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CASE REPORT

Thrombotic thrombocytopenic purpura as an initial presentation of systemic sclerosis: a case report

Musab Abdullah Bin Shuayl[1]

ABSTRACT

Background: Systemic Scleroderma is a rare disease of connective tissues. It involves rare complications including TTP and SRC, which are similar in presentation.

Case Presentation: The present case presents a 44-year-old female with SSc and combined complication. History was taken from the patient, several investigations and examinations were performed to confirm SSc, and to identify the accompanied complication. Lab investigations, kidney biopsy, CT scan, ECHO, X-ray, physical examination, cardiovascular, respiratory, and gastrointestinal examinations were performed, and SSc accompanied by idiopathic TTP was confirmed in the patient.

Conclusion: TTP is an indication of early SSc and identifying TTP from SRC requires accurate investigations.

Keywords: Systemic sclerosis, thrombotic thrombocytopenic purpura, idiopathic TTP, case report.

Introduction

SSc (systemic scleroderma) is a disease of connective tissues which is associated with autoimmunity, fibrosis, and vasculopathy. It is a rare disease with an annual incidence of 10–20 cases/1 million persons [1]. TTP is a rare clinical syndrome that is characterized by fever, fluctuating neurological signs, MAHA, thrombocytopenia, and renal dysfunction [2]. TTP is a rare complication of scleroderma [2]. Few reports were presented about TTP as a rare complication of SSc, hence, it is very important to distinguish between TTP combined with SSc and SRC for totally different pathogenesis and strategies of treatment [3]. Here, we present a case suspected of SSc with a combined complication.

Case Presentation

Our case is a 44-year-old female, who 5 months earlier developed pneumonia for which she received oral antibiotics, after 1 month she had developed PUD and gastric erosions. PUD was treated with triple therapy, which initiated nausea and vomiting (3–4 times/day) with decreased oral intake and weight loss (she lost almost 10 kg within 5 months); then, she started to lose hair and eyebrows, and also complained of bilateral progressive pre-orbital swelling. Two months earlier, the female was complaining of intermittent regular palpitations with shortness of breath NYHA class 2–3, PND, and orthopnea (slept on 3 pillows at home). Before presentation by 6 weeks, she suffered from progressive bilateral hand stiffness, pain, and blue discoloration of the right thumb sometimes extending to the index finger. On presentation, she denied any history of dysphagia for solid or liquid food, no oral or genital ulcers, no rashes, no joint pain or swelling, no scalp hair loss, no history of seizures before, or LOC.

Physical examination including hand examination, examination of the cardiovascular system, respiratory system and gastrointestinal system, lab investigations, kidney biopsy, pulmonary function tests, and measuring ADAMTS13 were performed for the case. The physical examination showed that she was conscious oriented and not pale, cyanosed, or suffering from jaundice. Her temperature was 36.9°C by oral method, respiratory rate was 20, blood pressure was 122/79, SpO2 was 98%, and her weight was 53 kg. Her hand showed tenderness at

Correspondence to: Musab Abdullah Bin Shuayl
*Department of Internal Medicine, College of Medicine, Prince Sattam Bin Abdulaziz University, Al-Kharj, Saudi Arabia.
Email: musab.binshuayl@gmail.com
Full list of author information is available at the end of the article.
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the 3rd, 4th, and 5th MCP and bilaterally PIP joints, blue discoloration of index finger of the right hand was found but with good perfusion, sensation and warmness, no other joints swelling or tenderness, and no rashes in upper or lower limbs were found. The lower limb examination showed that pulses were palpable starting from the femoral artery pulses down, radial artery was palpable bilaterally, but the brachial was difficult to palpate on both sides. The cardiovascular system examination revealed that JVP was not raised, S1 and S2 were normal, and there were no add or murmur sounds. Examination of the respiratory system detected bilateral breath sound and vesicular breathing only. High-resolution CT showed bibasilar reticulation with ground-glass changes concerning for underlying interstitial lung disease (NSIP). Chest X-ray showed increased reticular shadowing over the basal areas with normal cardiac shadow, while the ECHO showed normal LV function with impaired relaxation, EF: 58%, both lower limbs Doppler for DVT and upper limbs artery Doppler were negative. On gastrointestinal examination, the abdomen was soft, lax with no tenderness, ascites or organomegaly. The results of lab investigations were as follows: WBCS 8.700, Hgb 95, PLT 80.0, Na138, K3.8, Cl103, Cr354, BUN 4.1, glucose 5.5, Mg0.71, Phos2.3, Ca2.28, PT 14, INR 1.09, Lactic acid L 0.4, APTT34.7, ALT 23, ESR 9, CRP6, normal coagulation profile and complement level, and the lipid profile showed elevation in cholesterol, LDL and triglycerides. By autoimmune investigations, it was found that ANA was highly positive with the nuclear pattern, each of dsDNA, CRP, RF, SS-A Ab, SS B Ab, SM Ab and Jo-1Ab were negative, while Scl 70 Ab was positive. The peripheral blood smear (Figure 1) showed dimorphic blood picture with mild anisocytosis, and both schistocytes and spherocytes were significant (around 3%). Adequate neutrophil count was found with slight toxic changes and few reactive looking lymphocytes were noted. Occasional large and giant platelets were seen.

The kidney biopsy revealed that there was acute thrombotic microangiopathy with associated occlusive fibrin thrombi and fibrinoid necrosis in few arteries and arterioles, patchy acute tubular injury and mild interstitial edema were also found. There was no significant interstitial fibrosis or tubular atrophy, but mild arteriosclerosis was observed, and there was no arteriolar hyalinosis. The pulmonary function test of the patient is summarized in Table 1, ADAMTS13 activity was found to be low.

Discussion

Diagnosis of SSc is based on heterogeneous clinical findings, including a young or middle-age woman with Raynaud phenomenon as well as skin changes with musculoskeletal discomfort and gastrointestinal symptoms including dysphagia, and symptoms related to gastroesophageal reflux disease. The degree of skin thickening depends on the duration and subtype of the disease; hair loss is one of the cutaneous manifestations of SSc. With the progression of the disease, ulcerations may appear over joints, wrists and flexion contractures of the fingers [1]. Regarding the clinical presentation and skin manifestations, it seems that the case experienced several symptoms of SSc, while she did not experience others, the disease may be early in onset and can't be confirmed depending on the previous evidence, so further investigations were performed. Internal organ complications are common in SSc patients, and they are rarely symptomatic until the last stages of the disease [1]. SSc can affect lungs and pulmonary blood vessels, the

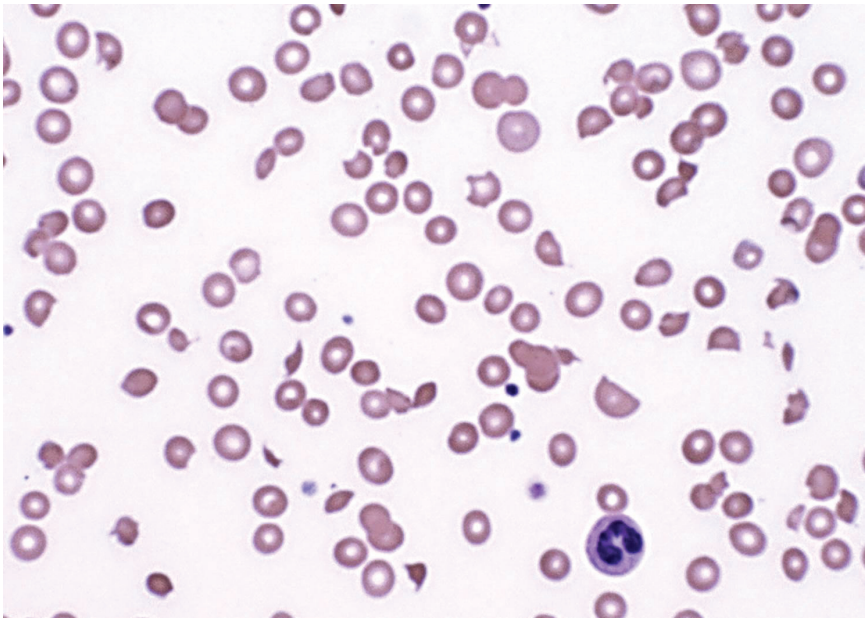

Figure 1. *Peripheral blood smear.*

Table 1. Pulmonary function tests.

Pulmonary Function Test	Pre-Branch (Pred Actual %)	Post-Branch (Pred Actual %)	Change
Spirometry			
FVC (L)	3.33	1.87	56
FEV1 (L)	2.69	1.66	61
FEV1/FVC (%)	81	89	109
FEF 25% (L/sec)	5.09	3.77	74
FEF 50% (L/sec)	4.09	3.04	74
FEF 75% (L/sec)	1.57	0.94	59
FEF 25–75% (L/sec)	2.83	2.19	77
FEF Max (L/sec)	6.51	3.79	58
FIVC (L)	1.8		
FIF 50% (L/sec)	3.5	1.85	52
FIF Max (L/sec)	2.06		
MEP (CmH2O)	150	57	37
MIP (CmH2O)	−83	−43	51
Lung Volumes			
SVC (L)	3.33	1.88	56
IC (L)	2.1	1.15	54
ERV (L)	1.23	0.73	59
TGV (L)	2.57	2.47	96
RV (Pleth) (L)	1.54	1.74	112
TLC (Pleth)(L)	4.67	3.63	77
RV/TLC (Pleth)(L)	33	48	145
Diffusion			
DLCOunc (mM/min/kPa)	6.96	1.9	27
DLCOcor (mM/min/kPa)	6.96	2.36	33
DL/VA (mM/min/kPa/L)	1.49	0.76	51
VA (L)	4.67	3.09	66
Airways Resistance			
Raw (kPa/L/s)	0.18	0.12	68
Gaw (L/s/kPa)	10.5	8.15	77
sRaw (kPa·s)	0.47	0.31	66
sGaw (1/(kPa·s)	2.09		

interstitial lung disease is suggested when pulmonary function tests reveal restrictive physiology such as vital capacity (FVC), reduction in forced expiratory volume in one second (FEV1) with normal FEV1/FVC ratio [1], it was reported that SSc patients with severe restrictive changes (FVC less than 50%) had a mortality rate of 42% in 10 years [4]. Dyspnea is a late manifestation of SSc, and it was stated that reticular or ground-glass opacification on CT refers to active alveolitis [1]. The present case showed bibasilar reticulation with ground-glass changes on CT examination with FVC change of 56%, the pulmonary tests are shown in Table 1. Both SRC and TTP are rare complications of SSc [3]. When TTP combined with SSc, it resembles SRC, where both of them can present with MAHA, thrombopenia, and acute renal impairment [3]. It was reported that idiopathic TTP results from the deficiency of a circulating metalloprotease, which is also known as ADAMTS13 [5]. TTP patients also experience renal dysfunction, seropositive for ANA about 20% and thrombi and fibrin depositions can be observed in microscopic findings [2]. In the present case, the activity of ADAMTS13 was lower, ANA was highly positive, low platelet count and schistocytes in blood smear which indicated the presence of MAHA. The kidney biopsy showed acute thrombotic microangiopathy with associated occlusive fibrin thrombi and fibrinoid necrosis in few arteries and arterioles, this evidence indicates to the TTP. It is difficult to distinguish between TTP and SCR in SSc patients, but TTP was reported previously in SSc cases. One case was reported to have TTP in progressive SSc patient [2], and the other was reported in a patient with diffuse SSc [6].

Conclusion

The present case was a woman with early SSc, as early symptoms were found by clinical examination, skin manifestation, and pulmonary evidence. TTP and SRC are similar rare complications of SSc, in the present case we excluded SRC, and we identified the case with idiopathic TTP as there was lower in ADAMTS13

activity, ANA was highly positive, low and MAHA was evidenced by presence of schistocytes, also thrombi and fibrin depositions were observed in kidney biopsy. TTP is an indication for early SSc and identifying TTP from SRC requires accurate investigations.

Acknowledgments

None

List of Abbreviation

ALT	Alanine aminotransferase
APTT	Activated partial thromboplastin time
BUN	Blood urea nitrogen
CT	Computed tomography
ERV	Expiratory reserve volume
ESR	Erythrocyte sedimentation rate
FEF	Forced expiratory flow
FVC	Forced vital capacity
IC	Inspiratory capacity
JVP	Jugular venous pressure
LOC	Loss of consciousness
LV	Left ventricular
MAHA	Microangiopathic hemolytic anemia
NSIP	Non-specific interstitial pneumonia
PND	Paroxysmal nocturnal dyspnea
PUD	Peptic ulcer disease
RV	Residual volume
SRC	Scleroderma renal crisis
SSc	Systemic Sclerosis
SVC	Static lung compliance
TLC	Total lung capacity
TTP	Thrombotic thrombocytopenic purpura

Funding

None

Conflict of Interests

None

Ethical Approval

Ethical approval is not required at our institution to publish an anonymous case report in a medical journal.

Consent for publication

Informed consent was taken from the patient to publish this case in a medical journal.

Author details

Musab Abdullah Bin Shuayl[1]
1. Department of Internal Medicine, College of Medicine, Prince Sattam Bin Abdulaziz University, Al-Kharj, Saudi Arabia

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