Neurol Sci (2015) 36:803–804
DOI 10.1007/s10072-014-1907-1

LETTER TO THE EDITOR

Efficacy of high dose methylprednisolone in a patient with cervical dystonia and blepharospasm and Sjögren's syndrome

Vittorio Mantero · Roberto Balgera · Andrea Rigamonti · Francesco Basso · Andrea Salmaggi

Received: 24 March 2014 / Accepted: 22 July 2014 / Published online: 25 July 2014

Dear Editor,

Dystonia is a term indicating a heterogeneous group of hyperkinetic movement disorders characterised by involuntary muscle contractions, resulting in twisting and repetitive movements and abnormal postures. Sjogren's syndrome (SS) is an autoimmune disorder characterised by lymphocytic infiltration and destruction of the salivary and lacrymal glands leading to xerostomia and xerophthalmia. Neurological complications have been reported in approximately 1.5–25 % of patients with SS, however, movement disorders like parkinsonism, chorea and dystonia are infrequent [1]. We herein report a patient with blepharospasm and cervical dystonia associated to primary SS.

A 67-year-old woman came to our observation complaining of difficulty in maintaining head posture, with anterior flexion of the neck, blinking and spasming of the eyelids since 3 months.

Five years previously she had been diagnosed with SS class III according to the Chisholm-Mason scoring system on the basis of xerophthalmia, xerostomia, positivity of rheumatoid factor (31 IU/ml) and a positive salivary gland biopsy. She had a copy of HLA-DRw52. Anti-Ro and anti-La antibodies were negative. She had been treated with pilocarpine chlorhydrate for 4 years which was then interrupted because of nausea. She also had hypothyroidism controlled with replacement therapy.

At entry, neurological examination disclosed blepharospasm and spasmodic anterocollis. Haematological investigations revealed increased title of myeloperoxidase-specific antineutrophil cytoplasmic antibodies (MPO-ANCA) 7 IU/ml, normal rheumatoid factor levels and absence of other autoantibodies (ENA, ANA, antithyroid, crioaglutinin, anticardiolipin, antiphospholipid). Brain and cervical magnetic resonance imaging showed no structural lesion, electroneurography and electromyography studies were normal.

The patient was initially treated with clonazepam (1 mg/day for 20 days) without benefit, so intravenous steroid treatment (500 mg methylprednisolone daily for 3 days) was introduced with a rapid and significant improvement of cervical dystonia whereas blepharospasm persisted. Because of a previously reported poor tolerance to chronic steroids treatment, the patient was not treated chronically with prednisone, but we performed monthly endovenous methylprednisolone administrations; moreover, botulin toxin (20 UI of Botox) was administrated to treat blepharospasm.

Presently, a year after onset, the patient is almost symptoms free.

A variety of movement disorders have been described in patients with SS but its have been reported very rarely [1]. Only six cases of dystonia associated with SS have been reported in the literature (Table 1). The first report was in 1999, in a patient with SS and dystonia of the left hand and foot [2]; in the same year three other patients with SS who developed intermittent tonic/dystonic limb spasms were reported [3]; more recently, a case of orofacial dystonia [4] and a case of right arm paroxysmal dystonia [5] were described. In three of these patients treatment with steroids led to clinical improvement [2–4]. A case of cervical dystonia in association with SS had not been described before.

Early recognition of the immunological pathogenesis of dystonia is important, as treatment, if initiated immediately

V. Mantero (✉) · R. Balgera · A. Rigamonti · F. Basso · A. Salmaggi
Neurological Department, A. Manzoni Hospital, Via dell'Eremo 9/11, 23900 Lecco, Italy
e-mail: vittorio.mantero@hotmail.com

Table 1 Clinical characteristics and administered therapy with improvement in the patients reported in literature affecting by SS in association with dystonia

References	Sex	Age	Symptoms	Therapy
[2]	M	36	Dystonic posture of the left hand and left foot	Prednisone and cyclophosphamide
[3]	M	53	Painful drawing and twisting postures of the left extremities	Prednisone and phenytoin
[3]	F	41	Involuntary, forceful movements of the right leg	Baclofen
[3]	M	36	Dystonic posture of right leg and right abdominal, back and leg spasms	Baclofen and carbamazepine
[4]	F	57	Orofacial dystonia	Endovenous methylprednisolone
[5]	F	57	Paroxysmal dystonia of the right arm	Clonazepam
Our case	F	67	Blepharospasm and spasmodic anterocollis	Endovenous methylprednisolone and botulin toxin (only for blepharospasm)

after diagnosis, can be effective in suppressing the autoimmune response [2]. Indeed, the interest of this report, as well as the infrequent association between SS and dystonia, lies in the important clinical improvement after immunosuppressive treatment. Even if we can not exclude a coincidental association between SS and dystonia, we think that response to immunosuppressive treatment, as in our patient, supports the hypothesis of a pathogenetic link; the lack of improvement in blepharospasm after steroid treatment, on the other hand, suggests that the pathogenesis of blepharospasm may not be the same as that of dystonia. As already reported, in SS central nervous system damage could be attributed to vasculitis or direct autoimmune damage to the neural tissue [4]. While proven cases of either primary CNS vasculitis or of CNS vasculitis in the context of systemic vasculitides nearly always result in abnormalities of brain/spinal cord imaging by MRI, antibody-mediated damage may result in CNS dysfunction without apparent morphological abnormalities; this is the case for some systemic lupus erythematosus-related and SS-related neurological complications such as lateralized parkinsonism, chorea or ballism [1].

In our case absence of structural lesion of brain and cervical spine and the good response to immunosuppressive therapy support the hypothesis of an immune antibody-mediated damage against striatal structures as the underlying mechanism for the pathogenesis of dystonia.

References

1. Baizabal-Carvallo JF, Jankovic J (2012) Movement disorders in autoimmune diseases. Mov Disord 27:935–946
2. Van den Berg JS, Horstink MW, van den Hoogen FH, Oyen WJ (1999) Dystonia; a central nervous system presentation of Sjögren's syndrome. Mov Disord 14:374–375
3. Jabbari B, Salardini A (1999) Painful tonic/dystonic spasms in Sjogren's syndrome. Mov Disord 14:860–864
4. Papageorgiou S, Kontaxis T, Bonakis A, Kalfakis N (2007) Orofacial dystonia related with Sjogren's syndrome. Clin Rheumatol 26:1779–1781
5. Alonso-Navarro H, Arroyo M, Parra A, Jiménez-Jiménez FJ (2009) Paroxysmal dystonia associated to primary Sjögren's syndrome. Mov Disord 24:788–790