Proposed diagnostic algorithm for patients with suspected mast cell activation syndrome

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Mast cell activation (MCA) accompanies diverse physiologic and pathologic processes and is one of the more frequently encountered conditions in medicine. MCA-related symptoms are usually mild and often transient. In such cases, histamine receptor blockers and other mediator-targeting drugs can usually control MCA. In severe cases, an MCA syndrome (MCAS) may be diagnosed. However, overt MCAS is an unusual condition, and many patients referred because of suspected MCAS are diagnosed with other diseases (autoimmune, neoplastic, or infectious) unrelated to MCA or suffer from MCA-related (eg, allergic) disorders and/or comorbidities without fulfilling criteria of an overt MCAS. These considerations are important as more and more patients are informed that they may have MCA or even MCAS without completing a thorough medical evaluation. In fact, in several instances, symptoms are misinterpreted as MCA/MCAS, and other clinically relevant conditions are not thoroughly pursued. The number of such referrals is increasing. To avoid such unnecessary referrals and to prevent misdiagnoses, we here propose a diagnostic algorithm through which a clinically relevant (systemic) MCA can be suspected and MCAS can subsequently be documented or excluded. In addition, the algorithm proposed should help guide the investigating care providers to consider the 2 principal diagnoses that may underlie MCAS, namely, severe allergy and systemic mastocytosis accompanied by severe MCA. Although validation is required, we anticipate that this algorithm will facilitate the management of patients with suspected MCAS.
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