

Review article

Mastocytoses

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Abstract

Mastocytoses are an important group of dermatoses characterized by histological infiltrates predominantly rich in mast cells. The present review focuses on the salient clinical, diagnostic and therapeutic features of different entities included in this group.

Introduction

Mastocytoses are benign or very rarely malignant proliferative disorders of the reticuloendothelial system. Mastocytosis is frequently self-limited especially in childhood, and represents an exaggerated accumulation of essentially normal looking mast cells in their regular distribution. The mast cell is the repository of a number of pharmacological substances, resulting in protean clinical manifestations. The skin is frequently involved with or without internal organ involvement. The present article presents an overview of the clinicopathological correlation and treatment options in mastocytoses in a simplified and tabulated version.

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Table 1
Mastocytoses

| Tumor | Clinical Features | Histopathology | Prognosis | Work up |
|-----------------------------------|---|--|---|--|
| Benign mastocytosis [1-23] | | | | |
| Cutaneous | | | | |
| Urticaria pigmentosa | <ul style="list-style-type: none"> • Children, adults • Macules, papules • Nodules, plaques • Monomorphic • Pigmented • Symmetrical • Trunk • Darier's sign elicitable • Systemic manifestations | <ul style="list-style-type: none"> • Mast cells • Oval, spindle shaped • Metachromatic granules | <ul style="list-style-type: none"> • Depends on the extent of disease • Usually self-limiting | <p>For diagnosis:</p> <ul style="list-style-type: none"> • Clinical evaluation • Skin biopsy • Increased plasma histamine and tryptase • Increased urinary histamine and PGD2 <p>For extent:</p> <ul style="list-style-type: none"> • Based on symptoms <p>Follow up:</p> <ul style="list-style-type: none"> • For disease state and opted therapy <p>Therapeutic options:</p> <ul style="list-style-type: none"> • Antihistamines • Antihistamine + aspirin (for GI symptoms) • Sodium chromoglycate • PUVA therapy • Topical and systemic steroids • Cytokines • Excision of solitary lesions |
| Solitary mastocytoma | <ul style="list-style-type: none"> • Children • Solitary nodules • Pink red, yellow • Systemic manifestations | <ul style="list-style-type: none"> • Mast cells • Oval, spindle shaped • Metachromatic granules | <ul style="list-style-type: none"> • Resolve in 2-3 years | <ul style="list-style-type: none"> • As for urticaria pigmentosa |

Table 2
Mastocytoses (contd....)

| Tumor | Clinical features | Histopathology | Prognosis | Work up |
|---|---|--|--|---|
| Telangiectasia macularis eruptiva perstans | <ul style="list-style-type: none"> • Adults • Telangiectatic macules • Persistent • Treatment resistant | <ul style="list-style-type: none"> • Mast cells • Oval, spindle shaped • Metachromatic granules | <ul style="list-style-type: none"> • Persistent | <ul style="list-style-type: none"> • As for urticaria pigmentosa |
| Diffuse cutaneous mastocytosis | <ul style="list-style-type: none"> • Infants, adults <p>Variants:</p> <ul style="list-style-type: none"> • Pseudoxanthomatous • Pachydermatous • Nodular, bullous • Systemic manifestation • Flexural involvement | <ul style="list-style-type: none"> • Mast cells • Oval, spindle shaped • Metachromatic granules | <ul style="list-style-type: none"> • Depends on the extent of disease | <ul style="list-style-type: none"> • As for urticaria pigmentosa |
| Systemic mastocytosis | <ul style="list-style-type: none"> • Adults <p>Systemic symptoms:</p> <ul style="list-style-type: none"> • Flushing, headache • Diarrhea, weight loss <p>Systemic signs:</p> <ul style="list-style-type: none"> • <i>Gastrointestinal tract:</i> <ul style="list-style-type: none"> • Hepatosplenomegaly • Peptic ulcer, malabsorption <p>Hematological system:</p> <ul style="list-style-type: none"> • Anemia, leucocytosis • Eosinophilia • Marrow infiltration <p>Skeletal system:</p> <ul style="list-style-type: none"> • Osteoporosis, osteosclerosis | <ul style="list-style-type: none"> • Mast cells • Oval, spindle shaped • Metachromatic granules | <ul style="list-style-type: none"> • Depends on the extent of disease | <ul style="list-style-type: none"> • As for urticaria pigmentosa |

Table 3
Mastocytoses (contd...)

| Tumor | Clinical features | Histopathology | Prognosis | Work up |
|---|---|--|--|---|
| Malignant mastocytosis [24-26] | | | | |
| Lymphadenopathic with eosinophilia | <ul style="list-style-type: none"> • Extremely rare • Depend on the system/extent of involvement | <ul style="list-style-type: none"> • Cytological atypia • Architectural atypia | <ul style="list-style-type: none"> • Depends on the extent of disease | <ul style="list-style-type: none"> • As for urticaria pigmentosa • Chemotherapy |
| Mast cell leukemia | | | | |
| Childhood mastocytosis | | | | |
| Childhood mastocytosis | <ul style="list-style-type: none"> • Half of all cases of mastocytosis • Urticaria pigmentosa is the commonest presentation • Frequent bullous lesions • Resolution by puberty • Aggressive disease and malignancy is rare | <ul style="list-style-type: none"> • Mast cells • Oval, spindle shaped • Metachromatic granules | <ul style="list-style-type: none"> • Good | <ul style="list-style-type: none"> • As for urticaria pigmentosa |

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