Telangiectasia macularis eruptiva perstans in the presence of liver cirrhosis

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INTRODUCTION

Telangiectasia macularis eruptiva perstans (TMEP), is a rare variant of cutaneous mastocytosis characterized by pruritic telangiectatic macules.[1,2] Cirrhosis is a relatively common disease of liver dysfunction associated with telangiectasias and pruritus.[3] Typical lesions of TMEP are blanchable, reddish-brown telangiectatic macules on a tan-brown background on the trunk. Cutaneous manifestations of cirrhosis include telangiectatic macules and spider angiomata, which can appear remarkably similar. Here we present a case of TMEP in a patient with underlying cirrhosis of the liver and underscore the importance of discerning different types of telangiectasia, both clinically and histologically, to make an appropriate diagnosis.

Abbreviation used:

TMEP: telangiectasia macularis eruptiva perstans

CASE REPORT

A 73-year-old Mexican man presented with a 3-month history of a worsening rash on his chest, back, and arms. He reported that the lesions were itchy and that after scratching, red bumps would develop. He denied any other symptoms, including nausea, vomiting, flushing, diarrhea, abdominal pain, or headaches. Detailed review of systems was negative. Medical history was notable for cirrhosis of the liver, *Candida* esophagitis, thrombocytopenia, hemorrhoids, rhinitis, hypertension, hyperlipidemia, and benign prostate hyperplasia. Medications included gabapentin, levothyroxine, loratadine, pantoprazole, and tamsulosin. Upon close visual inspection and under dermoscopy, physical examination found discrete, blanchable, reddish-brown macules with clearly visible telangiectasia (Fig 1, *A-C*). Darier sign was negative. Additionally, mild palmar erythema was noted bilaterally.

A biopsy of the back found a perivascular mononuclear cell infiltrate with increased mast cells confirmed by Leder stain, consistent with a diagnosis of cutaneous mastocytosis (Fig 2 *A* and *B*). Laboratory work was remarkable for elevated levels of alkaline phosphatase and total bilirubin at 169 IU/L and 2.3 mg/dL, respectively. A complete blood count found anemia (4.1 M/μL) and thrombocytopenia (48 K/μL). A serum tryptase level was within normal limits.

Management included the avoidance of alcohol and nonsteroidal anti-inflammatory drugs and the initiation of a regimen of antihistamines. The patient was started on cetirizine, 10 mg, in the morning; diphenhydramine, 50 mg, every night at bedtime; and Sarna lotion for control of pruritus. Two months after his initial presentation, he reported that he was feeling better on the prescribed regimen with significantly diminished pruritus. Despite a normal tryptase level, the patient was referred for further workup to evaluate for systemic mastocytosis because of the presence of the laboratory abnormalities described previously. However, he has yet to follow up.

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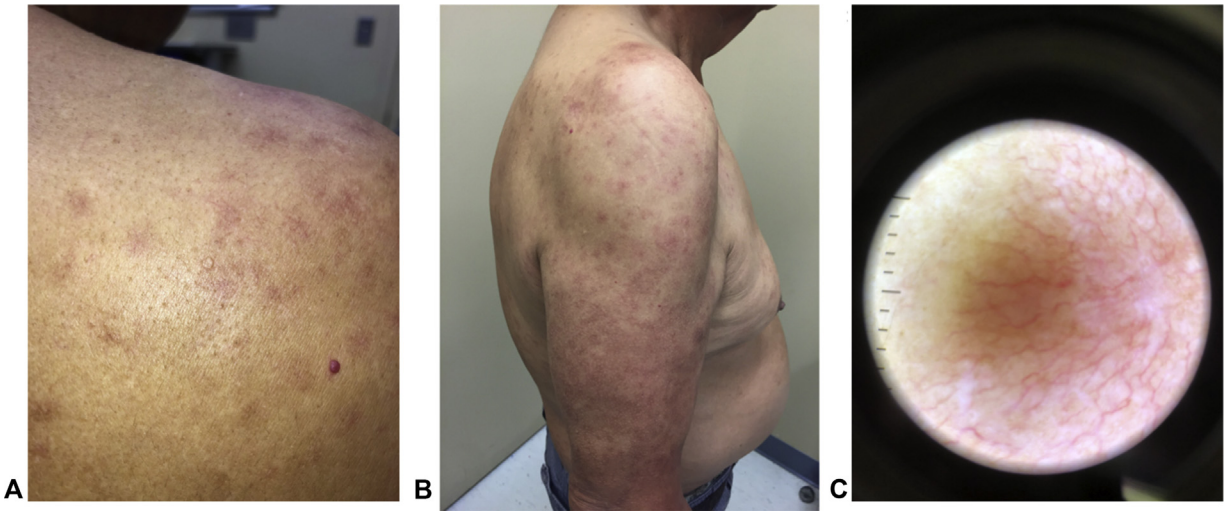

Fig 1. Numerous fine telangiectasias overlying erythematous patches on the (**A**) right posterior shoulder and (**B**) left lateral proximal arm. Dermoscopic image from a shoulder lesion depicting numerous telangiectasias (**C**).

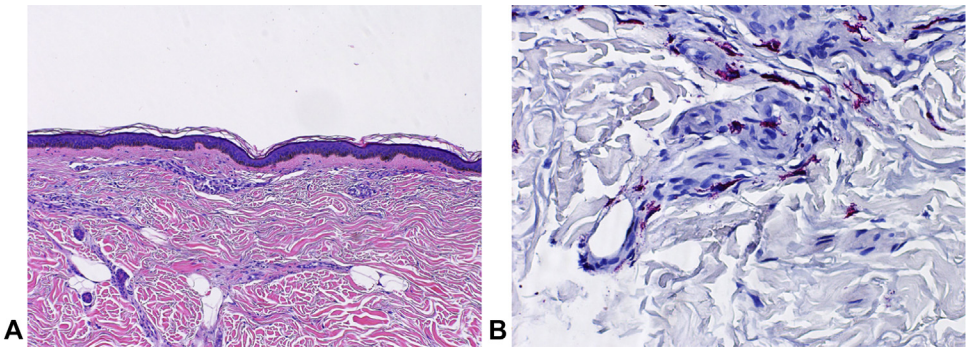

Fig 2. Biopsy of the upper left back showing (**A**) perivascular mononuclear cell infiltrate with increased mast cells and (**B**) Leder stain highlighting many mast cells. (Original magnifications: **A,** ×40; **B**, ×100.)

DISCUSSION

TMEP is a rare disorder of mast cell proliferation often limited to the skin but can rarely generalize and effect other organs. TMEP was initially thought to be a subvariant of urticaria pigmentosa but has since come to be recognized as its own type of cutaneous mastocytosis.[4,5] Urticaria pigmentosa, the most common form of cutaneous mastocytosis, and TMEP both present with a red-brown maculopapular rash.[6] Characteristic lesions of TMEP have telangiectasia, whereas those found in urticaria pigmentosa do not.[6] Another distinction is that TMEP frequently has a negative Darier sign, whereas urticaria pigmentosa has a positive Darier sign.[5,6]

Cirrhosis of the liver is a relatively common unrelated condition that similarly presents with telangiectasia and pruritus. The etiology of pruritus is thought to be the accumulation of bile salts under the skin, whereas the pathogenesis of the telangiectatic damage is a result of a hyperdynamic circulation.[7] The presence of these clinical findings in cirrhosis can easily obscure the identification of a rare, cutaneous, pruritic, telangiectatic disorder. As was the case in this patient, his pruritus was attributed to cirrhosis, and his rash was thought to be spider angiomata, a classic stigmata of liver disease.

Spider angiomata are vascular lesions that can appear exceedingly similar to the macules of TMEP (Fig 3).[8] However, subtle differences exist between the 2 types of telangiectasia, both clinically and histologically. Unlike TMEP, spider angiomata have a central arteriole from which numerous small, twisted vessels radiate.[7] Similar to TMEP, the lesions develop over the area of the superior vena cava, namely the upper trunk and arms, and overlap with areas commonly affected by TMEP.[1,7] Unlike lesions of TMEP, spider angiomata are not known to be pruritic. However, cholestasis of liver disease is

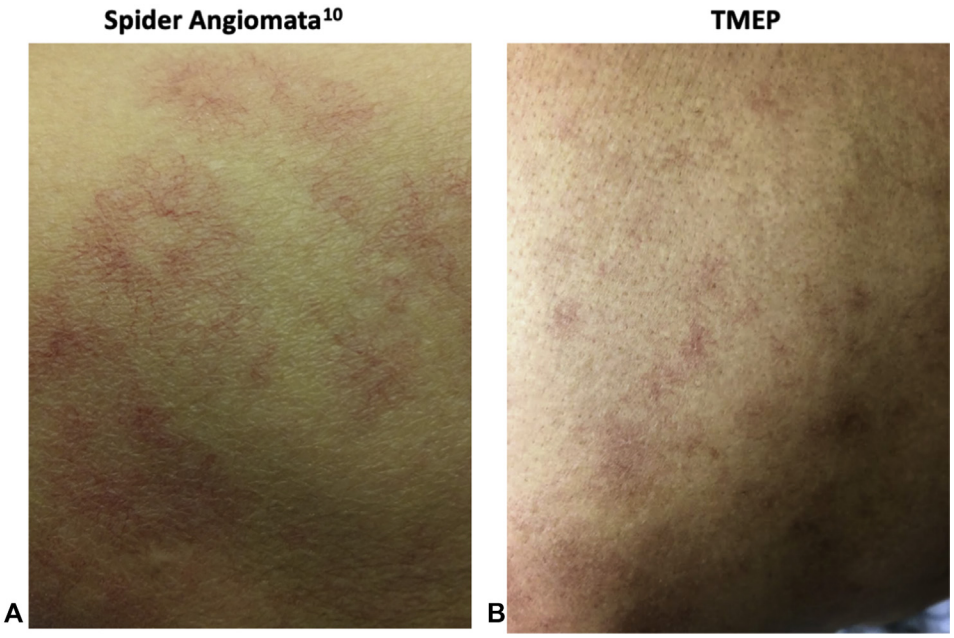

Fig 3. A central arteriole from which numerous small, twisted vessels radiate depicting (**A**) multiple spider angiomata. Numerous fine telangiectasias overlying erythematous patches characteristic of (**B**) TMEP.

associated with a generalized pruritus.[7] Patients with cirrhosis of the liver can also have increased blood flow in the upper extremities that may explain the palmar erythema in these patients.[7,9] Although patients with TMEP are not known to have similar palmer erythema, they may present with telangiectatic macules on the palmar surfaces of the hands.[1] Histologically, spider angiomata will show a central ascending feeding blood vessel with branching dilated blood vessels in the subepidermal region and would not show increased number of mast cells. In comparison, lesions of TMEP are characterized by the presence of a superficial perivascular infiltrate with an increased number of mast cells histopathologically.

Mast cell dysregulation in TMEP is thought to be the result of a mutation in the proto-oncogene c-*kit*, which encodes a mast cell growth factor receptor.[1] Interestingly, more recent research has also implicated mast cells and histamine as key players in the progression of liver damage in cirrhosis.[10] In fact, preventing mast cell migration and activation is found to be critical to mitigating inflammatory liver damage in alcoholic liver disease.[10] Although the incidence of TMEP in cirrhosis is unknown and likely uncommon, this case illustrates the importance of careful history taking, physical examination, and potentially biopsy to make an appropriate diagnosis in this circumstance.

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