Scabies masked by Systemic Mastocytosis

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INTRODUCTION

- Scabies is an intensely itchy dermatosis caused by the mite *Sarcoptes scabiei var hominis.* The infestation occurs at all ages and it is a common public health problem in poor communities, being widespread in many underdeveloped countries. Since the disease is transmitted by skin-to-skin contact, sexual transmission is common, as is non-sexual spread in family groups. Cutaneous lesions are usually characterized by papules, vesicles, pustules, and nodules. The pathognomonic sign is the burrow; a short, wavy, scaly, grey line on the skin surface. They are often missed if the skin has been scratched, has become secondarily infected, or if eczema is present.
- On the other hand , mastocytosis is a rare and heterougeneous clinical entity, characterized by mast cell (MC) infiltrattion of skin and organs (most commonly bone marrow) . Based on the WHO's classification 2016 can be divided: indolent systemic mastocytosis, aggressive systemic mastocytosis, mastocytosis associated with a hematologic non–mast-cell - lineage disease, and mast-cell leucemia. Skin involvement is observed in around 80% of patients with indolent systemic mastocytosis, but it is less present in aggressive systemic mastocytosis, and is rarely seen in mast-cell leukemia.

CASE REPORT

A 59-year-old woman has had a diagnosis of indolent systemic mastocytosis with cutaneous manifestation, with red-brownish maculopapular lesions (urticaria pigmentosa) for 20 years. However, the disease evolved to agressive systemic mastocytosis (bone lytic lesions) three years ago. She scheduled an extra appointment due to the worsing of her skin lesions in the last month, which were being treated with topical corticosteroid and oral H1 antihistamines, without improvement of the condition. On physical examination, the patient presented intensive itching and skin lesions characterized by papules and nodules, excoriation, and crusts widespread over red-brownish papules in her neck, thorax, abdomen and lower limbs **(*Fig.1* a,b,c,d).** The nodules were fixed and by rubbing them, an urtication with an erythematous halo (*Darier's sign*) could be visualized. The patient's husband also had similar symptoms. Skin scraping confirmed the diagnosis of Scabies (***Fig.2***) in both patient and partner. Treatment was done with 200 µg/kg of oral Ivermectin (patient and family contacts), a single dose, which was repeated after 14 days. Permethrin 5% lotion was also applied for 3 nights and repeated after 7 days.

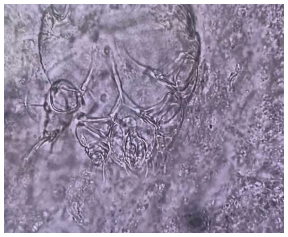

Figure 2. *Microscopic skin examination with 10% potassium hydroxide revealed Sarcoptes scabiei var. hominis.*

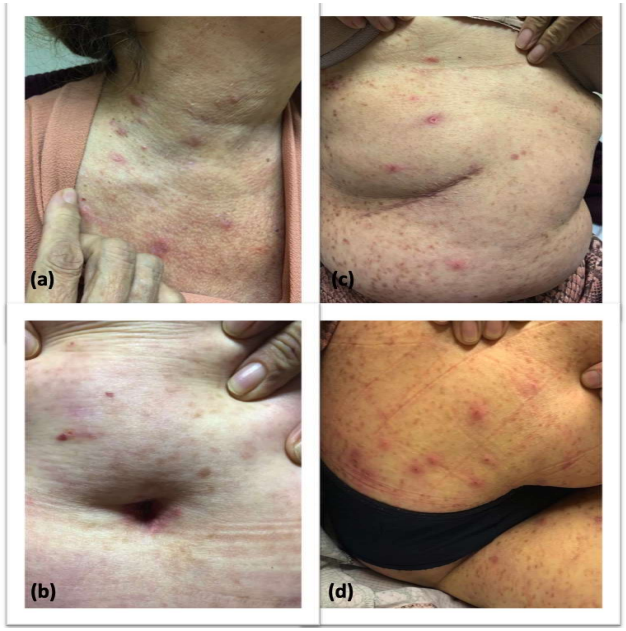

Figure 1*. Phototype III (Fitzpatrick,1988) with persitent eruption of macules and papules range from 3 to 4 mm in diameter, with red-brown hue.* ***(a)*** *Excoriated erythematous nodules in the right neck;* ***(b)*** *Crusty umbilicated erythematous papules in the abdomen;* ***(c)*** *Erythematous papules with vesicles around umbilicus area;* ***(d)*** *Excoriated erythematous crusted plaques in portions of the buttocks and lower limbs.*

DISCUSSION

Although our patient had a history of chronic pruritus and skin lesions due to aggressive systemic mastocytosis, the diagnostic suspicion was based on the observation of new lesions, which, although atypical, could be compatible with scabies, in addition to having a typical distribution and suggestive familial epidemiology.

- Two aspects made this case relevant and difficult to manage: the coexistence of mastocytosis and scabies lesions, and the atypical appearance of scabies lesions, much more erythematous-edematous than in patients without mastocytosis.
- Both types of her lesions share clinical findings and a positive Darier's sign. However, differences can be established since urticaria pigmentosa is more frequently observed as red to brown macules, papules and sometimes nodules and plaques over the trunk. The exclusive presence of nodular lesions is rare. Nodular scabies presents as reddish-brown pruritic, highly infiltrated nodules located mostly in intertriginous and covered areas. Our case is to highlight pitfalls that can be present in patients with Systemic Mastocytosis and others skin conditions.

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Comments from readers would be greatly appreciated.

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