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Mastocytosis: Review of natural history and recommended treatments

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Mastocytosis is a heterogeneous group of disorders characterized by the abnormal increase of mast cells in only the skin (cutaneous mastocytosis), in extracutaneous tissues involving multiple organs (systemic mastocytosis), or in solid tumours (mastocytoma and mast cell sarcoma). Recent studies estimate the 1 in 10,000 people are diagnosed with mastocytosis. The clinical course and treatments for mastocytosis vary depending on the time of onset and classification of the disease. However, the natural history and treatment guidelines have not yet been established. We conducted a systematic review to assess the natural history and management of different mastocytosis subtypes. A systematic review and meta-analysis were conducted using PubMed and Ovid database of studies published in English and French in the last fifteen years, from January 2001 to December 2016. The keywords "Cutaneous mastocytosis', 'Systemic mastocytosis', 'pathophysiology', 'clinical course', 'prognosis', 'drug therapy', and 'therapy' were searched. Rate of complete resolution was subjected to pooled analysis for different mastocytosis subtypes. Meta-analysis was conducted using Stata version 12.0. We reviewed 634 papers, of which 13 were included in the analysis of resolution and 124 were included in the assessment of management. Pooled estimate for rate of complete resolution varied depending on the mastocytosis subtype. In cutaneous mastocytosis, the complete resolution rate for mastocytoma was 10% per year (95% CI: 4.8%, 15.1%) while the rate for urticarial pigmentosa was 1.9% per year (95% CI: -0.5%, 4.3%). Diffuse cutaneous mastocytosis and other systemic subtypes did not show evidence of complete resolution in the studies reviewed. Treatment of cutaneous and systemic mastocytosis is purely symptomatic with topical corticosteroids, antihistamines, omalizumab and imatinib being common choices. Rate of resolution of mastocytosis is low especially in systemic forms and better management guidelines are required to improve the health of these patients.

Biography

Dr. Ben-Shoshan graduated from The Sackler School of Medicine, Tel-Aviv, Israel and completed his fellowship in Pediatric Allergy/Clinical Immunology at Montreal Children`s Hospital in 2009. Dr Ben-Shoshan has been granted his MSc degree in Epidemiology in McGill in 2011. In 2011 he was granted the Emerging Clinician Scientist fellowship award by AllerGen NCE and in 2013 the FRSQ junior 1 salary award. Dr Ben-Shoshan is currently a physician in the division of Allergy/ Immunology at Montreal Children's Hospital and is involved in research initiatives on anaphylaxis, chronic urticaria, matocytosis and immunodeficiency and has more than 60 publications related to these topics.

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Michelle Le is currently a 3rd-year Medical Student at McGill University Faculty of Medicine in Montreal, Canada. Prior to medical school, she has completed a Bachelor's degree in Biopharmaceutical Sciences at the University of Ottawa in 2014. She has previous research experience in Cellular and Molecular Medicine during her Undergraduate studies, where she was awarded with NSERC, Canadian Stroke Network, and Berlin-Brandenburg School for Regenerative Therapies studentships. She is very interested in pursuing a career in Dermatology and is actively involved in its research. Her medical and research interests surround immunodermatology including chronic urticaria, mastocytosis, atopic eczema, and microbial skin diseases.

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