Telangiectasia Macularis Eruptiva Perstans: A Case Presentation and Discussion

Tselangiektasia macularis eruptiva perstans (TMEP) is a rare subtype of cutaneous mastocytosis that tends to appear during adulthood. Cutaneous mastocytosis is a proliferation of mast cells limited to the skin that spares other organs. Dermatoscopy of the lesions show red-to-brown, telangiectatic macules diffusely spread over the trunk and upper extremities. We present a case of a 32-year-old male with TMEP who lacked systemic symptoms and discuss the clinical presentation, histopathology, and treatments.

**Case Report**

A 32-year-old Caucasian male presented complaining of new-onset skin lesions on his chest, back and left eye. He reported that the lesions began to appear about three months prior and since then had increased in number. The patient complained that the lesions felt pruritic, burning and very uncomfortable. He also reported that he had sudden onset of flushing and sweating with stress. A review of systems was negative for weight loss, constitutional symptoms, preceding illness, dyspnea, epistaxis, abdominal pain and diarrhea. Past medical and surgical history was insignificant. Family history included a first cousin on the maternal side with vitiligo and epistaxis and a mother who died of breast cancer. The patient had no known drug allergies.

Physical examination revealed no visible oral mucosal telangiectasias or lesions. Cutaneous findings included multiple brown-to-red, telangiectatic macules of varying sizes diffusely placed on the body, with the majority of the lesions present on the scapula (Figures 1 and 2). The remainder of the exam was essentially normal.

A total tryptase level, CBC with differential, CMP, BMP, and thyroid-hormone level were obtained, and all were within normal limits. A 6 mm biopsy was taken from the patient’s skin overlying the right scapula. The pathology report described dilated small blood vessels within the superficial dermis (Figure 3). A Leder stain highlighted a slightly increased number of mast cells within the dermis (Figure 4).

Another 3 mm punch biopsy was taken from the patient’s lower back on the right side. The pathology report described occasional small lymphocytes and scattered mast cells (Figure 5). Leder stain highlighted approximately 18 mast cells per high power field in the papillary and superficial dermis (Figure 6).

The patient was sent for genetic testing. No abnormalities were reported.

The constellation of physical and histological features pointed toward a diagnosis of telangiectasia macularis eruptiva perstans.

**Discussion**

Mastocytosis is a collection of rare disorders, all caused by the pathologic proliferation of mast cells. The disorders are typically categorized into two major subtypes based on whether or not the proliferation is localized. When limited to the skin, the term “cutaneous mastocytosis” is used. If the proliferation of cells is widespread throughout the organs of the body, it is termed “systemic mastocytosis.”

Mast cells play a role in inflammatory and allergic responses by releasing cytokines, histamines, tryptases, interleukins and other chemical mediators upon degranulation. The downstream effects of these mediators on their receptors cause the clinical manifestations seen in mastocytosis.

Systemic mastocytosis is marked by syncope, tachycardia, pruritus, dyspnea, abdominal pain, diarrhea or flushing. Cutaneous mastocytosis, on the other hand, does not manifest with systemic symptoms. It is subdivided into four categories: urticaria pigmentosa, mastocytoma, diffuse and erythrodermic cutaneous mastocytosis, and telangiectasia macularis eruptiva perstans (TMEP). TMEP was initially described by Parkes Weber in 1930 and is found in less than 1% of patients diagnosed with cutaneous mastocytosis. TMEP, unlike the other types of cutaneous mastocytosis, often presents in adulthood. Clinically, it is characterized by red-to-brown, telangiectatic macules. The lesions are usually...
between 2 mm and 4 mm in diameter and are commonly found on the trunk and proximal extremities, symmetrically. The palms, soles, and face are classically spared. There may be variable amounts of pruritus associated with the lesions. Darier's sign (urticaria after friction accompanied by erythema, pruritus, and swelling) is commonly absent in this form of cutaneous mastocytosis, but is found in other types.

TMEP has been found in the setting of systemic mastocytosis. Suspicion of systemic involvement should arise if patients have simultaneous symptoms of anaphylaxis, dysnea, diarrhea, syncope, tachycardia, pruritus, abdominal pain, and flushing. Tryptase is a large component of the granules contained within mast cells, and therefore measuring the total serum tryptase level is a good test to decipher if patients have systemic involvement.

Histopathologic studies of skin biopsies are used to confirm the diagnosis of TMEP. Histologically, TMEP demonstrates increased perivascular and interstitial mast-cell collections surrounding dilated telangiectatic blood vessels. The mast cells are usually located in the upper portion of the dermis, surrounding the dilated blood vessels. The number of mast cells is only slightly increased, and there may also be associated findings of epidermal hyperpigmentation.

There is no gold standard therapy for TMEP, and the goal is to alleviate symptoms. H1 antihistamine antagonists can be used to treat the pruritus and flushing symptoms, while H2 antagonists can be used in treating the gastric hypersecretion. It is important for patients to avoid triggers that can stimulate mast-cell degranulation. Triggers can include, but are not limited to: alcohol, bacterial toxins, stress, exercise, food, sunlight, temperature extremes, narcotics and anesthesia. Psoralen (oral), UVA photochemotherapy, high-dose UVA-1 and narrow-band UVB phototherapy have all been shown to improve symptoms and cosmetic appearance. Surgery via a flashlamp-pumped dye laser has also shown cosmetic improvement in the cutaneous lesions. The replacement of antihistamine therapy with montelukast therapy was shown to be effective in the treatment of TMEP. Most of the results from treatment are temporary unless therapy is continued indefinitely. A recent study used cabozantinib, a signal transduction inhibitor that blocked growth of mast cells with the D816V codon mutation.

References

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