not been previously described in English literature.

Histiocytic sarcoma shows homogenous hypoattenuation in CT images, not different from contrast enhancement patterns seen in bowel lymphomas. Features that help differentiate histiocytic sarcoma from adenocarcinoma include well-defined margins with preservation of fat planes, no invasion into adjacent structures, and perforation without desmoplastic reaction, which are also similar to lymphomas [8]. Despite the severe luminal narrowing, these lesions are less likely to cause obstruction because they do not elicit a desmoplastic response and submucosal infiltration weakens the muscularis propria of the wall. Lymphomas on the other hand show bulky lymphadenopathy and multifocal involvement, which is not common in histiocytic sarcoma.

Recognition of histiocytic sarcoma is important clinically and requires assessment of clinical, morphologic, and immunophenotypic features. Radiology plays an important role in early diagnosis of this rare malignancy especially at extranodal sites. When CT reveals minimal bowel wall thickening, without adenopathy, the diagnosis may be very difficult or may be confuse with various other gastrointestinal diseases, such as inflammatory bowel diseases. The rarity of histiocytic sarcoma among the general population also contribute to this diagnostic difficulty. However, histiocytic sarcoma should be included in the list of differential diagnosis at CT when ileum is a preferential site of intestinal involvement, there is mild segmental or circumferential thickening, preservation of fat planes, without obstruction or perforation.

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