Case Report

Interdigitating Dendritic Cell Sarcoma of the Small Intestine Presenting as Spontaneous Hemoperitoneum – A Rare Case Report

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INTRODUCTION

Dendritic cells are immune accessory cells derived from the hematopoietic stem cells. There are four types of dendritic cells present in lymph nodes: follicular, interdigitating, Langerhans, and fibroblastic/histolytic cells. Interdigitating cells are commonly located in the T-cell-rich areas of peripheral lymphoid tissue.[1] Interdigitating dendritic cell sarcoma (IDCS) is a rare neoplasm with only 100 cases reported in the English literature.[2,3] Although this tumor mostly arises in the lymph nodes, the extranodal sites such as spleen, small intestine, tonsil, nasopharynx, testis, and urinary tract are also noted.[1,2] To our knowledge, only few cases of the IDCS arising from the small intestine have been reported till date. Based on the findings from previous studies, patients with intestinal involvement tend to be presented with intestinal obstruction.[2,4] However, we report a first known case of IDCS of ileum presenting as spontaneous hemoperitoneum.

CASE REPORT

A 61-year-old male presented to the emergency department with complaints of lower abdominal pain, fever, and vomiting for 3 days and breathlessness for 1 day. There was no history of trauma. Medical history was significant for essential hypertension and diabetes mellitus which had been controlled with oral drugs. Family history was negative for malignancy. His physical examination revealed tenderness in the lower abdomen without any palpable abdominal mass. Bowel sounds were absent. Digital examination showed a normal fecal staining. The laboratory tests were normal except for anemia (4.5g/dl). Ultrasound abdomen revealed 15 cm × 7 cm heterogeneous collection with thick septations in the periumbilical region. Diagnostic peritoneal lavage revealed hemoperitoneum and gangrenous bowel. Emergency lapotomy was done and it revealed a friable mass arising from the ileum. Resection of 2 ft of the ileum and double-barrel ileostomy was done. Immunohistochemical analysis revealed a positivity of S100 and leukocyte common antigen). A diagnosis of IDCS was made. The patient had an uneventful postoperative recovery and planned for postoperative chemotherapy.

KEYWORDS: Immunohistochemistry, interdigitating dendritic cell sarcoma, spontaneous hemoperitoneum

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was done [Figure 2]. After adequate irrigation of the peritoneal cavity, the abdomen is closed by anatomical layers. Microscopically, the tumor cells were seen scattered in the subserosal aspect. Immunohistochemical analysis showed that the tumor cells were positive for leukocyte common antigen, CD 68, and S100 [Figure 3]. A diagnosis of the IDCS was made based on histological and immunohistochemical findings.

**Discussion**

Spontaneous hemoperitoneum is a rare but life-threatening complication that is defined as blood within the peritoneal cavity of nontraumatic etiology. It most commonly arises from hepatic, splenic, vascular, or gynecologic pathology, and usually in anticoagulated or coagulopathic patients.[5] Spontaneous hemoperitoneum caused by the rupture of tumors of the intestine is a rare entity.

Dendritic cell sarcomas should be differentiated from other histiocytic and dendritic neoplasms as well as from melanoma and fibroblastic reticular tumors. The diagnosis cannot be made with clinical and light microscopy alone. Immunohistochemistry helps in achieving the diagnosis. In our case, the tumor cells stained positive for S100, myeloid cell marker CD68, and the leukocyte common antigen which is consistent with the previous reports describing IDCS,[4,8] as well as negative for CD1a (LCS marker), CD3 (T-cell marker), CD20 (B-cell marker), HMB45 (melanoma marker), or cytokeratin (epithelial cell marker).

After a thorough review of the literature, we have found only eight cases of IDCS[4] involving the intestinal tract [Table 1]. The small intestine was the common site, and most of the cases were presented with intestinal obstruction. However, we report a first known case of spontaneous hemoperitoneum caused by the rupture of IDCS of ileum.

There are no standard treatment guidelines for the treatment of IDCS owing to its rarity. IDCS is treated with surgical excision, radiotherapy, chemotherapy, or a combination of these therapies. In localized IDCS, surgical resection is the main stay of the treatment. Adjuvant radiotherapy has shown to prolong disease survival rates in some studies.[6,7] Most of the advanced cases, which survived for several years, were treated with chemotherapy and/or radiation therapy.[4] Several chemotherapeutic agents have been tried, including cyclophosphamide, doxorubicin, vincristine, and prednisolone; doxorubicin, bleomycin, vinblastine, and dacarbazine; dexamethasone, cisplatin, and high-dose cytarabine; and ifosfamide, carboplatin, and etoposide.[6,7]

In our case, the patient was advised chemotherapy but he refused citing age as a factor. After 6-month follow-up, there was no recurrence and the patient was planned for the restoration of intestinal continuity.
Our case report describes the first known case of IDCS of the small intestine presenting as spontaneous hemoperitoneum. IDCS is a rare neoplasm, and extranodal IDCS is more uncommon than the nodal presentation. Besides, clinical picture and light microscopy, immunohistochemistry plays an important role in arriving at the diagnosis. Surgical resection is the mainstay of treatment for localized disease. The role of adjuvant, neoadjuvant chemotherapy, and radiotherapy in metastatic disease is still not clearly defined.

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Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/ her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.
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