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Painless lesions and an odd biopsy result

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A 41-year-old man presented with an 18-month history of asymptomatic macules on his central trunk. They had steadily increased in number, covering more and more area over time. The patient denied having been ill before the onset of the condition, had not taken any medicines, and was generally in excellent health. He reported no family history of similar conditions.

Examination revealed a rather dense, evenly distributed collection of ovoid 3- to 5-mm macules, tan to brown in color, covering most of his central trunk and spilling over onto his arms and upper legs (see Figure 1). The condition spared his face, palms, and soles entirely. Darier's sign was absent when the lesions were stroked. No nodes were palpable on the head, neck, axillae, or groin, and no abdominal organomegaly was present. Punch biopsy showed increased numbers of mast cells in superficial perivascular locations, as well as increased melanin in the basal layer of the epidermis.

What is your diagnosis?

- Telangiectasia macularis eruptiva perstans
- Dermatoheliosis
- Rosacea
- Liver disease

Discussion

The correct answer is telangiectasia macularis eruptiva perstans (TMEP), a rare manifestation of cutaneous mastocytosis. All subtypes of this disorder are idiopathic and include mastocytoma, urticaria pigmentosa, the diffuse and erythrodermic mastocytoses, and systemic mastocytosis. TMEP produces minimal symptoms and is self-limiting. Since all forms of cutaneous mastocytosis are caused by the accumulation of mast cells, degranulation can occur. This can lead to histamine release, which, in turn, can result in urtication, hives, or even anaphylaxis. Fortunately, TMEP seldom causes anything more than mild itching and cosmetic concern.

Dermatoheliosis is the generic term for chronic sun damage, which can include telangiectasias, but these will

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FIGURE 1

Asymptomatic macules on the central trunk



be confined to directly sun-exposed skin and will not produce lesions. Rosacea can manifest solely with telangiectasias, but these will likewise not be lesional and will be confined to the face, the neck, and—rarely—the scalp. Finally, liver disease can produce telangiectasias that again are not lesional and are confined to the face, neck, and chest.

Reassurance is the best treatment for TMEP, which almost always requires biopsy for confirmation. Even then, the requesting clinician must bring up the possibility of a mast cell disorder on the specimen requisition form, because a special stain is needed and because the changes are often subtle, even when suspected. In cases for which therapy is indicated, either because the patient desires it (usually for cosmetic reasons) or because histamine release produces significant itching and stinging, TMEP can be treated with phototherapy, with H1 and H2 blockers, and by avoidance of known triggers of mast cell degranulation, such as aspirin. Only rarely is enough histamine released that anaphylaxis is an issue. □