Mastocytosis is a clonal disorder characterized by the proliferation and accumulation of mast cells (MC) in different tissues, with a preferential localization in skin and bone marrow (BM). The excess of MC in mastocytosis as well as the increased releasability of MC may lead to a higher frequency and severity of immediate hypersensitivity reactions. Mastocytosis in adults is associated with a history of anaphylaxis in 22-49%. Fatal anaphylaxis has been described particularly following hymenoptera stings, but also occasionally after the intake of drugs such as nonsteroidal anti-inflammatory drugs, opioids and drugs in the perioperative setting. However, data on the frequency of drug hypersensitivity in mastocytosis and vice versa are scarce and evidence for an association appears to be limited. Nevertheless, clonal MC disorders should be ruled out in cases of severe anaphylaxis: basal serum tryptase determination, physical examination for cutaneous mastocytosis lesions, and clinical characteristics of anaphylactic reaction might be useful for differential diagnosis. In this position paper, the ENDA group performed a literature search on immediate drug hypersensitivity reactions in clonal MC disorders using MEDLINE, EMBASE, and Cochrane Library, reviewed and evaluated the literature in five languages using the GRADE system for quality of evidence.
and strength of recommendation.

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