respectively. Mast cell–mediated fibrotic changes may occur in liver, spleen, and bone marrow but not in gastrointestinal tissue or skin.

The cutaneous lesions of MPCM are reddish-brown macules, papules, or plaques that respond to trauma with urtication and erythema (Darier's sign). Two distinct forms of MPCM are recognized: polymorphic MPCM and monomorphic MPCM. Children with CM may present with MPCM, mastocytomas, or diffuse cutaneous mastocytosis (DCM). Mastocytomas are generally solitary elevated lesions that are yellow, brown, or red in color. Their size may vary from a few millimeters to several centimeters. Rubbing or irritation of the mastocytoma lesion may lead to systemic symptoms such as flushing and urticaria. Children with DCM present without distinct lesions, but rather a generalized thickening of skin and "peau d'orange" appearance due to diffuse mast cell infiltration. DCM may be associated with bullae formation and more severe systemic symptoms, including upper gastrointestinal irritation and vascular collapse in the first few years of life. Maculopapular skin lesions of mastocytosis may be present in patients with adult-onset systemic disease. The apparent incidence of cutaneous lesions is ≥80% in patients with ISM and <50% in those with SM-AHNMD or ASM. In the upper gastrointestinal tract, gastritis and peptic ulcer are significant problems. In the lower intestinal tract, the occurrence of diarrhea and abdominal pain is attributed to increased motility due to mast cell mediators; this problem can be aggravated by malabsorption, which can also cause secondary nutritional insufficiency and osteomalacia. The periportal fibrosis associated with mast cell infiltration may lead to portal hypertension and ascites. In some patients, anaphylaxis with rapid and life-threatening vascular collapse may occur. Anaphylaxis is most commonly induced by Hymenoptera stings, and patients often have evidence of venom-specific IgE. The neuropsychiatric disturbances are clinically most evident as impaired recent memory, decreased attention span, and "migraine-like" headaches. Patients may experience exacerbation of a specific clinical sign or symptom variably with alcohol ingestion, temperature changes, stress, use of mast cell–interactive opioids, or ingestion of nonsteroidal anti-inflammatory drugs (NSAIDs).