Telangiectasia macularis eruptiva perstans in an Indian man: Correlation between clinical, dermoscopic, histopathological, and immunohistochemical findings

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Telangiectatic macularis eruptiva perstans (TMEP) is the rarest type of cutaneous mastocytosis (CM). It presents on the trunk and upper limbs as telangiectatic macules and patches due to concentrated mast cells degranulation which are identified as reticular vascular pattern in dermoscopy. Histopathologically, they correlate with dilated capillaries and venules of the dermal superficial plexus surrounded by increased mast cells’ numbers. Giemsa and immunohistochemical stain CD117 highlight mast cells when the routine histopathology shows mild changes. Management of CM, aiming to avoid and treat symptoms of mast cell mediator release, includes systemic antihistamines as first line agents. We report a case of a 52-year-old Indian man with clinical, dermoscopic, histopathologic and immunohistochemical findings of TMEP in which intensity of erythema decreased after one month of treatment with Ketotifen 2 mg once every evening and Bilastine 20 mg once every morning.

Keywords: Telangiectatic macularis eruptiva perstans, dermoscopy, Giemsa, CD117, antihistamines

INTRODUCTION

Cutaneous mastocytosis (CM) is characterized by accumulation of mast cells confined to the skin. Telangiectatic macularis eruptiva perstans (TMEP) is the rarest type of CM, seen in <1% of mastocytosis patients, presenting as telangiectatic patches on the trunk and upper limbs. Although mostly confined to the skin, TMEP may involve bone marrow, lymph nodes, liver, spleen and gastrointestinal tract. Diagnosis is confirmed histopathologically by increased mast cells, dilated capillaries and venules of dermal superficial plexus. When mast cells’ numbers are within normal range, special stains are utilized to highlight these cells, including Giemsa, and immunohistochemical staining for c-kit (CD117), which is involved in the pathogenesis of mastocytosis.

CASE REPORT

A 52-year-old Indian man presented with six-month history of multiple asymptomatic telangiectatic macules and patches on the chest, abdomen, back and arms (Figures 1a, 1b), without associated systemic symptoms. Darier’s sign was negative. Lymphadenopathy and hepatosplenomegaly were absent. Dermoscopy showed reticular vascular pattern (Figure 2). Complete blood count and basic metabolic panel were normal. Hepatitis virus profiles were nonreactive. Histopathologic examination showed basket-woven stratum corneum, mild epidermal thinning, and basal cell layer hyperpigmentation (Figure 2). The dermis revealed a mild superficial perivascular inflammatory infiltrate of lymphocytes and mast cells, dilated blood vessels and pigment-laden macrophages. Giemsa and immunohistochemical CD117 stains showed increased number of mast cells. Clinical, dermoscopic, laboratory, histopathological, and immunohistochemical findings confirmed the diagnosis of TMEP. Ketotifen 2mg once every evening and Bilastine 20mg once every morning were given with noted decrease in intensity of erythema after one month (Figures 1c, 1d).

DISCUSSION

TMEP is the rarest CM variant. There were only 2 cases in our institution within the past 15 years, and 47 cases within the past 6 years in the database of Philippine Dermatological Society, predominantly affecting men (27 cases) and age of 18 years old and above (46 cases). Mast cells constitutively express type III receptor tyrosine kinase KIT (CD117), a useful marker in disorders of these cells. Encoded by c-kit proto-oncogene, it is involved in mast cell development and survival. Changes in KIT are important in pathogenesis of mastocytosis. Mutation in c-kit gene, causing pathologic KIT activation, results in mast cells’ accumulation, abnormal migration and activation in various tissues. Presence of this mutation in CM indicates more aggressive mastocytosis. However, this was not evaluated in our patient.

TMEP, a CM variant, manifests as telangiectatic patches on the trunk and upper limbs. Darier’s sign is frequently
negative due to mast cells’ scarcity. Reactive small blood vessels are results of persistent flushing due to concentrated mast cell degranulation. Thin tortuous linear vessels or reticular vascular pattern are seen with dermoscopy.

Histopathologically, TMEP shows increased number of mast cells, surrounding dilated capillaries and venules of the dermal superficial plexus. The increase may be mild as in our patient, hence special stains are used. The widely-available Giemsa stains mast cells’ metachromatic granules but may not detect hypogranulated cells. The more sensitive CD117 stains mast cells’ membrane strongly. CD117 is expressed in 66% (8 of 12 cases) of CM presenting as urticarial
pigmentosa, maculopapular cutaneous mastocytosis and mastocytoma. CD117 has also been used to confirm TMEP cases where routine histopathology showed normal mast cells’ numbers. Variability of mast cell count in consecutive sections may cause false negative results.Absent CD117 expression cannot exclude a diagnosis of CM. In our patient, both stains showed increased mast cells’ numbers, confirming the diagnosis of TMEP. Histopathological comparison of lesional and normal skin followed by CD117 staining can also confirm diagnosis.

Management of CM aimed to avoid and treat symptoms of mast cell mediator release. First line agents include systemic antihistamines. Ketotifen, an antihistamine and mast-cell stabilizer which has been shown as one of the best treatment for CM, was given once every evening due to sedative effect. Bilastine, a second generation H1 antihistamine, was given as substitute every morning due to its higher affinity for histamine H1 receptor than cetirizine, lack of cardiac toxicity, and sedation.

Second line treatments include topical calcineurin inhibitors, oral sodium cromolyn and phototherapy – inhibitors of mast cells’ degranulation or mediator release. Topical glucocorticoids improves pruritus, whealing, and infiltration. 585-nm flashlamp-pumped dye laser reduces vascularity of lesions.TMEP may involve other organ systems in 33-50% of cases within six years or more after onset. This risk increases with age, necessitating regular monitoring.

CONCLUSION

This report highlighted and correlated the clinical, dermoscopic, histopathologic and immunohistochemical findings of TMEP. Telangiectatic patches due to concentrated mast cells degranulation are identified as reticular vascular pattern in dermoscopy. Histopathologically, they correlate with dilated capillaries and venules of the dermal superficial plexus surrounded by increased mast cells’ numbers. Giemsa and immunohistochemical stain CD117 highlight mast cells when the routine histopathology shows mild changes. This report also supported systemic H1-antihistamines as first line agents for TMEP.