Bullous mastocytosis = اﻠﻔﻘاﻌﻲ اﻠﺒدﻴﻨﺔ داء mastocytosis

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 in the first form, and if any, usually develop later. However, in the second form, both skin and systemic manifestations are present and the disease is fatal.
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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 mastocytosis can be recognized. Although visceral lesions are common in the diffuse erythrodermic type,
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they usually improve and only very rarely progress to fatal systemic mast cell disease. On rare occasions,
In all five types of lesions, the histologic picture shows mast cell infiltration, which is characterized by the presence of mast cells in the dermis. The mast cells can be identified using Giemsa stain or toluidine blue. Additionally, the method using naphthol AS-D chloroacetate esterase, often called Leder's method, makes the 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...
Bullous mastocytosis = اﻠﻔﻘاﻌﻲ اﻠﺒدﻴﻨﺔ داء

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. If significant, the number of eosinophils is strongly indicative of a diagnosis of mastocytosis (327). The pigmentation of lesions of urticaria pigmentosa is due to the presence of increased amounts of melanin in the...
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 systemic mast cell disease, in which the lesions are symptomatic, widespread, and progressive.
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 differ from normal mast cells in that the former are densely packed with cellular granules. Studies of the cutaneous lesions of urticaria pigmentosa reveal a considerably higher level of histamine than is found in normal skin.

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

Even if numerous mast cells are present, an absolutely
The macular type of urticaria pigmentosa, especially telangiectasia maculosa eruptiva perstans, occasionally...
Bullous mastocytosis = "اﻠﻔﻘاﻌﻲ اﻠﺒدﻴﻨﺔ داء خلد"