

BRIEF ARTICLE

Treatment of Telangiectasia Macularis Eruptiva Perstans with an Intense Pulsed Light Device

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ABSTRACT

Telangiectasia macularis eruptiva perstans (TMEP) is a rare adult mastocytosis with prominent cutaneous manifestations including red to brown telangiectatic macules typically on the chest or limbs. TMEP can be a cause of morbidity with associated cutaneous findings (e.g. flushing, pruritus, ulcers) and extracutaneous symptoms (e.g. dyspnea, reflux, diarrhea, hypotension). Previous attempts to treat the cutaneous manifestations of TMEP include PUVA, topical steroids, electron beam radiation, and 585-nm flashlamp-pumped dye laser. We report the first case of successful treatment of the cutaneous findings of TMEP with intense pulsed light (IPL). The patient underwent 2 treatments with IPL spaced 3 months apart, and remained entirely clear at 14 months follow-up.

CASE REPORT

Telangiectasia macularis eruptiva perstans (TMEP) is a rare adult mastocytosis with prominent cutaneous manifestations. It was first described by Parkes Weber in the 1930s.¹ Irregular red to brown telangiectatic macules, usually on the chest or limbs, are seen clinically. TMEP can be a cause of significant morbidity with both cutaneous and extracutaneous symptoms. These include flushing, blisters, pruritus, ulcers, hypotension, dyspnea and gastrointestinal symptoms such as reflux and diarrhea.² Overall, TMEP is considered to have a good prognosis with the majority of cases lacking systemic involvement.^{3,4} If skin biopsies are performed in patients with TMEP, findings

typically include dilated upper dermal capillaries and venules with an upper dermal inflammatory infiltrate, largely comprised of mast cells around neurovascular bundles.⁵ Various approaches have been used to treat the cutaneous manifestations of TMEP including Psoralen plus ultraviolet light A (PUVA), topical corticosteroids, electron beam radiation and the 585-nm flashlamp-pumped dye laser.^{2,5-7} These approaches have shown variable success. There are no previous reports in the literature describing the treatment of TMEP with an intense pulsed light (IPL) device. However, the characteristic cutaneous lesions, with dilated capillaries of the superficial venous plexus of

the dermis, lend themselves to such an approach. We present the case of a patient with TMEP and the associated characteristic cutaneous manifestations who was successfully treated with IPL.

The patient was a 53-year-old gentleman who presented with a 2-year history of diffuse red telangiectatic blanching macules on both arms, hands and shins. Histology was consistent with TMEP. Over the previous 6 months he had two episodes of diffuse flushing and dizziness that lasted 1 hour. For these possible systemic components of disease, he was started on 10 mg of cetirizine nightly and advised to avoid histamine releasers like alcohol, anticholinergic medications, aspirin, nonsteroidal anti-inflammatory drugs, heat, friction and opioids. Medical history revealed that the appearance of his arms and legs had a large impact on his life. Incessant comments from others led him to become self-conscious, agoraphobic and he would not leave his home without full sleeve shirts and pants.

On examination we found numerous, confluent, blanching arborizing small vessels coalescing into large patches over the dorsal hands, forearms, posterior upper arms (Figures 1A-1E) and shins.

In an effort to address his cutaneous findings, a trial with IPL was conducted. The patient's bilateral arms underwent field treatment with an IPL device (Cutera Limelight, Brisbane, California). Using a handpiece size of 10 x 30 mm, an energy level of 14-18 J/s and a pulse width set to target vascular lesions (Program A). The entire affected area was treated in a confluent and sequential manner with a single pass with 10% overlap. Pre-treatment with 150 mg of ranitidine and 180 mg of fexofenadine was used to mitigate any potential treatment-induced mast cell histamine release. After a single treatment

there was a remarkable difference with only minimal remaining involvement. A second treatment to these areas was performed 3 months later with complete resolution. Treatment associated adverse events included mild erythema and edema and mild discomfort that resolved within 48 hours. His arms remain entirely clear and without recurrence 14 months later (Figure 2).



Figure 1. Extensive telangiectasias noted pretreatment A: left arm B: right arm, C: left elbow, D: right posterior upper arm, E: left posterior upper arm.



Figure 2. Post-treatment of bilateral arms showing complete resolution of telangiectasias 14 months after treatment.

DISCUSSION

The treatment of TMEP is largely based on the degree of systemic involvement or clinical symptoms the patient may have.⁴ Aside from the possible systemic symptoms of TMEP, its appearance can be unsightly and lead to significant social and emotional distress. Patients may report agoraphobia, social anxiety and low self-esteem which can significantly affect their quality of life and functioning.

Our patient had an astonishing response with one treatment session with only minimal day of treatment mild discomfort and erythema but no long-term side effects. The therapy was tolerated well without textural change. He is without recurrence 14 months after treatment. He continues to take 10 mg of cetirizine daily for any possible systemic involvement and has been free of symptoms.

There is no standard of care for the treatment of TMEP. The experimental treatments described previously include PUVA therapy, total body electron beam therapy and 585-nm flashlamp-pumped dye laser treatment.

Phototherapy with PUVA or UVB therapy can be helpful in terms of both pruritus and cutaneous appearance but requires the patient to come in several times per week and the lesions have been noted to recur after several months.^{6,7} Moreover, the patient is exposed to UV radiation.

A single case report of total body electron beam therapy demonstrates successful treatment of TMEP. The patient was free of his intense pruritus and cutaneous findings for up to one year. However, the treatment is expensive and required 24 treatment sessions over 6 weeks. Adverse effects of this therapy can include “mild erythema,

xerosis, temporary scalp alopecia, nail stasis, desquamation, anhidrosis, minor parotiditis, nosebleeds, and blistering or edema of hands and feet, as well as gynecomastia in males.”⁶

The 585-nm flashlamp-pumped dye laser has been reported to be effective for the treatment of TMEP in two cases. In both cases, the lesions begin to recur after 1 year. Moreover, one patient required general anesthesia and experienced immediate wheal and flare reaction to all treated lesions as well as swelling, pruritus and vesiculation of the lesions within 6-12 hours.² Our patient experienced only minimal erythema and tolerated the treatment with topical anesthesia. Previous histologic evaluation after treatment with a 585-nm flashlamp-pumped dye laser revealed fibrosis of the superficial vasculature, however, the number of mast cells were essentially unchanged. The authors suggested that the results were due to selective vascular fibrosis rather than destruction of mast cells.² This observation may imply that vascular laser therapy only treats the cutaneous appearance of the condition and does not address the underlying etiology and will not address any systemic symptoms that the patient may have. It is important that this be discussed with the patient as well as to inform them of the risk of eventual recurrence of their skin findings.

In summary, IPL is an effective treatment option for the cutaneous manifestations of TMEP. Further studies are warranted to fully explore this option.

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