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# Histiocytic Sarcoma of Rectum
## - A Case Report -

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We present a case of a 44 year-old man with histiocytic sarcoma arising from the rectum. Histiocytic sarcoma is a rare type of malignant lymphoma, comprising less than 0.5% of all non-Hodgkin's lymphomas (NHLs). In this case, the tumor was a polypoid mass, measuring 2.5 × 2 cm. On microscopic examination, it was composed of many bizarre multinucleated cells and sheets of xanthomatous histiocytes. There were also areas with spindle cells arranged in a storiform pattern. Extensive metastases to regional lymph nodes were found, and combined adjuvant chemotherapy was administered after complete resection.

**Key Words :** Sarcoma; Histiocytic lymphoma; Rectum

Histiocytic sarcoma is a rare hematopoietic malignancy characterized by histiocytic differentiation.[1] It comprises less than 0.5% of all non-Hodgkin's lymphomas (NHLs). The age range of patients is wide, with a mean age of 44 years. The most common extranodal sites are the skin, gastrointestinal (GI) tract, and soft tissues. We present a case of histiocytic sarcoma of the rectum, to our knowledge the second report of gastrointestinal histiocytic sarcoma in the Korean literature.

## CASE REPORT

A 44 year-old man with hematochezia and anal bleeding visited the Chungnam National University Hospital on 21 April 2003. On digital rectal examination, a mass was palpated 4cm above the anal verge. Colonoscopy revealed a polypoid tumor, measuring about 2 cm in size. It had an irregular surface with an area of mucosal ulceration (Fig. 1). A preoperative abdominal computed tomography (CT) scan showed a focal thickening of the lateral wall of the lower rectum, with submucosal extension and multiple enlarged lymph nodes (Fig. 2). The preoperative histopathologic diagnosis was 'consistent with malignant fibrous histiocytoma'. An operation was performed on 14 May 2003. There were adhesions around the rectum in the pelvis. Low anterior resection and wide en bloc lymph node dissection were performed.

The resected specimen consisted of a 43 cm segment of the large intestine with attached mesentery. The tumor was a polypoid mass, measuring 2.5 × 2 cm in size and protruded into the lumen. Overlying mucosal surface was partially eroded. The gross sections showed an intramural tumor. It was relatively well-circumscribed, solid, homogeneous, and yellowish to white in color. There were many enlarged lymph nodes in the resected specimen.

Microscopic examination showed a submucosal malignant tumor (Fig. 3). The overlying mucosa was infiltrated and partially destroyed by the tumor. The tumor was composed of many bizarre multinucleated cells and sheets of xanthomatous histiocytes. There were also areas of spindle cells arranged in a storiform pattern (Fig. 4). There were interspersed inflammatory cells, such as lymphocytes and plasma cells. The initial diagnosis was malignant fibrous histiocytoma. The tumor cells were

positive for leukocyte common antigen (LCA), CD45RO, CD68, CD31, and lysozyme, but were negative for CD34, smooth muscle actin (SMA), c-kit (CD117), desmin, CAM5.2, AE1/AE3, S100 protein, CD30, and melan A (Fig. 5). All of the regional lymph nodes and many retroperitoneal lymph nodes were involved. The final diagnosis was histiocytic sarcoma.

The patient was discharged 13 days after the operation. He was treated with postoperative adjuvant chemotherapy based on the misdiagnosis of malignant fibrous histiocytoma. Ifosfamide, mesna, doxorubicin, and dacarbazine were used. He received the same therapeutic regimen four times. Each treatment lasted 5 days, with an interval of 21 days. A postoperative CT was performed about 4 months after the operation. No evidence of metastasis was found in the thorax and upper abdomen. The patient also received a radiotherapy to the lower abdomen several times following the completion of the chemotherapy. The patient has been alive for 32 months since the operation without evidence of recurrence or metastasis.

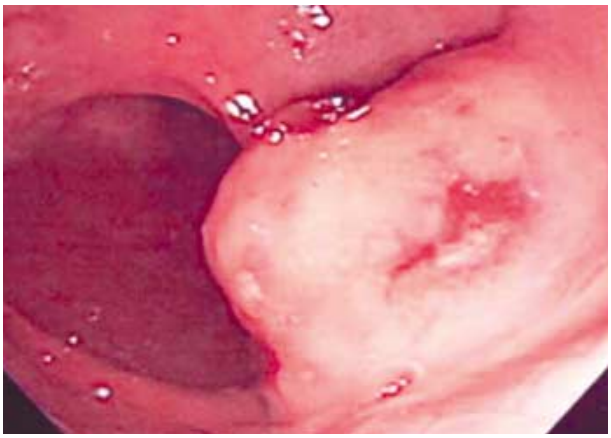

Fig. 1. The tumor is polypoid with mucosal erosion by colonoscopy.

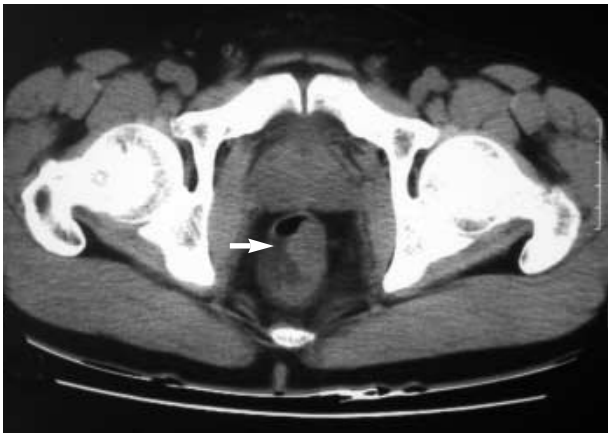

Fig. 2. Pelvic in CT scan, shows that the lateral wall of the lower rectum is focally thickened with submucosal extension (arrow).

## DISCUSSION

Histiocytic sarcoma is a rare malignant tumor of histiocytes that occurs in lymph nodes, skin, gastrointestinal tract, and soft tissues.[1] Misdiagnoses are common in extranodal lesions. Hornick *et al*.[2] reported that the diagnosis of histiocytic sarcoma was suggested by the referring pathologist in only 4 of 14 cases of extranodal histiocytic sarcomas. The referring diagnoses in the others were: malignant fibrous histiocytoma (3 cases); high-grade sarcoma, not otherwise specified (2 cases); epithelioid GI stromal sarcoma; epithelioid hemangioendothelioma or epithelioid sarcoma; and an inflammatory process. Our initial diagnosis was also malignant fibrous histiocytoma. The initial immunohistochemical study omitted a stain for lysozyme since we had leaned toward a diagnosis of malignant fibrous histiocytoma on the ground of any morphologic features. Another immunohistochemical studies, including for lysozyme, were performed at the Brigham and Women's Hospital, Boston, MA, and the final diagnosis of histiocytic sarcoma was made about one year later. In retrospect, the authors now note the fact that the tumor cells of malignant fibrous histiocytoma do not express CD68 and CD45,[3] which were found in our case.

The typical neoplastic cells of histiocytic sarcoma resemble those of large cell or anaplastic large cell lymphoma, but the cytoplasm is typically eosinophilic and voluminous.[1] In practice, however, the microscopic appearance is variable, so an immunohistochemical workup is required for confirmation. In this context, Hornick *et al*.[2] have said that the reports published more

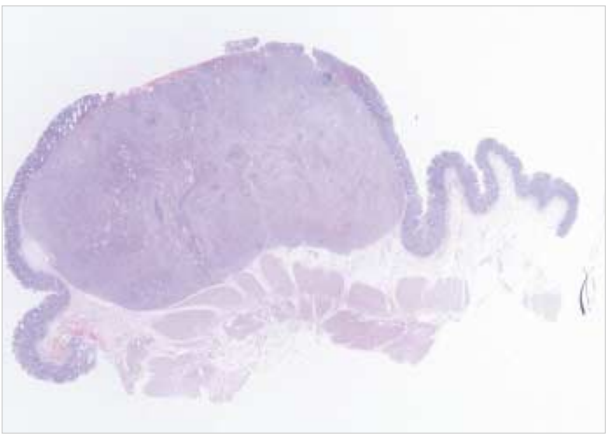

Fig. 3. The tumor is submucosal in location and homogeneous.

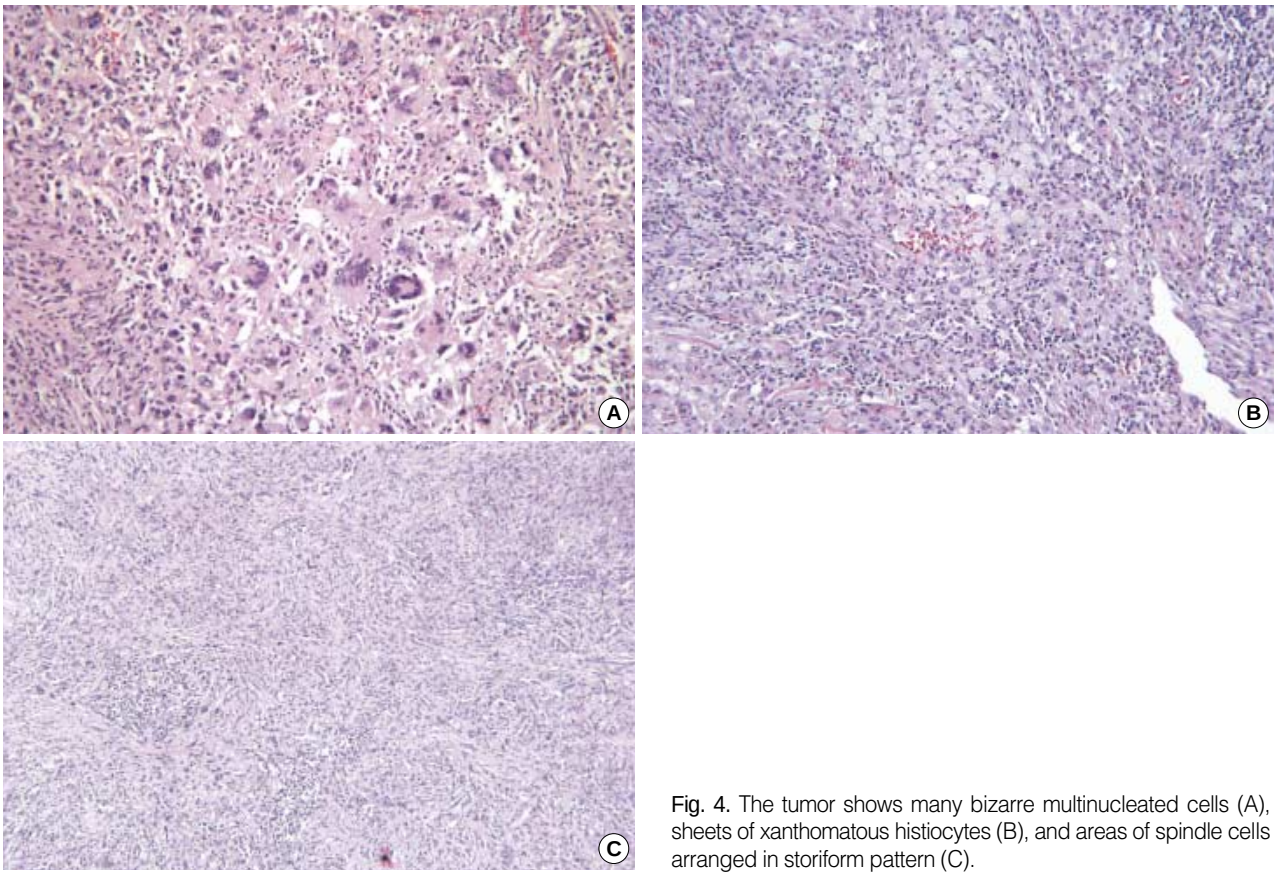


Fig. 4. The tumor shows many bizarre multinucleated cells (A), sheets of xanthomatous histiocytes (B), and areas of spindle cells arranged in storiform pattern (C).

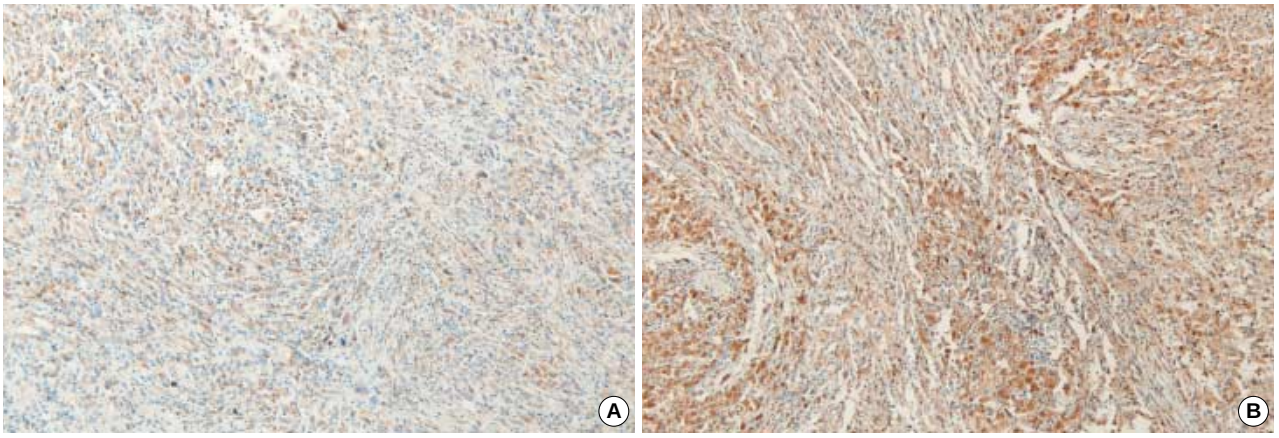


Fig. 5. The tumor cells are positive for CD68 (A) and lysozyme (B).

than 15 years ago are difficult to interpret.

There is only one report of a GI histiocytic sarcoma in the Korean literature. Jung *et al*.[4] reviewed 71 cases of GI lymphomas and identified 4 as being of a true histiocytic type. These four cases of true histiocytic lymphoma were positive for alpha-1-antichymotrypsin and for lysozyme. An immunohistochemical stain for CD68 was not performed. Involved sites were the stomach (1 case), small intestine (2 cases), and large intestine (1 case). Other locations of extranodal histiocytic sarcoma reported in the Korean literature have been the liver and the bone marrow.[5,6] The former occurred in a 55 years-old male and the latter in an 18 years-old male.

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