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Abstract Form

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Research Category (please check one):

☐	**Original Research**	☒	**Clinical Vignette**	☐	**Quality Improvement**	☐	**Medical Education Innovation**

Abstract

Mastocytosis is a group of rare disorders characterized by the cutaneous and/or systemic accumulation of mast cell. The most common skin findings are multiple, monomorphic, red-brown macules that may result in urticarial flaring when lightly rubbed, known as Darier's sign. Systemic mastocytosis can present with gastrointestinal disturbance, neurologic, hematologic, and musculoskeletal symptoms. The variable presentations of mastocytosis often lead to delayed diagnosis. We describe the case of a 33-year-old male with chronic tetrahydrocannabinol use who presented with a cyclic pattern of intractable nausea, vomiting, and diffuse hives.

The patient is a 33-year-old with history of multiple emergency room visits for cyclic vomiting and chronic tetrahydrocannabinol (THC) use. He presented with intractable nausea and was initially treated for suspected cannabinoid hyperemesis syndrome. The patient had minimal improvement despite treatment with antacid, proton pump inhibitor, antiemetic, Haldol and morphine as needed for epigastric pain. During the hospital course, the patient was noted to break out in a diffuse urticarial blanching rash that started on his trunk and spread to his extremities while sparing his face, palms, and soles. Initially, the rash was thought to be an adverse drug reaction. However, the rash noted to be worse in rubbed areas, such as the skin surrounding the blood pressure cuff. When asked if the patient had similar prior eruptions, he reported multiple emergency visits for similar rashes following episodes of emesis which were refractory to treatment with steroids and antihistamines. The patient was started on a histamine-2 blocker, topical hydrocortisone cream and a leukotriene inhibitor. Following administration of the leukotriene inhibitor, the patient has rapid improvement of both his skin and gastrointestinal symptoms. Serum tryptase level was within normal limits, likely secondary to the prior use of antihistamines.

Mast cells are a specialized portion of the immune system, which release histamine as an inflammatory response to foreign material. They serve as the primary immune response in anaphylactic reactions. Mastocytosis is a rare disorder with multiple different subtypes, affecting 1 in every 10,000 to 20,000 individuals. The diagnosis of systemic mastocytosis is generally delayed due to overlapping, variable symptoms. The World Health Organization outlines the diagnostic criteria based on histopathologic, morphologic, molecular, serologic, and clinical criteria. However early or low burden disease may not fully meet the diagnosis threshold. Clinical diagnosis is made by fixed, hyperpigmented red-brown ulcerated plaques, papules or macules less than 0.5 centimeters in diameter, and other organ involvement, such as the gastrointestinal tract. Patients generally present with episodic flushing, dyspepsia, diarrhea, abdominal pain, musculoskeletal pain, or hypotension. Physicians must have a high index of suspicion when treating a patient with recurrent, unexplained anaphylaxis chronic abdominal cramps. Treatment often involves a multidisciplinary team including an allergist, hematologist, and dermatologist. Patients with mastocytosis must be educated to avoid potential triggers such as heat, humidity, stress, alcohol, aspirin, opioids, or radiocontrast agents. Physicians must also be aware of perioperative management to limit stressors, premedicate, and provide prompt anaphylaxis treatment if necessary. The diagnosis of mastocytosis is challenging given its variable presentations and mimicry of many other conditions. With a detailed history, thorough physical exam and strong clinical suspicion, patients with mastocytosis can avoid unnecessary exams and delayed diagnosis.