Case report

A case of Sjögren's syndrome presenting as trigeminal nerve palsy

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summary

We herein report the case of a female with Sjogren's syndrome (SS) complicated with trigeminal nerve palsy. Although her sicca symptoms had been unnoticed, head magnetic resonance imaging (MRI) for detecting brain abnormalities revealed parotid gland changes associated with SS. SS should be considerd as a possible cause of trigeminal nerve disturbances. In addition, parotid gland changes related to SS should be aware in examination of cranial nerve disturbances with MRI.

Key words——Sjögren's syndrome; trigeminal nerve palsy

Introduction

Sjögren's syndrome (SS) is a chronic autoimmune disease characterized by sicca symptoms due to lymphocytic infiltration of exocrine glands, such as xerophthalmia and xerostomia[1]. Other than sicca symptoms, various extraglandular symptoms may develop in SS. Whereas lung diseases and renal disturbance are well -known complications of SS, neurologial symptoms are uncommon complications of SS. Because the etiology of neurological involvement of SS is not fully clarified, it is often difficult to prove association between SS and neurological symptoms. This fact makes SS associated neurological diseases difficult to diagnose.

We herein describe a 63-year-old female with SS complicated with trigeminal nerve palsy, found out on magnetic resonance imaging (MRI) findings.

Case report

A 63-year-old female developed persistent numbness and sensory disturbance in the right side of her face in mid January 2011. Although she visited a neighboring hospital, the causes of her symptoms remained unknown. She consulted a neurovascular surgery department at our hospital. MRI with gadolinium contrast did not demonstrate any marked abnormalities of the central nervous system. However, a mottled appearance of the parotids, which was compatible with Sjögren's syndrome (Fig. 1), was found. Therefore she was referred to our department for further examination and treatment.

A medical interview revealed that the patient had about one year history of xerophthalmia and xerostomia. Her blood pressure was 150/84 mmHg, pulse rate was 64 beats per minute, and temperature was 36.3℃. Her height was 156.2 cm and body weight was 62.7 kg. Whereas sensory disturbance in the right side of the face, loss of taste on the right side of the tongue and tenderness over the parotid areas

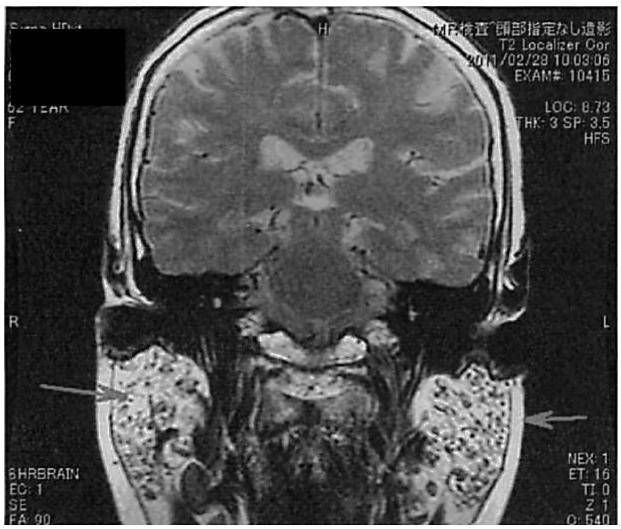

Fig. 1. MRI with gadolinium contrast revealed a mottled appearance of the parotids, which was compatible with Sjögren's syndrome.

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were found, there were no apparent asymmetries of expressive facial movements. No other abnormalities of cranial nerves were found. No neurological symptoms in the extremities were found. Atrophy of tongue papillae, cervical lymphadenomegaly, hepatosplenomegaly, arthralgia and swollen fingers were not apparent. Neither skin eruption nor erythema was found on her whole body. Schirmer's test and Saxon's test were positive（Schirmer's test demonstrated 5 mm in both eyes and Saxon's test demonstrated 0.24 g）. The results of laboratory tests on admission were : white blood cell count, 4,500/μl（neutrophils 49%, lymphocytes 44%, monocytes 3%, eosiniphils 3%, basophils 1%）; hemoglobin, 12.6 g/dl ; platelet count, 258,000/μl ; C-reactive protein（CRP）, 0.14 mg/dl ; CH50, 47.8 U/ml ; C3, 103.9 mg/dl ; blood urea nitrogen, 15 mg/dl ; serum creatinine, 0.68 mg/dl ; Na, 142 mEq/l ; K, 3.9 mEq/l ; and Cl, 109 mEq/l ; aspartate aminotransferase, 23 IU/l ; alanine aminotransferase, 7 IU/l ; lactate dehydrogenase, 235 IU/l ; prothrombin time international normalized ratio, 1.03 ; activated partial thromboplastin time, 27.2 seconds ; protein C antigen level, 106% ; protein S antigen level, 114% ; antithrombin III, 92% ; immunoglobulin G, 2,347 mg/dl ; immunoglobulin G4, 101 mg/dl ; soluble interleukin-2 receptor, 288 U/ml ; sialylated carbohydrate antigen（KL-6）(895 U/ml）. Tests for protein and occult blood in urine were both negative. The results of tests to detect autoantibodies were as follows : rheumatoid factor was positive（29.6 U/ml）, anti-citrullinated protein antibodies was negative（0.2 U/ml）, antinuclear antibody（ANA）was positive（×320, speckled pattern）, anti-Sm antibody was negative（0.4 U/ml）, anti-ribonucleoprotein antibody was negative（2.8 U/ml）, anti-SS-A-antibody was positive（127.1 U/ml）, anti-SS-B-antibody was negative（4.2 U/ml）, anti-Scl-70-antibody was negative（4.2 U/ml）, anti-double-stranded DNA antibody was negative（0.6 U/ml）. High-resolution computed tomography showed mild subpleural interstitial changes in the lower lobes. Sialography revealed apple tree lesion（Fig. 2）. The diagnosis of SS was established based on the presence of ocular symptoms, oral symptoms, ocular signs, tests for salivary gland involvement and anti-SS-A-antibody, meeting five of the six criteria for the classification of SS developed by American-European Consensus Group SS Classification Criteria in 2002[2]. She also fulfilled the Japanese SS criteria, meeting two of the four revised Japanese Criteria for SS[3]. In addition, we diagnosed

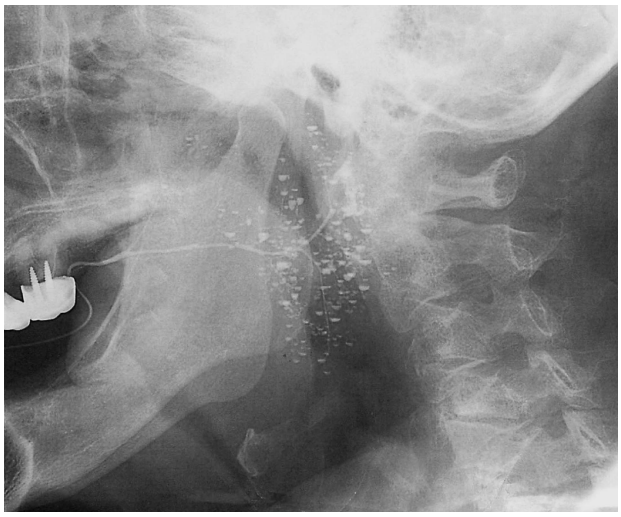

Fig. 2. Apple tree lesion revealed in sialography.

the patient with SS associated trigeminal nerve palsy on the fact that trigeminal nerve palsy occured with the occurrence with SS, and other causes of trigeminal nerve palsy were not found. Administration of 40 mg/day prednisolone（PSL）and 1.5 mg/day tacrolimus（TAC）was thus started. The patient's neurological symptoms and sicca symptoms thereafter gradually improved. However, some degree of sensory disturbance in the right side of the face persited. The dose of PSL was gradually reduced without the patient suffering an exacerbation.

Discussion

SS is a chronic autoimmune disorder in which mononuclear lymphocytes infiltrate into exocrine glands replace the glandular epitherium. Although SS is found on the development of dry eye or dry mouth,or both in many cases, SS is considered as a systemic inflammatory condition due to its various extraglandular complications including skin eruption, interstitial pneumonia, pericarditis and renal tubular acidosis.

Neurological involvement in SS is divided into two categories : central nervous system involvement in SS（CNS-SS）and peripheral nervous system involvement in SS（PNS-SS）. Clinical manifestations of CNS-SS vary among patients ; from mild headache to aseptic meningitis[4]. Massara A et al reported that 5.8% of patients with SS had CNS involvemnet[5]. On the contrary, common PNS-SS manifestations are symmetric pure-sensory or sensorimotor neuropathy. Gøransson LG et al reported that 27% of patients with SS had peripheral neuropathy[6]. Trigeminal nerve palsy is a rare manifestation of PNS-SS. Gono

Table 1. Cases of SS-associated trigeminal nerve disturbances in Japan.

Age, Sex	Unilateral or Bilateral	Treatment	Outcome
35F[11]	Bilateral	PSL 30 mg/day, satellite ganglion block, antipyretic analgesics	unchanged
45F[11]	Bilateral	Unknown	unknown
55F[11]	Bilateral	Antipyretic analgesics	improved
51F[11]	Unilateral	Steroid pulse, PSL 60 mg/day, Azathioprine 50 mg/day	repeated remission and deterioration
61F[11]	Unilateral	Antipyretic analgesics	improved
43F[11]	Unilateral	Carbamazepine	improved
52M[12]	Bilateral	Betamethasone 3 mg/day, 5 days, PSL 10→5 mg/day	improved
63F (presented case)	Unilateral	PSL 40 mg/day, TAC 1.5 mg/day	improved

Abbreviations : PSL, prednisolone : TAC, tacrolimus.

T et al reported that around 12% of PNS-SS were complicated with trigeminal neuralgia given that 2 of 17 cases of PNS-SS complicated with trigeminal neuralgia[7].

Head MRI is a useful tool for detecting central nervous system involvement in various connective tissue diseases including SS[8]. An interesting feature of the presented case is that head MRI was credited with the diagnosis of trigeminal nerve disturbance associated with SS. To the best of our knowledge, there is one report of SS complicating PNS-SS found out on head MRI findings : a 52-year-old woman revealing oculomotor palsy, diagonosed with SS on the presence of nodular appearance of parotid gland and sicca symptoms revealed on medical interview[9]. SS is occasionally found by chance in patients with unnoticed sicca symptoms, based on the positiveness of anti-SS-A-antibody. The presented case suggest head MRI may be useful for detecting parotid gland changes associated with undiagnosed SS. Therefore parotid gland changes seen on MRI related to SS should be explored in examination of cranial nerve disturbances.

Although the etiology of neurological involvement of SS is not fully clarified, it is likely that immune-mediated mechanism, such as direct mononuclear cell infiltration and vasculitis, take part in pathogenesis[10]. Trigeminal nerve involvement associated with SS has rarely been reported in Japan; there have only been seven reports of trigeminal nerve involvement in SS patients[11,12] (Table. 1). It is reported that 66% of SS patients with inflammatory vascular disease proven with biopsy complicated neurological involvement[13]. Therefore immune suppressant drugs including PSL are considered to be effective, despite there is no consensus on treatment. In this case, moderate dose of PSL gave relief from trigeminal nerve palsy and TAC was effective for reducing the dose of PSL without relapse.

In this study, we report a patient with SS found on the development of trigeminal nerve palsy found out with MRI. Although the association between SS and neurological involvement was not fully investigated, this case suggests that SS must be considered a possible cause of trigeminal nerve disturbances. The possibility of SS should be considered.

Conflict of interest statement None.

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