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Unusual Presentation of Lymphoma as Appendicitis

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Abstract
Two cases of lymphoma presenting as appendicitis are reported here. The diagnosis was made post operatively, mantle cell lymphoma in one appendectomy specimen in a known case of mantle cell lymphoma and high grade B cell lymphoma, NOS from other right hemicolectomy specimen complicating as appendicular abscess and cecal perforation. These cases highlight the importance of routine histopathological examination of appendix specimens.

Introduction

Gastrointestinal tract is the most common site of extranodal lymphoma, accounting for 30- 40% of all extranodal cases. The GIT is also a common site for post transplantation lymphoproliferative disorder. The most recent classification of small intestinal and colorectal lymphoma are listed here.[1]

WHO classification of small intestinal malignant lymphoma

1) immunoproliferative small intestinal diseases
2) MALT
3) Mantle Cell Lymphoma
4) DLBL
5) burkitt lymphoma
6) B cell lymphoma unclassifiable
7) follicular lymphoma
8) T cell lymphoma

Enteropathy associated intestinal T cell lymphoma
Monomorphic CD56 positive intestinal T cell lymphoma

WHO classification of malignant lymphoma of colon and rectum

1) MALT
2) Mantle cell lymphoma
3) DLBL
4) Burkitt lymphoma
5) B cell lymphoma unclassifiable

Appendicular lymphoma is a rare entity despite the fact that appendix is rich in lymphoid cells. Appendicular lymphoma comprises 0.015% of all gastrointestinal tract lymphoma. [2]

Mantle cell lymphoma is a B-cell lymphoma derived from inner mantle zone cells, that is composed of small to medium sized and slightly irregular lymphocytes (resembling centrocytes) without immunoblast, paraimmunoblast or proliferation centers. MCL are characterized by

t(11;14)(q13;q32) translocation involving CCND1 and immunoglobulin heavy chain (IgH) gene's and expression of cyclin D1.There are certain morphological variants of mantle cell lymphoma.

Aggressive variants

A) Blastoid

Cells resemble lymphoblasts with dispersed chromatin and a high mitotic rate (usually 20-30 mitoses per 10 high-power fields).

B) Pleomorphic:

Cells are pleomorphic, but many are large with oval to irregular nuclear contours, generally pale cytoplasm, and often prominent nucleoli in at least some of the cells.

Other variants

A)Small-cell:

Cells are small round lymphocytes with more clumped chromatin, either admixed or predominant, mimicking a small lymphocytic lymphoma.

B)Marginal zone-like:

There are prominent foci of cells with abundant pale cytoplasm resembling marginal zone or monocytoid B cells, mimicking a marginal zone lymphoma; sometimes these paler foci also resemble proliferation centres of chronic lymphocytic leukaemia I small lymphocytic lymphoma.[3]

DLBL (diffuse large B cell lymphoma) not otherwise specified is a heterogenous category that includes most diffuse lymphomas composed of large transformed B cells. Neoplastic large B cells are greater than twice the size of normal lymphocytes or greater than the nucleus of a macrophage. WHO classified lymphoma based on tumor cell cytology as: centroblastic, immunoblastic and anaplastic.

It's a high grade B cell lymphoma with MYC and BCL2 or BCL6 rearrangement. Often present as single rapidly growing nodal mass. 30-40% are extranodal like skin, GIT, genitourinary, CNS, liver and spleen.

High grade B cell lymphoma (HGBL) is an aggressive, mature B-cell lymphoma that for biological and clinical reasons should not be classified as DLBL or as Burkitt lymphoma. There are two categories of HGBL:

a) HGBL with MYC and BCL2 and/or BCL6 rearrangements.
b) HGBL, NOS encompasses cases having features intermediate between DLBL and Burkitt lymphoma.

Case-1

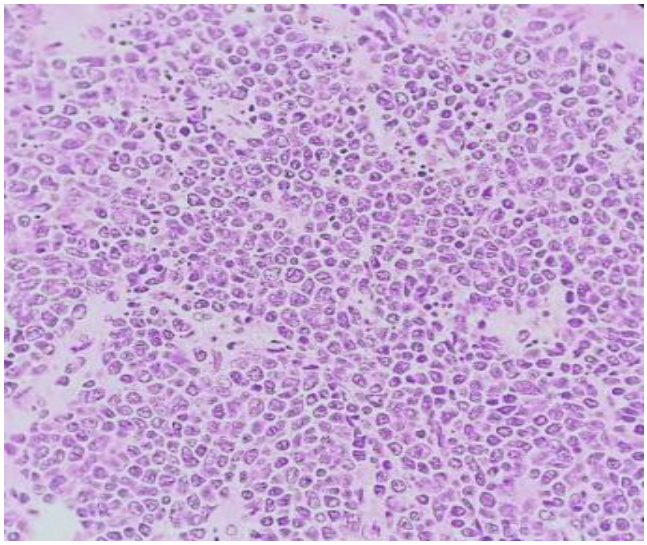
53 yrs old female presented with acute appendicitis, who was a known case of mantle cell lymphoma, post chemotherapy. Patient was evaluated with USG suggested abdominal lymphadenopathy along with spleenomegaly with multiple tiny hypoechoic nodules and acute appendicitis. CBP on peripheral smear showed leucocytosis with circulating blasts and thrombocytopenia. On gross appendix showed thickened wall and lumen was filled with hemorrhagic material. On histopathology appendix showed medium sized monomorphic population of lymphoid cells. The cells have irregular nuclear borders with coarse chromatin and inconspicuous nucleoli. Marked apoptosis and mitosis noted. Germinal centers not seen. Favouring appendix is involved by lymphoma. On IHC tumor cells are diffused positive for CD20 while overexpression cyclin D1, Mib-1 index was around 70- 80%. Thus on morphology and IHC a diagnosis of Blastoid variant of mantle cell lymphoma was done.

Case-2

A 41 yrs old Male presented with acute appendicitis for which he was evaluated by USG revealed perforated tip of appendix with adjacent phlegmon in right subhepatic region and hepatomegaly with grade1 fatty liver. Patient underwent appendectomy. Histopathology revealed acute appendicitis with periappendicular abscess formation. on follow up patient had pigtail catheter drainage was evaluated with CT scan revealed collection in right iliac floss in retro caecal region measuring 6.2x5.8cm, extending uptown parietal wall displacing the caecum

anterior, which was not resolving. so the patient was planned for right hemicolectomy. On gross the specimen showed caecal perforation along with thickened caecal and ascending colon wall. Histopathology revealed diffuse sheets of lymphocytes extending upto subserosal fat. individual cells are medium to large in size with vesicular nuclei. Frequent apoptosis and mitotic figures noted . Areas of necrosis noted and signs of serositis noted, suggesting atypical lymphoid proliferation with signs of sealed perforation. On IHC tumor cells were positive for CD20,CD10 and negative for CD3,BCL2 and MUM-1. MIB-1 index was 90%. Ultimately diagnosed as NHL high grade B cell lymphoma NOS.

Mantle cell lymphoma with CD20, cyclin D1 positive and high MIB



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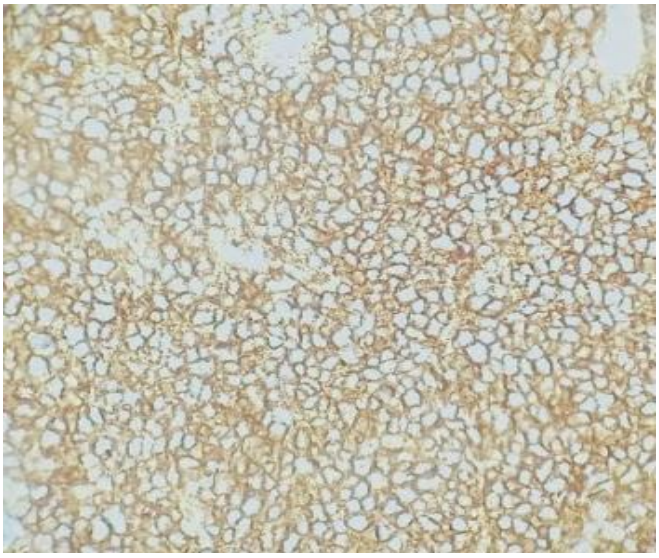
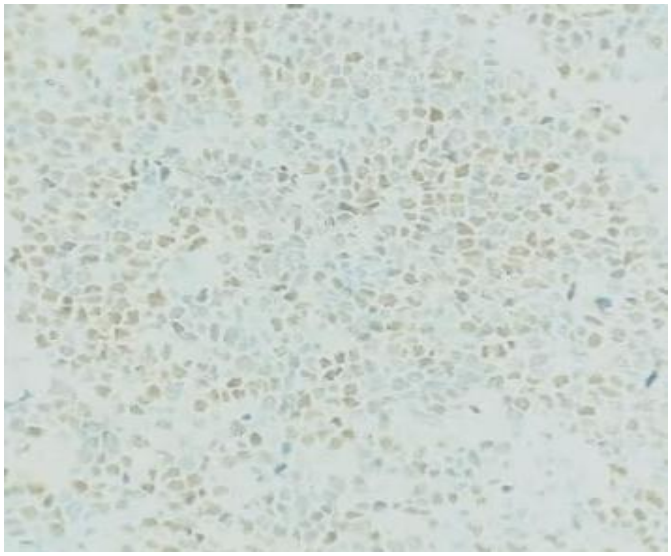
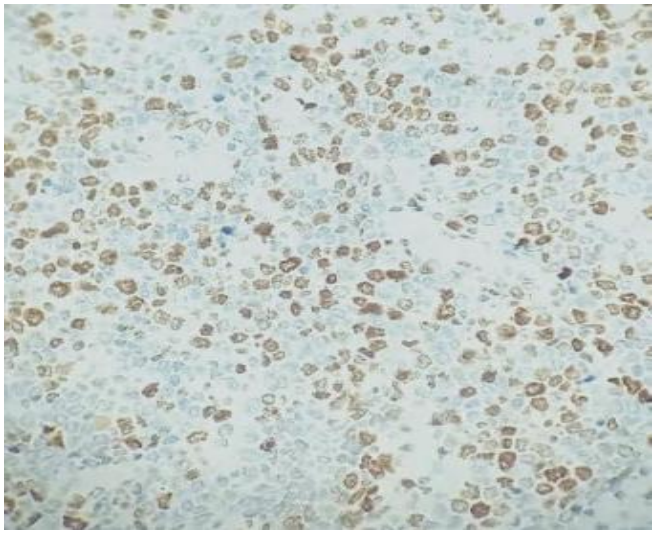


CD20

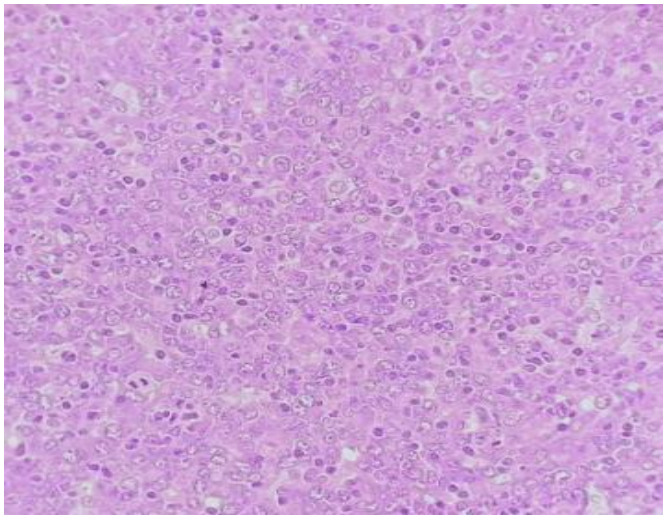


CYCLIN D1



MIB

NHL High grade B cell lymphoma NOS with CD20 and CD10 positive and negative for CD3, BCL2 and MUM-1. High MIB.



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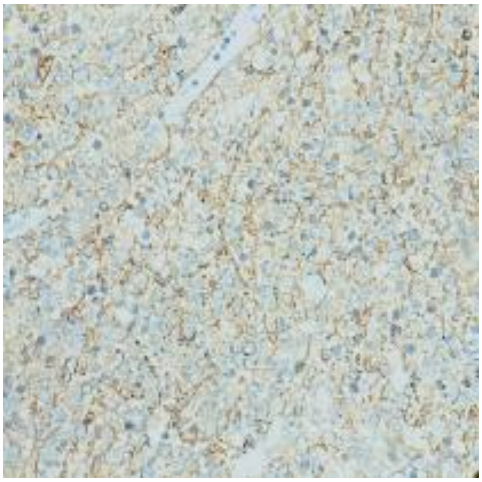
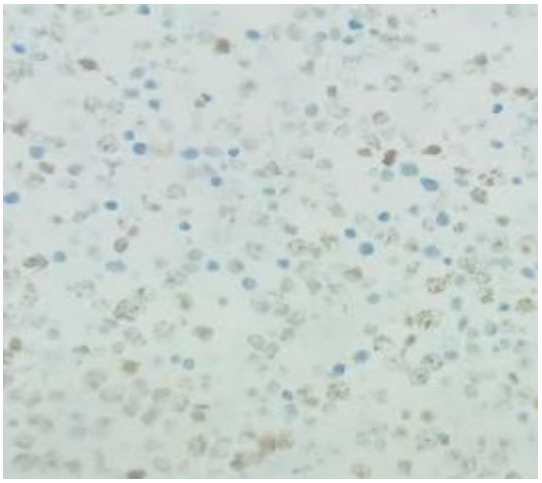
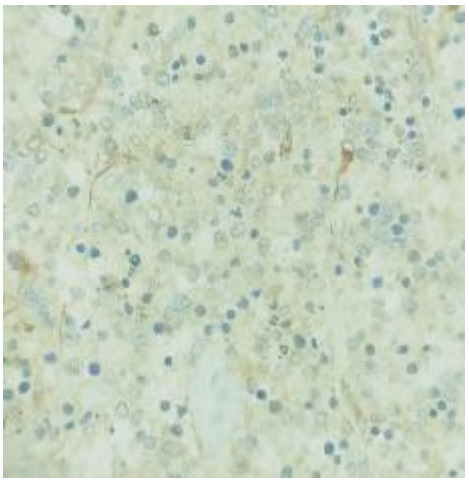


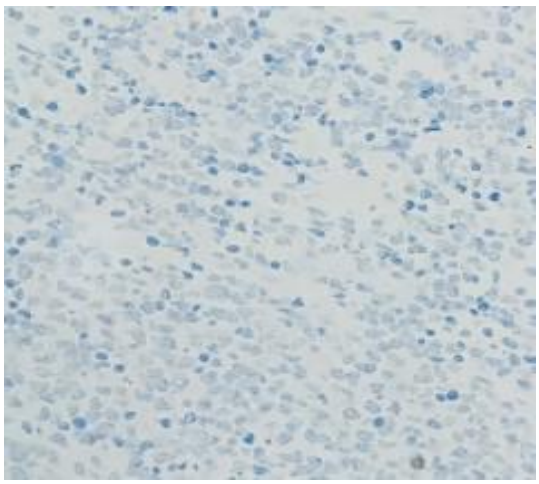
CD20



MIB%



CD10



MUM-1

Discussion

Primary GIT lymphomas are rare especially in appendix. Most common extranodal lymphoma is DLBL followed by burkitt lymphoma. Studies suggest a higher incidence in Male (81.9%) and white race (81%).[4] Secondary appendiceal tumors are rare,primary sites include like ovary followed by colorectal and stomach. In a study conducted by schmutzer et al in 8699 appendectomy specimens showed a total of 17 malignancies identified, in which 2 were primary adenocarcinoma, 12 were metastatic adenocarcinoma and 3 were lymphoma. [5] Patients presented with acute appendicitis, abdominal pain without appendicitis, palpable mass and constipation. Appendicitis is the initial manifestation of malignancies. Mode of appendicular involvement was predominantly metastasis.[6]

In MCL lymph nodes are most commonly involved followed by bone marrow and spleen with or without peripheral blood involvement. Other extranodal sites frequently involved are GIT, waldeyer ring, lungs and pleura.CNS involvement can occur at the time of relapse. Patients with extranodal and intra-abdominal tumors presents with symptoms of bowel obstruction, intussusception and appendicitis. [7] Here in both the cases patients typically gave the history of appendicitis especially regarding right iliac fossa pain and duration of pain. The differential diagnosis includes relapsing or chronic

appendicitis which is rare and difficult to diagnose. The diagnosis was made only post operatively on histopathological specimens.[8]

Burkitt lymphoma usually presents as rapidly growing abdominal masses with signs and symptoms of intestinal obstruction and if affecting the appendix presents as appendicitis. It is more likely that appendiceal and ovarian sites are areas of Secondary involvement, it is due to pre-existing lymphoid tissue in appendix and ovary that creates an at risk site for lymphoma development. [9]

Conclusion

We presented cases of high grade B cell lymphoma of colon and Mantle cell lymphoma of appendix in a known case of mantle cell lymphoma presenting as appendicitis. Our cases illustrates that this cancer type must be included in differential diagnosis of acute appendicitis.

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