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Iria Neri, Annalucia Viridi, Riccardo Balestri, et al.

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Diffuse cutaneous mastocytosis: a heterogeneous disease

A 6-month-old otherwise healthy girl was referred to us for a 3-month history of generalised persistent itching erythema.

Physical examination showed white transient linear oedematous lesions on widespread erythema caused by scratching (figure 1), leading to an initial diagnosis of physical urticaria.

After 1 month, during an acute febrile episode (38.3°C), tense vesicle-bullous lesions with a yellow-citrine fluid appeared on diffuse red-oedematous skin (figure 2).

Histological examination was consistent with mastocytosis.

Initial serum level tryptase was 111 ng/ml; however, instrumental investigations did not reveal any visceral involvement and C-kit molecular study did not show mutations, permitting the final diagnosis of diffuse cutaneous mastocytosis (DCM). After 2 weeks, the vesicle-bullous eruption regressed, but was followed by progressive skin infiltration and marked dermographism (figure 3).

DCM can present with three heterogeneous variants:¹ (i) reddish skin with widespread large bullae; (ii) yellow-orange infiltration and minimal small blisters; and (iii) pachydermia and extremely folded skin.

Dermographism may be the first sign of DCM and presents with a more marked and persistent oedematous reaction to scratching than dermographism related to allergy. Differential diagnosis between DCM dermographism and allergic dermographism is crucial since performing skin prick tests in DCM may induce an anaphylaxis.²

Although serum tryptase may be a marker of systemic involvement, in neonatal DCM it can be very high in the first 2 years of life and then progressively decreases, in spite of an exclusively cutaneous involvement.³ More invasive screening is recommended only if tryptase levels persistently exceed 100 ng/ml or increase and/or other signs of systemic mastocytosis appear.³



Figure 1 Multiple white transient linear oedematous itching lesions on widespread red skin.



Figure 2 Tense blisters containing a yellow-citrine fluid and arisen on erythemato-oedematous and mildly indurated skin.



Figure 3 Marked dermographism, progressive infiltration and thickening of the skin after bullous eruption.

Iria Neri, Annalucia Viridi, Riccardo Balestri, Annalisa Patrizi

Dermatology, Department of Specialised, Experimental and Diagnostic Medicine, Università di Bologna, Bologna, Italy

Correspondence to Dr Annalucia Viridi, Dermatology, Department of Specialised, Experimental and Diagnostic Medicine, Via Massarenti 1, Clinica Dermatologica, Bologna 40138, Italy; alviridi@libero.it

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