Telangiectasia Macularis Eruptiva Perstans (TMEP) Case report with review of the literature

Mohammed Mohy El-din Selim
Hassan M. A. Riad
Ahmad Hazem Takiddin
Hamda Al-Ansari

Abstract:

The most frequently affected organ in any form of mastocytosis is the skin [1]. Cutaneous lesions tend to appear early in life and include urticaria pigmentosa, mastocytoma, diffuse and erythematous Cutaneous mastocytosis and Telangiectasia Macularis Eruptiva Perstans[2, 3]. Telangiectasia Macularis Eruptiva Perstans is observed in less than 1% of cases of mastocytosis [4]. Although cutaneous mastocytosis appears to occur sporadic yet familial incidence was reported as four cases of telangiectasia macularis eruptiva perstans appearing in three generations of one family with an autosomal dominant mode of transmission with incomplete penetrance [5]. Telangiectasia macularis eruptiva perstans is characterized by telangiectatic reddish brown macules usually on the trunk, pruritus [6] with little tendency to urticate [7]. Telangiectasia macularis eruptiva perstans is mostly seen in adults and is occasionally seen in children or infants [8].

The classic symptoms in Telangiectasia macularis eruptiva persants include episodic flushing, gastrointestinal complaints, heart palpitation, syncope and may be confused with carcinoid [8].

We present a case of telangiectasia macularis eruptiva perstans in an adult and discuss its diagnosis and mastocytosis in general and its management.

Case Report:

H.S. a Jordanian female born 28.10.1981 was seen on 20.5.2003 working as computer programmer.

She has been suffering from diffuse redness of face, trunk and both upper and lower limbs and easy flushing of face. The condition dates to infancy and was asymptomatic. The family history revealed that a younger sister 7-year-old has been also suffering from a similar skin problem since early childhood as well.

On Examination:

General examination: patient was in Good general condition, pulse was regular 70 / minute, blood Pressure was 120/70, and temperature 37.4 0C

Examination of the Skin: Patient has scaly scalp easily flushed face. Her trunk and limbs showed Telangiectatic macular Erythema.Fig. 1,2,3,4,5, close up view in Fig. 6. Diascopy was demonstrated in Fig. 7. Her sister H.S. was photographed at the age of seven years and showed similar pattern, but to a lesser degree. Fig. 8.

The clinical impression was Telangiectasia macularis eruptive perstans.

Darier sign was negative and patient did not complain except from easy flushing.

Investigations:

A skin biopsy was done and confirmed the diagnosis. It showed dilated blood vessels with mast cells mainly perivascular. Fig. 9,10,11,12.

The mast cells were not tested for Tryptase or chymase.

Other investigations:

Other investigations done included Complete blood count and blood biochemistry including LFT, KFT, TG, Cholesterol, uric acid, and RBS.

- 24 hour urine for 5-hydroxyinodole acetic acid was normal (4.87 u mol/24 hours and the normal range is 0-52)
- Valinyl Mandelic Acid (VMA) was 25.7 u mol per 24 hours (normal range 35-45)
- Metanephrine was 0.37 u mol per 24 hours (normal range is up to 5.1 u mol/24 hours)
- X-ray chest
- Abdominal echography
- Bone scintigraphy

All results were within normal level, thus excluding systemic affection and carcinoid tumor.

Management:

On 09.6.2003 Patient was referred to PUVA therapy. (Group 1 Photos) minimum phototoxic dose (MPD) was done two hours after ingestion of 20 MG of 8 MOP (0.4 mg /KG) patient did not show any response, on all tested sites, throughout the following 48 hours and MPD was to be 14 joules.

Patient was started on 50% of MPD (7 joules) 3 sessions per week.

On 23.6.2003 patient got marked tanning with marked improvement on tanned skin.

Because of the marked improvement and tanning, doses were reduced gradually to 6 then to 5 joules per sessions and in spite of that there was more improvement.

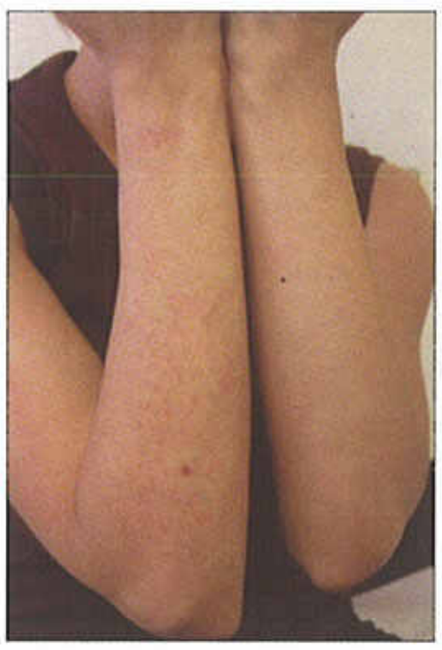

Fig.1a: before treatment

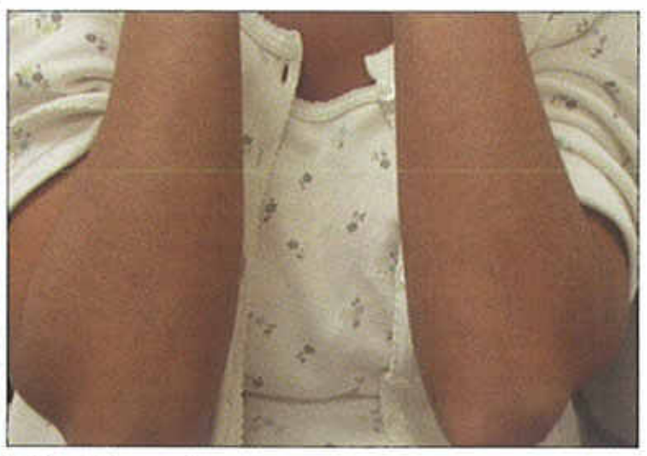

Fig.1b: after treatment

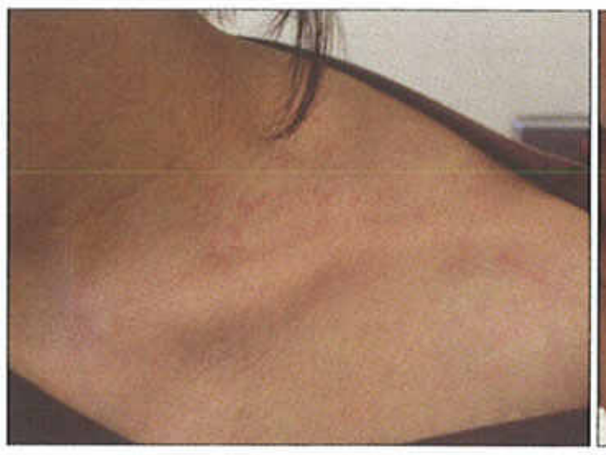

Fig.2a: before treatment

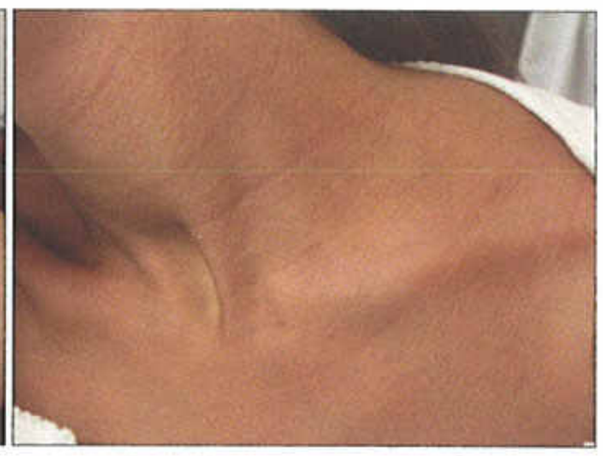

Fig.2b: after treatment

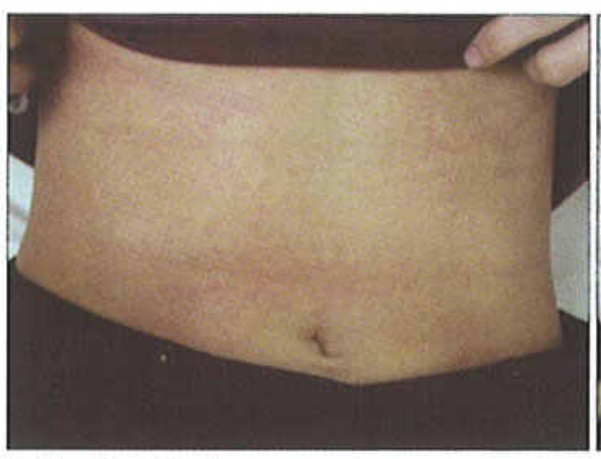

Fig.3a: before treatment

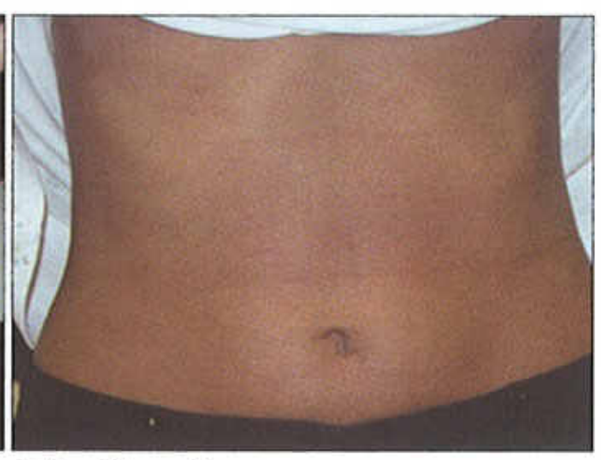

Fig.3b: after treatment

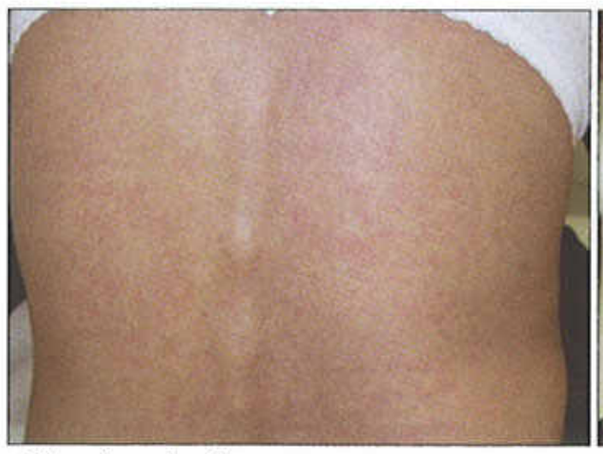

Fig.4a: before treatment

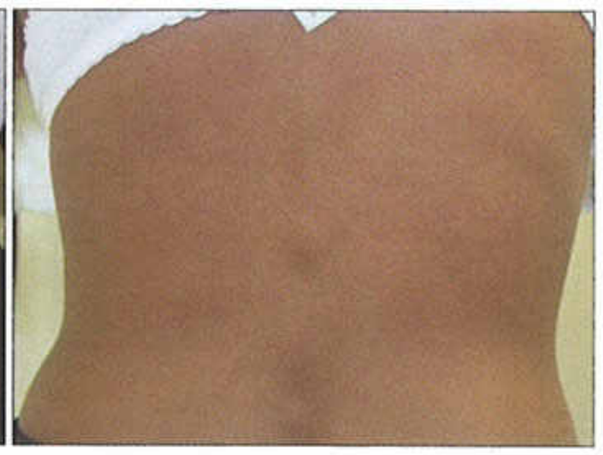

Fig.4b: after treatment

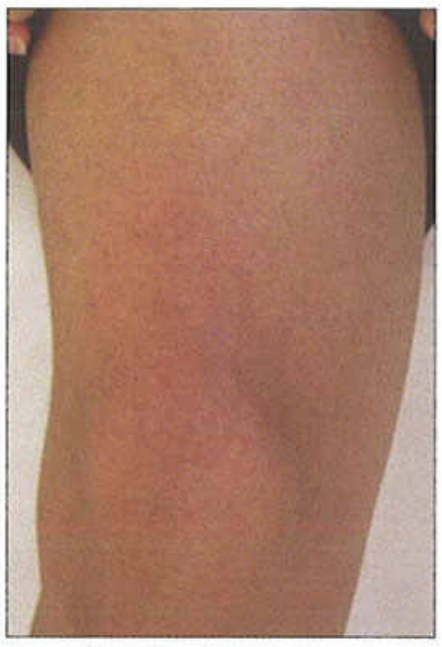

Fig.5a: before treatment

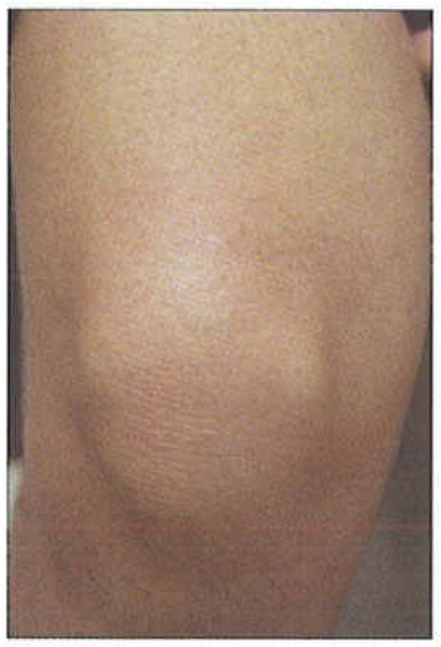

Fig.5b: after treatment

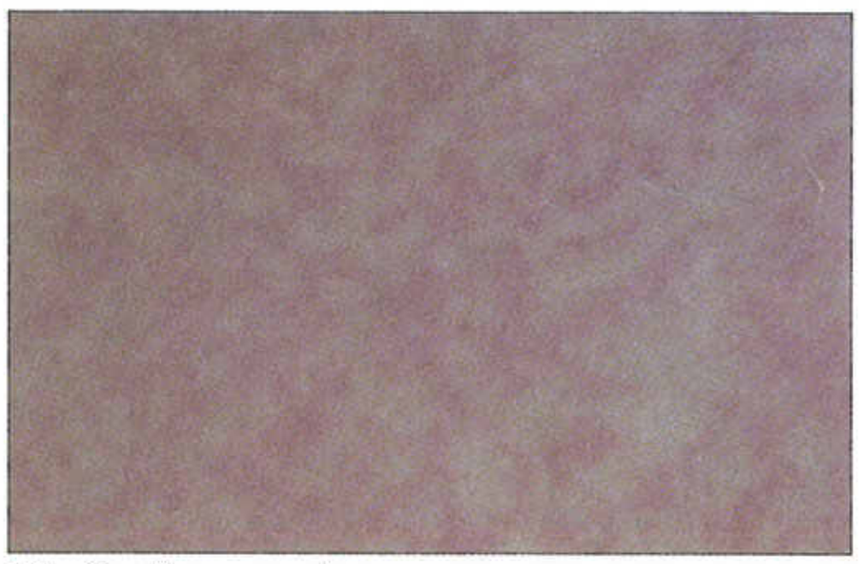

Fig.6: close up view

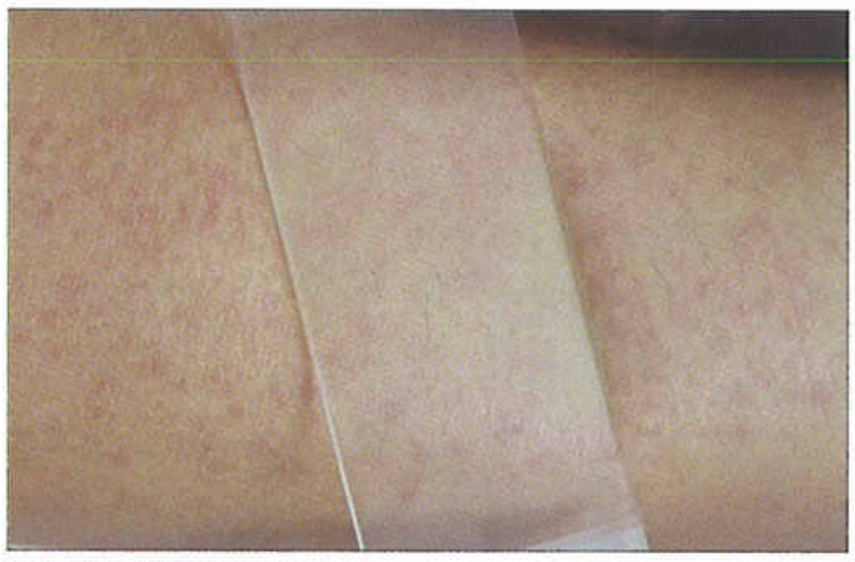

Fig.7: Diascopy

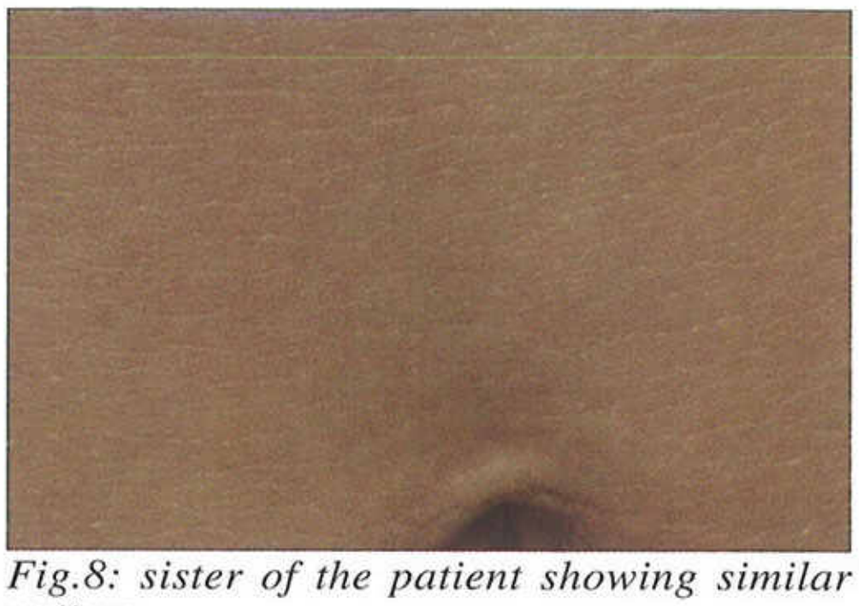

Fig.8: sister of the patient showing similar pattern

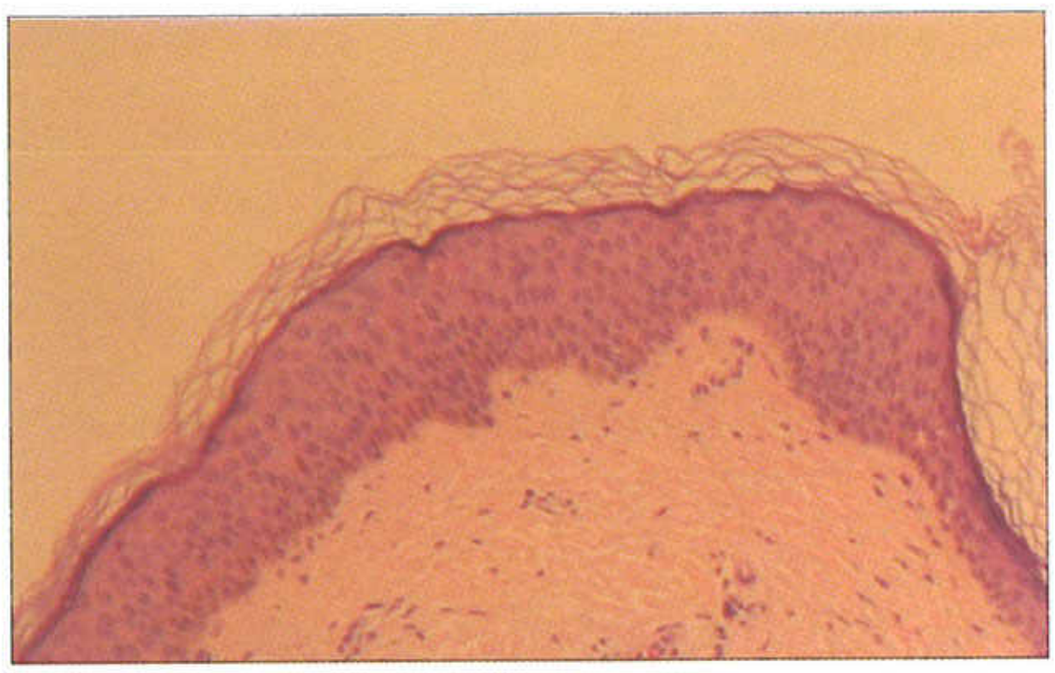

Fig.9: A medium power view of H&E stained section showing vague mild increase of perivascular cells in upper dermis

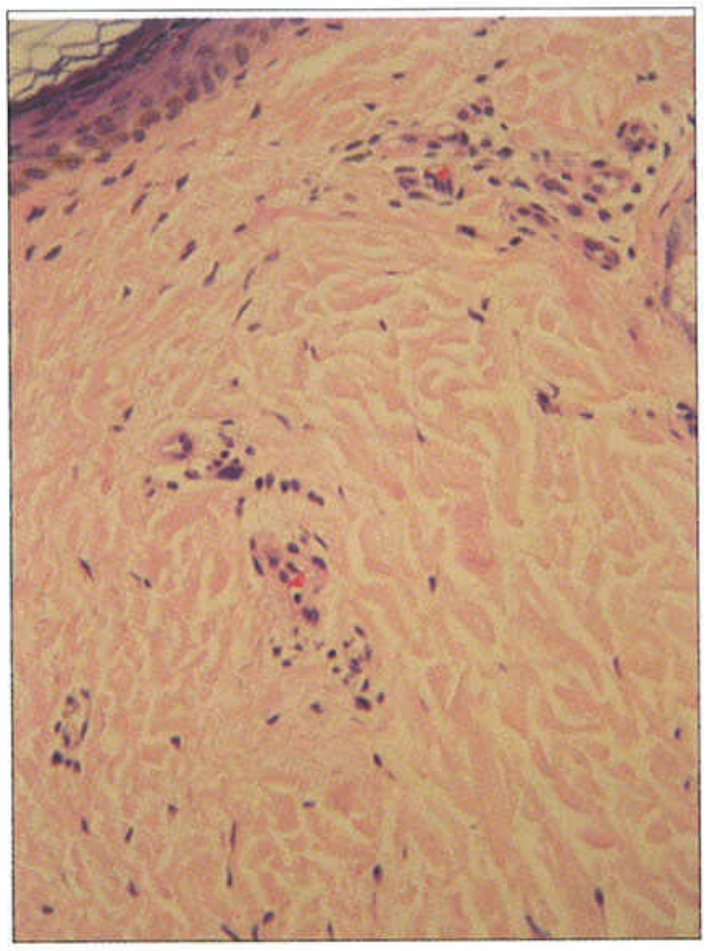

Fig.10: A higher power view showing sparse infiltrate around slightly telagiectatic upper dermal capillaries

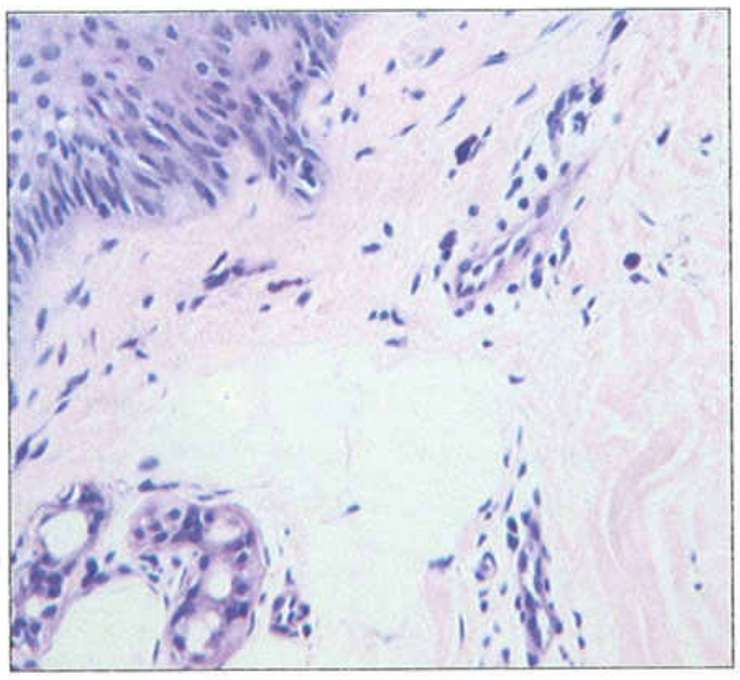

Fig.11: Geimsa stain visualising the mild increase of mast cells around upper dermal capillaries by staining the cytoplasmic granules. The sparse infiltrate could be easily missed in H&E.

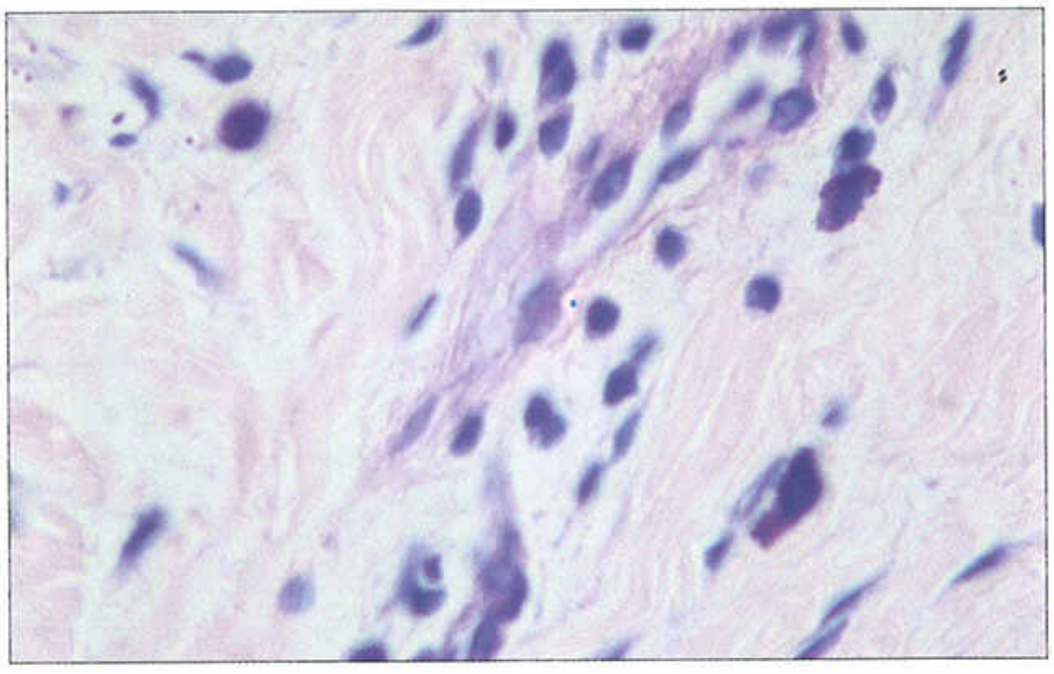

Fig.12: Oil immersion high power view of geimsa stained section showing metachromatic cytoplasmic granules of mast cells around slightly telangiectatic upper dermal blood capillary.

On 9.7.2003 Telangiectasia cleared and there were no more detected lesions, except for tanning and dryness of the skin.

Photochemotherpay was discontinued on 12.07.03 because of both the improvement and the annual leave.

Summary of PUVA therapy:

Total number of sessions was 10 sessions
Total cumulative dose was 60 joules
Duration of treatment was 27 days
Mean dose per session was 6 joules
Treatment was generally well tolerated

Discussion:

Mastocytosis is a disease characterized by abnormal increase in the number of mast cells. Mast cells originate from bone marrow stem cell which is CD34+ve. The stem cell differentiation to mast cell is

promoted by stem cell factor (SCF) (a mast cell growth factor) which acts on C-KIT receptor on surface of mast cell (KIT is a type III Tyrosine Kinase).

Activation of C-KIT receptor on surface of mast cell induces mast cells growth and prevents apoptosis. Mast cell disease or mastocytosis results form abnormal proliferation of mast cells with auto activation of C-KIT.

The onset of mastocytosis ranges from time of birth till late adult life. Most patients do not have familial incidence.

The currently presented case had TMEP in infancy and her younger sister is similarly affected since infancy.

Our patient presented with typical skin lesions with patchy Erythema and telangiectasia involving the trunk, extremities and face with flushing. She had no pruritus, urticaria pigmentosa and had a negative Darier sign. She had no constitutional symptoms, no gastrointestinal complaints as nausea, diarrhaea, abdominal cramps, malabsorption, gastric ulcer or intestinal hemorrhage. She also had no skeletal, cardiopulmonary or neurological manifestations as headache or depression.

TMEP is a rare clinical presentation and the rash consists of red telangiectatic macules with relatively little pigmentation and little tendency to urticate. It tends to persist and non-responsive to treatment. It is observed in less than 1% of mast cell disease. The association with multiple myeloma was reported [9]. Mastocytosis may predispose to anaphylaxis from wasp sting and is associated elevated serum level of mast cell tryptase[10]. Extracutaneous systemic manifestations as bone marrow involvement, gastrointestinal, hepatospleenomegaly, anemia, thrombocytopenia, cardiopulmonary or mastodysplastic syndrome [11] and proliferation of lymph nodes [12]. TMEP is also characterized by episodic flushing [13] which is quite manifest in the present case and is associated with elevated histamine excretion [14], [12].

Extra cutaneous manifestations may affect one system in 42%, 2 systems in 36% and 3 systems in 21%. Patients had generalized pruritus in 71% and wheal formation in 78.5% and flushing in 21%. Gastrointestional tract was affected in 57%, bone marrow involvement in 54%, hepatospleenomegaly in 14%, anemia in 14% and thrombocytopenia in 21% [11].

TMEP may have a unilateral facial [15, 16] or linear distribution [17] or may show poikelodermal pattern [18]. A very rare presentation of TMEP is prolonged losses of consciousness [19] and may present with pseudo-allergic reaction to food [4]. All patients with systemic mastocytosis and urticaria pigmentosa excrete increased amounts of histamine and its main metabolite 1-methyl-4-imidazaole acetic acid in urine [14, 12]. Two patients with TMEP had normal levels [20].

Erythrodermic mastocytosis or diffuse cutaneous mastocytosis is another rare variant of mastocytosis. The skin is red, thickened, lichenified with doughy consistency with multiple small papules on the surface giving the skin a leathery appearance [21].

Mastocytosis classification included four categories: indolent mastocytosis, mastocytosis associated with hematological disorders, mast cell leukemia, lymphadenopathic mastocytosis with Eosinophilia [22].

Treatment of TMEP:

TMEP is reported to be successfully treated with 585 nm flash lamp pumped dye laser. Patient is pretreated with doxepin and H1 and H2 receptor blockade to avoid complications from laser induced mediator release. Truncal lesions recurred up to 70%, 14-months after laser treatment [23].

One pregnant patient with TMEP was successfully treated with tocolytics [(a) adrenergic neurone blocking (b) alfa adrenoreceptors blocking (c) diazoxide (d) hydralazine hydrochloride (e) minoxidil (f) prozacin) and antihistamine [14]. TMEP associated with systemic mastocytosis was ameliorated by a combination of H2 receptor antogonist (cimetidine) and H1 receptor antogonist cyproheptadine [24].

Treatment of Cutaneous manifestation include the use of H1 and H2 antihistamines, oral disodium cromoglycate, Psoralen plus ultraviolet A photochemotherapy (PUVA). Medium or high doses of UVA1 phototherapy induces long term remission in most cases of urticaria pigmentosa[25] , and potent topical corticosteroid preparation. Our patient was treated with PUVA with marked general improvement.

Conclusion:

We present a rare case of telangiectasia maculares eruptiva with a positive family history of similar affection in a younger sister.

PUVA was tried as a first line of treatment with satisfactory result with relatively low doses and short duration of treatment.

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