Case Report

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Pathology Section

A Unique Presentation of Primary Intestinal MALT Lymphoma as Multiple Lymphomatous Polyposis

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ABSTRACT

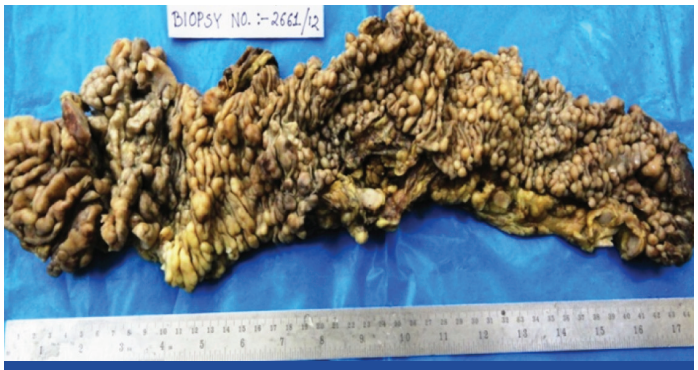
Multiple lymphomatous polyposis is considered to be a rare condition, with most of the cases being extranodal counterpart of mantle cell lymphomas. We report a rare case of multiple lymphomatous polyposis of the gastrointestinal tract in which the patient presented with abdominal pain and bloody diarrhea. Computer tomography of the abdomen showed circumferential wall thickening with intramural mass involving caecum & ascending colon with enlarged pericolonic lymph nodes. The patient underwent right hemicolectomy. Immunohistologic findings were characteristic of MALT lymphoma. Microscopic examination of polypoidal masses and mesenteric lymph nodes revealed infiltration by pleomorphic, atypical lymphoid cells which were CD20 positive and negative for CD3, CD10, Cyclin D1. Lymphoepithelial lesions were also noted. Careful endoscopic evaluation and histopathological review along with an immunohistochemical panel is extremely useful for accurately diagnosing such cases and avoiding unnecessary surgery and inappropriate therapy.

Keywords: CD 20 positivity, Lymphoepithelial lesion

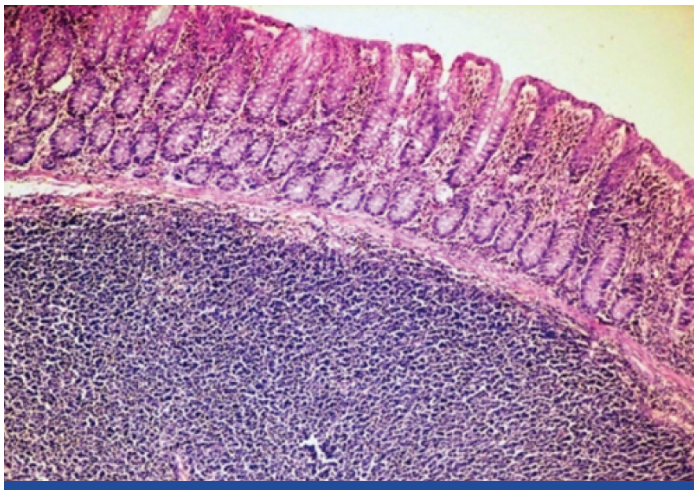
CASE REPORT

A 68-year-old male was admitted to Surgery department of the college with complaints of easy fatigability, pain abdomen & diarrhea since one year. He also complained of blood in stool, on and off, for last three months. His general physical and systemic examination & laboratory findings were normal except for an elevated ESR (=90 mm at the end of first hour) and anemia (hemoglobin 9.5 gm %). Chest X –ray was within normal limits. Colonoscopic biopsy was done at a different clinic, six months prior to his admission, and the microscopic examination showed “ulcerated colonic mucosa with granulation tissue, heavy mononuclear cell infiltrate and no evidence of malignancy.” Subsequently, computer tomography of the abdomen was advised. It revealed circumferential wall thickening with intramural mass involving caecum & ascending colon along with multiple enlarged pericolonic lymph nodes. There was no hepatosplenomegaly. A provisional diagnosis of colonic carcinoma was made and the patient underwent right hemicolectomy.

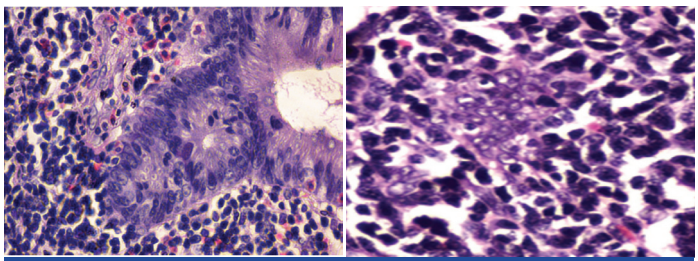
The resected specimen received in our pathology department was that of a colonic segment measuring 45cms in length, with the mucosal aspect being studded with a multitude of sessile polyps of varying sizes, largest measuring 3.5 x 2.5 x 1.8 cm [Table/Fig-1].



[Table/Fig-1]: Hemicolectomy specimen showing myriads of polyps and thickened mucosal folds.

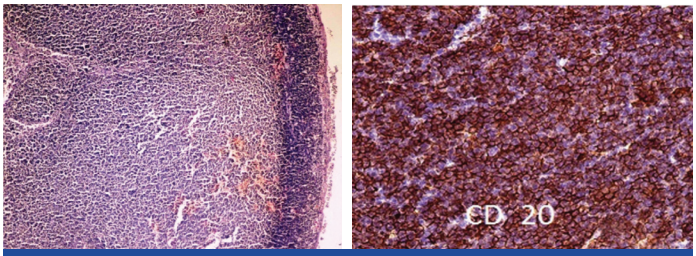


[Table/Fig-2]: Sections of polyp showing mucosal & submucosal infiltration with centrocyte-like cells (H&E, 40X).

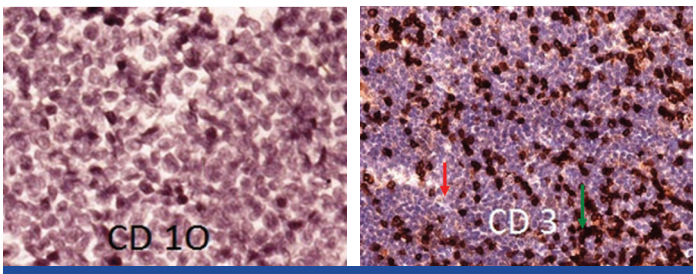


[Table/Fig-3]: (a):Lymphoepithelial lesion: infiltration of glandular epithelium with neoplastic lymphocytes (H&E, 200X). (b): Glandular destruction by centrocyte - like cells (H&E, 400X).

The largest polyps was situated in the ceacal area. These polyps were firm and homogenous grey-white on cut-surface. Mucosal folds were thickened at places. Multiple mesenteric lymph nodes were found enlarged, varying in diameter from 0.5 to 4 cm. A total of 32 lymph nodes were retrieved which were also solid grey-white on cut section. Appendix was measuring 4.5 cm and appeared normal grossly.



[Table/Fig-4]: Lymph node involvement by neoplastic lymphocytes (H&E, 40X).
[Table/Fig-5]: Neoplastic lymphoid cells show CD 20 positivity (200X).



[Table/Fig-6]: CD 10 negativity of the neoplastic lymphoid cells (400X).
[Table/Fig-7]: Neoplastic lymphoid cells are CD 3 negative (red arrow). The interspersed native benign T-cells show positivity for CD 3 (green arrow) (200X).

Microscopic examination of the cross-sections of the polypoidal masses revealed diffuse sheet-like infiltration of lamina propria, muscularis mucosae and submucosa by atypical lymphoid cells [Table/Fig-2]. The neoplastic cells were predominantly centrocyte-like with few monocytoid cells having pale to clear cytoplasm, small round lymphocytes and occasional centroblasts and plasma cells. Muscular coat and serosa were uninvolved. Partial epithelial destruction was present, with lymphoepithelial lesions [Table/Fig-3]. Surgical resected margins were positive. All the 32 mesenteric lymph nodes identified showed infiltration by similar neoplastic lymphoid cells [Table/Fig-4]. However, the appendix was uninvolved. A diagnosis of primary MALT lymphoma of colon was rendered. The immunoreaction of the neoplastic lymphoid cells confirmed the diagnosis. The cells were CD20 positive and negative for CD3, CD10 and Cyclin D1 [Table/Fig-5-7]).

Unfortunately, the patient succumbed to surgical complications on fifth day post-surgery.

DISCUSSION

Primary colorectal lymphomas are less frequent, accounting for about only 0.2% of all colonic neoplasms [1]. Although rare, they are very important clinical entities as the can have varied clinicopathologic presentations, mimicking reactive processes, benign conditions and colonic carcinomas. Diffuse large B cell lymphoma is the commonest type of colonic lymphoma and MALT type lymphoma accounts for less than 20% of such cases [2].

Here, we have presented the clinical, histopathological and immunohistochemical features of a case of primary colonic MALT lymphoma, presenting uniquely as multiple lymphomatous polyposis. Multiple Lymphomatous Polyposis (MLP) is an unusual form of gastrointestinal non-Hodgkin B-cell lymphoma manifesting itself by the presence of numerous lymphomatous polyp along one or more long segments of gastrointestinal tract [3]. The characteristic clinic-morphological manifestations were first brought into picture by Cornes in 1961 [4] and since then, no more than 100 cases have been reported. Most cases of MLP represent extranodal counterpart of mantle cell lymphomas which tend to have an aggressive clinical course. On the contrary, MALT lymphomas follow an indolent course. Hence, it is of utmost importance to render a correct diagnosis for appropriate management and prognostication. Other lymphomas that can present as MLP are diffuse large B-cell lymphoma [5,6] and follicular lymphoma [7].

WHO defines MALT lymphoma as "an extranodal lymphoma composed of morphologically heterogenous small B-cells including marginal zone (centrocyte-like) cells, cells resembling monocytoid cells, small lymphocytes, and scattered immunoblasts and centroblast-like cells" [8]. Lymphoepithelial lesions are typical and are characterized by aggregates of three or more neoplastic lymphoid cells along with distortion or destruction of the epithelium [8]. Stomach is the most common site. Colonic MALT lymphomas are very rare and their presentation as MLP is still rarer.

Colonoscopy plays a vital role in the detection of such polypoidal lesions of colon, as well as for obtaining biopsies. Differentiating lymphomatous polyposis from adenomatous or hamartomatous polyposis by colonoscopic or radiological evaluation alone is impossible and can often be misleading. Histopathological examination is the gold standard in such cases. Hence, generous sampling during colonoscopy is always preferable and advised. Otherwise, if the biopsy obtained is very superficial, it may be histologically inconclusive, as in the current case. The biopsy of this patient revealed heavy mononuclear cell infiltrate but did not show clear-cut features of MALT lymphoma, most probably because of scanty tissue. Also, optimal fixation and processing of the biopsy specimen are essential to produce high quality H&E sections. Critical histomorphologic evaluation of the biopsy has to be done. In our experience, presence of lymphoepithelial lesions and infiltrative growth pattern serves as important features in distinguishing MALT lymphomas from reactive lymphoid hyperplasia. Judicious use of immunohistochemical markers and sometimes, genetic work up is needed for confirmation and to distinguish it from other lymphomas. MALT lymphomas express pan-B cell immunomarkers and are often negative for CD5, CD 10 and Cyclin D1.

Due to rarity of the disease, there is controversy regarding the therapy of MALT type lymphomas of colon. They are not associated with H. pylori infection and so, there is no role of antibiotics. Surgical resection is generally performed, as colonic MALT-associated lymphomas, having low proliferation index, may show resistance to chemotherapy [9]. But, postoperative chemotherapy is often advocated. Patients with regional lymph nodal involvement are found to have a bad prognosis [10]. This patient had 32 mesenteric lymph node being involved, there by indicating a poor prognosis.

CONCLUSION

Not all MLP result from mantle cell lymphoma, rarely follicular lymphoma, diffuse large B-cell lymphoma & MALT lymphoma can also present as MLP. So, the clinicians as well as the pathologists should be aware of such a presentation, so that the appropriate therapeutic protocol can be planned.

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