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Increased Activated Plasma Cells in Inflammatory Bowel Disease When Compared to Ischemic Acute Colitis

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Indolent systemic mastocytosis or just ‘atypical enterocolic mast cell aggregates’?

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Introduction/Objective: Mastocytosis is a rare disease in which there are abnormal mast cell accumulation in one or more tissue sites. Multifocal dense mast cell aggregates with atypical morphology or immunohistochemistry are considered as systemic mastocytosis (SM) based on WHO criteria. SM usually involves bone marrow and majority of them also have KIT mutation. There are rare case reports of atypical enterocolic mast cell aggregates (EMCA) confined to gastrointestinal (GI) only with mild or no symptoms. Here we present a case with extensive atypical mast cell aggregates in lower GI tract yet no evidence of involvement of other organs.

Methods/Case Report: A 34-year-old woman presented with abdominal bloating, diarrhea along with pruritis but no cutaneous lesion. Biopsies from the ascending and descending colons, caecum and rectum consistently showed increased eosinophils and multifocal infiltrates of atypical spindle shaped mast cells which are positive for CD117/tryptase but negative for CD2 and CD25. This is consistent with SM by WHO criteria based on morphology. Bone marrow biopsy showed normal amount of mast cells with normal morphology. Upper gastrointestinal biopsy was unremarkable. Serum tryptase level was normal. No KIT mutation was detected in exon 9, 11, 13 or 17 from colonic mucosa. Patient has been treated with antihistamine and Montelukast and symptoms resolved.

Results (if a Case Study enter NA): N/A

Conclusion: This case met the criteria of SM based on the presence of multifocal mast cell aggregates and atypical spindle morphology >25%. Johncilla et al. previously reported 16 cases of EMCA with atypical morphology or immunohistochemistry, absent to mild localized symptoms, and negative KIT mutation. Based on lack of generalized disease, the authors preferred using descriptive terminology instead of ‘systemic mastocytosis’ for those cases. Our case has broader involvement of lower gastrointestinal tract than any reported case and the patient needs treatment for the symptoms. However, there is no ‘systemic’ involvement of bone marrow or any other organ. The diagnosis of ‘Systemic Mastocytosis’ would cause potential confusion and/or unnecessary anxiety. Further study of more cases is needed to better characterize and categorize the cases of atypical mast cell aggregates localized only to the GI.

Increased Activated Plasma Cells in Inflammatory Bowel Disease When Compared to Ischemic Acute Colitis

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Introduction/Objective: Inflammatory bowel disease (IBD) and acute ischemic colitis can both be involved by active colitis. IBD is characterized by crypt architectural distortion, basal lymphoplasmacytosis, and occasional granulomatous changes. However, diagnosis of IBDs is still largely by exclusion of other types of active colitis with similar changes. We previously demonstrated that glucose regulated protein 94 (grp94) is mainly expressed by activated plasma cells. We postulate that increased numbers of grp94-positive plasma cells may support diagnosis of IBDs. Here, we compared IBD and active ischemic colitis for grp94 expression in mucosal plasma cells of colectomy specimens

Methods/Case Report: Tissue sections from colectomy specimens with active IBD (n = 8) and ischemic colitis (n = 7) were examined for grp94 expression by immunohistochemistry (monoclonal antibody clone 9G10 at dilution of 1:200, Enzo Life Science, Inc Farmingdale, NY). The staining intensity and highest number of grp94 in plasma cells per high power field was counted and recorded for each case, and combined scores were calculated as # of plasma cells multiplied by staining intensity (ranging from 0 to 3+). Unpaired student T tests were used to compare these indices between the two groups for statistical significance (p value < 0.05 was considered significantly different)

Results (if a Case Study enter NA): Plasma cells in lamina propria identified by grp94 staining showed higher intensity in IBD than ischemic groups. The number of plasma cells and combined scores were also significantly higher in the IBD group than that of ischemic group

Conclusion: Our data indicates that active plasma cells are much more numerous in IBD than ischemic colitis, supporting the notion that active plasma cells are involved in the development of this disease process. Morphologically, active colitis with increased number of plasma cells appears to be another index favoring the diagnosis of IBD.

Duodenal Duplication Cyst in adult male; An Extremely Rare Entity and a Diagnostic Pitfall leading to Whipple procedure

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Introduction/Objective: Whipple procedure is a complex, invasive operation and has high morbidity and mortality.