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Case Report
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# A Case Report of Alopecia in a Patient with Cutaneous Mastocytosis

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## Abstract

Cutaneous mastocytosis is a condition characterized by increased numbers of mast cells in the skin. Atypical clinical features may mimic impetigo, Langerhans cell histiocytosis, and carcinoid syndrome; however, only 1 case of scarring alopecia associated with mastocytosis has been reported. We present a case of cutaneous mastocytosis associated with alopecia areata in a 9-year-old Iranian-boy. This case showed an atypical clinical presentation of mastocytosis, but histopathological results confirmed the diagnosis of cutaneous mastocytosis.

**Keywords: Alopecia; Cutaneous Mastocytosis; Urticaria pigmentosa**

## Background

Cutaneous mastocytosis is a condition characterized by increased numbers of mast cells in the skin. Mast Cells (MCs) are part of the immune system [1]. Mast cell proliferation and accumulation within different organs, most frequently the skin, are symptoms of the disorder mastocytosis [2]. The most common cutaneous forms are generalized Urticaria Pigmentosa (UP), solitary mastocytoma, diffuse erythrodermic form, telangiectatic macularis eruptive perstans (paucicellular), and pseudo xanthomatous form. Systemic mastocytosis (with or without skin involvement), mastocytosis linked to hematological disorders, lymphadenopathic mastocytosis with eosinophilia, and mast cell leukemia are other types [3]. A positive Darier's sign and typical lesions point to the diagnosis of cutaneous mastocytosis [4]. Atypical presentations may have constitutional symptoms resembling carcinoid syndrome, eruptive xanthomas, impetigo, Langerhans cell histiocytosis, or other conditions [4]. We present the instance of indolent mastocytosis masquerading as cicatricial alopecia.

## Case Presentation

The patient is a 12 years old boy with a 9 years history of alopecia on scalp and body (Figure 1). The lesions were red-brown and itchy. On physical examination he had a positive Darier's sign. There were no signs of involvement of other organs, including digestive, respiratory and blood pressure drop. Pathology report indicates that received specimen in formalin consisted of one fragment of skin tissue measuring 0.3 cm × 0.2 cm in thickness. The skin tissue was with unmarkable epidermis. The underlying dermis revealed mild interstitial edema with a mild infiltrate of lymphocytic inflammatory cells admixed with some eosinophils around small blood vessels with prominent endothelial cells mostly in superficial and mid dermis. There is spindle cell proliferation in dermis that show metachromatic granules in methylene blue stain (Figure 2, 3). According to the clinical suspicion, a skin biopsy was performed for him. Our diagnosis was cutaneous mastocytosis verified by methylene blue staining. The patient has been under FM and supportive care since then and is under control.

About six months ago, he experienced an increase in itching in the scalp area, which in the initial examination was in the form of a scaly macule, followed by an irregular and asymmetric area of scalp alopecia in subsequent visits.

## Discussion

Mast cells are mainly found in the skin and lining of the bronchopulmonary tree and gastrointestinal tract [5]. Mast cells are derived from multipotent myeloid progenitors and mature under the influence of c-kit ligands and stem cell factors, in the presence of diverse other growth factors provided by the tissue microenvironment in which they reside [6].

Cutaneous mastocytosis is a type of mastocytosis that primarily affects the skin. There are three

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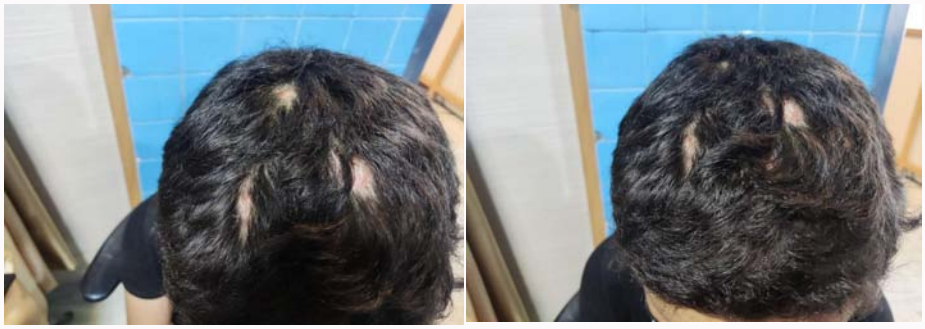

**Figure 1:** Clinical picture of patient's scalp showing scarring alopecia.

**Figure 2:** The underlying dermis reveals spindle cell proliferation in dermis that show metachromatic granules in methylene blue stain (a: print magnification, x105) (b: print magnification, x1050).

main forms of the condition: Maculopapular cutaneous mastocytosis (also called urticaria pigmentosa), solitary cutaneous mastocytoma, and diffuse Cutaneous mastocytosis [7]. There is also an extremely rare form called telangiectasia macularis eruptiva perstans. The signs, symptoms, and severity of the condition vary by subtype. Cutaneous mastocytosis is usually caused by alterations in the KIT gene [8].

As a result of certain stimuli, such as parasites or insect bites, mast cells release various chemicals, including histamine [9]. Histamine dilates blood vessels and can cause soft tissue swelling [9]. Cutaneous mastocytosis is a relatively rare condition. Cutaneous mastocytosis is caused by a genetic mutation [10]. Cutaneous mastocytosis constitutes 90% of cases of mastocytosis that are not associated with a hematologic disorder [11]. Urticaria pigmentosa is the most common variety, occurring in about two thirds of patients with cutaneous mastocytosis. Humans have about 30,000 to 40,000 different genes [12]. Some of these genes have defects that can affect body processes and functions [13]. Mastocytosis gene mutations increase the production of mast cells by the bone marrow [13]. They usually occur as sporadic or isolated changes. It is not normally passed from parent to child [13]. Mastocytosis occurs in all races, and there is no sex predilection [14]. The peak incidence is during infancy and early childhood, with a second peak occurring in middle age [14]. The disease may be benign, with minor transient symptoms and signs that never cause the patient to consult a physician, or it may be life-threatening [9]. It may be apparent at birth; it may appear during the first few months of infancy or it may have an onset at any age thereafter [9]. About one third of patients first develop the characteristic lesions in adulthood [9].

Previous studies have shed some light on the causes of mast cell accumulation in skin. Normal epidermal keratinocytes produce mast

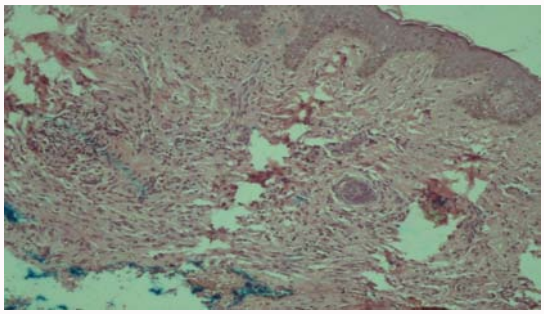

**Figure 3:** Dense dermal mast cell infiltration reveals spindle cell proliferation in dermis that show metachromatic granules (print magnification, x105).

cell growth factor, also known as stem cell factor and c-kit ligand, which binds to the c-kit receptor on mast cells and probably causes their proliferation and accumulation [12]. Xu et al. have suggested that alopecia and mast cell disease might be associated [4]. Mast cell granules are rich in cytokines, growth factors such as fibroblast growth factor, and vascular endothelium-derived growth factors [5]. The receptors of these growth factors have a role in regulating development and cycling of hair follicle; therefore, it is possible that alopecia may be consequence of mast cell disease [5]. According to the previous reports, CM patients may have mutations of c-kit a receptor of growth factor. The stem cell factor on mast cells, causes unregulated growth factor activation which results in increased proliferation and decreased apoptosis of mast cells [2]. And recently it was also reported that c-kit has a role in hair growth; therefore, upregulated c-kit in the hair follicle cells can trigger AA and androgenetic alopecia [2]. C-kit is also expressed in melanocytic lineage cells, so hyperactivation of c-kit can cause hyperpigmentation of affected skin, and this finding

can help to explain tan- to reddish brown–colored maculopapules, which is a typical finding in CM, especially in urticaria pigmentosa [2]. In summary, cicatricial alopecia with increased numbers of mast cells on scalp biopsy guarantees the diagnosis of mastocytosis. This can be confirmed by testing urine for elevated levels of histamine metabolites [7].

## Conclusion

The scalp biopsy from this patient revealed mild interstitial edema with a mild infiltrate of lymphocytic inflammatory cells admixed with some eosinophils around small blood vessels with prominent endothelial cells mostly in superficial and mid dermis and increased numbers of mast cells. In conclusion, scarring alopecia with increased mast cell numbers on scalp biopsy justifies the diagnosis of mastocytosis.

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