

Diffuse telangiectasia: A clue to the TEMPI syndrome



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Key words: bortezomib; monoclonal gammopathy; telangiectasia; TEMPI syndrome.

INTRODUCTION

TEMPI syndrome (a syndrome related to monoclonal gammopathy) is a rare disorder; only 22 cases have been reported in the literature. It is characterized by the pentad of telangiectasias, erythrocytosis with elevated erythropoietin, presence of monoclonal gammopathy, perinephric fluid collection, and intrapulmonary shunting. The first letter of acronym TEMPI stands for telangiectasia, which is the most common feature of this disease and which can be identified by dermatologists. Here, we report a case of TEMPI syndrome with typical cutaneous manifestations.

CASE REPORT

We present the case of a 75-year-old woman with an 8-year history of diffuse telangiectasia involving the face, trunk (Fig 1), upper extremities, and oral mucosa. Dermoscopy revealed dilated vessels and red lacuna (Fig 2), consistent with the dilated venules observed by histologic examination. Nailfold capillary abnormalities were not observed. The patient also complained of dyspnea (when climbing the stairs) and fatigue for the last 2 years without diarrhea, loss of weight, myalgia, or cold intolerance. She was previously diagnosed with polycythemia vera at another hospital 4 years earlier and treated with hydroxyurea. Initial physical examination showed no abnormalities.

Her blood work showed erythrocytosis (red blood cell count, $5.52 \times 10^9/L$; hemoglobin, 156 g/L). Further investigation revealed elevated erythropoietin (EPO > 750 mIU/mL). Hepatic function and renal function were normal. Abdominal and pelvic

ultrasound examination was unremarkable, except for bilateral perinephric fluid collections (Fig 3). Chest computed tomography was normal. Serum protein electrophoresis and immunofixation revealed IgA-lambda paraproteinemia. Further pulmonary ventilation/perfusion scan revealed an intrapulmonary shunt. Bone marrow biopsy showed that less than 10% of all nuclear cells were plasma cells with lambda light-chain restriction. Abdominal computed tomography did not show organomegaly. Nerve conduction test findings were normal. The hypothalamic–pituitary–adrenal axis and thyroid function were also normal. Based on these findings, we diagnosed the patient with TEMPI syndrome. After 2 cycles of bortezomib (1 mg/m² on days 1, 4, 8, 11) and dexamethasone (40 mg on days 1, 4, 8, 11), the telangiectasia improved (Fig 4), the hemoglobin level increased, and M-protein and erythropoietin levels decreased.

DISCUSSION

The TEMPI syndrome, first described by Sykes in 2011,¹ is characterized by the pentad of telangiectasias, erythrocytosis with elevated EPO, monoclonal protein, perinephric fluid collection, and intrapulmonary shunting. To date, 22 cases of TEMPI syndrome have been reported.² Previous reports suggest no gender-specific differences, and the average age at disease onset was 49 years. The associated monoclonal proteins seen in the TEMPI syndrome were IgG-kappa, IgA-lambda, and IgG-lambda.³ As TEMPI syndrome has been classified under plasma cell dyscrasia, bortezomib remains the only first-line therapeutic option.⁴ Other alternative

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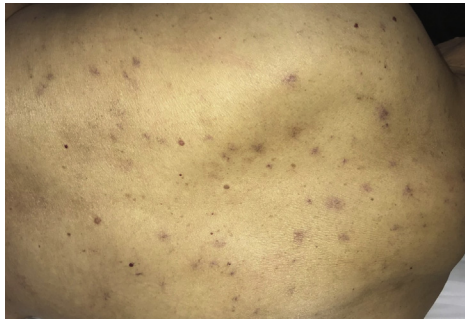


Fig 1. Diffuse telangiectasia on the trunk.

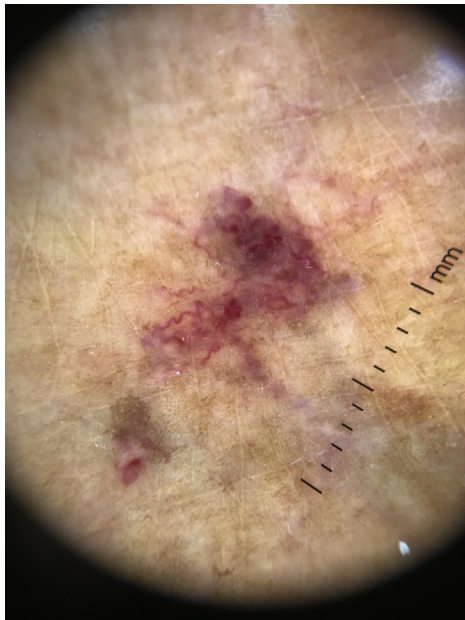


Fig 2. Dilated vessels and red lacuna observed by dermoscopy.

therapeutic options include daratumumab,⁵ lenalidomide,⁶ and autologous stem cell transplantation.⁷

Our patient presented with telangiectasia on the face, trunk, and upper extremities, but sparingly on the lower extremities. We also observed hemangioma-like lesions on the face and red macules on the mucosa. Telangiectasia and intrapulmonary shunt improved in most patients as well as the disappearance of the M spike, which indicated that telangiectasia was likely the consequence of the angiogenic process due to monoclonal gammopathy.² The angiogenic process may be related to the production of angiogenic cytokines released by plasma cells and microenvironmental factors.⁸ However, the precise mechanism of action remains unclear. We performed total-body skin examination and used dermoscopy to evaluate these lesions for the first time, which may provide information for further exploration of the TEMPI syndrome.



Fig 3. Perinephric fluid collections observed by ultrasound.



Fig 4. The diffuse telangiectasia disappeared after treatment.

There are many cutaneous conditions associated with monoclonal gammopathy, which are strongly suggestive of the diagnosis. For example, raccoon eyes in patients with light-chain amyloidosis; necrobiotic xanthogranuloma, scleromyxedema, scleroedema without diabetes, and diffuse hyperpigmentation in the POEMS syndrome, as well as febrile urticaria in the Schnitzler syndrome. The differential diagnosis of diffuse telangiectasia in this patient includes hereditary disorders, several hormonal abnormalities (iatrogenic excess of steroid or estrogen, tumors overproducing hormones, failure of the liver to inactivate estrogen, etc.), autoimmune diseases (scleroderma; especially, CREST syndrome), carcinoid syndrome, and mastocytosis.

Even though the TEMPI syndrome is a newly named rare disorder, telangiectasia and monoclonal gammopathy are not rare in clinical practice. Therefore, for patients with diffuse telangiectasia and erythrocytosis, the clinician should measure plasma erythropoietin levels and recommend serum protein electrophoresis as well as screening for TEMPI syndrome.

Conflicts of interest

None disclosed.

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