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Bilateral facial paralysis as a rare neurological manifestation of primary Sjögren's syndrome: case-based review

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Abstract

Primary Sjögren's syndrome (pSS) is a chronic autoimmune disorder associated with a variety of systemic presentations. Varied neurological dysfunctions of newly diagnosed adult patients with pSS have been observed in recent years. We aimed to describe a rare case of acute bilateral facial paralysis diagnosed with pSS for the first time and review the previous studies including similar cases. A 69-year-old female, who had experienced ocular and oral dryness for more than 10 years, presented with bilateral facial palsy. Her laboratory test results showed positive anti-Ro (SSA) and anti-Ro-52 antibodies. Ophthalmic examination and test of saliva secretion verified xerophthalmia and xerostomia, respectively. Other possibilities of Lyme disease, Möbius syndrome, tumor, bilateral temporal bone fracture, Guillain–Barré syndrome, central nervous system lymphoma and HIV infection were ruled out. A diagnosis of pSS associated with bilateral facial paralysis was made. The literature review revealed one article describing a similar patient. Our case was the only one suffering from acute bilateral facial palsy without other nerve involvement. The presence of such patients reveals that pSS is an underlying cause of acute bilateral facial paralysis.

Keywords Bilateral facial paralysis · Sjögren syndrome · Diagnosis · Treatment

Introduction

Primary Sjögren's syndrome (pSS) is a systemic autoimmune disorder of unknown aetiology characterized by lymphocytic infiltration of the exocrine glands, leading to xerophthalmia and xerostomia [1]. However, extraglandular involvement is not rare. Peripheral neuropathy (PN) is frequently seen in patients with rheumatic disease including pSS, which can be clinically classified into polyneuropathy, mononeuropathy, mononeuropathy multiplex, polyradiculoneuropathy and cranial neuropathy on the basis of spatial distribution patterns [2–4]. Most patients present with polyneuropathy or mononeuropathy multiplex, and cranial nerve involvement is less common [4]. In one case series of pSS-associated PN, 20 of 92 patients suffered from cranial neuropathy, of which 15 patients had trigeminal neuropathy [5].

The facial nerve is the seventh cranial nerve, which emerges from the pons and then enters the internal auditory canal of the temporal bone [6, 7]. Three branches leave the facial nerve within the temporal bone. The first, greater superficial petrosal nerve, controls tear secretion. The other two branches include one innervating the stapedius muscle within the middle ear and chorda tympani, receiving fibers of taste and sending fibers to the sublingual and submandibular glands. The facial nerve becomes extracranial at the stylomastoid foramen, where the nerve divides into the posterior auricular branch supplying sensation to the ear. Other terminal branches control muscles of facial expression.

Very few cases of peripheral facial nerve paralysis associated with pSS have been described previously [5, 8–11]. Most of them were unilaterally affected and facial palsy was often accompanied by other cranial nerve damage [5, 11, 12]. There has been no report on pure bilateral facial palsy with pSS. Here, we report a case of pSS with acute bilateral facial paralysis and without other nerve involvement.

Zhang Wei and Shi Jiaying contributed equally to this work.

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Case report

A 69-year-old female developed stabbing pain in her left ear after a cold. Two days later, she noticed symptoms of bilateral facial weakness including drooling, slurred speech and incomplete eye closure without double vision and facial numbness. The patient did not suffer from facial palsy before and the hearing of her left ear was impaired since young. She had experienced ocular and oral dryness for more than 10 years.

On admission, physical examination revealed dental caries, which had existed for several years. Neurological examination showed bilateral facial palsy of House–Brackmann scale grade 3 (Fig. 1a). Other cranial nerves were normal except decreased hearing in left ear. Her muscle strength, muscle tone, motor coordination, tendon reflexes, plantar responses and gait were normal, with intact sensation.

Clinical laboratory testing were normal, including rheumatoid factors, anti-GBM, serum immunoglobulin (IgA, IgG, IgM, IgE), anti-CCP, anti-DNA, anti-Sm and antineutrophil cytoplasmic antibodies (ANCA). Viral screening tests for HBV, HCV, and HIV were negative. Cerebrospinal fluid (CSF) cytology, CSF protein and CSF glucose were normal. Complete blood count revealed decreased leukocyte (3.0×10^9/L) and neutrophil (1.2×10^9/L). Antinuclear antibodies (ANA) at a titre of 1/40 (speckled pattern) were detected, along with increased erythrocyte sedimentation rate (25 mm/h). Anti-Ro (SSA) and anti-Ro-52 antibodies were positive. Anti-La (SSB) antibodies were negative. Complement component 3 (0.78 g/L) was decreased.

Salivary flow-rate was 0 ml within 15 min. Tear break-up time (TBUT) was 2 s, Schirmer's test result was bilaterally 0 mm in 5 min, and corneal fluorescent staining was positive. Electromyography and nerve conduction studies (EMG/NCS) in the four limbs were normal. NCS showed decreased compound muscle action potential (CMAP) amplitudes in left facial nerve and normal amplitudes in right facial nerve. Bilateral facial nerve latencies were normal with left longer than right. Brain magnetic resonance imaging (MRI) ruled out brainstem lesion.

The patient started rehabilitation after hospitalization. Methylprednisolone at a dose of 40 mg/day was given for 1 week, followed by methotrexate at a dose of 7.5 mg per week. 2 months later, bilateral facial weakness was slightly improved with oral dryness unchanged (Fig. 1b).

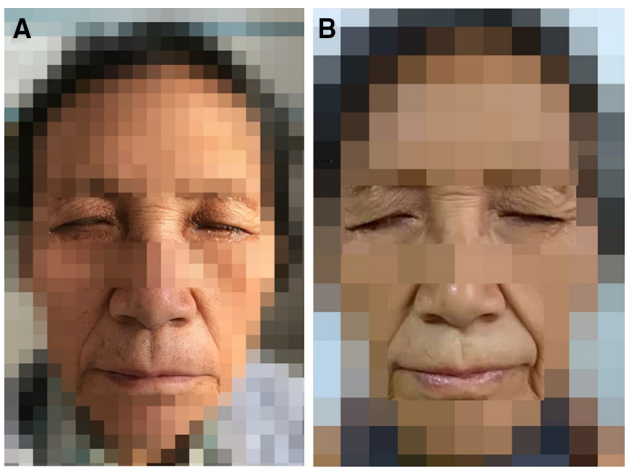

Fig. 1 The patient attempting to close her eyes at admission (**a**) and 2 months after discharge (**b**)

Search strategy

According to the published guidance on narrative reviews [13], the authors conducted a literature search in PubMed, Scopus and Web of Science up to May 6th, 2019, combining the main terms "facial paralysis", "Bell's palsy" or "facial palsy", and "Sjögren's syndrome" or "sicca syndrome". Clinical studies, case reports, and case series reporting pSS patients with acute bilateral facial paralysis published in English were included. Two authors (ZW and SJ) independently screened titles, abstracts, and full texts of all relevant articles.

Literature review and discussion

In addition to ours, we found only one detailed description of bilateral facial palsy in patients with pSS [12]. The case was a 48-year-old Japanese female who complained of bilateral facial and right forearm weakness. She had noticed dryness of the eyes and mouth for several years and the diagnosis of pSS was confirmed by laboratory tests and lip biopsy. In this case, concomitant neurological findings in the right forearm comprise mononeuritis multiplex, which is often caused by vasculitis. Vasculitis of vasa nervorum has been proposed as a pathogenic mechanism of peripheral neuropathy associated with pSS [14].

Our patient had positive autoantibody against SSA and typical pSS symptoms, including dryness of the eyes and mouth. Xerophthalmia and xerostomia was verified by ophthalmic examination and test of saliva secretion, respectively. Dental caries was also a sign of xerostomia [15]. There was no evidence of rheumatoid arthritis and systemic lupus erythematosus. Therefore, the case was diagnosed as pSS [16]. Although facial palsy itself can lead to xerophthalmia, other manifestations of our case excluded the possibility. Symptoms including ear pain, taste change, hearing change or deceased tearing can also present in patients with facial palsy, which help to determine the location of facial

nerve lesion. Normal taste without hyperacusis in the present patient indicated that nerve damage was distal to stylomastoid foramen and the concomitant xerophthalmia resulted from pSS (not from facial palsy).

In addition, the present patient exhibited acute bilateral facial paralysis. Bilateral facial paralysis is a rare condition, with an occurrence of 0.3–2% in facial palsy cases [17]. In a case series, Bell's palsy was the most common diagnosis among patients with bilateral facial paralysis [17]. However, all cases presented in an asynchronous fashion. Other causes of simultaneous bilateral facial paralysis were also ruled out, including Lyme disease, Möbius syndrome, tumor, bilateral temporal bone fracture, Guillain–Barré syndrome, central nervous system lymphoma and HIV infection [17]. A final diagnosis of pSS associated with bilateral facial paralysis was made.

The prevalence of PN in pSS patients ranges from 10 to 60% [18]. Several forms of peripheral nerve dysfunction occur. In 1 case series, 10 of 46 patients (21.7%) with pSS had clinical and electrophysiological signs of PN, including 9 cases with polyneuropathy, and 1 with mononeuritis multiplex [19]. Less commonly, cranial nerves are affected with a predisposition to trigeminal nerve. In pSS, facial nerve involvement is rare [8]. However, facial paralysis occasionally is the presenting symptom of pSS [8]. For most cases, facial paralysis associated with pSS was unilateral and often presented as multiple cranial neuropathy or mononeuritis multiplex [8]. To our knowledge, the present patient is the first description of a case with pSS suffering from bilateral facial paralysis without other nerve involvement.

Conventional disease-modifying therapies do not appear to be effective for xerophthalmia and xerostomia in pSS [1]. Similarly, the course of pSS-associated PN was slowly progressive or stable, except for a few patients who may improve with immune therapies [19–21]. The prognosis of pSS-associated facial paralysis without other nerve involvement was not well described in the literature [5, 22–24]. In other cases, facial paralysis presenting as multiple cranial neuropathy were generally responsive to steroid treatment [11]. Recurrent episodes of facial weakness and subsequent facial spasm can be seen occasionally [9]. Our patient received methotrexate treatment for 2 months and the facial weakness did not recover, which did not support the diagnosis of Bell's palsy. For most patients with Bell's palsy, important improvement occurs within 3 weeks [25]. In addition, it is worth noting that long-term methotrexate therapy can cause cranial or optic neuropathy [26, 27]. Methotrexate-induced neuropathy needs to be considered in the differential diagnosis of facial palsy, if neuropathy presents after methotrexate treatment.

In conclusion, bilateral facial paralysis is a rare clinical entity caused by myriad disparate conditions [17]. This case indicates that pSS is an underlying cause of acute bilateral facial paralysis and serves to remind the physicians to take pSS in consideration when seeing a patient complaining of bilateral facial weakness without other neurological manifestations.

Author contributions ZW: writing of manuscript including editing and revision and the literature review at all stages of its production, and clinical management of patient. SJ: writing of manuscript including editing and revision and the literature review at all stages of its production, and clinical management of patient. GJ: manuscript editing and revision at all stages of production, final approval of manuscript, and clinical management of patient.

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Compliance with ethical standards

Conflict of interest All the authors declare no conflict of interest.

Ethical approval All the procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed consent Written informed consent was obtained from this patient prior to submission of this article for consideration as a case-based review.

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