



Case Report

Sjögren's syndrome with bipolar disorder, case report

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ABSTRACT

Introduction: Sjögren's syndrome is an autoimmune disease affecting exocrine glands and other organs. At the onset of the disease and during its course, it most commonly manifests as neuropsychiatric symptoms, most frequently, depression and anxiety. However, the association with bipolar disorder seems rare and not frequently documented.

Case report: Our case report presents a 43-year-old male complaining from neuropsychiatric symptoms including but not limited to periods of elevated mood, increased energy, headaches. Examination revealed xerophthalmia and xerostomia, thus Sjogren's Syndrome was diagnosed. A psychiatric consultation further diagnosed the patient with Bipolar Disorder. Successful treatment consisted of Carbamazepine and Azathioprine.

Discussion: People with immunological diseases like Sjogren's Syndrome have a higher chance of developing Bipolar Disorder with an unclear etiology. Clinical symptoms, MRI findings, and cerebral fluid analysis can lead to a proper diagnosis. Treatments include usual treatment for Sjogren's such as corticosteroids and immunosuppressants, in the addition of mood stabilizers for Bipolar.

Conclusion: Psychiatry assessments should be done systematically in patients with autoimmune diseases to avoid delays in diagnosis and to make better treatment decisions.

1. Introduction

Sjögren's syndrome is an autoimmune disease affecting exocrine glands and other organs, by inducing inflammation and the lymphocytic infiltration [1,2]. At the onset of the disease and during its course, it most commonly manifests as neuropsychiatric symptoms [3,4]. Depression, anxiety, psychiatric features, and cognitive impairment are usually the most frequent [5,6]. However, the association with bipolar disorder seems rare and not frequently documented [7,8].

Bipolar Disorder affects around 1–2% of the population [9], with an unknown etiology. Some factors such as genetic, biological, and psychosocial, have been implicated in its etiology [10,11]. BD is characterized by a depressive cycle and recurrent manic or hypomanic episodes [12,13].

BD has been reported to be associated with systemic diseases like autoimmune thyroiditis, Inflammatory Bowel Disease, autoimmune hepatitis, systemic lupus erythematosus, rheumatoid arthritis, and psoriasis [14,15]. However, the number of published studies about Sjögren's syndrome (SS) remains limited [16].

This article aims to highlight that bipolar disorder could be

commonly associated with Sjögren's syndrome and may lead to a major therapeutic impact.

1.1. Case presentation

A 43-year-old Syrian male was admitted in 2021 into the Neurology department for episodes of irritability, agitation, sleeping problems (sleep difficulty, decreased need for sleep), poor concentration, periods of elevated mood, increased energy, boxing movements in the upper limbs, and pedaling movements in the lower ones over the previous 2 years (2–3 times a month), accompanied by headaches centered around the occipital region, diplopia, and distal sensory loss in extremities.

Neurological examination revealed both posterior chordal and pyramidal signs, sensory abnormalities involving the extremities, and diplopia.

Anti-nuclear antibodies, anti-DNA test, Rheumatoid factor, anti-Ro, and anti-La tests were all negative. Total complement was normal. The viral serology (HIV and Hepatitis B and C virus), and VDRL were also negative.

The MRI showed periventricular white matter lesions on T2-

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weighted images. The cerebrospinal fluid analysis was normal. When the medical history was retaken, the patient revealed xerophthalmia and xerostomia during the previous 8 months. He considered these symptoms to be a consequence of his job, as he works with his hands for about 12 hours a day.

A Schirmer's test was positive and salivary gland biopsies were also positive showing a lymphocytic infiltration, scoring 4 using Chisholm's criteria (focus score >1) [17].

According to the ACR criteria, and to the absence of findings of other connective tissue diseases, he was diagnosed as having a primary Sjögren's syndrome [18]. Corticosteroid therapy 35 mg/day (1/2mg/kg) followed by immunosuppressive treatment in the form of Azathioprine 70mg/day (1mg/kg) was administered for 15 days without improvement. The prednisolone dose was tapered to 5 mg per week. A psychiatric consultation was ordered because of his symptoms at admission, persisting episodes of agitation, and depressed mood.

According to the Diagnostic and Statistical Manual of Mental Disorders, fifth edition (DSM5) criteria, "Bipolar Disorder" was diagnosed [19].

Carbamazepine 600 mg/day was initiated, and the patient showed significant improvement within 4 weeks displayed by a decrease in episodes of agitation and the disappearance of instability and altered mood. He was discharged on 70 mg/day Azathioprine, and 600 mg/day Carbamazepine. Three months later, the patient had no complaints, so the Carbamazepine dose was decreased to 400 mg/day. The patient continued receiving 70 mg/day Azathioprine and 400 mg/day Carbamazepine for the next six months, with lab tests monitoring.

2. Discussion

Central nervous system (CNS) involvement in SS includes focal neurological deficits, vascular encephalopathy, transverse myelitis, neuromyelitis, aseptic meningitis, dementia, and psychiatric disorders, which are more frequent in secondary Sjogren [5,20].

HIV and Hepatitis B and C virus serology, and VDRL tests were done to exclude viral and infectious encephalitis, and other connective tissue disease.

A depressive or anxious reaction to a chronic illness, either caused by the disease itself or secondary psychological distress, can explain the mental disorders in SS(6,7). People with autoimmune disease have a higher capacity to developing Bipolar Disorder than the normal population [14].

The direct immunological activity of SS on the central nervous system, may be directly responsible for the pathogenesis of psychiatric symptomatology. T cells, autoantibodies, cytokines, and apoptosis, or to autoantibodies reacting with adrenocorticotrophic hormone and a melanocyte-stimulating hormone are the accountable substances for this immunological reaction [10].

The pathoetiology of BD in SS is also unclear. Genetic association, and immunological effects, due to autoantibodies against the neurons, and the ganglionic acetylcholine receptor, and the classification of BD, itself, as an autoimmune disease are the main suspected factors [11,12,21].

Anxiety, depression, and some personality disorders were the most common psychiatric manifestations in Sjögren as reported by several studies [13–15]. Some studies confirmed that depression is the most frequent comorbidity in SS patients [3,22]. However, based on the role played by autoimmunity in the etiology of bipolar disorder, the association between BD and Sjögren's syndrome is uncommon [6,7,23].

The symptoms of BD in SS include mania, hypomania, and depressive symptoms in a relatively healthy period [13].

The MRI abnormalities include non-enhancing T2 hyperintensities in the periventricular and subcortical regions. The cerebrospinal fluid analysis may be normal, or showed elevated IgG index [23].

Treatment of SS includes: topical ocular and oral anti-inflammatory drugs, topical steroids, intravenous and/or oral steroids,

immunosuppressants, and biologics [4].

Corticosteroid therapy could induce psychiatric manifestations like manic episodes, so it should be used with caution in Bipolar-associated cases [24].

In untreated patients, mania episodes may last from 3 to 6 months, meanwhile depressive episodes can persist between 6 and 12 months, but these episodes usually improve within 3 months when treated [25]. Carbamazepine is an anticonvulsant, used in BD as a mood stabilizer, beginning with low dose and then gradually increased. Based on the above, Carbamazepine was our drug of choice. Blood tests should be used to monitor the side effects on blood white cells, liver, and kidneys [26].

The difference between this case and Salem Bouomrani et al. [8] case is our patient's younger age and difference in clinical manifestations. As Bouomrani's patient presented depressive symptoms and did not respond to antidepressants while improving on Methylprednisolone pulse treatment followed by 1 mg/kg oral prednisolone. Also, their patient's headache complain had begun 3 months ago, while in our case, the patient has been experiencing headaches for 2 years.

We had a diagnostic challenge as bipolar disorders was not in mind from the beginning. The patient was satisfied with all what we did for him including the procedures and the prescription.

3. Conclusion

Psychiatry assessments should be done systematically in these patients to avoid delays in diagnosis and to make better treatment decisions, especially corticosteroid therapy, improving their quality of life.

Availability of supporting data

The data supporting the results of this article is included within the article's references.

Provenance and peer review

Not commissioned, externally peer reviewed.

Ethical approval

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Authors' contributions

Dr. Naram Khalayli wrote the case presentation, discussion, and conclusion. Dr. Maysoun Kudsi wrote the abstract and introduction. Both authors reviewed the manuscript. Slight revisions and edits were later made by Dr. Naram.

Please state any conflicts of interest

None of the authors state any conflict of interest.

Registration of research studies

Name of the registry:

Unique Identifying number or registration ID:

Hyperlink to your specific registration (must be publicly accessible and will be checked):

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Guarantor

Dr.Naram Khalayli Is the guarantor for this particular study.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.amsu.2022.104243>.

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