

4.7.3 Cutaneous mastocytoses

Definition Localized or diffuse cutaneous proliferations of mast cells, sometimes associated with systemic disease

Clinic

- Mastocytoma: Single or several red-brown papules or nodules, usually in children
- Urticaria pigmentosa (UP): Numerous symmetrical macular, papular or plaque-like, usually red-brown lesions
- Telangiectasia macularis eruptiva perstans (TMEP): Disseminated small tan macules admixed with telangiectases

All of these lesions may urticate after mechanical irritation (Darier sign)

Histopathology

Mastocytoma

- *Dense circumscribed infiltrate of mast cells* in upper and mid-dermis
- Mast cells have round nuclei and basophilic cytoplasmic granules if not degranulated

UP/TMEP

- Epidermis with increased basal layer melanin
- Discrete lymphocytic perivascular infiltrate in upper dermis with *increased numbers of perivascular mast cells* and admixed eosinophils
- Ectatic capillaries

Additional studies

Metachromatic granules seen better with Giemsa or toluidine blue stain. Mast cells stain with chloroacetate esterase (Leder stain), CD117 (c-kit), and mast cell tryptase

Differential diagnoses

- **Mastocytoma**
 - Lymphoproliferative disorders: No granular cytoplasm, no c-kit reactivity. Expression of lymphocytic markers
 - Granular cell tumor: Tumor cells are S100-positive and granules are not Giemsa-positive
- **UP/TMEP**
 - Urticaria: More eosinophils, also found between collagen bundles and in capillaries
 - Drug reaction: Lymphocytes and eosinophils, but fewer mast cells

Comment

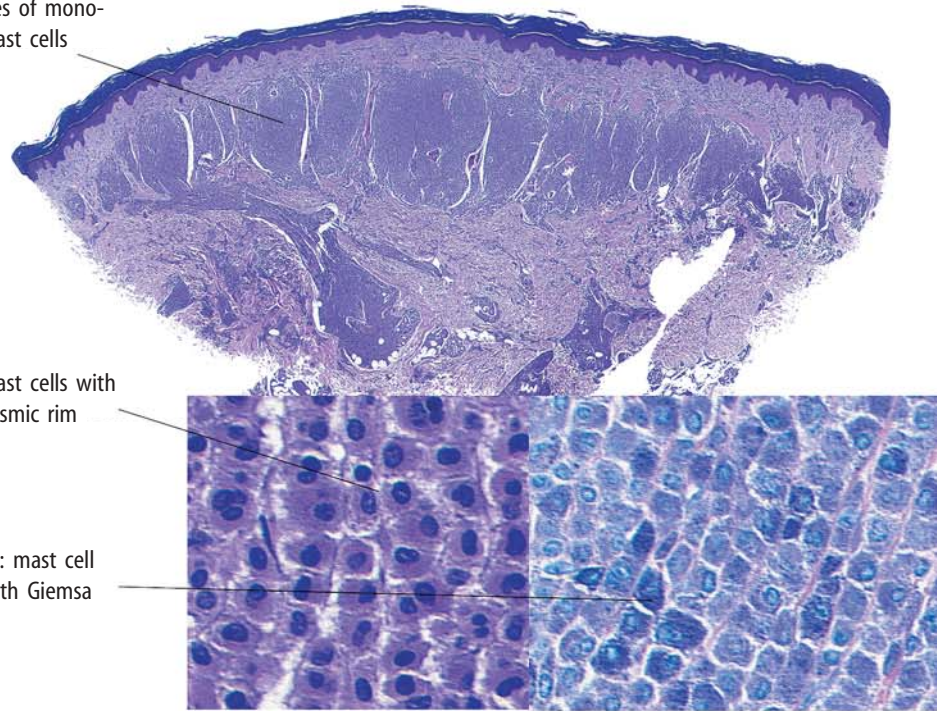
In UP/TMEP mast cells are very hard to see in H&E sections, especially if degranulated by manipulation or biopsy technique. There is no absolute number of mast cells per high-power field required for the diagnosis – numbers between 6 and 10 have been proposed. When diffuse clusters of mast cells are associated with basal layer hyperpigmentation, eosinophils or telangiectases, mast cell disease is likely.

Mastocytoma

dense nodules of monomorphous mast cells

polygonal mast cells with wide cytoplasmic rim

Right panel: mast cell granules with Giemsa stain



Urticaria pigmentosa

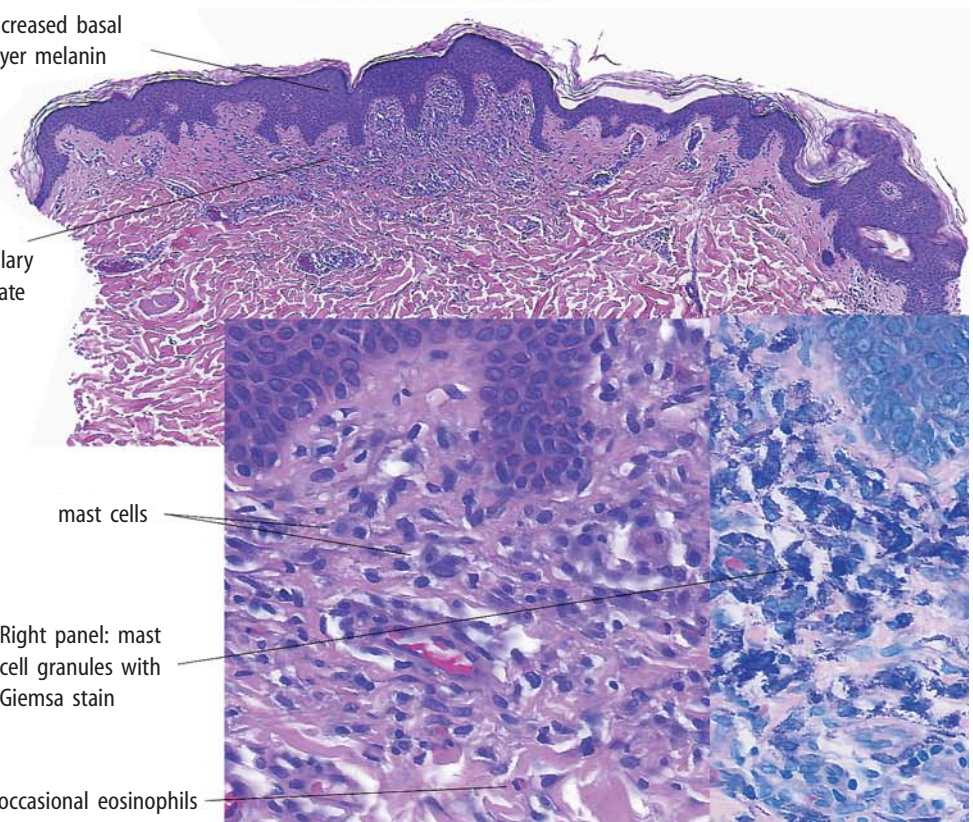
increased basal layer melanin

discrete papillary dermal infiltrate

mast cells

Right panel: mast cell granules with Giemsa stain

occasional eosinophils



Dermatopathology

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