

Unveiling the connection: exploring the link between rash and diarrhea

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Introduction

Diarrhea is a commonly encountered problem in Hospitalist Medicine. We present a case of intractable abdominal pain with diarrhea, Later diagnosed as progression of a rare preexisting condition

Case Presentation

- A 74-year-old male presented with intractable nausea, vomiting abdominal pain, abdominal distention, diarrhea, and loss of appetite for the past 2 weeks. Abdominal pain was spasmodic, generalized, moderate intensity, without any precipitating or relieving factors and brown-yellow non-bloody diarrhea.
- Interestingly he had a similar episode of diarrhea 3-4 weeks ago, at that time was evaluated in ER and discharged on antibiotics for presumed gastroenteritis. Patients denied travel history or outside food consumption on both presentations.
- Physical examination revealed lethargy, generalized abdominal tenderness on superficial palpation along with macular-papular rash over the entire back. Initial workup including WBC count, blood cultures, C difficile, stool parasite/ova, stool WBC count, Anti-Tissue Transglutaminase antibody were negative. Chemistries were within normal limits.
- Imaging significant for small bowel wall thickening, with no obstruction. Patient was started on empiric antibiotics. CTA abdomen revealed less than 50% celiac artery stenosis thus ruling out acute bowel ischemia.
- Upon further interviewing regarding the patient's rash, he reported past medical history of Cutaneous Mastocytosis, which had been confirmed with skin

biopsy and bone-marrow biopsy, but currently was not taking any treatment.

- While de-escalating antibiotics, we started H1/H2 blockers (Loratadine and Famotidine). A progressive improvement in diarrhea was observed. Patient was discharged home, and outpatient EGD, and colonoscopy were scheduled to rule out gastric/duodenal ulcers.

Discussion

- Mastocytosis is the clonal proliferation of mast cells (MC) characterised by multifocal clusters of abnormal MC within multiple organ systems.
- Depending on tissue/Organ involvement (skin and/or internal organs), it is classified as Cutaneous mastocytosis (CM) or Systemic mastocytosis (SM) respectively. Increased cytokinin secretion from mast cells (including histamine, heparin, leukotrienes, prostaglandins, and PAF) activates H1/H2 receptors causing not only cutaneous manifestations but also increased gastric acid secretion, (predisposing to gastric and duodenal ulcers, Upper GI bleeding, abdominal pain)[6]. Thus SM can frequently present as pruritic maculopapular rash/urticaria[2], and chronic recurrent diarrhea, nausea, and weight loss[1].
- A study comparing histopathological samples for SM, inflammatory bowel disease, eosinophilic colitis, etc, found that in all SM patients, D816V KIT mutation was observed [4]. Another study found that adult-onset CM more frequently progressed to SM[9][10]

- The definite diagnosis of SM involves biopsy of affected tissues including immunohistochemical stains (CD117and CD25), serum tryptase(>20 ng/mL) [7], and MC aggregates in gastrointestinal lamina propria [8]. Treatment depends on the disease subtype. H1 antagonists treat skin symptoms whereas H2 antagonists are generally used as 1st-line therapy for GI symptoms[3]

Conclusion

Diarrhea is a common symptom of systemic mastocytosis. However systemic mastocytosis is a rare cause of diarrhea. There should be a high degree of suspicion in patients with otherwise unexplained symptoms of abdominal pain and diarrhea, especially if pruritic maculopapular skin rash is also present

References

- [1]Sokol H., Georjin-Lavialle S., Canioni D., Barette S., Damaj G., Soucie E., Bruneau J., Chandris M.-O., Suarez F., Launay J.-M., et al. Gastrointestinal manifestations in mastocytosis: A study of 83 patients. *J. Allergy Clin. Immunol.* 2013;132:866–873. doi: 10.1016/j.jaci.2013.05.026.
- [2]Cherner JA, Jensen RT, Dubois A, O'Dorisio TM, Gardner JD, Metcalfe DD. Gastrointestinal dysfunction in systemic mastocytosis. A prospective study. *Gastroenterology.* 1988 Sep;95(3):657-67. doi: 10.1016/s0016-5085(88)80012-x. PMID: 3396814.
- [3]Pardani A. Systemic mastocytosis in adults: 2021 update on diagnosis, risk stratification and management. *Am. J. Hematol.* 2021;96:508–525. doi: 10.1002/ajh.26118.
- [4]Kirsch R., Geboes K., Shepherd N.A., de Hertogh G., Di Nicola N., Lebel S., Mickys U., Riddell R.H. Systemic mastocytosis involving the gastrointestinal tract: Clinicopathologic and molecular study of five cases. *Mod. Pathol.* 2008;21:1508–1516. doi: 10.1038/modpathol.2008.158.