Unilateral Telangiectasia Macularis Eruptiva Perstans of the Breast

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We report a case of a 22-year-old female with an asymptomatic telangiectatic rash involving her left breast of 10 years' duration. Biopsies revealed findings consistent with telangiectasia macularis eruptiva perstans (TMEP). Telangiectasia macularis eruptiva perstans most often presents in a symmetric fashion; our patient represents an unusual case of unilateral TMEP involving the breast. Therefore, TMEP should be considered when a patient presents with telangiectasia, even if the presentation is unilateral.

Cutis. 2012;90:26-28.

Case Report

A 22-year-old white female presented to our institution with an asymptomatic rash of 10 years' duration involving her left breast that had not appreciably changed since its initial appearance. Other than occasional localized flushing, the rash was asymptomatic. She denied flushing in other areas, pruritus, gastrointestinal symptoms, syncopal episodes, weight loss, or bone pain. Prior treatment with mild topical steroids and antifungal agents did not result in improvement, and she presented to our institution for further diagnostic and treatment considerations.

Physical examination revealed multiple discrete pink, erythematous, and yellow-brown macules scattered over her left breast with a background of telangiectasia (Figure 1). Dermoscopy revealed a reticulated network of telangiectasia on both the involved and uninvolved skin of her left breast (Figure 2). The discrete lesions did not urticate with gentle stroking. Aside from mild preexisting microcytic anemia, complete blood cell count with differential and thyroid function tests were unremarkable.

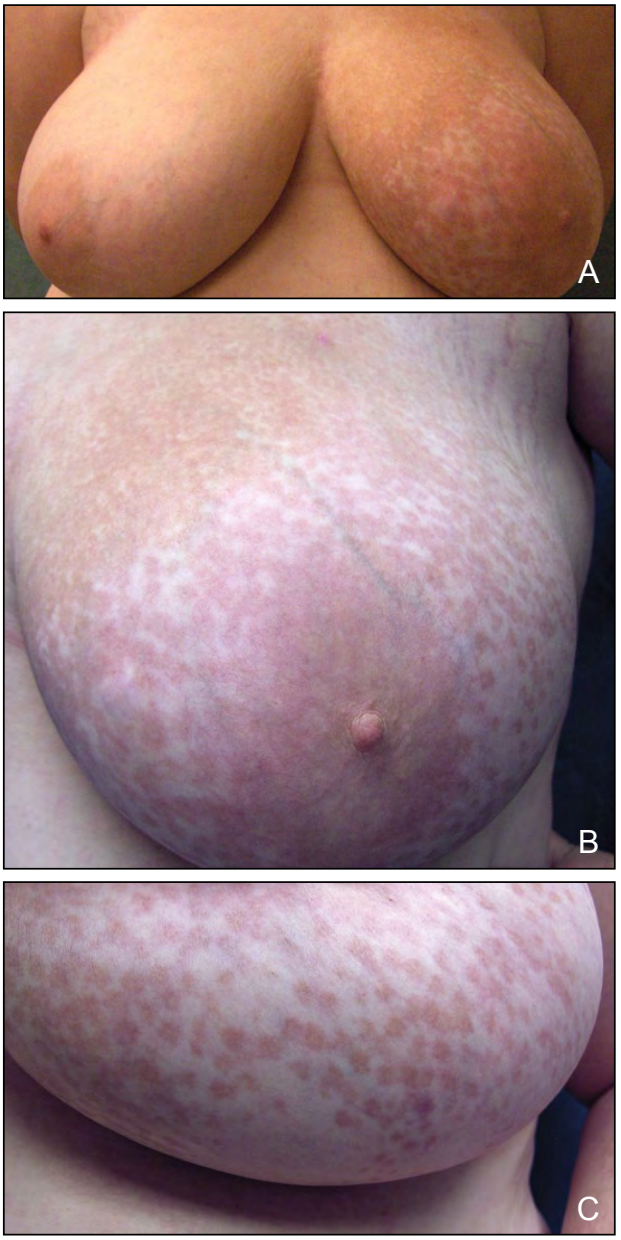

Figure 1. Multiple discrete pink, erythematous, and yellow-brown macules scattered over the left breast with a background of telangiectasia (A–C).

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The authors report no conflict of interest.
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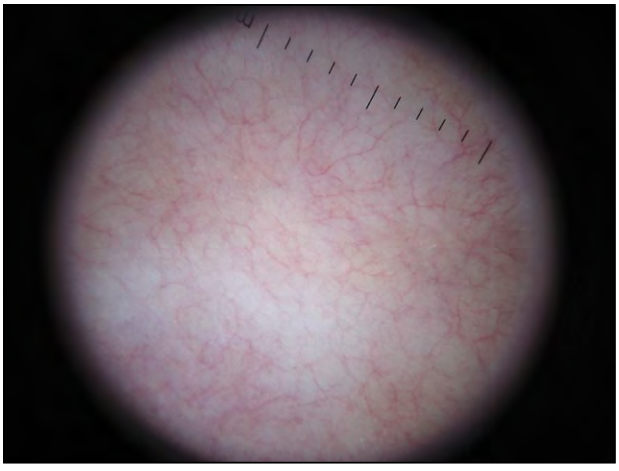

Figure 2. Dermoscopy showed a reticulated network of telangiectasia on involved skin of the left breast.

Two biopsies of her left breast previously had been performed; one biopsy was inconclusive in relation to her patient history and the other showed slightly increased dermal mast cells. The cells were considered to represent telangiectasia macularis eruptiva perstans (TMEP), though a definitive diagnosis could not be made. Two additional punch biopsies were performed at our institution, which revealed innumerable cells in the upper papillary dermis staining positive for expression of tryptase in the mast cells. Considering the clinical and histopathologic analysis, the patient was diagnosed with unilateral TMEP. Because the rash was asymptomatic and the patient lacked systemic symptoms or notable laboratory abnormalities, the patient opted to conceal the rash with cosmetic cover-up as needed.

Comment

Telangiectasia macularis eruptiva perstans is a rare type of cutaneous mastocytosis (CM), which is a subtype of mastocytosis. Mastocytosis is a heterogeneous neoplastic disease that is defined by an abnormal proliferation of normal active mast cells.[1] The World Health Organization adopted a consensus classification system first recorded by Valent et al[2] in 2001 that divided mastocytosis into 3 categories: systemic mastocytosis, extracutaneous mastocytoma, and CM. Cutaneous mastocytosis is a benign disease that most commonly affects children and is defined by the absence of criteria for systemic mastocytosis, histologic findings of focal infiltrates of mastocytosis in the dermis, and typical clinical skin lesions.[3] The most common form of CM is urticaria pigmentosa (UP), also known as maculopapular CM, which represents two-thirds of patients with CM. Other forms of CM include diffuse erythrodermic mastocytosis, solitary mastocytoma, and TMEP.[1] The consensus classification system places TMEP as a subvariant of UP alongside a nodular and plaque type.

Telangiectasia macularis eruptiva perstans most commonly affects adults, as opposed to UP, which has a predilection for children. It is aptly named because its clinical presentation consists of erythematous telangiectatic macules that often are confluent and smaller than macules seen in UP.[1,4] Telangiectasia macularis eruptiva perstans generally involves the trunk and extremities symmetrically, has a scarcity of systemic symptoms, and has a notable absence of Darier sign.[4] Although uncommon, cases of TMEP with systemic symptoms including gastrointestinal symptoms, tachycardia, dyspnea, headaches, bone pain, pruritus, and flushing have been reported.[5,6] Histologically, TMEP has mast cells surrounding the capillary venules and the superficial venular plexus with notable vessel dilatation secondary to the release of angiogenic factors and other mast cell mediators.[5-7]

Although classical TMEP generally is a symmetric eruption, 2 unilateral cases have been reported.[4,5] In the 2 cases, lesions were noted to involve the face. In addition, both cases lacked systemic symptoms and both were diagnosed by biopsy.[4,5]

The differential diagnosis for TMEP includes unilateral nevoid telangiectasia, carcinoid syndrome, and other forms of CM, all presenting with telangiectasia. In unilateral nevoid telangiectasia, which was the initial primary diagnostic consideration in our patient, there are patches of superficial telangiectases in a unilateral dermatomal or linear distribution that can be congenital or acquired. The underlying mechanism is believed to be somatic mosaicism, which is unmasked by a systemic increase in estrogen levels exhibited during pregnancy, puberty, and alcoholic cirrhosis.[8] Histologically, there is a notable absence of mast cells and there are multiple dilated, thin-walled vessels in the papillary and upper reticular dermis.[9]

In our patient, there was a complete absence of systemic symptoms or gastrointestinal concerns, thus excluding carcinoid syndrome. Unilateral nevoid telangiectasia was our initial primary diagnostic consideration, especially given the onset of eruption near puberty. However, histologic examination revealed the presence of mast cells, which eliminated the diagnosis of unilateral nevoid telangiectasia. Furthermore, the age of onset in our patient, the absence of Darier sign, the appearance of the lesions, and the lack of systemic symptoms or laboratory abnormalities also helped to eliminate other forms of CM.

Telangiectasia macularis eruptiva perstans is a benign disorder without a definitive treatment. Therapeutic options include H_1- and H_2-receptor

antagonists when symptoms are present and recurrent treatment with a 585-nm flashlamp-pumped pulsed dye laser.[7] Our patient declined further treatment because she was asymptomatic, and instead she chose to conceal her rash with cosmetic cover-up as needed.

Conclusion

Our case illustrates an unusual presentation of TMEP, a rare variant of CM. Although TMEP usually presents in a symmetric fashion, cases of unilateral involvement have been reported; 2 prior reports involved the face[4,5] and our case involves the breast. We further present our case to raise awareness of unilateral TMEP as a diagnostic consideration when telangiectasia presents unilaterally and appears to be most consistent with unilateral nevoid telangiectasia. In these cases, skin biopsy is necessary for diagnosis.

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