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Clinical Image

A Cutaneous Mastocytosis with an Unusual Presentation

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Clinical Image

A 13 year-old patient presented with a 4 years history of skin lesions on the trunk and upper arm. The patient reported episodic increase in redness without any identified trigger factor. Physical examination showed erythematous ill-defined macula with negative Darier sign on the right side of the chest (Figure 1A) and the upper arm (Figure 1B). Differential diagnoses were acquired port wine stain, angioma serpiginosum and the telangiectatic variant of cutaneous mastocytosis (telangiectasia macularis eruptiva perstans).

Histological analysis on Haematoxylin and eosin stain (X20) showed perivascular mast cell infiltrates surrounding slightly dilated blood vessels (Figure 2C1). Immunohistochemical analysis with anti-CD117 antibodies identified a monomorphic dermal mast cell infiltrate (130 cells per mm^2) (Figure 2C2). Serum tryptase was normal.

A diagnosis of cutaneous mastocytosis with unilateral distribution was established.

Cutaneous mastocytosis (CM) is divided into maculopapular CM (MPCM), also termed urticaria

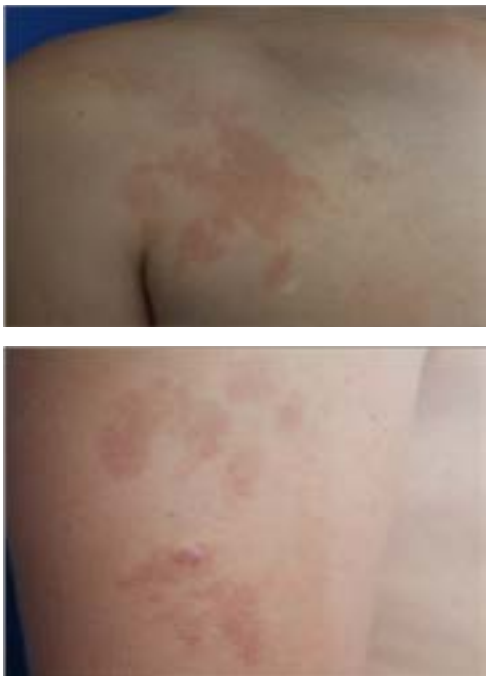

Figure 1: Erythematous ill-defined macula of the trunk (a) and upper arm (b)

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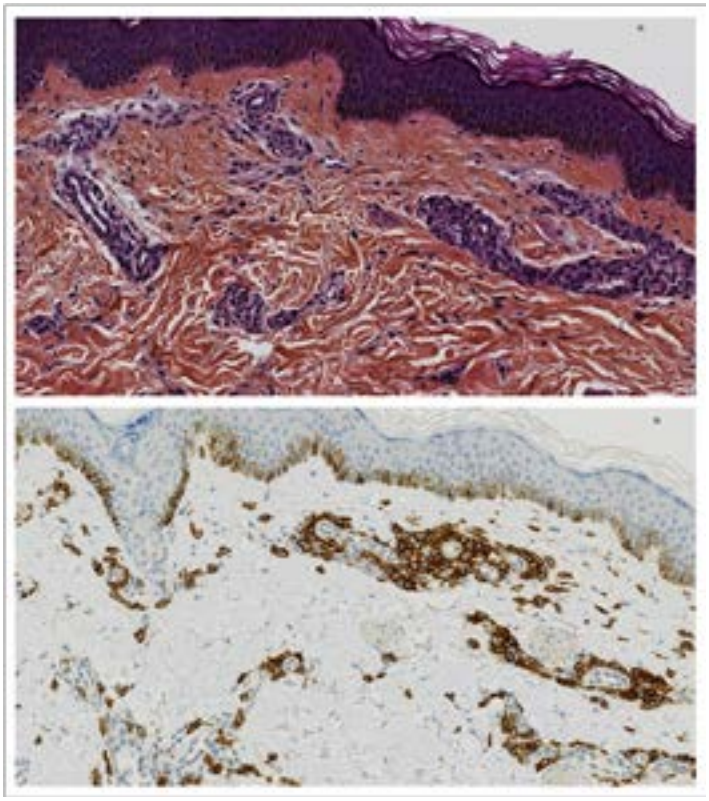

Figure 2: Perivascular mast cell infiltrates surrounding slightly dilated blood vessels (C1) with positive anti-CD117 staining (C2)

pigmentosa, diffuse CM and localized mastocytoma of skin [1].

The latest task force recommended eliminating the telangiectasia macularis eruptiva perstans form. However, the term MPCM does not cover all patients in this category in an optimal manner [2].

The most frequent form of CM in children is the MPCM. CM is usually diagnosed in childhood and has a good prognosis. In many cases, the skin lesions fade away and disappear during puberty [3].

Differential diagnoses are angioma serpiginosum and port wine. Angioma serpiginosum is a rare, naevoid benign vascular proliferation, which starts in early childhood. Clinically it is characterized by multiple red to purple macules grouped in a serpiginous pattern. Histologically, angioma serpiginosum consists of clusters of dilated capillaries lined by thick walls in dermal papillae with no inflammatory cells [4].

Port wine stain is a cutaneous vascular proliferation, which appears clinically as unilateral deep red or purple macules. Histologically, it shows an increased number of dilated and thin-walled capillaries and venules [5].

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