

# Extranodal Histiocytic Sarcoma Rectum: A Case Report

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## Abstract

Histiocytic sarcoma (HS) is an exceedingly rare but aggressive hematopoietic tumor, showing morphologic and immunophenotypic features of mature tissue histiocytes. Diagnosis of HS is based on histological and immunohistochemical evidence of histiocytic differentiation supported by an extensive immunophenotypic analysis that excludes other large cell malignancies in the differential diagnosis. Search of world literature revealed only three previous reports of extranodal histiocytic sarcoma (ENHS) involving rectum specifically. We report a case of ENHS of rectum in a 65 year old man presenting with hematochezia. Radiological findings were indicative of polypoidal soft tissue mass lesion in rectum. Histopathology and immunohistochemical studies of the resected specimen confirmed the uncommon diagnosis ENHS.

**Keywords:** Histiocytic sarcoma, extranodal histiocytic sarcoma, magnetic resonance imaging

## Introduction

Histiocytic Sarcoma (HS) is a malignant proliferation of cells showing morphological and immunophenotypic features of mature tissue histiocytes.<sup>[1]</sup> Lymph nodes are the most common site of presentation, although a variety of extranodal sites may be affected, particularly the gastrointestinal tract, spleen, soft tissue and skin. Other sites of involvement include head and neck regions, salivary gland, lung, mediastinum, breast, liver, pancreas, kidney, uterus, central nervous system, bone and bone marrow.<sup>[1,2]</sup> Search of world literature revealed eight previous reports of extranodal histiocytic sarcoma (ENHS) involving the large intestine and only

three previous reports of ENHS involving rectum specifically (Table 1).<sup>[3-7]</sup> We report here a case of ENHS of rectum with typical morphological and immunophenotypic features.

**Table 1: Case reports showing demographic profile and treatment with outcomes of extra nodal histiocytic sarcoma (ENHS) involving the rectum.**

Reference	Age/sex	Tumor location	Treatment	Metastasis	Survival
Hornic <i>et al</i> <sup>[3]</sup> 2004	40/Female	Rectum	Surgical + Chemotherapy	No	Alive-21 months
	27/Female	Rectum	Surgical + Chemotherapy	Yes	Alive-10 years
Park <i>et al</i> <sup>[5]</sup> 2006	44/Male	Rectum	Surgical + Chemotherapy+ Radiotherapy	No	Alive-32 months
This Report	65/Male	Rectum	Surgery + Chemotherapy	No	Alive

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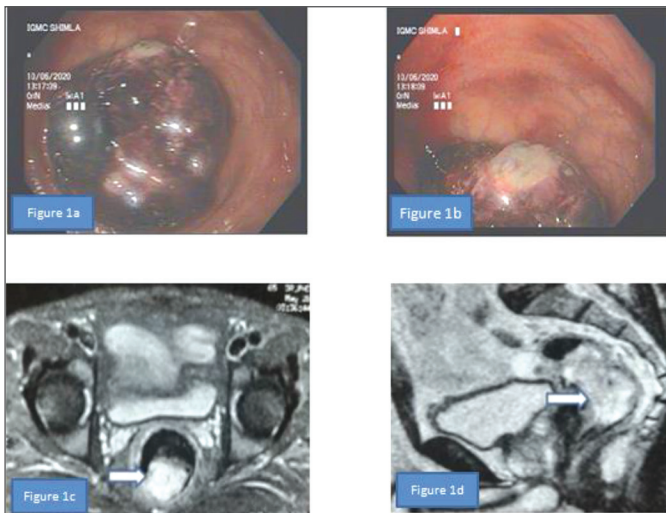
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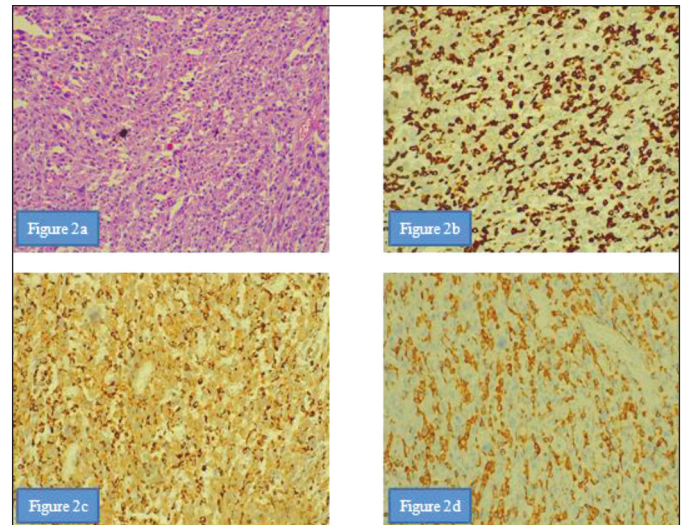
## Case Report

A 65 year old male presented to our gastroenterology outpatient department with complaints of bleeding from rectum for the past 2 months. There was history of feeling of incomplete evacuation with stool frequency of 4-5 per day. Stool was semisolid in consistency and contained fresh and altered blood mixed with stool. There was no history of fever or weight loss. The patient was a smoker and occasionally consumes alcohol. On digital rectal examination, a mass was palpated 6 cm above the anal verge. Colonoscopy revealed a polypoid tumor, measuring about 4 x 4 cm in size. It had an irregular reddish vascular surface, with areas of mucosal ulcerations (Figure. 1a, 1b). A preoperative abdomino-pelvic magnetic resonance imaging (MRI) scan showed heterogeneously enhancing intraluminal polypoid soft tissue mass lesion in rectum involving all layers up to serosa with associated fat stranding in mesorectum (Figure. 1c,1d). The presacral space and pelvic walls were normal. The preoperative histopathologic diagnosis was 'consistent with malignant melanoma'. An operation was performed after discussion with surgeons. 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 19 cm segment of the large intestine (rectum and adjacent sigmoid colon) with attached mesentery and lymph nodes. The tumor was a grey brown polypoidal mass, measuring 4x3x2 cm in size and protruded into the lumen.



**Figure 1a:** Colonoscopic image showing polypoidal vascular rectal mass lesion compromising lumen; **Figure 1b:** rectal mass showing surface ulceration; **Figure 1c,1d:** MRI axial and sagittal image showing heterogeneously enhancing intraluminal polypoidal soft tissue mass in rectum with associated fat stranding in mesorectum.



**Figure 2a:** Histopathological image of rectum showing large pleomorphic cells with abundant eosinophilic cytoplasm; **Figure 2b:** shows neoplastic cells positive for CD163; **Figure 2c:** shows neoplastic cells positive for CD68; **Figure 2d:** shows neoplastic cells positive for CD4.

Overlying mucosal surface was partially eroded. The tumor was 11 cm away from proximal resected margin and 2 cm away from distal resected margin. The detailed histopathological and immunohistochemistry examination of the resected specimen was suggestive of histiocytic sarcoma of the rectum and resected margins were free of malignancy. The neoplastic cells were immunoreactive for CD4, CD68, CD163, CD31 and vimentin suggestive of histiocytic sarcoma (Figure 2a, b, c, and d).

Neoplastic cells were non-immunoreactive for S100, HMB-45, Melan-A and SOX-10 excluding melanoma, which was initially suggested on colonoscopic biopsy histopathological examination. The patient was discharged 11 days after the operation. He was attached to oncology department for postoperative adjuvant chemotherapy and further management.

## Discussion

HS is an exceedingly rare but aggressive hematopoietic tumor of mature histiocytes, accounting for less than 1% of all hematolymphoid neoplasms.<sup>[1]</sup> It can be present as a primary malignancy or, less commonly, a secondary malignancy. There are multiple reports of presumed transdifferentiation from low-grade B-cell lymphoma to histiocytic sarcoma; this accounts for approximately one-fourth of cases.<sup>[2]</sup> The median age at diagnosis is 51 years (range, 1–89 years), with a slight male predominance, our patient was 65-years old male.<sup>[8]</sup> Histiocytic sarcoma commonly presents as a painless solitary mass at an extranodal site and ma-

ny patients have systemic (B) symptoms such as fever, night sweats, or weight loss.<sup>[2]</sup> Search of world literature revealed eight previous reports of ENHS involving the large intestine and only three previous reports of ENHS involving rectum specifically (Table 1).<sup>[3,7]</sup>

Misdiagnoses are common in extranodal lesions. Hornick *et al.* reported that the diagnosis of histiocytic sarcoma was suggested by the referring pathologist in only 4 of 14 cases of extranodal histiocytic sarcomas.<sup>[3]</sup> Our initial diagnosis was also malignant melanoma. We did not perform immunohistochemical study on the initial colonoscopic biopsy specimen and diagnosis of malignant melanoma was considered only on the grounds of morphologic features alone.

Histologically, the tumor shows diffuse infiltration of large, round to ovoid pleomorphic cells with round to oval often eccentrically placed nuclei and abundant eosinophilic often foamy, vacuolated or clear cytoplasm.<sup>[1,2,7]</sup> Occasional bizarre cells with pleomorphic hyperchromatic nuclei and coarse chromatin are identified. Mitotic activity is a consistent finding, but varies in degree from case to case. A variable number of reactive cells including small lymphocytes, plasma cells, benign histiocytes, neutrophils and eosinophils may be seen. In practice, however, the microscopic appearance is variable, so an immunohistochemical workup is required for confirmation.

The diagnosis is based on histological and immunohistochemical evidence of histiocytic differentiation supported by an extensive immunophenotypic analysis that excludes other large cell malignancies in the differential diagnosis.<sup>[1]</sup> The differential diagnosis includes reactive histiocytic proliferations, dendritic cell neoplasm, large cell non-Hodgkin lymphoma, especially anaplastic large cell lymphoma and diffuse large cell lymphoma, malignant melanoma, undifferentiated large cell carcinoma and monocytic leukemia.<sup>[1,2]</sup> By definition in HS, there is the expression of one or more histiocytic markers, including CD163, CD68 (KP1 and PGM1) and lysozyme, with typical absence of B-cell- and T-cell related markers, and Langerhans cell (CD1a, langerin/ CD207), follicular dendritic cell (CD21, CD23, CD35, CAN. 42), epithelial (pancytokeratin, EMA), melanocytic (HMB45, Melan A) and myeloid cell (CD13, CD33, myeloperoxidase) markers.<sup>[1,2]</sup> CD163, a new immunohistochemical marker of monocytes and histiocytes, is more specific than other macrophage/histiocytic markers such as CD68 and may have significant diagnostic utility.<sup>[1,2]</sup>

HS most often presents at an advanced clinical stage, with limited response to chemotherapy and a

high mortality. There is no consensus on the prognostic factors and the standard treatment approach due to the rarity of the disease. The most popular therapy for advanced histiocytic sarcoma appears to be cyclophosphamide, hydroxydaunorubicin, oncovin, and prednisone.<sup>[2]</sup> The median overall survival for patients who receive adjuvant or neoadjuvant therapy is approximately 2.5 years, but there is no clear survival advantage.

## Conclusion

Histiocytic sarcoma is a rare histiocytic neoplasm with broad morphologic differentials including histiocytic/dendritic cell neoplasms, myeloid neoplasms, lymphomas, carcinoma, and melanoma. The knowledge of key morphologic and immunohistochemical features allows for accurate classification and management.

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