

## Telangiectatic Mastocytosis Treated with Intense Pulsed Light.

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# Telangiectatic Mastocytosis Treated with Intense Pulsed Light

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## Case Presentation

**Patient:** 42 year-old Hispanic female.

**History of Present Illness:** Our patient presents with a rash and persistent redness of the face. The rash has been present since childhood and is described as being located all over. It is occasionally associated with pruritus and “hives.” She denies previous treatment and history of anaphylactic reaction.

**Medical/Surgical History:** Diabetes mellitus type II, hypertension, hyperlipidemia, anemia, anxiety, depression, gastroesophageal reflux disease, cholecystectomy

**Current Medications:** Insulin, liraglutide, metformin, simvastatin, lisinopril, aspirin, methylphenidate, monteleukast, omeprazole, clonazepam, trazodone

**Laboratory Data:** (10/27/2015) RDW 17.2% (12-16%), lactate dehydrogenase 94 (100-250 U/L), tryptase 129 ug/L (<10.9 ug/L), (11/02/2015): blood- positive for KIT (D816v) point mutation by PCR; remainder CBC, CMP, B-2 microglobulin, IgA, IgG, IgM WNL

**Physical Examination:** Scattered 0.5-1.5cm thin, red-brown papules and plaques on the trunk, arms, and legs with telangiectasias on dermoscopy. There is a 1cm red, waxy plaque at the left elbow. Multiple telangiectasias on the cheeks.

**Biopsy:** *Advanced Dermatology Associates, LTD* (AD15-05556, 05/18/2015) Left elbow, shave biopsy: “Mastocytoma. ‘By immunohistochemistry stain, there is a fairly dense band-like population of CD117 positive, CD163 negative, and CD68 negative cells that is somewhat obscured by a patchy lymphohistiocytic infiltrate.’”

*Health Network Laboratories* (9/8/2015, S15-28545, Bone marrow, clot and biopsy left iliac crest): Systemic mastocytosis. “The marrow findings meet criteria for systemic mastocytosis... multifocal dense infiltrate of mast cells (>15 mast cells in aggregates), scattered throughout the biopsy. The majority of mast cells demonstrates spindle cell morphology. The mast cells express CD25. No c-KIT mutation in exons 8,9,11,13 and 17 detected.”

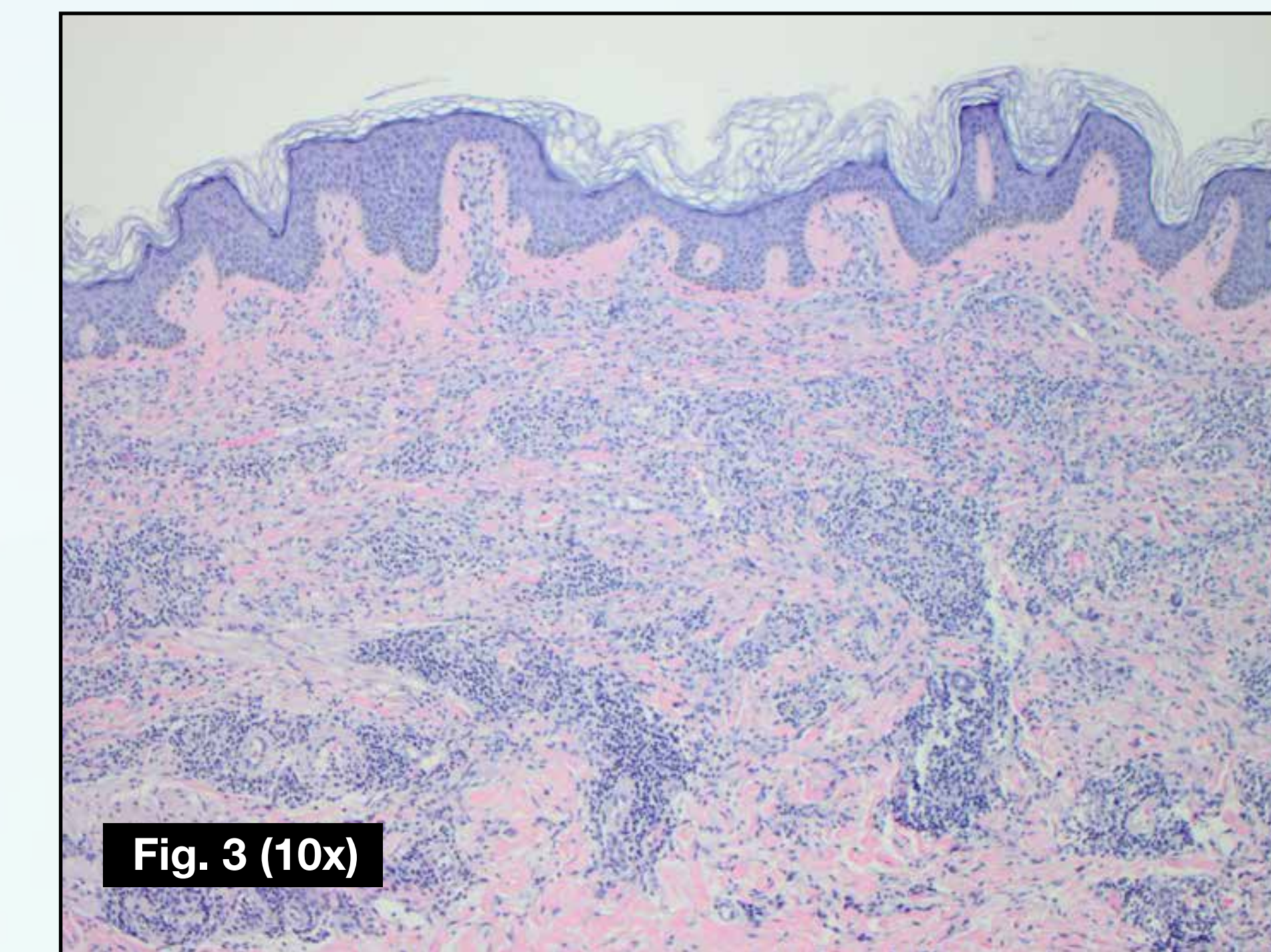
**Treatment:** After a test spot was performed with no urticarial response, multiple sessions of intense pulsed light at wavelength 570nm and fluence of 45.0 J/cm<sup>2</sup> were performed to lesions on the cheeks with improvement of telangiectatic lesions.



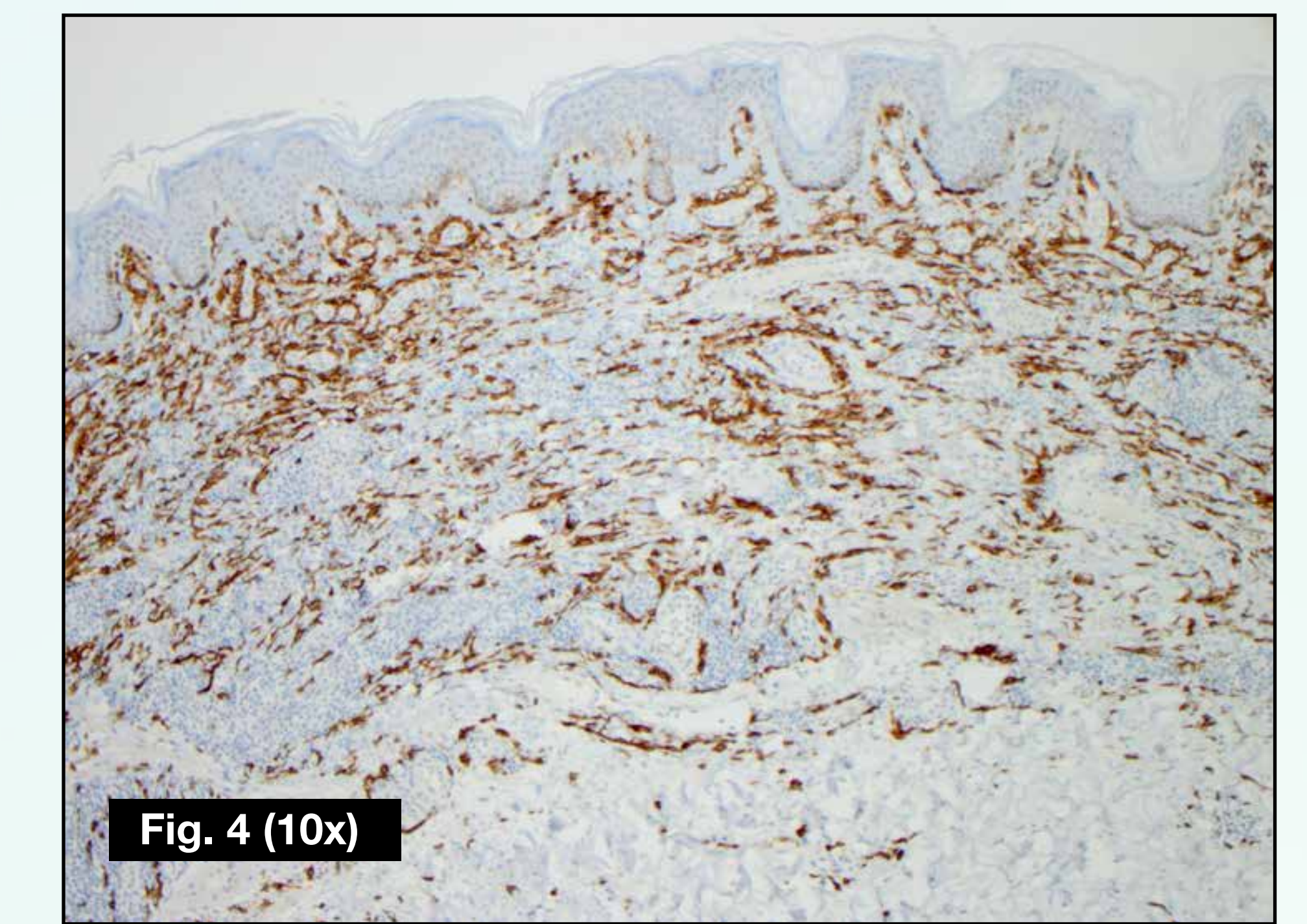
**Figure 1:** Discrete and coalescent telangiectasias prior to initiation of intense pulsed light (IPL) treatment.



**Figure 2:** Same patient after three treatments with IPL at wavelength 570nm and fluence of 45.0 J/cm<sup>2</sup>.



**Figure 3:** (10x): On low power a lymphohistiocytic infiltrate can be observed. On higher power the histiocyte-appearing cells are found to sometimes contain subtle cytoplasmic granules.



**Figure 4:** (10x): Fairly dense band-like population of CD117+ mast cells that is somewhat obscured by a patchy lymphohistiocytic infiltrate.

## Discussion

Mastocytoses encompass a group of disorders that result from accumulation of abnormal mast cells in both the skin and other organs. Mastocytosis can present in both children and adults. The time of development has prognostic implications in regard to systemic involvement. Childhood disease is typically limited to cutaneous lesions and includes maculopapular cutaneous mastocytosis (urticaria pigmentosa), diffuse cutaneous mastocytosis or mastocytoma. These patients often have spontaneous resolution by puberty and rarely have life threatening anaphylaxis. Adults who present with new cutaneous lesions are more likely to have systemic involvement, although an indolent disease course is the rule.

Serum tryptase levels often reflect the burden of neoplastic mast cells and help determine when bone marrow biopsy should be pursued to assess presence of systemic involvement. Criteria for diagnosis of systemic mastocytosis include dense infiltrates of mast cells in bone marrow or other tissue (major) and atypical mast cell morphology, aberrant mast cell surface phenotype, serum/plasma tryptase >20 ng/mL, codon 816 c-KIT mutation in peripheral blood, bone marrow, or lesional tissue (minor). Fulfillment of either one major and one minor criterion or three minor criteria is consistent with systemic mastocytosis and should prompt workup to rule out an associated hematologic non-mast cell lineage disorder.

KIT is a type III receptor tyrosine kinase encoded by the proto-oncogene c-KIT that is expressed on mast cells. A subset of mastocytosis patients have alterations in KIT structure and activity leading to constitutive activation of KIT and mast cell proliferation. Higher amounts of mast cell mediators released from abnormal mast cells cause the symptoms of mastocytosis. These mediators include those that are preformed, newly formed and various cytokines. Symptoms are variable and include headaches, flushing, pruritus, gastrointestinal distress, dizziness, and bone pain.

Treatment of mastocytosis is aimed at symptom relief primarily with antihistamines. Cutaneous disease can be treated effectively with psoralen and ultraviolet A. Avoidance of mast cell degranulating stimuli is important. Telangiectatic lesions have been successfully treated with modalities that target hemoglobin including pulsed dye laser and intense pulsed light. Intense pulsed light device is a non-coherent light source that delivers multiple wavelengths of visible and near-infrared light simultaneously. Filters are used to specify desired wavelengths that correspond with a target for treatment. When treating patients with multiple telangiectasias, pretreatment with antihistamines should be used to ensure large scale mast cell degranulation does not occur with resultant anaphylaxis. This patient represents a case of indolent systemic mastocytosis with diffuse telangiectasias that were improved dramatically with use of intense pulsed light. Long term follow up will be required to determine whether these results are maintained or there is reappearance of the telangiectasias.

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