CASE REPORT

An Unusual Case of Telangiectasias

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Abstract

Background: Telangiectasias are defined as persistent dilatation of small capillaries in the superficial dermis

Case: A 26-year-old woman presented with red lesions, epistaxis, joint pains, color changes of the hands, and breathlessness. On clinical examination and investigations, a final diagnosis of mixed connective tissue disease (MCTD), with interstitial lung disease (ILD), with telangiectasias, and epistaxis was made. Telangiectasias and epistaxis are rare presentations of MCTD.

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Introduction

Telangiectasias are defined as persistent dilatation of small capillaries in the superficial dermis that is visible as fine, bright, nonpulsatile red lines, or net-like patterns on the skin. The dilated vessels are commonly seen on the face around the nose, cheeks, and chin. Telangiectasias can be congenital or acquired. Congenital causes are nevi flammeus, Von Hippel–Lindau syndrome, ataxia–telangiectasia, Sturge–Weber syndrome, hereditary hemorrhagic telangiectasias (HHT), etc. Acquired causes are Cushing’s syndrome, venous hypertension, acro rosea, connective tissue disease, carcinoid syndrome, radiation exposure, or chemotherapy. Epistaxis is a common complaint, the etiologies of which are mostly local nasal lesions, arteriovenous malformations, bleeding, coagulation disorders, leukemias, and Wegener’s granulomatosis. Connective tissue diseases (CTD’s) form a rare cause of both these complaints. Telangiectasias mostly occur in systemic sclerosis (SSC) (calcinosis, Raynaud’s phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia syndrome or localized SSC). Epistaxis can occur due to nasal telangiectasias or as concomitant Von Willebrand disease (VWD). MCTD, a separate entity, which comprises features of systemic lupus erythematosus (SLE), SSC, rheumatoid arthritis (RA), and polymyositis (PM), is a relatively rare cause of both these complaints. In most studies on such patients, only periangual telangiectasias were found. Here, we report a rare case of a young woman who presented with telangiectasias all over the face and upper body, along with epistaxis that occurred together.

Case Description

A 26-year-old female patient came with complaints of reddish colored lesions over face and palms since 10 months. She had joint pain and swelling involving small joints of hands with early morning stiffness lasting for 40 minutes since 9 months. She complained of color change (white to reddish blue) in the fingers of both hands on working in cold water since 9 months, which was suggestive of Raynaud’s phenomenon. She also had multiple oral ulcers and epistaxis since 9 months. Bouts of epistaxis occurred at least thrice weekly with a loss of a quarter teaspoon of blood. There was a history of breathlessness on climbing stairs since 3 months. She had no history of (h/o) photophobia, hematuria, decreased urine output, or thrombotic episodes. There was no history of bleeding from any other site. There was no history of bleeding or coagulation disorders in the family. No h/o similar lesions over the body in any family member.

On general examination, she was moderately built and nourished. Her vitals were stable, mild pallor was present. Multiple oral ulcers 1–2 mm were present over buccal mucosa and inside of lips. Red lesions, small, around 1 mm in diameter, blotchy not raised, blanching on pressure suggestive of telangiectasias were present over the face, hands, and including palms. Similar red lesions were seen inside the oral or nasal cavity; on preliminary examination, no h/o similar lesions over the body in any family member.

On musculoskeletal examination, her skin appeared stretched, and could not be pinched up over her fingers, up till wrist. Tenderness over all metacarpophalangeal joints and proximal interphalangeal joints. No swelling was noted.

On bathing her hands with cold water, Raynaud’s phenomenon was induced.

On nose and throat examination, telangiectasias were noted in the nasal cavity. There were no local lesions or arteriovenous malformations. On ophthalmology examination, no uveitis or telangiectasias were seen (Figs 1 to 4).

On account of multisystem involvement—Raynaud’s, numerous telangiectasias, skin tightening, inflammatory joint pains, dyspnea, and dyspnea, the patient appeared to have scleroderma, but for oral ulcers and epistaxis, further investigations were warranted. Her hemoglobin was 9.8 gm%. Total leukocyte count was 7,800/cumm, platelets were 2.6 lakh/cumm, and peripheral blood smear was normocytic and normochromic. Erythrocyte sedimentation rate was 24 mm. Her Liver and renal functions were normal. Her blood sugar level was 91 mg%. Her urine showed no albumin and sugars. Her bleeding time, clotting time, prothrombin time, and activated partial thromboplastin time were normal. Iron studies and vitamin B12 were normal. Her pulmonary function test showed moderate restriction, and the 6-minute walk test was normal (no significant desaturation). The helium dilution study (diffusing capacity study) were normal.

Based on the clinical features and investigations, she was finally diagnosed with MCTD, with extensive ILD, and epistaxis. She was treated with prednisolone 1 mg/kg/day tapered gradually and maintained at 10 mg/day, azathioprine 50 mg once a day (OD), hydroxychloroquine 200 mg OD, and nifedipine 10 mg were started. She reported improvement in her symptoms. Her telangiectasias had reduced in visible number, while no further lesions appeared. Her epistaxis episodes were reduced in frequency.

Keywords: Telangiectasias, Epistaxis, MCTD, ILD

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Criteria are used for diagnosis—Kasukawa (Table 1), Sharp, and Alarcon-Segovia. Characteristic initial presentation is Raynaud’s phenomenon associated with puffy fingers. Skin rashes suggestive of SLE-like malar rash or of dermatomyositis like heliotrope rash on eyelids and oral ulcer. Pulmonary fibrosis and isolated pulmonary arterial hypertension (PAH) or secondary PAH may develop. Other features include periangual telangiectasias, esophageal dysmotility, arthritis, pericarditis, and renal disease. Such extensive telangiectasias are relatively rare in MCTD. Telangiectasias have many causes, of which MCTD forms a rare part. HHT is the most important differential for a patient with multiple telangiectasias. Our patient did not have a family history, and so this differential was ruled out, as were others. Patients of MCTD can sometimes have epistaxis due to telangiectasia in the nasal mucosa. There are also reports of acquired VWD in patients with CTD, which may cause bleeding (epistaxis).

Glucocorticoids remain the mainstay of treatment. ILD mostly responds to low to moderate dose steroids and if needed cyclophosphamide pulses are used.

Table 1: Kasukawa criteria for MCTD

<table>
<thead>
<tr>
<th>Disease category</th>
<th>Symptom</th>
</tr>
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<tbody>
<tr>
<td>SLE</td>
<td>Polyarthritis</td>
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<tr>
<td></td>
<td>Lymphadenopathy</td>
</tr>
<tr>
<td></td>
<td>Malar rash/facial erythema</td>
</tr>
<tr>
<td></td>
<td>Mericarditis or pleuritis</td>
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<tr>
<td></td>
<td>Thrombocytes of &lt;100–10^9/1 or leukocytes of &lt;4.0–10^9/1</td>
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<tr>
<td>Progressive SSC</td>
<td>Sclerodactyly</td>
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<tr>
<td></td>
<td>Long fibrosis or restrictive lung disease (VC &lt; 80%) or carbon monoxide diffusion of &lt;70%</td>
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<tr>
<td></td>
<td>Esophageal hypomotility or dilatation</td>
</tr>
<tr>
<td>PM</td>
<td>Muscle weakness</td>
</tr>
<tr>
<td></td>
<td>Elevated serum levels of myogenic enzymes</td>
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<tr>
<td></td>
<td>Myogenic pattern on electromyography</td>
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</tbody>
</table>

MCTD criteria are met in case of Raynaud’s phenomenon or presence of swollen hands or fingers or positivity for anti-U1 small nRNP-antibodies and at least one symptom in two of three disease categories. VC, vital capacity

Kasukawa, R. Excerpta Medica. 1987

Fig. 1: Oral ulcers and oral telangiectasias

Fig. 2: Telangiectasias over the face

Fig. 3: Telangiectasias over the hands

Fig. 4: Telangiectasias in the nose causing epistaxis

Discussion

Mixed connective tissue disease (MCTD) is a distinct rheumatic disorder with features of SLE, RA, SSC, and PM in varying proportions and high titers of uridine (U1) nuclear ribonucleoprotein particle (NRP). Three references