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mastocytosis
Urticaria pigmentosa, although occasionally showing an autosomal dominant mode of transmission (with...
In the first form, the cutaneous lesions often improve or even clear at puberty. Systemic lesions are absent as a rule...
Patients with extensive mast cell infiltration of the skin or the internal organs commonly have attacks of flushing, palpitation, or diarrhea as a result of degranulation of mast cells and the release of histamine.

Five types of cutaneous lesions are seen in urticaria pigmentosa. Two types can occur in both the infantile and the chronic cutaneous types: urticarial type (erythematous papules) and the telangiectatic or telangiectatic type (small telangiectatic vessels). If bullae are a predominant clinical feature, the term bullous mastocytosis has been applied. Although visceral lesions are common in the diffuse erythrodermic type,
they usually improve and only very rarely progress to fatal systemic mast cell disease. On rare occasions,
Histopathology

In all five types of lesions, the histologic picture shows an infiltrate composed chiefly of mast cells, which are often seen in large numbers, especially in the upper dermis. The mast cells can be demonstrated by various stains, such as a Giemsa stain or toluidine blue. Another method, called Leder's method, uses naphthol AS-D chloroacetate esterase to make mast cell granules appear red and thus quite conspicuous.
In the maculopapular type and in telangiectasia macularis eruptiva perstans, the mast cells are limited to the upper third of the dermis and are generally located around capillaries. In some mast cells, the nuclei may be round or oval, but in most mast cells, they are spindle shaped. Because the mast cells may be present only in small numbers, the diagnosis may be missed unless special staining is employed.
In cases with multiple nodules or plaques or with a solitary large nodule, the mast cells lie closely packed.
In the diffuse, erythrodermic type, a dense, bandlike infiltrate of mast cells with a rather uniform appearance showing round to oval nuclei and a distinctly outlined cytoplasm is observed in the upper dermis.

Eosinophils may be present in small numbers in all types of urticaria pigmentosa with the exception of telangiectasia. In some cases, mast cell granules are observed, which is an indication that granules have been released by the cells (327).

The bullae that may occur in infants with multiple or solitary nodules or with the diffuse erythrodermic type...
basal cell layer and occasionally also of melanophages in the upper dermis.

**Systemic Lesions**

It is important to distinguish between asymptomatic systemic involvement of limited degree and true system...
Asymptomatic systemic involvement of a limited degree may occur in urticaria pigmentosa of children, but...
In true systemic mast cell disease, massive infiltration of the bones may cause collapse of several vertebrae.
Histogenesis. As seen by both light microscopy and electron microscopy, the mast cells of urticaria pigmentosa resemble those of bullous mastocytosis. Lesional skin biopsies reveal that the mast cells of urticaria pigmentosa contain microgranules which differ from those of bullous mastocytosis.

The increased melanin pigmentation in lesions of urticaria pigmentosa is the result of stimulation of epidermal Langerhans' cells and melanocytes by mast cell mediators.
Differential Diagnosis

Even if numerous mast cells are present, an absolutely
The macular type of urticaria pigmentosa, especially telangiectasia maculosa eruptiva perstans, occasionally...
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