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## Iguratimod in treatment of primary Sjögren's syndrome concomitant with autoimmune hemolytic anemia: A case report

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### Abstract

#### BACKGROUND

Primary Sjögren's syndrome (pSS) concomitant with autoimmune hemolytic anemia (AIHA) but without eye and mouth dryness is exceedingly rare. Iguratimod (IGU) has been widely used in the treatment of pSS. However, there are few reports about the application of IGU in pSS concomitant with AIHA.

#### CASE SUMMARY

Here, we present the case of a patient with pSS concomitant with AIHA but without eye and mouth dryness. The patient was initially diagnosed with hyperplastic anemia and AIHA while pSS was missed, and was finally diagnosed with pSS concomitant with AIHA. The patient was treated with IGU along with prednisone and hydroxychloroquine, and her hemoglobin, reticulocytes and IgG returned to normal levels.

#### CONCLUSION

IGU was effective for and well tolerated by our patient with pSS concomitant with AIHA, and may be a promising therapy for the treatment of this disease.

**Key Words:** Autoimmune hemolytic anemia; Iguratimod; Primary Sjögren's syndrome; Case report

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**Core Tip:** Primary Sjögren's syndrome (pSS) concomitant with autoimmune hemolytic anemia (AIHA) but without eye and mouth dryness is exceedingly rare. Iguratimod

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(IGU) has been widely used in the treatment of pSS. However, there are few reports about the application of IGU in pSS concomitant with AIHA. Here, we present the case of a patient with pSS concomitant with AIHA who was successfully treated with IGU. IGU was effective for and well tolerated by our patient with pSS concomitant with AIHA, and may be a promising therapy for treatment of this disease.

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## INTRODUCTION

Primary Sjögren's syndrome (pSS) is one of the most common autoimmune diseases, and is characterized by salivary and lacrimal gland dysfunction and lymphocytic infiltration[1,2]. Autoimmune hemolytic anemia (AIHA), a rare autoimmune disease, is characterized by hemolysis mediated by autoantibodies directed against red blood cells (RBCs)[3]. The most common manifestation of hematologic involvement in pSS is leukopenia, while pSS concomitant with AIHA is rare[4]. Currently, there is no standardized treatment regimen for AIHA, although the First International Consensus Group recommended second-line treatment for AIHA in 2017, including azathioprine, mycophenolate or ciclosporin[5]. Igruratimod (IGU), a novel modified anti-rheumatic drug, has shown both good efficacy and safety in the treatment of pSS[6,7]. However, whether IGU can be used in the treatment of pSS concomitant with AIHA remains to be further elucidated. Here, we present the case of a patient with pSS concomitant with AIHA who was successfully treated with IGU.

## CASE PRESENTATION

### Chief complaints

In 2015, a 31-year-old Chinese female presented with dizziness and fatigue without obvious inducement, and received Chinese medicine treatment (the specific medication was unknown) at a local Chinese medicine clinic. However, her symptoms did not remit, and she was admitted to the emergency department of Lanzhou Military Region General Hospital on November 23, 2015.

### History of present illness

There was no history of present illness.

### History of past illness

There was no history of past illness.

### Personal and family history

There was no personal and family history.

### Laboratory examinations

Laboratory tests results revealed abnormal decreases in hemoglobin (Hb, 46 g/L; reference range, 120-160 g/L) and RBCs ( $0.91 \times 10^{12}$  cells/L, reference range,  $3.5-6.0 \times 10^{12}$  cells/L), and abnormal increases in the erythrocyte sedimentation rate (ESR: 78 mm/h; reference range, 0-20 mm/h) and hs-CRP (1.64 mg/L; reference range, 0-0.5 mg/L). Biochemical examination results revealed 41.4 g/L IgG (reference range, 8-16 g/L), 4.19 g/L IgA (reference range, 0.7-3.3 g/L), 0.63 g/L IgM (reference range, 0.5-2 g/L), 0.37 g/L complement C3 (reference range, 0.9-1.8 g/L), and 0.1 g/L complement C4 (reference range: 0.1-0.4 g/L). Urine Bence-Jones protein electrophoresis was negative. Bone marrow aspiration results indicated hyperplastic anemia. She was treated with ferrous sulfate oral solution, folic acid tablets and vitamin B12 (the specific dose was unknown).

As the symptoms did not improve, she was admitted to the hematology department of Lanzhou Military Region General Hospital on November 27, 2015. In addition to abnormal Hb, ESR, hs-CRP, IgA, IgG and complement C3, laboratory test results also revealed abnormal increases in rheumatoid factor (RF: 399 IU/mL; reference range: 0-80 IU/mL), total bilirubin (32.0  $\mu$ mol/L; reference range: 1.5-20  $\mu$ mol/L) and indirect bilirubin (22.5  $\mu$ mol/L; reference range: 1-20  $\mu$ mol/L). The patient was positive for antinuclear (ANA; titer > 1:320; speckled pattern), anti-Sjögren's syndrome A (SSA) and anti-SSB antibodies, and negative for serum immunofixation electrophoresis. The direct Coombs test results were strongly positive (anti-IgG and anti-C3d). Bone marrow aspiration results still indicated hyperplastic anemia. She was diagnosed with AIHA and connective tissue disease, and was treated with oral prednisolone (1 mg/kg/d), and leflunomide (20 mg/d) and a washed red frozen blood cell transfusion. Thereafter, her condition improved, and she was discharged 10 d later. However, due to the abnormal decrease in Hb when the dose of prednisone was reduced to less than 20 mg/d, she was admitted to the hematology department again on September 17, 2018. The symptoms improved after treatment with prednisone (40 mg/d), hydroxychloroquine (HCQ; 0.2 g twice a day) and a washed frozen red blood cell transfusion.

Considering the repeated recurrence of the above symptoms and the abnormal increase of RF in the rare case, the patient was referred to the rheumatology department of Lanzhou University Second Hospital on September 27, 2018. In addition to being positive for ANA, anti-SSA and anti-SSB antibodies, her abnormal examination results also included 105 g/L Hb, 4.9% reticulocytes (RET; reference range: 0-1.5%), 46.71 g/L IgG, 4.15 g/L IgA, 82 Ru/mL RF-IgA (reference range: 0-20 Ru/mL) and > 200 Ru/mL RF-IgM (reference range: 0-20 Ru/mL). The Schirmer test result was positive, and minor salivary gland biopsy revealed focal lymphocytic infiltration of the exocrine glands.

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## FINAL DIAGNOSIS

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The patient was diagnosed with pSS concomitant with AIHA.

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## TREATMENT

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Immunosuppressants such as cyclosporin A or mycophenolate were rejected because the patient could not afford these drugs. She was initially treated with IGU (25 mg twice a day) in addition to the combination therapy of prednisone (40 mg/d) and HCQ (0.2 g twice a day).

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## OUTCOME AND FOLLOW-UP

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The patient was followed up every 1-2 mo, and the dose of prednisolone was gradually tapered (10% every 2 wk until reaching 5 mg/d for maintenance). During the 24 mo of follow-up, her Hb, RET and IgG returned to normal levels (Table 1).

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## DISCUSSION

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Here, we present a rare case of pSS concomitant with AIHA but without eye and mouth dryness that was successfully treated with IGU. To our knowledge, this is the first case report describing the efficacy of IGU in the treatment of pSS concomitant with AIHA, and the findings from this case report indicate that IGU might broaden the treatment options available for patients with pSS concomitant with other rare diseases.

AIHA is a rare but clinically significant complication of pSS, only 2.8% of patients with pSS were explicitly diagnosed with AIHA in a large cross-sectional study[4]. In certain patients, the typical symptoms (sicca symptoms) of pSS do not appear, and the symptoms of AIHA develop before the diagnosis of pSS, which might result in a delay in diagnosis[8]. In this rare case in which the patient did not experience eye and mouth dryness, the initial diagnosis was hyperplastic anemia and AIHA while pSS was missed; however, the patient was finally diagnosed with pSS concomitant with AIHA

**Table 1** The changes in hemoglobin, reticulocytes and IgG during the 24 mo of follow-up

Indicator	Baseline	M1	M3	M5	M12	M18	M24
Hb (g/L)	105	132	155	150	139	148	134
RET (%)	4.9	1.6	1.2	1.0	1.2	0.9	0.9
IgG (g/L)	46.71	23.47	15.33	14.2	13.22	15.53	12.29

Hb: Hemoglobin; RET: Reticulocytes; M: Month.

based on abnormally elevated RF. In addition, a previous report revealed that ANA, anti-SSA, anti-SSB antibody positivity and lower complement levels were common in pSS concomitant with AIHA[9], which was consistent with our case. Taken together, these findings indicate that more comprehensive clinical examination and evaluation should be carried out to improve the diagnostic accuracy of pSS concomitant with AIHA.

Currently, there is no standardized treatment regimen for the treatment of pSS concomitant with AIHA. IGU is a novel anti-rheumatic drug approved only in China and Japan[6,7]. Evidence has shown that IGU can be considered as an effective and safe drug for the clinical therapy of pSS[7]. However, whether IGU can be used for the treatment of pSS concomitant with AIHA remains unknown. In our case, due to the cost of treatment and preventable adverse reactions, the patient received IGU as a second-line treatment under background treatment with a glucocorticoid combined with HCQ. The results showed that the patient responded well to IGU, and her Hb, RET and IgG returned to normal levels during the 24 mo of follow-up. The following mechanisms might explain the clinical efficacy of IGU in the treatment of pSS concomitant with AIHA. AIHA is a rare autoimmune disease in which autoantibodies directed toward RBC antigens lead to RBC accelerated destruction[3]. Several immunologic mechanisms are involved in the pathogenesis of AIHA, including autoantibodies, antibody-dependent cell-mediated cytotoxicity, phagocytes, B and T lymphocytes, Tregs, cytokines, and the complement system[10]. IGU is an anti-inflammatory and immunomodulatory compound[11]. IGU can significantly inhibit the production of inflammatory cytokines (such as interleukin-6, interleukin-8, and tumor necrosis factor- $\alpha$ ) in animal models of arthritis or autoimmune diseases[12]. In addition, IGU plays a significant immunomodulatory role in the synovial tissue of patients with rheumatoid arthritis by regulating T and B lymphocyte subsets and inhibiting the production of cytokines and immunoglobulins[6,13]. Therefore, the improvement in symptoms of patient with pSS concomitant with AIHA might result from the immunomodulation of B lymphocytes. However, the mechanism underlying the clinical efficacy of IGU in the treatment of pSS concomitant with AIHA needs to be determined in further investigations. The present study revealed that IGU was effective for and well tolerated by our patient with pSS concomitant with AIHA.

Clinicians should be reminded that when hemolytic anemia occurs in young women, they should be alert to the possibility of autoimmune diseases. More comprehensive clinical examination and evaluation, including autoantibodies, immunoglobulins, and complement levels, should be carried out to improve the diagnostic accuracy. The treatment of AIHA should take into account the primary disease, and the dose of glucocorticoids should be gradually tapered. For patients with poor glucocorticoid responses, immunosuppressants should be added as soon as possible.

There are some limitations in this case report that should be kept in mind. First, the classification of AIHA was not performed in our patient due to hospital condition limitations. Second, the proportion of B lymphocytes was not dynamically monitored during the treatment. Whether B lymphocytes are involved in the possible therapeutic mechanism of IGU in pSS concomitant with AIHA is not clear. Finally, this case report involved experience with a single patient. Prospective studies with a large sample size are needed to provide more information about the safety and efficacy of IGU in patients with pSS concomitant with AIHA.

## CONCLUSION

The findings from our case report showed that AIHA may occur in pSS, and the diagnosis of pSS should be considered even in patients with a diagnosis of AIHA. For

our patient with pSS concomitant with AIHA, IGU was effective and well tolerated. The findings from this case report also indicate that IGU might broaden the treatment options available for patients with pSS concomitant with other rare diseases.

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