

# Treatment of Telangiectasia Macularis Eruptiva Perstans With Total Skin Electron Beam Radiation

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*We describe the treatment of a 60-year old man with severely symptomatic telangiectasia macularis eruptiva perstans (TMEP) with a poor response to several standard therapeutic strategies. At that time, the patient underwent total skin electron beam (TSEB) radiation, based on the theory that by decreasing cutaneous mast cell infiltration, his pruritus would be relieved. He received a total dose of 4000 cGys given in 40 fractionated treatments. The patient had complete resolution of both his cutaneous lesions and pruritus, which has continued through one year of follow-up.*

## Case Report

A 60-year-old man with a history of flushing presented with a pruritic, reddish-brown macular eruption on his trunk and proximal extremities (Figure 1). A punch biopsy of his thigh was performed, and the results suggested cutaneous mastocytosis. Results of a Giemsa stain showed a modest increase in mast cells with overlying epidermal spongiosis, which is consistent with the diagnosis of telangiectasia macularis eruptiva perstans (TMEP).

A workup for systemic mast cell disease was conducted. Results of a complete blood cell count, chemistry profile, liver-associated enzymes, antinuclear antibody, and erythrocyte sedimentation rate were normal or negative. A 24-hour urine level test of histamine metabolites—*N*-methylhistamine and

*N*-methylimidazole acetic acid—was within normal limits. Results of a bone marrow biopsy showed normocellular bone marrow without increased numbers of mast cells. Results of a bone density scan revealed that the patient had mild osteopenia, for which he was prescribed alendronate, vitamin D, and calcium.

The patient was instructed to avoid substances known to elicit mast cell degranulation, such as aspirin, alcohol, opiates, and intravenous contrast dye. He was started on oral cromolyn sodium for diarrhea and gastrointestinal cramping. Various treatments were tried to control the patient's severe pruritus. Clobetasol cream 0.05% and several oral medications with antihistaminic properties, including cetirizine, hydroxyzine, cimetidine, fexofenadine, doxepin, and cyproheptadine, were prescribed but provided little relief. Colchicine and oral prednisone, starting at 60 mg and tapering slowly over several weeks, also were ineffective. The patient then underwent UVB phototherapy because of reports that it may be beneficial for cutaneous mast cell disease.<sup>1,2</sup> His lesions improved, and his pruritus subsided with UVB phototherapy but recurred 6 weeks after discontinuing the therapy. At that time, photochemotherapy with psoralen-UVA (PUVA) was instituted. Although photochemotherapy with PUVA resulted in a significant reduction in the patient's lesions and pruritus, severe pruritus and TMEP lesions recurred 2 months after discontinuing treatment. A trial of total skin electron beam (TSEB) radiation then was considered and discussed with the patient.

TSEB radiation has long been standard therapy for inducing long-term remission in cutaneous T-cell lymphoma.<sup>3,4</sup> We theorized that TSEB radiation could be equally effective in inducing long-term remission of TMEP, although a search of the literature revealed no reports of such. Studies of the effect of radiation on various tissues in rats have shown that mast cells are radiosensitive,

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**Figure 1.** Telangiectasia macularis eruptiva perstans lesions before treatment (A and B).

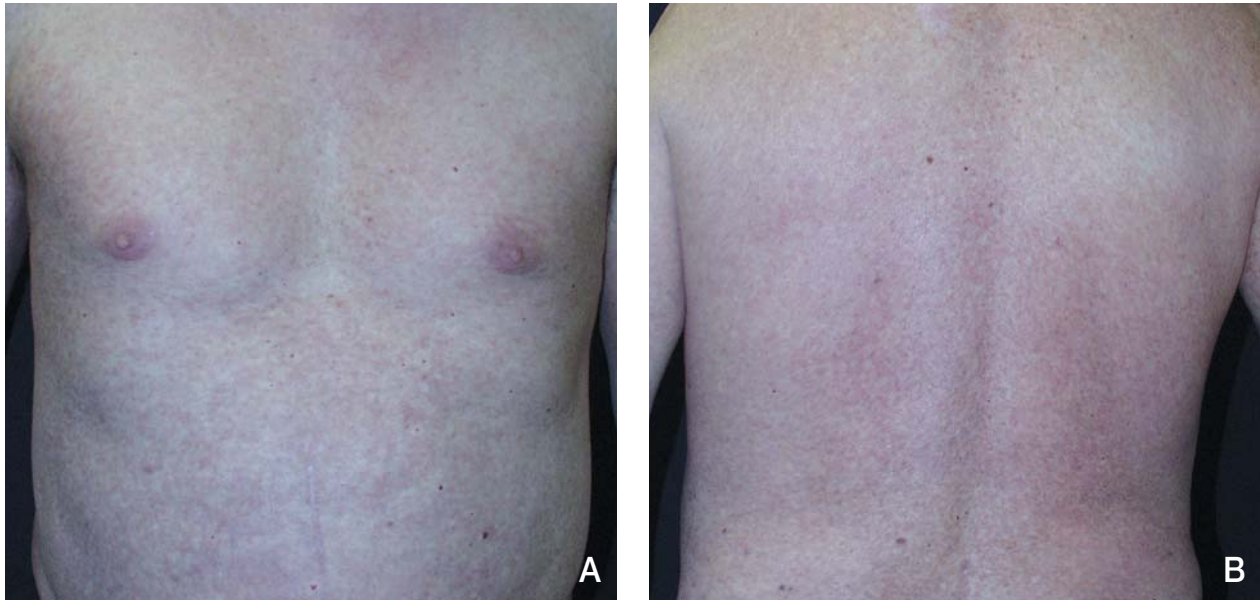
though regeneration of mast cells in the tissue eventually occurs.<sup>5-8</sup> Decreasing the number of mast cells in the cutaneous tissue should have the beneficial effect of decreasing the patient's severe pruritus. Our major concern for using TSEB therapy was that it might inadvertently cause massive histamine release if mast cells were destroyed during treatment. A report on the use of palliative radiation therapy to the spine in patients with severe bone pain due to systemic mastocytosis, however, did not demonstrate an elevation in plasma histamine levels or significant morbidity.<sup>9</sup>

The patient agreed to TSEB therapy and was referred to a radiation oncologist. Because of the potential for massive liberation of histamine from mast cell irradiation, the patient continued to take fexofenadine to help mitigate any symptoms. Initially, a test dose of 3000 cGys, given in 10 fractionated treatments, was administered to a 25-cm<sup>2</sup> area of the patient's left shoulder. At follow-up, the patient reported a significant decrease in pruritus and clearing of TMEP lesions at the test site. The patient then underwent TSEB therapy over the next 6 weeks, receiving whole body radiation 4 days a week and radiation to his feet 1 day a week, for a total dose of 4000 cGys given in 40 fractionated treatments. He continued to take fexofenadine daily. Over the course of the radiation treatments, all cutaneous signs and symptoms of TMEP resolved (Figure 2). Our patient reported no adverse side effects from TSEB radiation. One

year after treatment, his lesions were still in remission, and he reported no pruritus. On his own accord, the patient discontinued taking antihistamine.

### Comment

TMEP is an uncommon form of cutaneous mastocytosis, occurring in fewer than 1% of persons with mastocytosis. This disorder primarily affects adults and is manifested mainly by telangiectatic tan-to-brown macules on the trunk and extremities, as well as by pruritus. Symptoms from systemic involvement include headaches, flushing, diarrhea, and palpitations. Although TMEP has no potential for malignant development, symptoms may be disabling. Current standard treatment is intended to provide symptomatic relief with antihistamines or steroids. Photochemotherapy with PUVA has been successful, but, as in our patient, lesions recurred after cessation of therapy. Other experimental therapies reported in the literature include interferon alfa and the 585-nm flashlamp pumped dye laser.<sup>10-12</sup> TSEB radiation has been used for decades, mainly for the treatment of cutaneous T-cell lymphoma. Adverse effects from TSEB radiation tend to be self-limited and 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. Possible long-term complications include chronic nail dystrophy, chronic xerosis, partial but permanent alopecia of the scalp, as well



**Figure 2.** Telangiectasia macularis eruptiva perstans lesions cleared after treatment (A and B). (Note: The patient also lost 30 pounds).

as fingertip dysesthesias. No acute or late mortality has been reported with TSEB radiation.<sup>13,14</sup>

This case report demonstrates that TSEB radiation therapy was effective in achieving long-term remission in a severely symptomatic patient with TMEP who had not responded to standard management.

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