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Histiocytic Sarcoma of Colon Mimicking Lymphoma: A Diagnostic Challenge and a Rare Case Report

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Abstract **Case Report**

We received a specimen of right hemicolectomy in a case of 35 years male, presented with persistent abdominal pain for last 6 months. Patient was admitted with sevre abdominal pain and frequent episodes of haematochezia for last 15 days. On gross examination of right hemicolectomy specimen, no obvious growth was found, only thickening of caecal wall was noted. Microscopical features showed atypical discohesive cells involving muscle fibers, serosal fat and subserosal lymph node. These discohesive cells having pleomorphic nuclei, nuclear grooving, prominent nucleoli surrounded by foamy to eosinophilic cytoplasm. Coagulative tumor necrosis was also present in the wall of the colon. Morphologically our initial diagnosis was non-Hodgkin's lymphoma or poorly differentiated carcinoma or neuroendocrine tumor. Diagnosis of Histiocytic Sarcoma was confirmed by the immunohistochemistry study using CK, Chromogranin, Synaptophysin, CD20, CD3, S100, CD 45 and CD 68. In our case the tumor cells were diffusely positive for CD45 and CD 68. Tumor cells were CK, Chromogranin, Synaptophysin, CD20, CD3, and S100 negative.

Keywords: Histiocytic Sarcoma, Colon, non-Hodgkin's lymphoma, Diagnostic dilemma, CD 45, CD68.

INTRODUCTION

Histiocytic sarcoma [HS] of colon is a very rare malignant neoplasm originating from histiocytes. It is not a true sarcoma; it is named HS because the neoplastic cells have a morphological & immunophenotypic resemblance to mature tissue histiocytes [1]. The prevalence rate is 1 in 1 million. It most commonly occurs in 30 to 50 years age group with a male predominance [2]. Most common affected sites are extranodal sites like skin, soft tissue and intestinal tract [3,4,5] It causes diagnostic dilemma because morphologically it mimics lymphoma especially large B-cell lymphoma and anaplastic large cell lymphoma [6].

CASE REPORT

We received a specimen of right hemicolectomy in a case of 35 years male, presented with persistent abdominal pain for last 6 months. Patient was admitted with severe abdominal pain and frequent episodes of haematochezia for last 15 days. On gross examination of right hemicolectomy specimen, no obvious growth was found, only thickening of caecal wall was noted. Microscopical features showed atypical discohesive cells involving muscle fibers, serosal fat and subserosal lymph node. These discohesive cells having pleomorphic nuclei, nuclear grooving, prominent nucleoli surrounded by foamy to eosinophilic cytoplasm. Coagulative tumor necrosis was also present in the wall of the colon. Morphologically our initial diagnosis was non-Hodgkin's lymphoma or poorly differentiated carcinoma or neuroendocrine tumor. Diagnosis of Histiocytic Sarcoma was confirmed by the immunohistochemistry study using CK, Chromogranin, Synaptophysin, CD20, CD3, S100, CD 45 and CD 68. In our case the tumor cells were diffusely positive for CD45 and CD 68. Tumor cells were CK, Chromogranin, Synaptophysin, CD20, CD3, and S100 negative.

DISCUSSION

Histiocytic Sarcoma is not a true sarcoma as the neoplastic cells have a morphological & immunophenotypic resemblance to mature tissue histiocytes and tumor of macrophages- dendritic cell lineage [7]. It is a extremely rare malignant aggressive disease that generally appears in adults and leads to symptoms of B cell lymphadenopathy, hepatosplenomegaly and peripheral blood cytopenias. Lymph nodes may show expansion of sinuses by

neoplastic histiocytes that spare follicles or islands of lymphoid tissue. Tumor cells display varying degrees of pleomorphism and often have reniform nuclei and abundant eosinophilic cytoplasm. Tumor giant cells may be seen. Necrosis is common and erythrophagocytosis may be present [8]. Rare cases of HS have been shown to result from trans differentiation of a preexisting B-cell neoplasm. Most frequent genomic alterations are mutations in the mitogen activated protein kinase (MAPK) pathway, including KRAS, NRAS, BRAF, MAP2K1, PTPN11, NF1 and CBL. Mutations in tumor suppressors gene such as CDKN2A and TP53 are common. Less usual alterations include mutations in the phosphoinositide 3-kinase (PI3K) pathway [such as PTEN, MTOR, PIK3R1, and PIK3CA], CSF1R mutations and rearrangements involving BRAF and NTRK1 [1,8,9].

Diagnosis of HS is difficult only by microscopical examination as its histologic overlap with diverse mimics. The Differential Diagnosis of HS lymphoma [especially large B-cell lymphoma and anaplastic large cell lymphoma] is most common. Neuroendocrine tumor, Langerhans cell sarcoma, follicular dendritic cell sarcoma, melanoma, poorly differentiated carcinoma and undifferentiated pleomorphic sarcoma are also important differential diagnosis. Morphological examination and proper immunohistochemical study are crucial for diagnosis [10]. Wide margin surgical resection is the mainstay of treatment. Post operative radiotherapy and chemotherapy are other treatment modalities especially in case of incomplete resection of tumour and multiple system involvement. The prognosis for disseminated HS is generally poor, with a median survival of several months [11,12].

CONCLUSION

Histiocytic Sarcoma of colon is a rare aggressive malignant neoplasm. Multi-systemic disease often requires chemotherapy, but the optimal treatment regimen is not well-defined due to the rarity of the disease. It shows differentiation towards macrophages lineage and with or without a component of non-Hodgkin lymphoma. Due to its aggressive behavior, it often involves multiple systems. As histopathological features mimics with lymphoma, very often diagnostic dilemma occur. Therefore, immunohistochemical study plays a very important role to differentiate Histiocytic Sarcoma of colon from colorectal Lymphoma.

We conclude our diagnosis of Histiocytic Sarcoma of colon by history, clinical examination, gross and histopathological examination. And finally, we confirmed our diagnosis of Histiocytic Sarcoma of colon by immunohistochemical study of positive staining of CD45, CD68 and negative staining of CK, Chromogranin, Synaptophysin, CD20, CD3, S100.

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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