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Review Article

A Case of Gougerot Sjögren Syndrome (SGS) Discovered by Chronic Pulmonary Impairment at the Nianankoro Fomba Hospital in Segou

Camara, B. D[1*], Dramé Boubacar, M[11], Coulibaly, O[10], Dao, K[2], Coulibaly, A[5], Sylla Mala[10], Keïta Kaly[9], Sy Djibril[9], A. Koné[3], Drago, A. A[4], Dollo, I[5], Kamissoko, C. O[8], Maiga, A[6], Diakité, M[7], Guindo, H[5], D. S. Sow[3]

[1]Department of Internal Medicine Nianankoro Fomba Hospital in Ségou, Mali
[2]Department of Internal Medicine, CHU Gabriel Touré, Bamako, Mali
[3]Medicine and Endocrinology Department of Mali Hospital, Bamako, Mali
[4]Endocrinology and Diabetology Department of the Reference Health Center of Commune I of the Bamako District, Mali
[5]Gao Regional Hospital, Mali
[6]Hepato-Gastroenterology Department of Point-G Hospital, Bamako, Mali
[7]Bocar Sidy Sall University Hospital Center in Kati, Bamako, Mali
[8]Sélingué Reference Health Center, Mali
[9]Department of Internal Medicine, CHU Point G, Bamako, Mali
[10]Bougouni reference health center
[11]Odonto-Stomatology and Maxillofacial Surgery Department at the NF Hospital Segou.

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***Corresponding Author:** Dr Camara Boua Daoud
Internist Research Fellow, Nianankoro Fomba Hospital in Ségou, Bamako, Mali

Abstract

Introduction: Sjögren's syndrome (SS) is a systemic autoimmune disease whose target is the epithelium of the exocrine glands and in particular the salivary glands. SS affects women more often with a sex ratio of 9 women to 1 man and the peak frequency is around age 50. It is mainly described in Western literature where it seems to come second after RA. In Africa, it is mainly reported in the Maghreb. We report a case, revealed by pulmonary damage at the Nianankoro Fomba hospital in Ségou. ***Observation*:** This was a patient, aged 50, without any particular ATCD, who had consulted for chronic cough with mucco-purulent sputum and dyspnea, which had been present for more than four (4) months. Faced with the failure of a trial anti-tuberculosis treatment despite the negativity of the sputum testing for BARR in the Pneumophthisiology department, she decided to consult internal medicine. Clinical examination revealed NYHA stage III dyspnea. A chest CT concluded with bilateral pulmonary parenchymal interstitial syndrome associated with inflammatory bronchopathy with bronchiectasis. The immunological assessment revealed positive anti–SSA and anti–SSB Abs. Clinical improvement was obtained after initiation of corticosteroids and immunosuppressants. ***Conclusion*:** This observation illustrates the interest of looking for latent SS in the etiological assessment of a persistent cough.

Keywords: Sjögren's syndrome, salivary glands, Maghreb, pulmonary damage, internal medicine.

INTRODUCTION

Sjögren's syndrome (SS) is a systemic autoimmune disease whose target is the epithelium of the exocrine glands and in particular the salivary glands. This pathology is therefore called autoimmune epithelitis [1].

SS manifests as a symptomatic triad associating dryness, pain and fatigue and can be complicated, in 30 to 50% of patients, by systemic complications [2].

The disease can be primary or associated with other systemic diseases (rheumatoid arthritis (RA), systemic lupus erythematosus, inflammatory myopathies or scleroderma). The associated forms appear to be as severe as the primitive forms [3].

It has been described that patients with RA and secondary SS presented more severe forms of RA with more frequent systemic involvement [4, 5].

SS affects women more often with a sex ratio of 9 women to 1 man; the peak frequency is around age 50.

The American-European AECG (American European Consensus Group) diagnostic classification criteria dating from 2002 have recently been replaced by a new series of criteria according to a consensus of the European (EULAR) and American (ACR) rheumatology societies [6].

Inclusion Criteria

- At least one symptom of dry eye or mouth (according to AECG 2002 criteria) or suspicion of SS based on an ESSDAI activity score / = 0 Items
- Focus score ≥ 1: 3 points Anti-SSA/Ro+: 3 points Ocular staining score ≥ 5: 1 point Schirmer ≤ 5 mm/5 min: 1 point Unstimulated salivary flow ≤ 0.1 mL/min: 1 point →Diagnosis of Primary SS if score ≥ 4 points

NB: exclusion criteria those of the AECG 2002 criteria with 3 modifications:

Added Hyper IgG4 syndrome;
Disappearance of a pre-existing lymphoma;
Restriction of hepatitis C to active hepatitis C (i.e. with positive PCR).

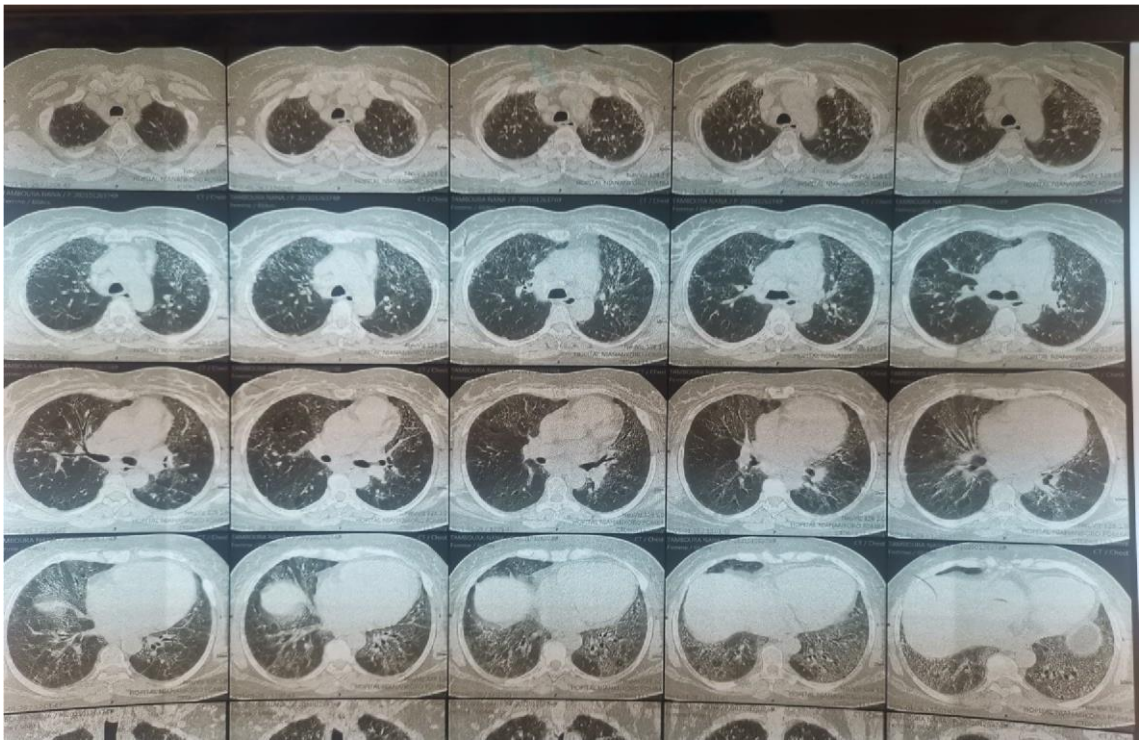
CETA: American European Consensus Group;
SS: Sjögren's syndrome;
ESSDAI: EULAR Sjogren's Syndrome Disease activity Index;
PCR: Polymerase Chain Reaction.

Lung involvement is common, affecting between 9 and 75% of patients, especially in cases of positive anti-SSA antibodies, but it is rarely severe [7].

Clinical Case

It was Ms. NT, aged 57, who consulted for a chronic cough with purulent mucous sputum that had been present for more than 4 months associated with increasingly severe dyspnea preventing any physical effort.

She first consulted the pneumophthisiology department where sputum samples looking for BK or other germs came back negative.



Figure

Chest CT revealed bilateral pulmonary parenchymal interstitial syndrome associated with inflammatory bronchopathy with bronchiectasis (See Figure above).

A trial anti-tuberculosis treatment for a period of six months was instituted without success. Given the persistence of the signs, the parents decided to consult the internal medicine department.

On clinical examination: the patient had an altered general condition, with NYHA 3 dyspnea, she reported the notion of polyarthralgia with an inflammatory appearance that she had been carrying for at least two years.

Pulmonary examination showed intercostal drawing and diffuse crackling rales. The osteoarticular examination revealed pain when mobilizing the large

joints without swelling or joint deformation. The remainder of the examination was unremarkable.

Paraclinical examinations:

CBC: normal, CRP increased to 41 mg/l, blood sugar, serum creatinine, transaminases, blood ionogram all unremarkable.

The immunological assessment shows a high anti-SSA Ab level at 199 AU/ml, an anti-anti-SSB Ab level at 400 AU/ml, anti-U1 RNP Ab negative, Anti-Sm, Scl70 negative. The diagnosis of primary sicca syndrome was made.

A treatment based on Methotrexate 10 mg/week, folic acid 5 mg/day (6 days a week), cortancyl 5 mg/day allowed a clear improvement in clinical signs with the disappearance of cough, dyspnea and polyarthralgia at the end. of 4 months of treatment which allowed the patient to have a better quality of life and to resume her daily activities.

DISCUSSION

SS affects women more often with a sex ratio of 9 women to 1 man; the peak frequency is around the age of 50 [4, 5]. The S. El Kettani S study [8] in Morocco included 26 patients including 24 women and 2 men with a sex ratio of 12. Our only case was a woman. Also in this Moroccan study [8], the average age was 50.0 ± 9.3 years with extremes of 36 to 70 years. Our patient was 57 years old, which is similar to the data in the literature and that of the Moroccan study.

The main attacks encountered during SS are ILD (diffuse infiltrative pneumonia), bronchiolitis, pulmonary cysts, pulmonary lymphomas, pulmonary amyloidosis and pulmonary hypertension.

Interstitial damage is seen in 3 to 11% of SS cases (5). The age at diagnosis of PID is around 55-60 years (range: 14-78 years) [6, 9]. This is comparable to the age of our patient who was 57 years old. Lung involvement usually sets in in the years following sicca syndrome.

Cough is a common manifestation during SS, which can affect 10 to 50% of patients [10, 11, 13, 14]. This is a dry cough, which can precede the diagnosis of SS by several years as was the case in our patient associated with bronchial hyperreactivity.

Recurrent respiratory infections constitute a frequent complication of SS, with 10% to 35% of patients with SS presenting recurrent bronchitis or pneumonia [12, 14, 15].

The systematic performance of chest scans confirms the high frequency of bronchial abnormalities during SS. The systematic study of patients, without prior selection based on respiratory warning signs, shows thickening of the bronchial walls in 8% to 22% of patients, dilation of the bronchi in 6% to 38%, and bronchiolar nodules in 6. % to 23% of patients [13, 16, 17].

Dilatation of the bronchi is frequently detected on the scanner during SGS. The frequency of symptomatic bronchial dilatation is poorly known and seems less common than in rheumatoid arthritis.

Thus, in the English series of 150 patients suffering from bronchial dilation described by Pasteur *et al*., [18], there were no cases of SS while there were three cases of rheumatoid arthritis.

These extra glandular manifestations were mainly represented in the Moroccan study [8] by arthralgia, myalgia and bronchopulmonary damage with respectively 91.7%, 15.3% and 11.5% of cases. In our patient, the main reason for consultation was persistent cough.

A biological inflammatory syndrome was noted with an ESR of 32.7 ± 17.5 and extremes ranging from 3 to 107 in the Moroccan series [8], but in our patient, it was rather the CRP which was at 41 mg/ L.

In the Moroccan series [8], 70% of patients had positive anti-SSA Ab, while 44% had positive anti-SSB. In our patient, both Abs (anti-SSA, anti-SSB) were positive.

The diagnosis of primary sicca syndrome was retained in our case, unlike the Moroccan case [8], where SS was primary in 61.5% of cases and secondary in 35.2% of cases. These associated pathologies consisted mainly of systemic acute lupus erythematosus (SLE), rheumatoid arthritis (RA) and scleroderma.

U, treatment based on an immunosuppressant (Methotrexate) and corticosteroid therapy was started, which allowed for a favorable outcome. In the Moroccan case [8], it consisted of Chloroquine (Nivaquine), hydroxychloroquine (Plaquénil), corticosteroid therapy and finally Methotrexate in one case.

CONCLUSION

The respiratory manifestations of SS are particularly common and put the pulmonologist in a position to make the diagnosis of the disease in very varied contexts: diagnosis of chronic cough, assessment of diffuse infiltrative pneumonia, recurrent respiratory infections. Given the low prevalence of this pathology in our country, the diagnosis can often escape our pulmonologist colleagues.

The ACR/EULAR 2016 diagnostic classification criteria for primary SS associated with the search for anti-SSA and anti-SSB autoantibodies, and the

performance of a biopsy of the accessory salivary glands can confirm the diagnosis of this pathology.

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