

# Dermatoscopic findings of urticaria pigmentosa\*

## Achados dermatoscópicos na urticária pigmentosa

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**Abstract:** Mastocytosis is a rare disease characterized by proliferation and accumulation of mast cells in various organs. The maculopapular cutaneous mastocytosis is divided into three subtypes: papular/plaque variant, urticaria pigmentosa and eruptive macular telangiectasia perstans. Dermoscopic may help to better characterize the different forms of cutaneous mastocytosis. We report a 55 year-old female with urticaria pigmentosa and its dermoscopy.

**Keywords:** Dermoscopy; Mastocytosis; Mastocytosis, cutaneous; Urticaria pigmentosa

**Resumo:** Mastocitose é uma doença rara caracterizada pela proliferação e acúmulo de mastócitos em vários órgãos. A mastocitose cutânea maculopapular divide-se em três subtipos: variante máculo-papular, urticária pigmentosa e telangiectasia macular eruptiva perstans. A dermatoscopia pode ajudar a caracterizar melhor as diferentes formas de mastocitose cutânea. Relatamos um caso de paciente feminina de 55 anos com urticária pigmentosa e sua dermatoscopia.

**Palavras-chave:** Dermoscopy; Mastocitose; Mastocitose cutânea; Urticaria pigmentosa

### INTRODUCTION

Mastocytosis is a rare disease characterized by proliferation and accumulation of mast cells in various organs (skin, bone marrow, skeletal system, gastrointestinal tract, spleen, lymph nodes and liver). It is classified as cutaneous or systemic mastocytosis. Actually, cutaneous mastocytosis is subclassified into three clinical variants: maculopapular cutaneous mastocytosis, diffuse cutaneous mastocytosis, and solitary mastocytoma. Furthermore, the maculopapular cutaneous mastocytosis is divided into three subtypes: papular/plaque variant, urticaria pigmentosa (UP) and eruptive macular telangiectasia perstans (TMEP).<sup>1,2,3</sup>

Dermoscopy is a noninvasive technique designed to evaluate colour and structures of the epidermis, the dermoepidermal junction and the papillary dermis, which are not seen with the naked eye. One of its main applications is to classify melanocytic and non-melanocytic lesions.<sup>4,5</sup> Several algorithms

have been designed to diagnose melanocytic lesions, and one of the main criteria is the presence of pigmented network. However, this dermoscopic structure is also seen in non-melanocytic lesions such as dermatofibroma, solar lentigo, seborrheic keratosis, accessory nipple, Kaposi's sarcoma and even in the normal skin.<sup>4,7</sup> Recently, two authors reported the dermoscopic patterns of mastocytosis.<sup>8,9</sup> The pigmented network was also described in urticaria pigmentosa, a subtype of maculopapular cutaneous mastocytosis.<sup>8,9</sup>

### CASE REPORT

We report a 55 year-old female with asymptomatic brownish macules and papules in the neck, inframammary area, lower abdomen and thighs with a positive Darier's sign since a year ago (Figure 1). She reported sporadic episodes of epigastric pain, sweating, dizziness, diarrhea and headache. There was no hospitalization history for anaphylaxis and drug

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allergy. Dermoscopy showed a pigment network and a light-brownish blot (Figures 2 and 3). We performed complete blood count, hepatogram and kidney function, tryptase and abdominal ultrasonography which were all normal. A biopsy taken from an abdominal lesion showed hyperpigmentation of the basal layer and dermal grouping of lymphocytes and mast cells, identified with the Giemsa staining, around vessels (Figures 4 and 5).



FIGURE 1: Urticaria Pigmentosa. Asymptomatic brownish macules and papules in inframammary area



FIGURE 2:  
Dermoscopy  
10x. Brownish  
lesions with a  
pigmented  
network



FIGURE 3:  
Dermoscopy  
10x.  
Pigmented  
network and  
light-brown  
blot

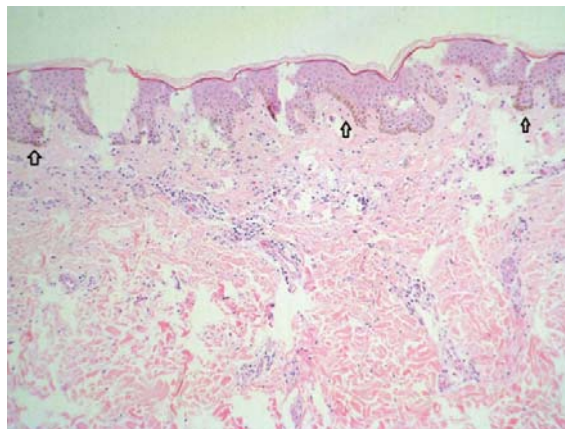


FIGURE 4: Histopathology. Arrow points to hyperpigmentation of the basal layer which correlates with the pigmented network

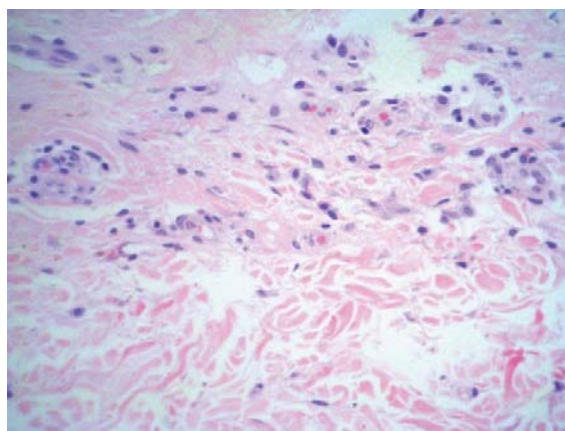


FIGURE 5: Histopathology. Dermal infiltrate composed of mast cells

## DISCUSSION

Urticaria pigmentosa is the most common form of cutaneous mastocytosis. Usually it begins in childhood, however it can affect adults. It is characterized by monomorphic and symmetrical macules, papules, plaques or nodules, without sex predilection. It can affect any area of the body, mainly trunk and thighs. Palms, soles and scalp are usually spared. The complaints of pruritus, erythema, abdominal pain, diarrhea, palpitations, dizziness and syncope are due to the physiological effects of secreted mast cell mediators such as histamine. Thus, patients with systemic complaints should be investigated to rule out involvement of other organs. Urticaria pigmentosa in adults is more likely to progress to systemic disease and its severity is related to the age of onset and to the size of cutaneous involvement.<sup>1,2,3</sup>

Our patient had no systemic involvement so far. She was oriented to avoid anaphylaxis situations that lead to mast cell degranulation such as friction,

exercise, emotional stress, general anesthesia, alcohol, radiological contrast, ionizing radiation and certain medications (nonsteroidal anti-inflammatory, narcotics, polymyxin B and dextran). Furthermore, gastrointestinal symptoms may be controlled by H2 receptor antagonists (sodium cromoglycate and ketotifen), which are also used to reduce the flushing and muscle pain.<sup>1,2</sup>

The diagnosis of cutaneous mastocytosis is confirmed by a dermal infiltrate composed of mast cells that can be better seen with special stains such as Giemsa, toluidine blue or Astra blue.<sup>1,2,3</sup> At present, the subclassification of urticaria pigmentosa is based on clinical history, lesions features and the presence of Darier's sign.<sup>1,2</sup> However, the dermoscopic analysis can be useful to distinguish clinical forms of maculopapular cutaneous mastocytosis. Akay and cols described for the first time two dermoscopic patterns: vessels with linear and reticular pattern in TMEP and pigmented network in UP.<sup>8</sup> Then, Vano-Galvan *et al* identified four dermatoscopic patterns in a study of 127 patients with cutaneous mastocytosis: light-brown blot, pigment network, reticular vascular pattern (consisting of thin reticular telangiectasias) and yellow-orange blot. They emphasized that a predomi-

nant dermatoscopic pattern was observed in patients with multiple lesions. This study found light-brown blot and pigment network more prevalent in patients with maculo-papular mastocytosis, while reticular vessels were mostly seen in patients with TMEP and the yellow-orange blot was more prevalent in mastocytoma.<sup>9</sup>

It is believed that the pigment network seen with dermoscopy is due to a high concentration of mast cell growth factor that stimulates melanocyte proliferation and melanogenesis which leads to hyperpigmentation of basal keratinocytes.<sup>3,8,9</sup>

The dermoscopic findings in our patient with urticaria pigmentosa were similar to those reported by Akay<sup>8</sup> and Vano-Galvan.<sup>9</sup> The dermoscopic patterns found on the brownish lesions were a pigmented network and light-brown blot. Moreover, the biopsy showed the presence of basal hyperpigmentation which correlates with the pigmented network. Dermoscopy is easy to be performed and the features of mastocytosis are reproducible, which helps to better characterize the different forms of CM and set them apart from other exanthematous skin diseases. □

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