

Case Report

Telangiectasia macularis eruptiva perstans

持久發疹性斑狀毛細血管擴張症

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A 58-year-old man presented with asymptomatic erythematous telangiectatic macules over the back, and upper limbs sparing the face and hands. A diagnosis of telangiectasia macularis eruptiva perstans was made based on clinicopathological findings. No clinical evidence of systemic involvement was present. Regular follow-up will be given to detect any development of systemic mastocytosis.

男患者，58歲，於背部及上肢出現無症狀性毛細血管擴張紅斑，面及手部未累及。臨床及病理學診斷為持久發疹性斑狀毛細血管擴張症。無臨床跡象顯示有內臟系統受累。患者將接受定期隨診，以發現任何系統性肥大細胞增生症。

Keywords: Systemic mastocytosis, telangiectasia macularis eruptiva perstans

關鍵詞：系統性肥大細胞增生症，持久發疹性斑狀毛細血管擴張症

Introduction

Telangiectasia macularis eruptiva perstans (TMEP) is a rare form of cutaneous mastocytosis which was first described by Parkes Weber in 1930.¹ It is generally considered to be limited to the skin

but there has been reports of systemic involvement and progression to systemic mastocytosis. A case of TMEP affecting a Chinese man is reported.

Case report

A 58-year-old ethnic Chinese man presented with a two year history of an erythematous macular rash over his back and upper limbs which became more prominent with heat and sweating. He was otherwise asymptomatic with no systemic complaints. Past medical history included hypertension and gastro-esophageal reflux which was controlled with felodipine and esomeprazole. He was a chronic smoker and social drinker and there was no significant family history.

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Physical examination showed coalescing erythematous macules which demonstrated fine telangiectases on a background of multiple faint tan macules. The lesions were distributed over the back and upper limbs sparing the face, chest, abdomen and hands (Figures 1 & 2). Darier's sign and dermatographism were not elicitable. Systemic examination was unremarkable with no stigmata of chronic liver disease and without lymphadenopathy or hepato-splenomegaly. The differential diagnoses included generalised essential telangiectasia, telangiectasia macularis eruptiva perstans, chronic alcoholism, chronic liver disease.

Investigations including complete blood picture, liver and renal function tests showed normal



Figure 1. Coalescing erythematous macules with fine telangiectases over the arm with faint tan macules in the background.



Figure 2. A Close up of the macular lesions on the arm showing fine telangiectases.

results. A skin biopsy from a back lesion without using adrenaline showed telangiectases with a sparse perivascular infiltrate consisting of oval and spindle shaped mast cells with basophilic granules on haematoxylin and eosin stain (Figures 3 & 4). Special staining with chloroacetate esterase highlighted an increase in mast cells (Figure 5). The clinical and histological features were consistent with telangiectasia macularis eruptiva perstans.

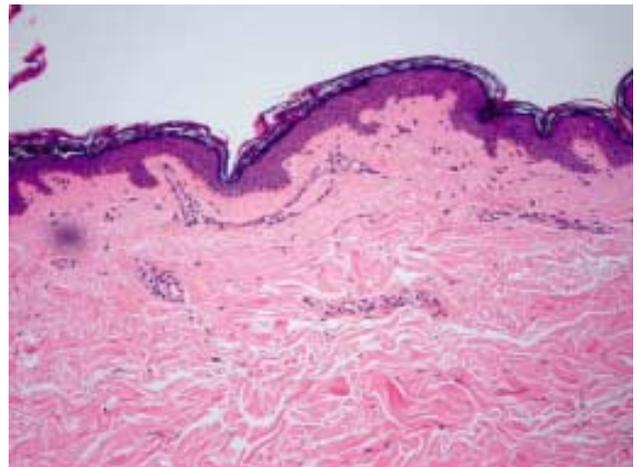


Figure 3. There are telangiectases in the upper dermis with a sparse perivascular infiltrate. (H&E, Original magnification x 10)

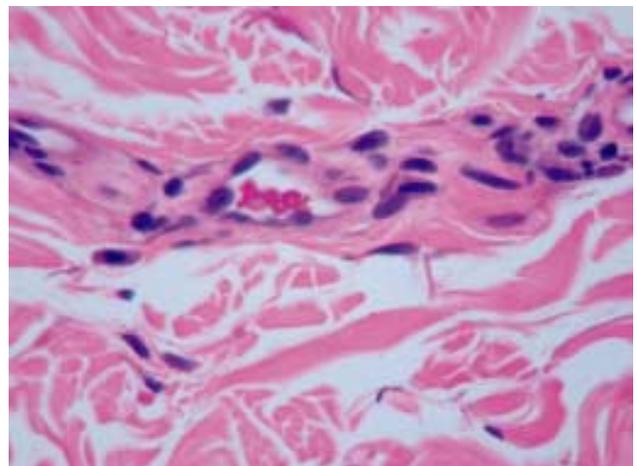


Figure 4. High magnification showing oval and spindle shaped mast cells with basophilic granules. (H&E, Original magnification x 40)

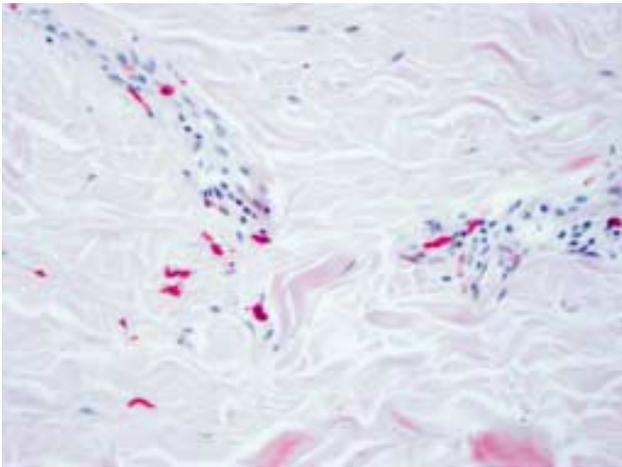


Figure 5. An increase in mast cells number is highlighted by the chloroacetate esterase stain (CAE, Original magnification x 40).

Discussion

TMEP is characterised by the permanent vessel expansion secondary to the local release of mast cell mediators and angiogenetic factors.² It occurs more commonly in adults than in children.³ Familial cases of TMEP have been reported in four children in three generations of one family.⁴

Clinically TMEP is characterised by coalescing erythematous and tan coloured macules measuring 2 to 10 mm in diameter with fine telangiectases. The lesions are typically distributed over the trunk and limbs bilaterally, although a unilateral variant has been reported.⁵ Darier's sign is usually absent or slight due to the paucity of mast cell numbers in the skin. This may be of importance to distinguish true TMEP from a different group of patients with adult onset cutaneous mastocytosis with telangiectases associated with lesions of urticaria pigmentosa with significant systemic involvement.⁶

Histologically, TMEP comprises of a perivascular infiltrate of mast cells in the upper third of the dermis with proliferation of blood vessels within

the papillary dermis. The mast cells are spindle shaped and may be very subtle. They are better visualised with special stains such as giemsa, toluidine blue, or by histochemical techniques to reveal chloroacetate esterase, or immunocytochemical stains of tryptase or chymase. Eosinophils are generally absent.⁷ The number of mast cells is marginally increased in TMEP which may pose difficulties in diagnosis. Weedon has suggested that normal skin may contain up to 15 mast cells per high power field,⁸ whilst in our patient a total of 19 mast cells per high power field were present. There however exists no reliable numerical cut off point that allows a decision whether a mast cell count is within the limits of normal skin or whether it is increased.⁷

TMEP is considered to be benign and limited to the skin, but some opionate that it is merely a macular form of maculopapular cutaneous mastocytosis which has high association with systemic involvement.⁷ Features of systemic involvement include headaches, flushing, diarrhoea, splenomegaly, abnormal skeletal radiographs, and bone-marrow involvement. Other associations include multiple myeloma⁹ and polycythemia rubra vera.¹⁰ A case of TMEP has been reported to progress rapidly to systemic mastocytosis after a five year history of purely cutaneous involvement.¹¹ Apart from baseline workup, additional investigations should be done and a bone marrow examination is necessary if systemic symptoms occurred. Assay for serum tryptase level is useful which would be greater than 20 ng/ml in most patients with SM and correlate with the burden of neoplastic mast cells.¹²

Counselling on the avoidance on possible triggering factors is important for patients with mastocytosis: temperature, physical exertion, emotional stress, general anaesthetics, alcohol (may potentiate the telangiectasia in our patient), non-steroidal anti-inflammatory drugs, narcotic analgesics. Otherwise the treatment of TMEP is

symptomatic with H1 and H2 histamine receptor antagonists which help with pruritis, flushing and GI complaints. Oral sodium cromoglycate may help alleviate abdominal pain, nausea or diarrhoea. Phototherapy utilising UVA plus psoralen (PUVA) or UVA1 has been shown to be effective in reducing mast cells and symptoms in those with severe pruritis but lesions will almost invariably recur.¹³ Effective treatment for the skin lesions has been reported with surgery with 585 nm flashlamp-pumped Dye laser which reduces the vasculature with no apparent effect on mast cells.^{6,14} One patient with severely symptomatic and refractory TMEP was treated with total skin electron beam irradiation (TSEB) with resolution of skin lesions and pruritis.¹⁵

In summary, our patient has asymptomatic TMEP. Regular follow-up should be undertaken to watch out for sinister features. Meanwhile, patient is advised to abstain from alcohol.

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