


## CASE REPORT

# First report of rheumatoid arthritis and secondary Sjögren's syndrome complicated with heart failure

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**Funding information**

There is no funding support for this study

**Abstract**

A 70-year-old woman with rheumatoid arthritis for 45 years developed secondary Sjögren's syndrome. She had a long-term low-salt and low-fat diet and did not adhere to long-term hormone and rheumatic immunotherapy, which led to heart failure.

**KEYWORDS**

heart failure, rheumatoid arthritis, secondary Sjögren's syndrome

## 1 | INTRODUCTION

This is the first report about heart failure caused by secondary Sjögren's syndrome (SS) in a 70-year-old female patient with rheumatoid arthritis (RA). She was not treated with long-term hormone and rheumatic immunotherapy after diagnosed with RA and SS at the age of 50. She had hyponatremia and chlor-emia due to low-salt and low-fat diet, and lacked outdoor exercise. At the age of 67, she had intermittent chest tightness with shortness of breath with persistent asthma and edema of both lower limbs. She could not lie on her back at night for nearly 2 months at the age of 70 and died of heart failure.

Rheumatoid arthritis (RA) is a common systemic auto-immune disease.<sup>1</sup> Sjögren's syndrome (SS) is also a chronic

autoimmune disease that mainly involves exocrine glands, such as lacrimal and salivary gland. SS can be divided into two types: primary and secondary. The occurrence of the latter is closely related to RA, another autoimmune disease.<sup>2</sup> This type of RA secondary to SS is more common and manifests as a complex overlapping manifestation of symptoms.<sup>3</sup> Clinically, a combination of multiple drugs is used for treatment.<sup>4</sup> SS can accumulate multiple systemic damage throughout the body, but cardiovascular involvement is rare. However, SS with heart failure as the main manifestation is even rarer. But so far, there is no report about the eventual heart failure in patient with RA secondary to SS without long-term use of hormones and rheumatic immunotherapy.

Zhang and Kong contributed equally to this work.

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## 2 | CASE REPORT

A 70-year-old female patient began to suffer from multiple joint swelling and pain from the age of 30 and was diagnosed with RA. In 1998 (49-year-old), she developed dry mouth and needed water to take dry food without oral ulcer or dry eye. In 2000, she showed the swelling and pain of right parotid gland with rampant teeth (Figure 1), the left parotid gland was swollen and painful, and the left parotid gland was gradually subsided, but the right side was aggravated (Figure 2).

In April 2013, she showed the swelling and pain of right parotid gland with rampant teeth (Figure 1), the left parotid gland was swollen and painful, and the left parotid gland was gradually subsided, but the right side was aggravated (Figure 2). On physical examination, she had a temperature (T) of 36°C, pulse (P) 78 beats/min, respiratory (R) 18 beats/min, blood pressure (BP) 120/70 mmHg. Laboratory evaluation showed erythrocyte sedimentation rate 58.00 mm/h (normal 0–20), IgM 3.74 g/L (0.63–2.77), positive antinuclear antibodies 1:1000 (+) centromere pattern B (+). No other laboratory abnormalities were observed. Parotid ultrasound showed bilateral parotid gland enlargement and echo changes. Cysts were found in bilateral parotid glands, and multiple anechoic areas were found in the right parotid gland, with a maximum of 1.9 × 1.2 cm. In the left parotid gland, an anechoic area was showed with the size of 0.4 × 0.4 cm. The labial gland biopsy showed 4 lymphocytic infiltration foci in two pieces of small salivary gland tissue (about 8 mm<sup>2</sup>), which was consistent with the pathological changes of salivary gland in SS. Chest computed tomography (CT) demonstrated interstitial changes in both lungs, small nodules in the right upper lobe (a high density of 7 × 6 mm), revealed pneumonia in the right middle lobe, and a small amount of pleural thickening on both sides, which were related to the primary disease. No fever, cough, expectoration, chest tightness, and



**FIGURE 1** The Rampant teeth of the patient. (Please consider for our online covers)

other symptoms were observed. The treatments for SS are oral tanshinone for improving circulation, leflunomide, hydroxychloroquine, and total glucosides of paeony for regulating immunotherapy. She was discharged from the hospital 9 days later due to changes in symptoms.

She took these medicines at home for 3 months and re-examined once a month. Because white blood cell dropped obviously, she stopped the medicines and began to use traditional Chinese medicine. From May 2017, she had intermittent chest tightness with shortness of breath, no chest pain, syncope, hemoptysis, blackness, hematemesis, and melena. From June 2019, she had chest tightness, shortness of breath gradually aggravated, and could not lie on her back at night for nearly 2 months, with persistent asthma and edema of both lower limbs. She slept in a sitting position during the day and often fell from the chair involuntarily.

On February 25, 2020, she had more severe chest tightness, shortness of breath, significantly reduced activity tolerance. She was unable to lie on her back at night and had to sit on her back, significantly showed edema of both lower limbs. She was diagnosed as “cardiac insufficiency” in department of emergency. She appeared dispirited spirit, thick breath sounds of both lungs, dry and wet rales, edema of lower limbs, and normal muscle strength of limbs. Electrocardiogram (ECG) examination demonstrated old myocardial infarction. Laboratory evaluation showed D-dimer (gold standard) 0.64 mg/L (0.00–0.50), sodium (Na) 109 mmol/L (135–145), chlorine (Cl) 74 mmol/L (98–107), creatinine 37 μmol/L (46–110), aspartate aminotransferase 57 U/L (15–46), r-Glutamyltransferase 76 U/L (12–58), alkaline phosphatase 180 U/L (38–126), glucose 10.70 mmol/L (3.60–6.10), creatine kinase 523 U/L (30–135), creatine kinase isoenzyme 67.0 U/L (0.0–16.0); N-terminal pro brain natriuretic peptide (NT proBNP) 4400 pg/ml (< 125). Blood gas analysis showed lactic acid 3.2 mmol/L (1.0–1.8), arterial partial pressure of oxygen 146.9 mmHg (80.0–100.0), arterial oxygen saturation 98.7%, actual carbonate concentration 19.2 mmol/L (22.0–27.0), total carbon dioxide 20.2 mmol/L (24.0–32.0). Blood routine examination showed white blood cell count  $9.77 \times 10^9/L$  (3.50–9.50), neutrophils (%) 92.4 (40.0–75.0), lymphocyte (%) 3.4 (20.0–50.0), eosinophil (%) 0.1 (0.4–8.0), absolute value of neutrophil  $9.03 \times 10^9/L$  (1.80–6.30), absolute value of eosinophil  $0.01 \times 10^9/L$  (0.02–0.52), absolute value of lymphocyte  $0.33 \times 10^9/L$  (1.10–3.20), hemoglobin: 107 g/L (115–150), hematocrit 31.8% (35.0–45.0). Bilateral lung CT showed pulmonary edema was the first consideration for changes in both lungs; multiple patchy shadows in both lungs and changes near the right lung hilum were not excluded, and appeared bronchiectasis of right middle lobe and left lingual segment, a small amount of pleural effusion on both sides, mediastinal lymph node enlargement, and calcification of aorta and coronary artery.



**FIGURE 2** The swollen parotid gland (right) of the patient

Admission diagnosis results included (1) cardiac insufficiency, cardiac function grade IV; (2) pneumonia; (3) ion disorder, hyponatremia, and hypochloremia; (4) Sjogren's syndrome. After admission, she was given vasodilator, diuretic, and anti-inflammatory treatment.

But, at night, she showed nonsense, delirious, and always grabbed things. The next morning, at 7:06, when urinating in bed, she suddenly lost consciousness, could not breathe, and had no spontaneous breathing, bilateral pupils were equal in size and equal in circle, 3.5 mm in diameter, and the light reflex and orbital baroreflex disappeared. ECG demonstrated borderline rhythm, blood pressure, and finger pulse oxygen could not be detected. She was immediately put on a supine position with the pillow removed, with the frequency of 100 times/min continuous chest cardiac compression, artificial balloon assisted breathing (10 L/min), was contacted the MICU for tracheal intubation, stopped the infusion of isosorbide, gave epinephrine 1 mg intravenously, dopamine pump pressure, and normal saline at full speed. At 7:20, ECG demonstrated sinus rhythm, heart rate (HR) 58 beats/min, BP 113/52 mmHg, finger pulse oxygen could not be detected, great artery pulse could be touched, adrenaline 1 mg was given intravenously. Tracheal intubation was smoothly performed at 7:26, spontaneous breathing was still not allowed at 7:28, great artery pulse could be touched, ECG monitored sinus rhythm, HR 140 beats/min, BP 159/95 mmHg, finger pulse oxygen 92%, and was stopped cardiac compression, was given continuous artificial balloon assisted breathing (10 L/min). At 7:42, she still did not respond to the call and had no spontaneous breathing. The big artery was palpable, the bilateral pupils were equal in size and circle, the diameter was 2 mm, the light and orbital pressure reflex disappeared. ECG demonstrated sinus rhythm, HR was 123 beats/min, BP was 151/93 mmHg, pulse oxygen was 97%. She was given continuous artificial balloon assisted breathing (10 L/min) and was transferred to ICU for further treatment.

She suffered from limb flexion after pain stimulation. In endotracheal intubation ventilator assisted ventilation, she had a T 36.6°C, P 96 beats/min, R 18 beats/min, BP 90/64 mmHg, and 85% pulse oxygen. Bilateral pupils were equal in size and circle, 4 mm in diameter. She showed slow in light response, symmetrical in breath sounds of both lungs, no dry and wet rales, soft abdomen, no muscle tension, edema of both lower limbs, and suspicious positive in bilateral bibinsky sign. She was treated with hypothermia brain protection, endotracheal intubation, mechanical ventilation, norepinephrine pumping to maintain BP, anti-inflammation, and brain cell nutrition. CT scan did not demonstrate cerebrovascular disease or pulmonary embolism. After the above treatment (24 days), her consciousness gradually improved. Norepinephrine was stopped, the ventilator condition was lowered, and the extubation was successful. After extubation, the oxygenation was good, the hyponatremia was corrected to normal. But fever was still found, positive cocci were found in blood culture, vancomycin anti-inflammatory treatment was given, and T gradually decreased. On March 22, the back waist bed sore appeared. On March 24, the obvious sputum sounds appeared in the morning. The effect of sputum suction was not good. The small respiratory tract sputum embolism was considered. Unfortunately, her family did not agreed with tracheotomy and later died of acute left heart failure and respiratory failure at 17:00.

### 3 | DISCUSSION

To our knowledge, this is the first report about heart failure caused by secondary SS in patient with RA. Specifically, this RA patient suffering from secondary SS was not treated with long-term hormone and rheumatic immunotherapy after she was diagnosed with RA and SS. More specifically, the patient only relied on low-salt and low-fat diet (hyponatremia and chloremia) and lacked outdoor exercise.

On further questioning, the patient wanted to treat the disease through self-regulation of the constitution ("Essence", "Qi" of Traditional Chinese Medicine) and reduce the side effects of drugs. Sicca symptoms are the common manifestation of secondary SS, and the key to the diagnosis of SS,<sup>5</sup> but other symptoms are quite uncommon and usually mild,<sup>6</sup> moreover, these symptoms are easy to be covered up by the symptoms of RA and ignored by some patients.<sup>7</sup> When the symptoms of SS are concerned, the course of SS may worsen.<sup>8</sup>

The patient showed dry eyes, dry mouth, and Raynaud's symptoms as the first symptoms. Physical examination showed that the knuckles of the middle and ring fingers were white, and red subcutaneous bleeding points could be seen at the fingertips. There were intermittent purpura-like rashes on both lower limbs, with the manifestation of vasculitis. In

general, patients with vasculitis do not accumulate a specific vessel alone and may show diffuse vascular damage.<sup>9</sup> So the patient was damaged in the whole blood vessels including coronary artery and had no traditional risk factors for coronary heart disease.<sup>10</sup> Therefore, heart failure may be secondary to SS. In addition, this patient had hyponatremia, which is also a risk factor for congestive heart failure.<sup>11</sup> But, for this patient, secondary SS may be the main cause of heart failure. Because the family history of this patient showed that her mother suffered from RA with low-fat and low-salt diet, without outdoor activities and using long-term hormone and rheumatic immune drug treatment. But her mother did not have secondary SS, she died of respiratory failure at 93 years of age.

## 4 | CONCLUSION

We reported a case showing the importance of early diagnosis and long-term immunosuppressive therapy for patients with RA and secondary SS. Because this RA patient with secondary SS did not use long-term hormone and rheumatic immune drugs for treatment. On the other hand, she had a long-term low-salt, low-fat diet, and lacked outdoor exercise, ultimately led to heart failure.

## ACKNOWLEDGEMENTS

We thank the patient's family for their collaboration.

## CONFLICT OF INTEREST

None declared.

## AUTHOR CONTRIBUTIONS

Study concept and design: FY and SH. Acquisition of data: HZ, FX K, FY, and SH. All authors read and approved the final manuscript.

## ETHICAL APPROVAL

The project was approved by the Medical Ethics Committee of Dalian Central Hospital. Written informed consent was obtained from the son of patient for publication of this Case Report and any accompanying images.

## DATA AVAILABILITY STATEMENT

Data available on request from the corresponding author. All data related to this case report are contained within the manuscript.

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**How to cite this article:** Zhang H, Kong F, Yu F, Hao S. First report of rheumatoid arthritis and secondary Sjögren's syndrome complicated with heart failure. *Clin Case Rep*. 2021;9:e04581. <https://doi.org/10.1002/ccr3.4581>