

Primary Sjögren syndrome that initially presented with repeated hypergammaglobulinemic purpura after prolonged sitting

A case report

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Abstract

Rationale: Purpura is a common dermatologic manifestation in Sjögren syndrome (SS). When a patient presents with sicca symptoms, the diagnosis of SS is not difficult.

Patient concerns: Here, we reported a case of a 52-year-old Chinese woman who initially presented with nonpalpable purpura on both lower extremities, and these lesions had developed soon after prolonged sitting. In the past 2 years, she had repeated cutaneous nonpalpable purpura 4 times. She had no sicca symptoms, dry eyes, or dry mouth.

Diagnoses: Combining the laboratory findings, Schirmer test, and labial gland biopsy, primary SS was confirmed.

Interventions: The patient was placed on a trial of hydroxychloroquine (200 mg once daily).

Outcomes: The purpura on both lower extremities had faded at the sixth day after onset and at the third day after hydroxychloroquine treatment.

Lessons: These case was not easy to diagnosis primary SS because she had no sicca symptoms. A patient with primary SS who initially presented with recurrent purpura associated with prolonged sitting. Prolonged sitting had been a possible aggravating factor for the cutaneous purpura of this patient with primary SS.

Abbreviations: CT = computed tomography, HGP = hypergammaglobulinemic purpura, SS = Sjögren syndrome.

Keywords: primary Sjögren syndrome, prolonged sitting, purpura

1. Introduction

Sjögren syndrome (SS) is a chronic, systemic, autoimmune disease that mainly affects the extraglandular anatomy, such as salivary and lacrimal glands. SS can be isolated (primary SS [pSS]) or associated with other autoimmune diseases such as rheumatoid arthritis, systemic lupus erythematosus, and systemic sclerosis (secondary SS). pSS has wide clinical manifestations. Cutaneous manifestations of SS have been thoroughly reviewed.^[1] Patients with SS usually report persistent sicca symptoms of dry eyes (xerophthalmia) and dry mouth (xerostomia), and other symptoms, including diverse general symptoms and cutaneous

symptoms, such as purpura.^[2] Here, we report a case of a 52-year-old woman who had no obvious symptoms of dry eyes and dry mouth, and who initially presented with recurrently nonpalpable purpura on both lower extremities, and these lesions had developed soon after prolonged sitting.

2. Case report

A 52-year-old Chinese woman was admitted to our hospital because of recurrent purpura on both lower extremities for 2 days (Fig. 1). In the past 2 years, she reported that similar lesions had developed 4 times after prolonged sitting. The longest time of sitting was 1.5 hours in an airplane, and the shortest time was 4.5 hours by high-speed rail. The purpura on both lower extremities lasted 5 to 7 days and subsided completely each time.

The rest of her medical history was not remarkable. The patient denied a history of dry eyes, dry mouth with occasional painful sores, intermittent swollen and painful lymph nodes, and perpetual nausea. She occasionally felt fatigue. She reported no history of Raynaud phenomenon, swelling of the parotid glands, swollen or joint pain, fever, headache, abdominal pain, shortness of breath, weakness, blood clots, miscarriage, or photosensitivity. On admission, her vital signs were stable, with a temperature of 36.5°C, blood pressure of 120/75 mm Hg, and pulse rate of 70/min. Except for the purpura on both lower extremities, physical examination for chest and abdomen showed no abnormalities.

Her laboratory findings at that time were significant for mild anemia, elevated antinuclear antibody, anti-SS-A/SS-B/Ro-52

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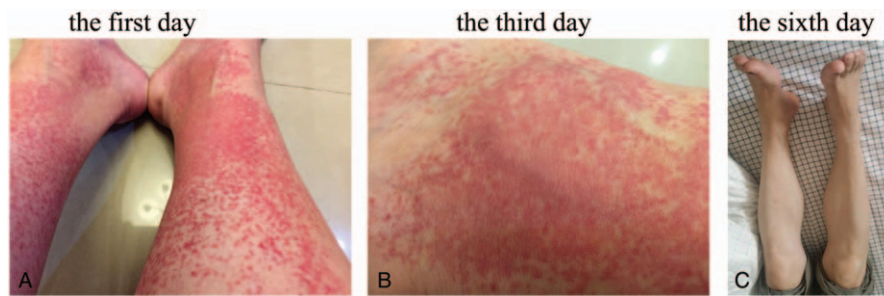


Figure 1. Pictures demonstrating purpura on both extremities. (A) demonstrated the purpura on both lower extremities on the first day, (B) demonstrated purpura on the right medial malleolus at the third day, and (C) showed the purpura subsided completely at the sixth day.

Table 1

Laboratory tests of the patient.

Laboratory tests	Results	Normal range
Urine protein	Negative	Negative
Urine latent blood test	Negative	Negative
Urea Bence-Jones protein	Negative	Negative
White blood cells count	$4.35 \times 10^9/L$	$3.5-9.5 \times 10^9/L$
Red blood cells count	$3.75 \times 10^{12}/L$	$3.8-5.1 \times 10^{12}/L$
Hemoglobin	111 g/L	115-150 g/L
Platelets count	$262 \times 10^9/L$	$125-350 \times 10^9/L$
Total bilirubin (TBil)	8.7 $\mu\text{mol/L}$	5.1-20.5 $\mu\text{mol/L}$
Direct bilirubin (DBil)	1.9 $\mu\text{mol/L}$	0-6.0 $\mu\text{mol/L}$
Alanine aminotransferase (ALT)	54 IU/L	5-40 IU/L
Aspartate aminotransferase (AST)	47 IU/L	5-40 IU/L
Total protein	88.1 g/L	60.0-83.0 g/L
Albumin	46.0 g/L	35.0-55.0 g/L
Globulin	42.1 g/L	20.0-30.0 g/L
Prothrombin time (PT)	12.2 s	11.0-15.0 s
Activated partial thromboplastin time (APTT)	33.8 s	28.0-45.0 s
Thrombin time (TT)	17.4 s	14.0-21.0 s
International normalized ratio (INR)	0.92	0.8-1.2
Serum immunoglobulin G (IgG)	34.0 g/L	7.2-16.8 g/L
Serum immunoglobulin A (IgA)	3.96 g/L	0.7-3.8 g/L
Serum immunoglobulin M (IgM)	1.81 g/L	0.6-2.8 g/L
Complement 3 (C3)	1.60 g/L	0.85-1.93 g/L
Complement 4 (C4)	0.41 g/L	0.12-0.36 g/L
Rheumatoid factor (RF)	374.0 kU/L	0-30 kU/L
Erythrocyte sedimentation rate (ESR)	85 mm/h	0-37 mm/h
Hypersensitivity C-reactive protein (hsCRP)	2.57 mg/L	0.21-0.53 mg/L
Kappa light chain	7.34 g/L	1.7-3.7 g/L
Lambda light chain	3.26 g/L	0.9-2.1 g/L
Antinuclear antibody (ANA)*	>1:320	<1:100
Anti-SS-A antibody	+++	—
Anti-SS-B antibody	+	—
Anti-Ro-52 antibody	+++	—

* Antinuclear antibody (ANA) speckled pattern.

antibody titers, and serum globulin of 42.1 g/L (20.2-30.0 g/L). Serum immunofixation electrophoresis was negative. Anti-Sm/Scl-70/PM-Scl/Jo-1 antibody and anti-CENP-B/PCNA/ds-DNA/Anu/AH/ARP/AMA-M2 antibody were negative. Detailed laboratory results are shown in Table 1.

No malignant tumor was observed by ultrasonography and computed tomography (CT) scan. Hepatic ultrasonography showed steatosis in the liver. The Schirmer test showed that the strips were wet only 2 mm in 5 minutes on each eye. Tear break-up time was 2 seconds in each eye.

The patient underwent a minor labial gland biopsy under local anesthesia. Histopathologic examination revealed partial atrophy of the salivary glands and hyperplasia of interstitial fibrous tissue (Fig. 2A, B). There were infiltrations of many lymphocytes and plasmacytes, and no malignant changes were observed (Fig. 2C, D).

Based on these findings, the diagnosis of pSS was confirmed. The patient was placed on a trial of hydroxychloroquine (200 mg once daily), and the purpura on both lower extremities had faded at the sixth day after onset and at the third day after hydroxychloroquine treatment.

3. Discussion

Purpura is a common dermatologic manifestation of SS,^[3] and the most common palpable and nonpalpable purpura, which is associated with pSS, occurs on the lower extremities.^[4] This patient's clinical manifestation is nonpalpable purpura on both lower extremities. The patient had no obvious symptoms of dry eyes and dry mouth, but the diagnosis of pSS was confirmed based on the positive findings, Schirmer test, anti-SS-A/SS-B antibody, and biopsy.

In Chinese patients with pSS, the ratio of purpura at initial presentation is 7.7%.^[5] When this patient was confirmed as having pSS, she had presented obvious sicca symptoms, such as

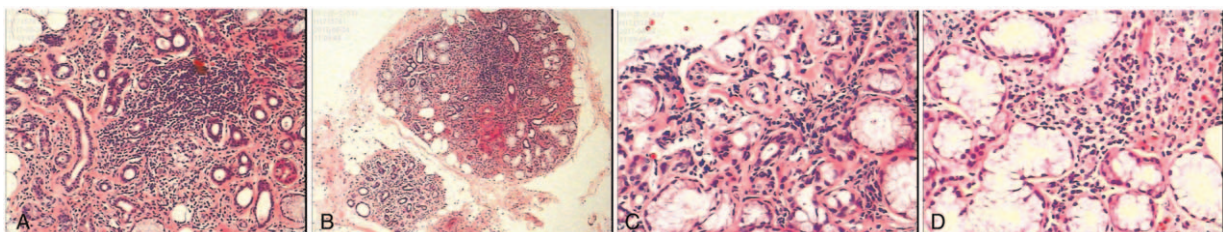


Figure 2. Right lower labial gland biopsy. Histopathologic examination revealed a partial atrophy of the salivary glands (A) and hyperplasia of interstitial fibrous tissue (B). There were infiltrations of many lymphocytes and plasmacytes (C, D), and no malignant changes were observed (hematoxylin and eosin, $\times 100$).

dry eyes and dry mouth. Hypergammaglobulinemic purpura (HGP) is associated with SS.^[6] Laboratory findings indicated that the patient had hypergammaglobulinemia. So, the clinical manifestation and abnormal laboratory findings indicated that this patient's purpura was caused by hypergammaglobulinemia. In addition, abnormal liver function was also found in pSS.^[5] However, because the patient's hepatic ultrasonography showed steatosis in the liver, we could not conclude that the patient's abnormal liver function was caused only by pSS.

Our patient had a peculiar history of repeated presentation of the nonpalpable purpura in association with prolonged sitting. Prolonged standing can induce purpura, and the author speculated that prolonged standing increased the capillary pressure and caused purpura.^[7] In our case, prolonged sitting seemed to trigger purpura.

4. Conclusions

Thus, for patients who have only palpable and nonpalpable purpura, early diagnosis of pSS is not easy. Moreover, pSS has variable clinical manifestations, which partially explains delays in diagnosis of up to 10 years from the onset of symptoms.^[8] Therefore, it is an interesting case of pSS with HGP. This case may suggest that prolonged sitting could have potentially

triggered the recurrent purpura in patients who suffered from SS with HGP. Further, it reminds the patient with pSS to engage in intermittent activity during prolonged sitting.

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