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Histiocytic Sarcoma of Colon - A Rare Entity

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ABSTRACT

INTRODUCTION: Histiocytic sarcoma (HS) is a very rare malignant neoplasm of unknown etiology which is not a true sarcoma. It shows similar morphological and immunohistochemical features of mature histiocytes. Most cases involve the extra nodal sites, most commonly the skin, soft tissue, intestinal tract. The clinical presentation of HS is related to the organs involved. HS is an aggressive neoplasm with poor prognosis and lack of standard treatment guidelines.

CASE REPORT: We are presenting a case of HS of sigmoid colon in a 33-year-old male patient. The patient had a 3 month history of abdominal pain, bloating and intermittent constipation associated with several episodes of hematochezia. He underwent low anterior resection with wide en bloc lymph node dissection. Detailed histopathological and Immunohistochemical examination of resected specimen led to the diagnosis of Histiocytic sarcoma of sigmoid colon.

CONCLUSION: HS is a rare malignant neoplasm with histiocytic differentiation. Patients with localized resectable disease treated with surgery with or without adjuvant radiotherapy while patients with multifocal disease are treated with combination systemic chemotherapy regimens.

Keywords: contrast enhanced computed tomography, dendritic cell neoplasm, hematochezia, Histiocytic sarcoma, histiocytic proliferation, melanoma, monocytic leukemia, non Hodgkin lymphoma, undifferentiated large cell carcinoma.

INTRODUCTION

Histiocytic sarcoma (HS) is a very rare malignant neoplasm of unknown etiology. It is not a true sarcoma and is named so because it shows similar morphological and immunohistochemical features of mature tissue histiocytes. It shows differentiation towards macrophage lineage and may occur with or without a component of Non-Hodgkin Lymphoma [1]. It is commonly diagnosed in the fourth and fifth decades of life. Males are slightly more commonly affected than females. HS can affect any organ system and clinical presentation is related to the organs involved. Majority of the cases involve the extra nodal sites, most commonly the skin, soft tissue and intestinal tract [2,3,4]. HS has an aggressive clinical course and often involves multiple systems. Localized disease is usually treated with surgery with or without adjuvant radiotherapy while disseminated disease is

treated with chemotherapy. Due to its aggressive nature, poor prognosis and lack of standard treatment guidelines, its management becomes a challenge to the clinicians.

CASE REPORT

A 33-year-old male presented history of abdominal pain, bloating and intermittent constipation for the period of three months. He also had several episodes of hematochezia. There was no history of fever or significant weight loss. The patient was a smoker and occasionally consumed alcohol.

Contrast enhanced computed tomography (CE-CT) scan of whole abdomen was done which revealed an enhancing broad based polypoidal lesion measuring 4.5 X 4.3cm with surface ulceration in the distal part of the sigmoid colon with luminal narrowing. (Fig.2) He underwent low anterior resection (LAR) with wide en bloc lymph node dissection and the specimen were sent to the department of Pathology for histopathological examination (HPE).

The resected specimen consisted of 18cm long segment of large intestine (sigmoid colon and adjacent rectum) with attached mesentery and lymph nodes. The tumor was a grey brown polypoidal mass measuring 4.5 x 4 x 3 cm in size protruding into the lumen. Overlying mucosal surface showed erosions and ulcerations. Cut section of the tumor was greyish white with areas of haemorrhage and necrosis. (Fig.1) HPE of the tumor revealed transmural infiltration of colonic tissue by poorly cohesive large cells with pleomorphic nuclei, abundant eosinophilic often foamy or vacuolated cytoplasm resembling mature histiocytes. The background showed mixed inflammatory cells composed of variable numbers of reactive lymphocytes, plasma cells, benign histiocytes and eosinophils. (Fig. 3-5). There were numerous abnormal mitotic figures.

Tumor necrosis was present. The sections from the sigmoid colon shows infiltration by the similar types of cells (Fig. 6-9). The accompanying lymph nodes showed histological features of reactive hyperplasia.

On immunohistochemistry (IHC), the tumor cells were found to be diffusely positive for CD 163 (Fig.10), CD 68 (Fig.11) and CD 45 (Fig.12), indicating histiocytic differentiation and hemato-lymphoid origin. Immunostaining with epithelial markers (pancytokertain), lymphoid markers (CD3, CD20, CD 15, CD 30), neuroendocrine markers (synaptophysin chromogranin) were negative. The melanoma markers (HMB-45, Melan-A, S100) were all negative. So the diagnosis of HS of sigmoid colon was made. The patient was discharged after 10 days and referred to the Oncology department. He is now under monthly follow up. Six months have been passed. He is now doing well.

DISCUSSION

HS is an extremely rare malignant tumor of macrophage-dendritic cell lineage [5]. Although its pathogenesis is still unknown, few cases of HS are reported to be arising by trans differentiation from low-grade B cell lymphoma or after chemotherapy from mediastinal or gonadal germ cell tumors [6,7]. Majority of the cases of HS occur in adults with a male predominance [2], as seen in our case. HS usually involves the gastrointestinal tract, soft tissue, skin, spleen, liver rarely the lymph node or brain [2,4,8,9].

In our case the sigmoid colon was involved. Clinical presentation of HS ranges from localized disease to widely disseminated disease [2,4]. In the present case, it was a localized HS involving the sigmoid colon.

On histological examination, HS shows diffuse infiltration of large round to ovoid pleomorphic cells with round to oval eccentrically placed nuclei with abundant eosinophilic often foamy, vacuolated or clear cytoplasm [10.11,12]. Mitotic activity including atypical mitotic figures is a consistent finding in HS. A variable number of reactive cells including small lymphocytes, plasma cells, benign histiocytes, neutrophils and eosinophils are

seen in the background [10-12]. We found similar histological features in our case.

The diagnosis of HS requires histological and immunohistochemical evidence of histiocytic differentiation supported by an extensive immunophenotypic workup that excludes other large cell malignancies which comes in the differential diagnosis (DD) [10]. The DD of HS includes reactive histiocytic proliferation, dendritic cell neoplasm, large cell NHLs especially anaplastic large cell lymphoma and diffuse large cell lymphoma, malignant melanoma, undifferentiated large cell carcinoma and monocytic leukemia. HS expresses one or more histiocytic markers including CD 163, CD 68 and lysozyme with typical absence of B-cell and T-cell related markers, Langerhans cell markers (CD 1a, langerin), follicular dendritic cell (CD 21, CD23, CD 35 CAN.42), epithelial (pan cytokeratin, EMA), melanocytic (HMB-45, Melan-A) and myeloid cell markers (CD 13, CD 33, myeloperoxidase) [10.11]. In our case the tumor cells were diffusely positive for CD 45, CD 68 and CD 163 while negative for B-cell and T-cell markers, epithelial, melanocytic and neuroendocrine markers.

The gold standard treatment of HS is surgical resection with wide margins. Other treatment options include chemotherapy and radiotherapy in case of incomplete tumor resection [13]. In our case the patient underwent complete surgical resection of the tumor in terms of LAR with wide end bloc lymph node dissection. The site of the tumor, the tumor size and the stage of the disease are considered to be the most important factors in the prognosis [9].

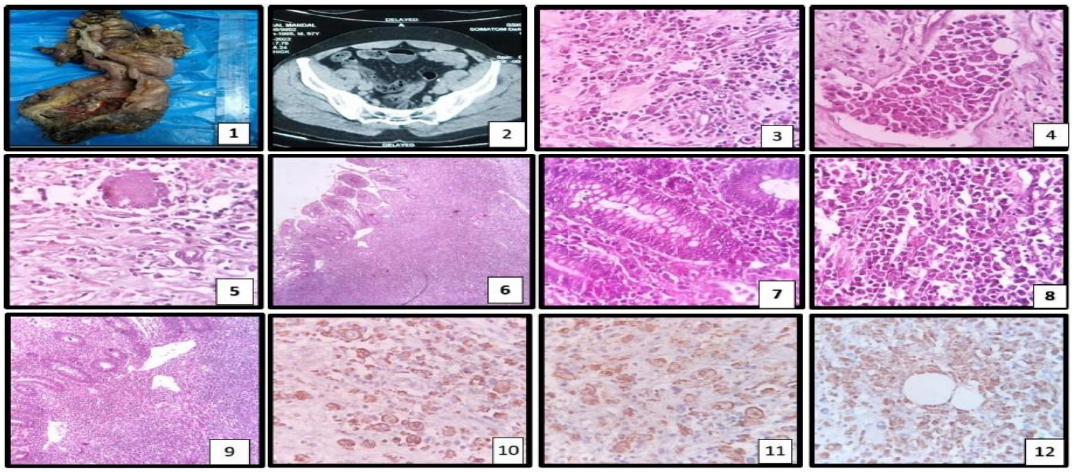

Figure 1: Gross picture of HS of sigmoid colon showing grey brown mass with surface erosions and ulcerations.
Figure 2: CE-CT whole abdomen showing enhancing broad based mass with luminal narrowing.
Figure 3: H&E (400X) HS showing poorly cohesive large cells with pleomorphic nuclei abundant eosinophilic to foamy cytoplasm.
Figure 4: H&E (400X) HS showing cluster of cells resembling tissue histiocytes.
Figure 5: H&E (400X) HS showing tumor cells with histiocytic differentiation admixed with mixed inflammatory cells and giant cell.
Figure 6: H&E (40X) HS of sigmoid colon showing sheets of tumor cells admixed with mixed inflammatory cells.
Figure 7: H&E (400X) HS of sigmoid colon showing tumor cells admixed with mixed inflammatory cells.
Figure 8: H&E (400X) HS showing histiocytes admixed with reactive lymphocytes, plasma cells, eosinophils.
Figure 9: H&E (100X) HS of sigmoid colon showing tumor cells admixed with mixed inflammatory cells.
Figure 10: H&E (400X) HS of sigmoid colon showing positive expression of CD 163 in the tumor cells.
Figure 11: IHC (400X) Positive expression of CD 68 in the tumor cells in HS of sigmoid colon.
Figure 12: IHC (400X) Positive expression of CD 45 in the tumor cells in HS of sigmoid colon.

CONCLUSION

HS is a rare histiocytic neoplasm with no standard treatment regimen. Due to its rarity and histologic overlap with diverse mimics, the recognition of the histological features and proper immunohistochemical analysis is crucial for the diagnosis. Patients with localized resectable disease treated with surgery with or without adjuvant radiotherapy, while patients with multifocal disease are treated with combination systemic chemotherapy regimens. Close follow up is recommended for such cases.

Declaration by Authors

Consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Conflict Of Interest: The authors declare no conflict of interest.

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