



Short communication

Pseudotumoral brain lesion as the presenting feature of primary Sjögren's syndrome



Samia Ben Sassi^{a,*}, Fatma Nabli^a, Adnene Boubaker^a, Imed Ben Ghorbel^b, Sonia Neji^a, Fayçal Hentati^a

^a National Institute Mongi Ben Hmida of Neurology, Faculty of Medicine of Tunis, University Tunis El Manar, Tunis, Tunisia

^b Department of Internal Medicine, La Rabta Hospital, Tunisia

ARTICLE INFO

Article history:

Received 27 November 2013

Received in revised form 5 January 2014

Accepted 23 January 2014

Available online 30 January 2014

Keywords:

Sjögren's syndrome

Connective tissue disease

Auto-immune disease

Brain magnetic resonance imaging

Pseudotumoral

Central nervous system

ABSTRACT

Background: The frequency and type of central nervous system involvement in primary Sjögren's syndrome (pSS) remain controversial. Brain magnetic resonance imaging (MRI) abnormalities in pSS are usually discrete hyperintense areas in the white matter. Tumefactive brain lesions have been rarely reported.

Case report: We describe a 31-year-old woman who exhibited transcortical motor aphasia, hemiparesis and partial motor seizures as the initial manifestation of pSS. Brain MRI revealed a large frontoparietal lesion extending into the corpus callosum. The patient had spontaneous recovery and developed sicca symptoms 6 months after onset. Primary SS was diagnosed on the basis of clinical features, abnormal Schirmer test findings, high levels of anti-La/SSB antibodies and positive salivary gland biopsy results.

Conclusion: The present case suggests that a pseudotumoral brain lesion can occur as an initial symptom of pSS.

© 2014 Elsevier B.V. All rights reserved.

1. Introduction

Sjögren's syndrome (SS) is a common autoimmune disease that may be primary (pSS) or secondary to other connective tissue diseases. It is characterized by lymphocytic infiltration involving mainly salivary and lacrimal glands and resulting in xerostomia and xerophthalmia. Approximately one-third of patients with primary Sjögren's syndrome (pSS) present with systemic manifestations related to extraglandular mononuclear infiltrates or vasculitis lesions [1,2]. While peripheral neuropathy is a well-defined complication of pSS, the prevalence of central nervous system (CNS) involvement remains controversial and its clinical presentation varies considerably leading to delay in diagnosis [3–5]. We herein report a patient with primary SS who presented with a pseudotumoral brain lesion as the initial manifestation.

2. Case report

A previously well left-handed 31-year-old woman at 12 weeks' gestation developed rapidly progressive speech difficulty, left-sided weakness, left partial motor seizures and global headache over a 15-day

period. On admission, she was afebrile and conscious and she had transcortical motor aphasia, dysarthria and left hemiparesis. Magnetic resonance imaging (MRI) revealed a large right frontoparietal lesion extending into the splenium of the corpus callosum and a small left subcortical parietal lesion (Fig. 1). Cerebral angiography was normal. Stereotactic brain biopsy was performed, but only reactive gliosis was observed.

The patient was treated with carbamazepine. She had a complete spontaneous recovery within 2 months and an uncomplicated delivery. Six months after onset, the patient complained of arthralgias and dry mouth and eyes. Blood analysis showed hypergammaglobulinemia (IgG, 17.1 g/L), antinuclear antibody titers of 1/1600, positive rheumatoid factor and anti-La/SSB antibodies.

The following parameters were all normal: complete blood counts, biochemical screening, erythrocyte segmentation rate, C-reactive protein, serum C3, C4, and CH50, anti-ds-DNA, p and c ANCA, antiphospholipid antibodies and lupus anticoagulant. Serological tests for syphilis, HIV, hepatitis C and hepatitis B were negative.

A salivary gland biopsy revealed inflammatory changes with a focus score of 4, using Chisholm's classification. The result of Schirmer's test was strongly positive. The patient was diagnosed with pSS according to the American and European criteria [6]. Oral prednisone 1 mg/kg daily was started and subsequently tapered to 10 mg daily over the ensuing months. The patient recovered without any residual neurological deficit and remained seizure free.

A 12-month follow-up MRI showed brain atrophy and residual frontoparietal and corpus callosal lesions (Fig. 1).

* Corresponding author at: Institut National Mongi Ben Hmida de Neurologie, La Rabta, 1007 Tunis, Tunisia. Tel.: +216 52564544; fax: +216 71963799.

E-mail address: bensassisam@yahoo.fr (S.B. Sassi).

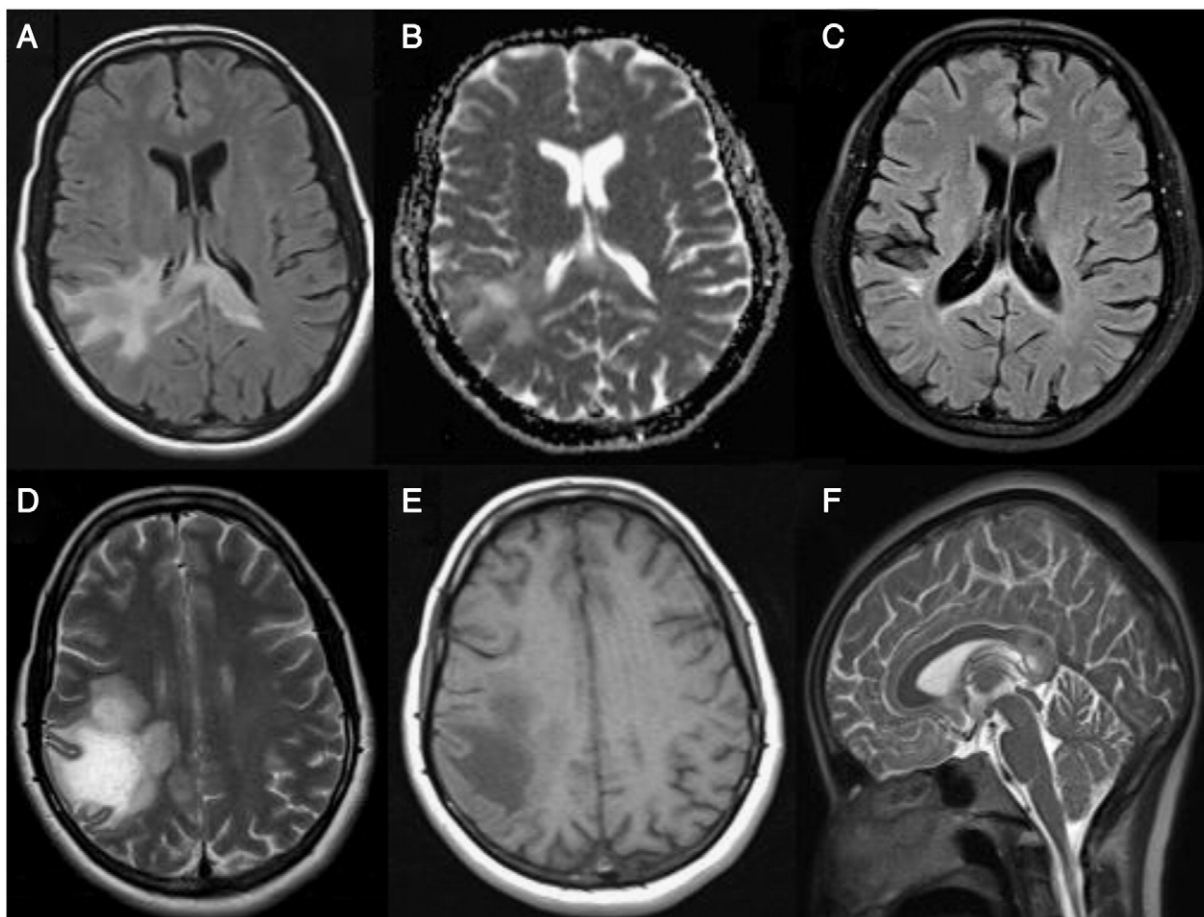


Fig. 1. Axial fluid-attenuated inversion recovery (FLAIR) (A), T2-weighted (D) and T1-weighted (E) MR images show a large cortico-subcortical frontoparietal lesion with a necrotic component extending into the splenium of the corpus callosum. The lesion is hyperintense on the apparent diffusion coefficient image (B). Sagittal FLAIR image (F) shows tumefactive lesion in the splenium of the corpus callosum. Axial FLAIR (C) image at the 12-month follow-up shows brain and corpus callosum atrophy with residual parietal and corpus callosal hyperintensities.

3. Discussion

The exact prevalence of CNS manifestations in unselected pSS patients remains unclear and ranges widely from 0% to 60% [3–5]. CNS involvement varies from diffuse compromise, manifested as cognitive deficits, psychiatric abnormalities or migraine, to focalized compromise with myelitis, optic neuropathy, subarachnoid hemorrhage or, as in our case, focal encephalitic involvement [3].

The great majority of patients with focal CNS disease exhibit multiple hyperintensities in the subcortical and periventricular white matter on T2-weighted and fluid-attenuated inversion recovery (FLAIR) sequences [4,5]. However, the cerebellum, corpus callosum and basal ganglia can also be involved [3]. Brain lesions are generally less pronounced in pSS than in multiple sclerosis (MS) and hyperintensities are usually less than 1 cm and are often not enhanced with gadolinium [4].

Our patient with established pSS exhibited a pseudotumoral lesion. Differential diagnosis including lymphoma and progressive multifocal leukoencephalopathy, which are potential complications of pSS [7,8], was excluded particularly with regard to the spontaneous favorable outcome. Although described in other autoimmune CNS diseases like MS, tumefactive lesions have been rarely reported in pSS [9,10]. Sanahuja et al. [9] depicted a 50-year-old woman with pSS and a one-year history of arthralgias and dry mouth who developed hemiparesis and hemihypoesthesia. She exhibited a large frontoparietal lesion with complete improvement under oral prednisone treatment. Michel et al. [10] reported the case of a 63-year-old woman with pSS and sicca symptoms who had Broca's aphasia and hemiparesis due to

a large white matter lesion in the corona radiata. Full recovery was observed under corticoids and immunosuppressive treatment.

It is worth noting that in contrast to the previously described cases [9,10], the neurological complication of pSS did occur in our patient as the initial manifestation without clinical sicca symptoms. The CSF profile in SS is nonspecific and strongly resembles that described in MS patients with mild lymphocytosis and increased IgG index in up to 50% of active cases. Nevertheless, the presence of oligoclonal bands which can be helpful in the diagnosis of pseudotumoral MS is usually lacking in SS [11].

The pathogenesis of CNS involvement in pSS is still unknown. Our patient had hypergammaglobulinemia with positive anti-La/SSB antibodies. Abnormalities in humoral and cellular immunity, including antineuronal antibody, seem to be involved in the CNS damage seen in pSS. However, the immunological profile associated with CNS manifestations remains controversial. Some studies have shown the direct involvement of anti-Ro/SSA antibodies while in others anti-SSA and anti-SSB antibodies were found in less than 50% of patients [12,13].

On the other hand, previous studies have suggested that the main pathophysiologic mechanism was ischemia related to an immunologically mediated small vessel vasculopathy [14]. However, in our case, the aspect of MRI lesions with no restricted diffusion and the normal cerebral angiography were not consistent with a vasculitic process.

In pSS patients, lymphocytic infiltration causing pseudotumoral extraglandular lesions has been described in several organs like the lung, kidney, liver and pancreas. A direct infiltration of the CNS by lymphocytic cells has also been reported by Bakchin et al. [15]. In our

patient, the type of MRI findings and the spontaneous recovery suggest an inflammatory-demyelinating mechanism.

In conclusion, this case illustrates that Sjögren's syndrome should be considered in case of pseudotumoral brain lesion even in the absence of sicca symptoms.

Conflict of interest statement

The authors declare no conflict of interest.

References

- [1] Ramos-Casals M, Font J. Primary Sjögren's syndrome: current and emergent aetiopathogenic concepts. *Rheumatology* 2005;44:1354–67.
- [2] Garcia-Carrasco M, Ramos-Casals M, Rosas J, et al. Primary Sjögren syndrome: clinical and immunologic disease patterns in a cohort of 400 patients. *Med (Baltimore)* 2002;81:270–80.
- [3] Delalande S, de Seze J, Fauchais AL, et al. Neurologic manifestations in primary Sjögren syndrome: a study of 82 patients. *Med (Baltimore)* 2004;83:280–91.
- [4] Morgen K, McFarland HF, Pillemer SR. Central nervous system disease in primary Sjögren's syndrome: the role of magnetic resonance imaging. *Semin Arthritis Rheum* 2004;34:623–30.
- [5] Soliotis FC, Mavragani CP, Moutsopoulos HM. Central nervous system involvement in Sjögren's syndrome. *Ann Rheum Dis* 2004;63:616–20.
- [6] Vitali C, Bombardieri S, Jonsson R, et al. Classification criteria for Sjögren's syndrome: a revised version of the European criteria proposed by the American–European Consensus Group. *Ann Rheum Dis* 2002;61:554–8.
- [7] Hayashi Y, Kimura A, Kato S, Koumura A, et al. Progressive multifocal leukoencephalopathy and CD4+ T-lymphocytopenia in a patient with Sjögren's syndrome. *J Neurol Sci* 2008;268:195–8.
- [8] Itoh T, Shimizu M, Kitami K, et al. Primary extranodal marginal zone B-cell lymphoma of the mucosa-associated lymphoid tissue type in the CNS. *Neuropathology* 2001;21(3):174–80.
- [9] Sanahuja J, Ordoñez-Palau S, Begué R, Brieva L, Boquet D. Primary Sjögren's syndrome with tumefactive central nervous system involvement. *AJNR Am J Neuroradiol* 2008;29:1878–9.
- [10] Michel L, Toulgoat F, Desal H, et al. Atypical neurologic complications in patients with primary Sjögren's syndrome: report of 4 cases. *Semin Arthritis Rheum* 2011;40:338–42.
- [11] Alexander EL, Malinow K, Lejewski JE, Jerdan MS, Provost TT, Alexander GE. Primary Sjögren's syndrome with central nervous system disease mimicking multiple sclerosis. *Ann Intern Med* 1986;104(3):323–30.
- [12] Anaya JM, Villa LA, Restrepo L, Molina JF, Mantilla RD, Vargas S. Central nervous system compromise in primary Sjögren's syndrome. *J Clin Rheumatol* 2002; 8:189–96.
- [13] Alexander EL, Ranzenbach MR, Kumar AJ, et al. Anti-Ro (SS-A) autoantibodies in central nervous system disease associated with Sjögren's syndrome (CNS-SS): clinical, neuroimaging, and angiographic correlates. *Neurology* 1994;44: 899–908.
- [14] Alexander E. Central nervous system disease in Sjögren's syndrome. New insights into immunopathogenesis. *Rheum Dis Clin North Am* 1992;18:637–72.
- [15] Bakchin S, Duyckaerts C, Hassine L, et al. Central and peripheral neurologic lesions in primary Gougerot–Sjögren syndrome. Clinicopathological study of a case. *Rev Neurol* 1991;147:368–75.