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Rare presentation of adult-onset cutaneous mastocytoma of the breast

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Abstract

Mastocytosis is a rare condition in which mast cells accumulate throughout various organs of the body—the most common subtype being confined to the skin. We present an unusual case of cutaneous mastocytosis localized to the unilateral breast of a young woman with partial involvement of the areola. Previously diagnosed as nipple eczema, the patient failed appropriate treatment with class III and IV topical corticosteroids. Given it was adult onset, failed appropriate treatment, and had an atypical clinical appearance, a biopsy was pursued that revealed mastocytosis in skin. This is another clinical diagnosis dermatologists may consider in their differential diagnosis of nipple dermatitis.

Keywords: cutaneous mastocytosis, eczema of breast

Introduction

Mastocytosis is a rare condition in which mast cells proliferate in an uncontrolled manner in various areas of the body. Accumulated mast cells are able to release various vasoactive and inflammatory substances when triggered by physical stimulation or heat. Various mechanisms of pathogenesis have been proposed, such as activating *c-kit* mutations, alteration of growth factors, or dysregulation of apoptosis, but the exact cause for increased mast cell count is largely unknown. The prevalence of this condition in the general population is also unknown. Mastocytosis can be broadly divided into systemic mastocytosis (SM) with involvement of multiple organs or cutaneous mastocytosis (CM) when confined to the skin. The further classification of CM is continually evolving but was most recently identified as maculopapular CM, diffuse CM, mastocytoma, and telangiectasia macularis eruptive perstans [1]. Across all subtypes, the most common clinical presentation is pruritus of mastocytic lesions and the diagnostic Darier sign—the development of localized erythema and whealing following physical irritation of lesions owing to mast cell degranulation [2]. Cutaneous mastocytosis can be diagnosed based on clinical signs and symptoms but should be confirmed through skin biopsy to rule out secondary causes of increased mast cells. Adult-onset mastocytosis in skin is generally considered a manifestation of SM, as isolated CM occurrence is rare outside of childhood; some guidelines advise bone marrow evaluation for all such cases [3,4]. Mastocytosis of the breast has been rarely associated

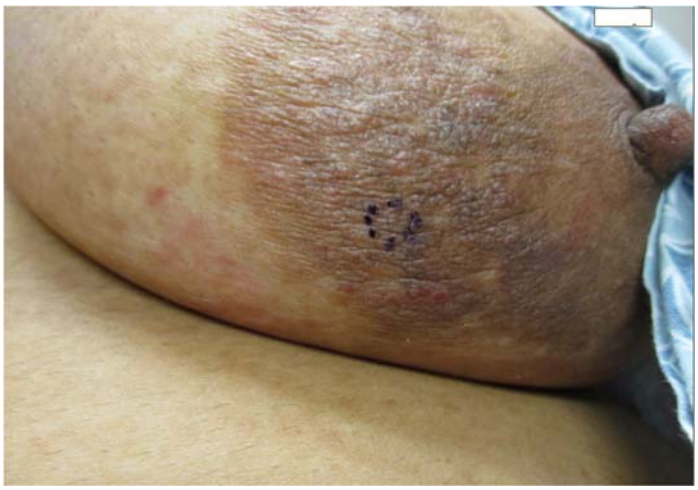

Figure 1. *Left breast rash with biopsy site indicated.*

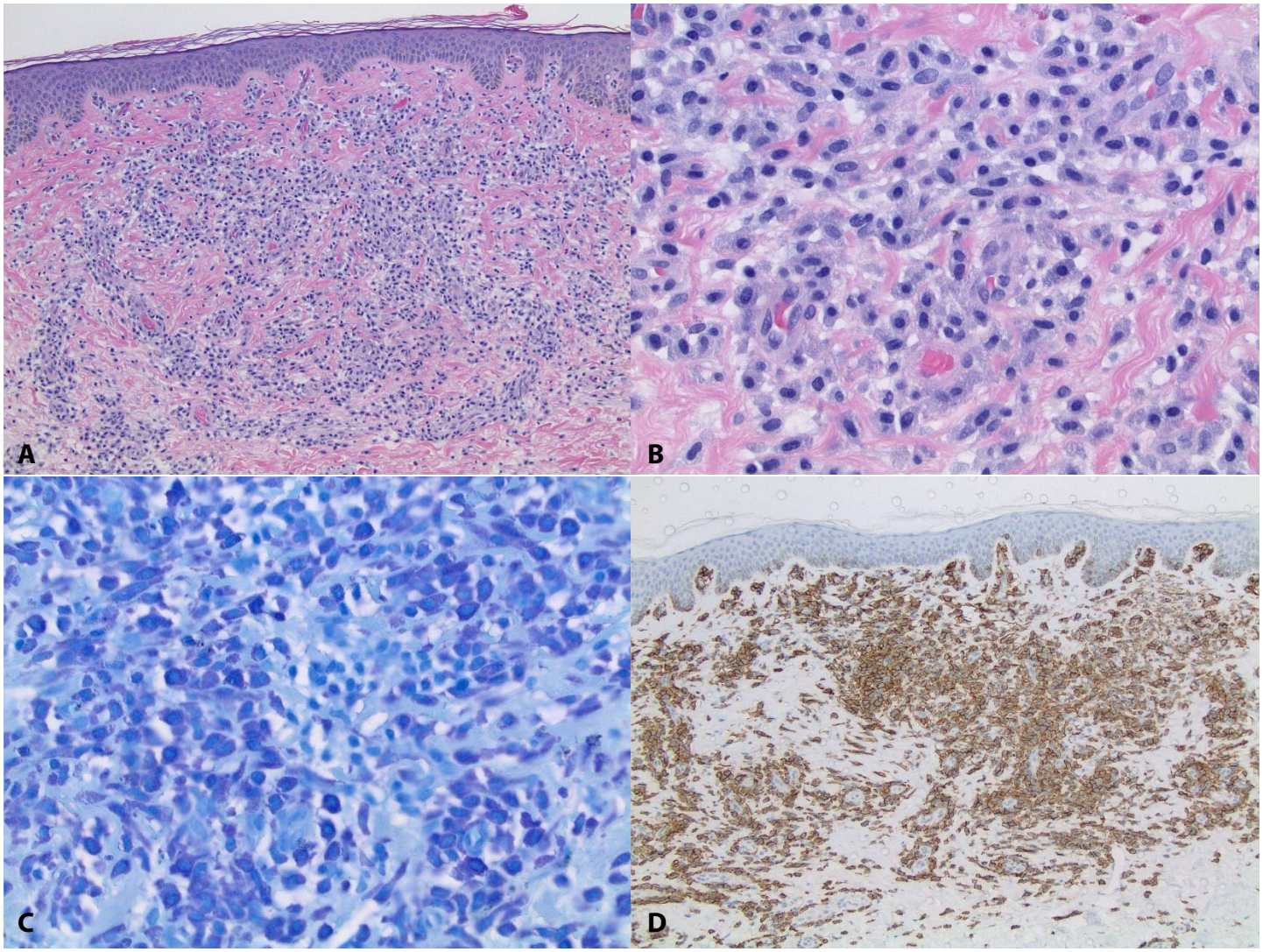

Figure 2. *Dermatopathology results from left breast punch biopsy, consistent with cutaneous mastocytosis.* ***A)*** *Perivascular and interstitial infiltrate of discohesive cells with monotonous appearance. H&E, 100×.* ***B)*** *Mast cells showing characteristic central nucleus and granular cytoplasm. H&E, 400×.* ***C)*** *Giemsa stain highlights metachromatic granules in mast cells, 400×.* ***D)*** *CD117 highlights perivascular and interstitial mast cells in clusters, 100×.*

with underlying conditions of the breast including underlying malignancy and breast hypertrophy, but also has been reported following radiotherapy [5-9]. Herein, we present an unusual presentation of CM of the unilateral breast previously treated as eczema.

Case Synopsis

A 35-year-old woman presented to the dermatology clinic with a rash on her left breast that began a few years prior. She reported pruritis of the lesion with subsequent blistering. The rash was initially believed to be a form of eczema but had a poor response to topical corticosteroid application. The patient had a family history of breast cancer in both grandmothers, with one diagnosed in her 50s. Upon skin examination, the patient had a large brown-pink smooth infiltrative dermal plaque involving areolar skin on her left breast (**Figure 1**). The patient failed prior treatment with class three and four topical corticosteroids. Owing to the atypical clinical presentation of an infiltrative plaque and failure to clear with appropriate moderate strength topical corticosteroids, a punch biopsy was performed. The differential diagnosis included atopic dermatitis, contact dermatitis, erosive adenomatosis of the nipple, and Paget disease.

Microscopic findings and clinical course

Microscopic evaluation demonstrated perivascular and interstitial infiltrates of discohesive cells with a

monotonous appearance. CD117 immunostaining and Giemsa special staining highlighted the increased clusters and metachromatic granules of mast cells (**Figure 2**). The tissue had negative Cam5.2 and pan-keratin cocktail stains. These findings were consistent with CM and diagnostic labs to rule out SM consisting of complete blood count, thyroid panel, lipid panel, and tryptase level were all within normal limits. The patient also denied any systemic symptoms such as hives or flushing. Further evaluation with bone marrow evaluation and mammogram were recommended to the patient, though she declined. Treatment for CM was started and the patient was instructed to apply fluocinonide 0.05% ointment in combination with tacrolimus 0.1% ointment. She was instructed to continue clinical breast examinations with primary care and follow up a mammogram.

Case Discussion

Adult-onset mastocytosis in skin is generally considered a manifestation of SM, as isolated CM occurrence is rare outside of childhood. Therefore, some guidelines advise bone marrow evaluation for all such cases [3, 4]. In a case report from 2016, Cohen describes a 38-year-old woman found to have a solitary cutaneous mastocytoma on her right abdomen, a subtype of CM that is almost exclusively seen in children [10]. The plaque was described as asymptomatic, mimicking a dysplastic nevus. Further diagnostic studies did not fulfill criteria for SM. Cohen summarizes 16 total adult-onset cutaneous mastocytoma cases that have been described throughout the literature, none of which were associated with SM [10]. Given that the current treatment guidelines that do not indicate systemic workup for childhood mastocytoma and the scarcity of documented adult-onset mastocytoma cases, Cohen concludes with recommendations for a conservative work-up of adult-onset solitary mastocytosis [11, 10, 4]. Cutaneous mastocytomas are generally described as sharply defined nodules that demonstrate urticaria with rubbing [12]. Although our patient presented with an isolated cutaneous breast lesion demonstrating mastocytosis, the clinical appearance of the lesion was not consistent with previously described cases of cutaneous mastocytoma and was deemed a very unusual presentation of CM that does not fit pre-existing categories. The biopsy of the plaque was obtained because of the inconsistency with classic eczema and was not expected to demonstrate mastocytosis in the skin. Given the lack of systemic findings and the patient's refusal of bone marrow evaluation, staging was kept to a minimum in alignment with isolated CM recommendations. The patient was treated topically for her CM [11, 4].

A literature search for previous cases of isolated adult onset CM localized to the breast yielded a very limited number of documented cases involving the female breast. One previous case describes a 33-year-old woman with mastocytosis associated with bilateral mammary hypertrophy and no cutaneous lesions [7]. Molderings et al. describe a 70-year-old woman with a history of treated breast carcinoma who presented years later with SM involving her vertebrae [9]. In addition, Setia et al. describe a 59-year-old woman who had a macular rash appear over her breast and chest years after radiotherapy, which led to the diagnosis of SM with bone marrow involvement [8], and Soilleux et al. describe a similar occurrence of telangiectasia macularis eruptiva perstans lesions in a 62-year-old woman over a radiotherapy field involving bilateral breasts [6]. To our knowledge, a patient with a solitary plaque of the breast has not been described in the literature and provides an interesting diagnostic consideration for the differential diagnosis of nipple dermatitis.

Conclusion

This is a unique presentation of CM localized to one breast with partial involvement of the nipple. The condition was adult onset and caused pruritis that did not improve with standard topical corticosteroid treatment. Given this documented rare case of CM over the breast, the diagnosis may be considered in lesions that appear similarly to eczema but are atypical or do not resolve with corticosteroids. It is important to note that any pruritic eruptions of the breast, particularly involving the nipple, should be investigated to rule out Paget disease or metastatic

breast cancer before considering more benign conditions such as chronic eczematous dermatitis, allergic contact dermatitis, or CM.

Potential conflicts of interest

The authors declare no conflicts of interests.

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