Aseptic meningitis as an initial presentation of Sjögren syndrome: a report of two cases and literature review

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ABSTRACT

Sjögren syndrome (SS) is one of several collagen vascular diseases that occasionally involve the central nervous system. We report two cases of SS involving young patients who initially presented with aseptic meningitis. A male with recurrent AM was found to have anti-Ro/SSA and La/SSB antibodies in a screening test for autoimmune process. A minor salivary gland biopsy revealed lymphocytic infiltrations compatible with SS, although the patient did not exhibit sicca symptoms. A female presenting with AM and polyarthritis also reported xerophthalmia. Anti-Ro/SSA antibody testing and a positive result in a minor salivary gland biopsy led to the diagnosis of SS. In the literature review, we found that AM or aseptic meningoencephalitis (AME) preceded or had a concomitant onset with SS in approximately 70% of cases. Screening for anti-Ro/SSA antibody, as well as systemic assessment for rheumatic symptoms, may be useful for diagnosing AM/AME of unknown etiology.

Keywords: Sjögren syndrome, aseptic meningitis, aseptic meningoencephalitis, anti-Ro/SSA antibody, sicca symptoms

INTRODUCTION

Sjögren syndrome (SS) is a classic collagen vascular disease that usually causes xerophthalmia and xerophthalmia due to lymphocytic infiltration in the exocrine glands, but occasionally involves the skin, lung, heart, kidney, and nervous system.1 Central nervous system involvement
is observed in 14% to 19% of cases, and includes transverse myelitis, encephalopathy (i.e., psychosis, seizure), and AM (aseptic meningitis). AM is defined by cerebrospinal fluid (CSF) pleocytosis with a negative CSF bacterial culture, while aseptic meningoencephalitis (AME) refers to inflammation involves the brain parenchyma as well as the meninges. AM occur at any age but are most common in children. Most cases of AM caused by viruses (i.e., enterovirus) have a self-limited course with a symptom duration of 1–2 weeks. On the other hand, certain drugs, neoplasms, or rheumatic diseases can be a cause of AM, and diagnosis and treatment of underlying etiologies may thus be required in selected cases.

In 1983, Alexander et al first reported five cases of acute meningoencephalitis (AME) in SS, and reported that four (80%) experienced spontaneous improvement. On the other hand, four (80%) relapsed at least three times, and three (60%) subsequently required glucocorticoid therapy. Herein, two cases of AM with an unknown background of SS are described, followed by a literature review of SS cases associated with AM and/or AME.

CASE 1

A 19-year-old man was admitted to our hospital with a 3-day history of fever, headache, and sore throat. Relevant medical history included three episodes of self-limiting AM at 13, 14, and 16 years of age. During the third episode of AM at 16 years of age, the patient was evaluated for recurrent meningitis, and was found to have anti-Ro/SSA at 64-fold dilution and anti-La/SSB antibodies at 32-fold dilution (using the immunoprecipitation method: normal < 1-fold dilution for each). Results from a minor salivary gland biopsy revealed at least one focus defined as ≥ 50 lymphoid cells infiltrations per surface area of 4 mm², leading to the diagnosis of SS, although he did not exhibit sicca symptoms or superficial punctate keratitis on ophthalmological examination.

On this admission, he was alert, with a body temperature of 36.8°C, a heart rate of 79 beats/min, blood pressure of 110/58 mmHg, and a respiration rate of 16 breaths/min. Neurological examination revealed normal function, except for positive jolt accentuation. On laboratory investigation, complete blood count, electrolytes, renal and liver function, and urinary tests were normal, except for an elevated level of C-reactive protein (1.37 mg/dL). Antinuclear, anti-Ro/SSA, and anti-La/SSB antibodies were positive at 640-, 256-, and 32-fold dilution (normal, < 40-fold dilution for each), and rheumatoid factor of 42.9 U/mL (normal, ≤ 15 U/mL). Results for anti-double stranded DNA, anti-RNP, anti-Sm, anti-β2 glycoprotein I and anti-aquaporin-4 antibodies, and lupus anticoagulant were negative. The results of magnetic resonance imaging (MRI) were normal. CSF analysis revealed mild pleocytosis (14/µL [monocytes, 7/µL]), protein 59 mg/dL, glucose 57 mg/dL, and negative results for oligoclonal bands and myelin basic protein. Polymerase chain reaction amplification of herpes simplex virus DNA and culture of CSF samples were both negative, leading to the diagnosis of SS-associated AM. He was observed without therapy; nevertheless, fever and headache persisted. On day 5 of hospitalization, CSF analysis revealed pleocytosis (530/µL [monocytes, 445/µL]), protein 165 mg/dL, and a glucose level of 45 mg/dL. T2 fluid-attenuated inversion recovery sequence on gadolinium-enhanced MRI of the brain revealed diffuse enhancement of the meninges, suggesting worsening of AM (Fig. 1). After methyl-prednisolone pulse therapy (1 g/day for three days), his symptoms improved within one week.

One year later, he had the fifth episode of AM and received pulse steroid therapy followed by oral prednisolone. However, the patient developed the sixth episode two month later, while taking daily 10 mg of prednisolone. He again improved with pulse steroid therapy and is now taking prednisolone and azathioprine as maintenance therapy.
CASE 2

A 23-year-old woman with a three-day history of headache and fever was admitted to our hospital. She also experienced vomiting for 1 day, and polyarthralgia for 2 days. She denied any relevant medical history but took over-the-counter analgesic medication for menstrual cramps one week before her visit. Physical examination revealed normal consciousness, a body temperature of 39.2°C, a heart rate of 107 beats/min, a blood pressure of 123/68 mmHg, and respiration rate of 16 breaths/min. There was tenderness at the wrists, knees, and sternoclavicular and temporomandibular joints, while the left wrist and the right knee were swollen. Neurological examination showed neck stiffness and jolt accentuation. Laboratory investigations did not show remarkable results, except for elevated CRP level (3.37 mg/dL). Examination of CSF revealed mild pleocytosis (11/µL; on the next day, 31/µL, with polynuclear cells at 23/µL and monocytes at 8/µL), and negative results for myelin basic protein. T2 fluid-attenuated inversion recovery sequence on MRI of the brain demonstrated white matter lesions in the temporal and parietal lobes (Fig. 2). After starting therapy with acyclovir and ceftriaxone, her symptoms resolved within one week. Polymerase chain reaction amplification of herpes simplex virus DNA and culture of CSF samples were both negative, and antibody tests for herpes group virus suggested negative results or past infection.

When she recovered, the patient reported a one-year history of xerophthalmia. Antinuclear and anti-Ro/SSA antibodies, and rheumatoid factor were found to be present in her sera at 40- and 256-fold dilution, and 54.9 U/mL, respectively, while anti-La/SSB, anti-double stranded DNA, anti-RNP, anti-Sm, or anti-phospholipid antibodies were absent. Result of minor salivary gland biopsy revealed several foci of lymphoid cells infiltration, leading to the diagnosis of SS, while
ophthalmological examination did not show superficial punctate keratitis. Thereafter, she remained well at least for 11 months. The white matter lesions on the T2 fluid-attenuated inversion recovery imaging remained unchanged 10 months after presentation, suggesting asymptomatic white matter lesions in SS.7

DISCUSSION

We encountered two cases of AM in young patients with an unknown background of SS. A diagnosis of SS was suspected from anti-Ro/SSA antibody as a screening test for recurrent meningitis in one, and from simultaneous onset of polyarthritis in the other. Both cases did not have any laboratory findings of systemic lupus erythematosus or mixed connective tissue disease. Both experienced a self-limiting course of AM, while the male subsequently required glucocorticoid therapy for worsening meningitis. As a possible cause of SS-AM, the role of autoantibodies, other than anti-Ro/SSA or anti-La/SSB, has been suggested,8 while exogenous factors, such as drugs, may also have a role in triggering AM in SS.9-12 Recurrent episodes of AM in our male case may suggest that SS, by itself, has a role in the development of AM, whereas a single episode of AM in the other case may have been triggered by a drug.

To clarify the clinical characteristics of AM associated with SS, we searched the PubMed and ICHUSHI databases for case reports and case series addressing AM or AME associated with SS, without language restrictions. Inclusion criteria were cases of AM or AME associated with SS in peer-reviewed articles, published up to September 18, 2019. Publications lacking patient details, such as CSF examination, those mainly associated with encephalitis (i.e., psychosis, dementia), and those probably associated with other collagen vascular disease, were excluded. Clinical data were extracted for these cases.

The literature review retrieved 33 cases of SS-AM and AME (Table 1).2,6,8-30 There were 30

Fig. 2 MRI findings in Case 2
MRI at the fifth episode of AM. The T2-fluid-attenuated inversion recovery sequence of magnetic resonance imaging of the brain revealed white matter lesions (white arrows) in the temporal and parietal lobes (A and B, respectively).
## Table 1  Clinical characteristic of the published cases of aseptic meningitis or meningoencephalitis associated with Sjögren syndrome

| Age/Sex | Preceding disease | times | FUP (mo) | Dry eyes | Dry mouth | αRo | αLa | CSF cells/mm³ (Monocyte,%) | Therapy |
|---------|-------------------|-------|----------|----------|-----------|------|-----|--------------------------|---------|
| 52F⁰   | NA                | 3     | 18       | (+)      | (+)       | (+)  | (–) | 900 (40)                 | (–)→(–)→GC |
| 50F⁰   | NA                | 1     | NA       | (+)      | (+)       | (+)  | (–) | 33 (18)                  | GC      |
| 46F⁰   | NA                | 4     | 1.75     | (+)      | (+)       | (–)  | (–) | 70 (60)                  | (–)→(3→GC) |
| 33F⁰   | NA                | 3     | 64       | (+)      | (+)       | (+)  | (–) | 245 (0)                  | (–)→(–)→GC |
| 33F⁰   | NA                | 3     | 3.5      | (+)      | (+)       | (–)  | (–) | 238 (79)                 | (–)→(–)→(–) |
| 25F¹³  | Sim.              | 1     | NA       | NA       | NA       | 256  | 1   | 33 (100)                 | GC      |
| 56F¹³  | SS                | 1     | 48       | NA       | (+)      | 256  | (–) | 104 (100)                | IVIg     |
| 42F¹³  | SS                | 1     | 12       | NA       | NA       | 256  | 2   | 161 (100)                | GC      |
| 45F⁰   | SS                | 1     | 0.4      | NA       | NA       | NA   | NA | 341 (3)                  | (–)     |
| 59F¹⁴  | SS                | 1     | 9        | NA       | NA       | (–)  | 64  | 23 (22)                  | GC      |
| 41F¹²  | SS                | 2     | 1        | NA       | NA       | (–)  | (–) | 640 (NA)                 | (–)→GC |
| 27F¹⁵  | Sim.              | 1     | 2        | (–)      | (–)      | 128  | 1   | 363 (30)                 | GC      |
| 18F¹⁵  | Sim.              | 1     | 6        | (+)      | (–)      | (–)  | (–) | 69 (90)                  | GC, CY  |
| 29F¹⁷  | AM                | 2     | 9        | NA       | NA       | 256  | (–) | 140 (80)                 | GC      |
| 15F¹⁸  | Sim.              | 1     | 2        | (–)      | (–)      | (–)  | (–) | 77 (75)                  | GC      |
| 48F²⁹  | Sim.              | 2     | 96       | (+)      | (+)      | NA   | NA | 224 (38)                 | (–)→GC |
| 8F²⁰   | AM                | 3     | 72       | NA       | NA       | (+)  | (–) | 16 (70)                  | NA→(–)→GC |
| 76F²¹  | Sim.              | 1     | 4        | NA       | NA       | 128  | 128 | 699 (60)                 | (–)     |
| 14M²²  | Sim.              | 1     | 1        | (–)      | (–)      | 16   | 32  | 37 (NA)                  | (–)     |
| 26F²²  | SS                | 1     | 4        | (–)      | (–)      | 4    | 8   | 603 (38)                 | GC      |
| 11M²⁸  | AM                | 2     | 26       | (–)      | (–)      | (+)  | (–) | 40 (NA)                  | GC→GC |
| 8F²⁴   | Sim.              | 1     | 6        | (–)      | (–)      | (–)  | (–) | 30 (83)                  | GC+CyA→GC |
| 35F²⁹  | Sim.              | 1     | 3        | NA       | NA       | (–)  | (–) | 381 (3)                  | GC      |
| 62F¹¹  | SS                | 1     | NA       | (+)      | (+)      | NA   | (–) | 38 (18)                  | (–)     |
| 50F²⁵  | Sim.              | 1     | 48       | (–)      | (–)      | (+)  | (–) | 43 (NA)                  | GC      |
| 29F²⁶  | AM                | 4     | 72       | NA       | NA       | (+)  | (–) | 153 (95)                 | GC→GC→GC |
| 58F²⁷  | Sim.              | 1     | 24       | (+)      | (+)      | NA   | NA | 192 (40)                 | GC      |
| 19F³   | AM                | 3     | 9        | NA       | NA       | 256  | 8   | 85 (81)                  | NA→NA→GC |
| 16F²⁰  | SS                | 1     | 5.7      | (+)      | (+)      | (+)  | (–) | 43 (98)                  | GC      |
| 32F²⁹  | NA                | 1     | 84       | (+)      | (+)      | (–)  | (–) | 162 (90)                 | GC      |
| 42F²³  | Sim.              | 1     | NA       | NA       | NA       | NA   | NA | 382 (45)                 | GC      |
| 19M    | AM                | 6     | 78       | (–)      | (–)      | 64   | 32  | 88 (95)                  | (–)→(3→GC→GC→GC→AZP |
| 23F    | Sim.              | 1     | 11       | (+)      | (–)      | 256  | (–) | 11 (NA)                  | (–)     |

The results of anti-Ro/SSA and anti-La/SSB antibody analyses are expressed as positive (+) or negative (–). Results for the immunodiffusion method are expressed as multiple numbers (normal, < 1-fold).

M: male, F: female, FUP: follow-up period, αRo: anti-Ro/SSA antibody, αLa: anti-La/SSB antibody, CSF: cerebrospinal fluid, NA: not available, Sim: Simultaneous onset, SS: Sjögren syndrome, AM: aseptic meningitis (or meningoencephalitis), IVIg: intravenous immunoglobulin, CY: cyclophosphamide, CyA: cyclosporine A, GC: glucocorticoids.
females and three males, with a mean (± standard deviation) age of 34.8 ± 17.4 years. In five (15%) patients, drugs were considered to possibly trigger or cause AM. Of the 27 cases that were evaluable, the diagnosis of SS and AM/AME simultaneously onset in 13 (48%), while SS or AM/AME preceded one another in eight (30%) and six (22%), respectively. This finding means that 70% (19/27) of SS-AM/AME developed in an unknown background of SS, as reflected in our cases. Initial symptoms of AM included headache (n = 28 [at least 85%]), fever (n = 26 [79%]), nausea or vomiting (n = 15 [45%]), disturbance of consciousness (n = 11 [33%]), and neck pain or rigidity (n = 6 [18%]). Other neurological complications included seizure (n = 3 [9%]), cranial nerve disorder (n = 3 [9%]), myelitis (n = 2 [6%]), coma (n = 1 [3%]) and peripheral neuropathy (n = 1 [3%]). Xerophthalmia and xerostomia was described in 12 (36%) cases each, respectively, while either was absent at least in nine (27%). Other rheumatic manifestations included parotiditis (n = 6 [18%]), joint pain (n = 5 [15%]), lymphadenopathy (n = 2 [6%]), renal tubular acidosis (n = 2 [6%]), interstitial lung disease (n = 2 [6%]), and Raynaud phenomenon (n = 1 [3%]). Anti-Ro/SSA, anti-La/SSB, and antinuclear antibodies were found in 27 (82%), 19 (70%), and 26 (79%) cases, respectively. At least 12 (36%) patients experienced relapse of AM/AME, although relapse in one was likely due to restart of a causative drug. For the relapsing cases, a mean of 2.9 ± 0.80 events of AM occurred during a mean of 37 ± 35 months. On the results of CSF analysis that were initially available, median total cell counts were 104/μL (interquartile range, 40–245), monocytes 63.5% ± 39.7, protein, median 55 mg/dL (interquartile range, 5–84 mg/dL) and glucose, mean 60.0 ± 15.7 mg/dL. Of the 28 cases not related to drug use, eight (29%) experienced a self-limiting course, but 25 (89%) subsequently required therapy. Only one (3%) case died of respiratory failure during therapy.

Of the 19 cases in which AM/AME preceded or onset with a diagnosis of SS, eight (42%) did not have xerophthalmia or xerostomia, suggesting that it may be difficult to suspect underlying SS in some AM/AME cases. Delay of diagnosis of underlying SS was found in five (26%) of such cases by 0.75 to 3 years. However, at least 89% (17/19) of the cases had anti-Ro/SSA antibodies. Screening underlying SS by anti-Ro/SSA antibody may, thus, be suggested for cases of AM/AME of unknown etiology, especially for recurrent cases. It is also important for indications for immunosuppressive therapy to recognize underlying autoimmune mechanisms early because some SS-AM/AME subsequently may result in cerebral and spinal cord involvement that would be more likely associated with neurological sequelae.

In conclusion, AM or AME in SS often onset with or preceded the diagnosis of SS, as seen in our cases. To prevent diagnostic delay of underlying SS and neurological sequelae, we suggest that screening for anti-Ro/SSA antibody and systemic assessment of autoimmune features including sicca, parotiditis, or joint pain may be useful for AM or AME cases of unknown etiology. Our literature review was clearly at risk for bias, and a larger study focusing on this rare disorder is required to confirm these results.

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CONFLICT OF INTEREST

None to declare.
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