

A Middle-aged Man with Diffuse Red Papule: A Quiz

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A 47-year-old male presented with a 2-month history of diffuse, asymptomatic skin eruption involving the face, upper and lower extremities, abdomen, chest, and back. An extensive laboratory workup including complete blood cell count, comprehensive metabolic profile, ANA spectrum, erythrocyte sedimentation rate, serum protein electrophoresis, lipid profile, and angiotensin-converting enzyme level did not reveal any abnormalities. In addition, an age-related malignancy screen including a physical examination, colonoscopy, computed tomography scan of chest and abdomen, chest radiograph, and prostate-specific antigen level were then conducted, which were all within normal limits, and there was no lymphadenopathy or splenomegaly.

On presentation, the patient had hundreds of soft red papules, ranging in size from 0.5 to 1 cm, without scale

crust, symmetrically scattered over the face, trunk, neck, and proximal upper extremities (Fig. 1a, b). The mucous membranes and cutaneous appendages were not involved. Biopsy of skin from the back demonstrated diffuse histiocytic infiltration in the dermis, with a few lymphocytes and neutrophils, and no foam cells or Touton giant cells (Fig. 1c). Immunohistochemically, cells expressed CD68 (Fig. 1d) and lysozyme, but were negative for S-100 protein.

What is your diagnosis?

1. Rosai-Dorfman disease
2. Langerhans cell histiocytosis
3. Generalized eruptive histiocytoma

See next page for answer.

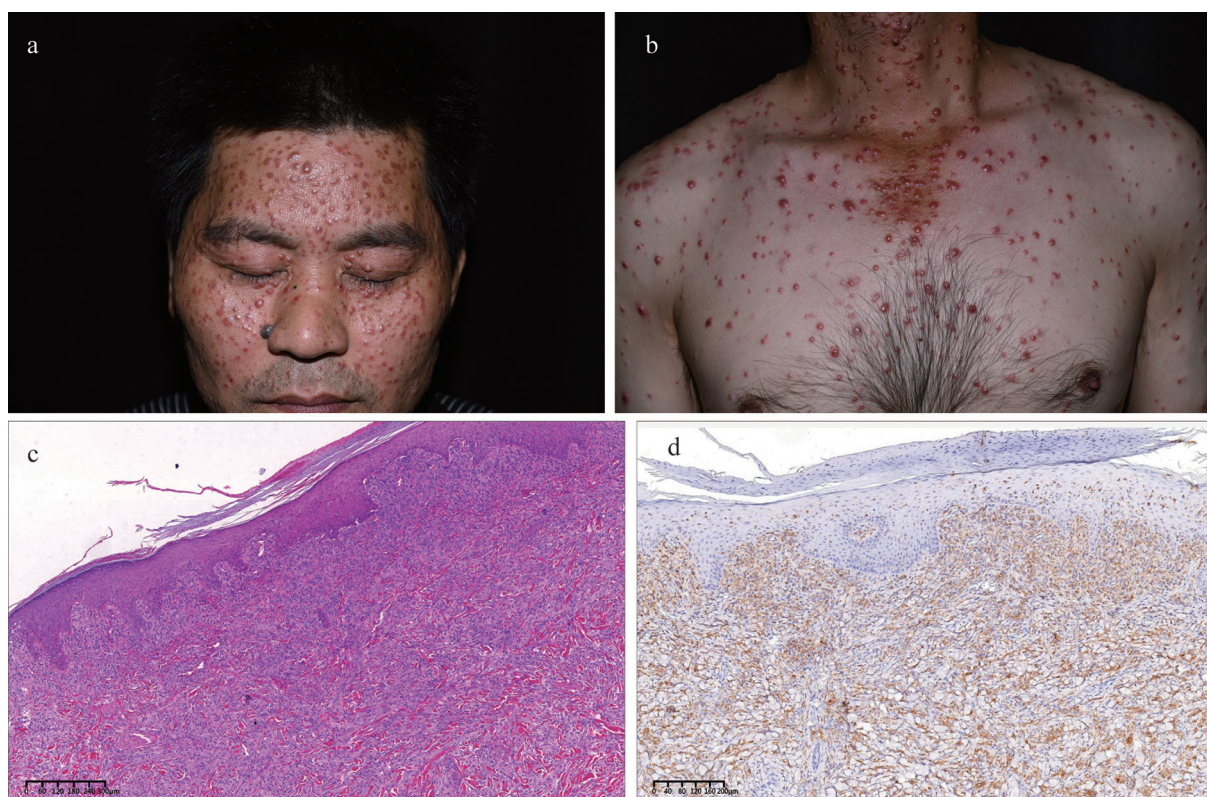


Fig. 1. (a, b) Dense red papules are visible on the patient's face and trunk, (c) shows the hema-toxylin-eosin stain, and (d) displays a CD68 stain.

ANSWERS TO QUIZ

A Middle-aged Man with Diffuse Red Papule: A Commentary

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Diagnosis: Generalized eruptive histiocytoma

Generalized eruptive histiocytoma (GEH), also called non-Langerhans cell histiocytosis, was first reported by Winkelmann and Muller in 1963 (1). Its clinical features include systemic, asymptomatic, multiple red or reddish-brown papules. It usually occurs in the face, trunk, and the proximal limbs, and will not induce systemic damage generally. Moreover, its histopathological findings are monomorphic tissue and cell infiltration within the dermis, cytoplasmic acidophily, clear nuclear membrane and nucleolus, with no foam cells or Touton giant cells. In immunohistochemistry analysis, the tissue-like cells are positive for CD36, but negative for S-100 and CD1a. No Birbeck granule is observed under the electron microscope.

GEH may be an early undifferentiated stage of a different histiocytosis, which can evolve into multiple juvenile type or adult type xanthogranuloma, disseminated xanthoma, progressive nodular histiocytosis, or multicentre reticular histiocytosis (2). Therefore, long-term follow-up is required

for GEH. Most GEH cases are self-limiting, and the rash can spontaneously resolve within months to years, with no trace or only hyperpigmentation after resolution, and generally does not require treatment. Nonetheless, some patients may actively seek treatment due to mental stress. Corticosteroids, PUVA, and isotretinoin have been reported to be effective (3–5), but mainly in case reports. Therefore, whether treatment should be applied must be decided cautiously after sufficient physician–patient communication and benefit–risk balance.

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