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Unilateral hypertrophic pachymeningitis in Sjögren syndrome

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Abstract

Hypertrophic pachymeningitis is a diffuse cranial inflammatory disease that causes thickening of the dura mater. It is associated with underlying conditions such as autoimmune disease, infection, and malignancy. Patients with Sjögren syndrome can develop extraglandular manifestations, but the manifestation of hypertrophic pachymeningitis in Sjögren syndrome is very rare. Here we report a case of Sjögren syndrome that is associated with hypertrophic pachymeningitis with unilateral involvement.

Keywords: Hypertrophic pachymeningitis, Sjögren syndrome, dry eye, dry mouth

INTRODUCTION

Hypertrophic pachymeningitis (HP) is a rare disease characterized by localized or diffuse thickening of the cranial or spinal dura mater.[1,2] It may occur in association with various conditions such as autoimmune disease, malignancy, and infection.[1,2] In an autoimmune disease, HP mainly appears diffusely on bilateral dura mater, and is rarely reported to manifest only on one side.[1,2] Herein we report HP in a patient with Sjögren syndrome, with an unusual unilateral involvement.

CASE REPORT

A 57-year old woman developed a generalized headache and paresthesia on hand and foot for a few weeks. Physical examination showed mild dysarthria, hypesthesia and paresthesia on both hands and feet, and mild proximal weakness on upper and lower extremities. Deep tendon reflexes were decreased in lower limbs. She had been suffering from joint pain in hands for three years and was suspected to have undifferentiated connective tissue disease.

Magnetic resonance imaging of the head showed pachymeningeal thickening in left cerebral convexity (Figure 1A), consistent with pachymeningitis, and diffuse enlargement of the lacrimal gland and parotid gland (Figure 1B). The cerebrospinal fluid analysis showed normal pressure (100 mmH_2O), absent leukocytes response, increased protein of 225.2 mg/dL, and slightly low glucose 42 mg/dL (serum glucose level was 97 mg/dL). Nerve conduction studies showed axonal sensorimotor polyneuropathy in all extremities. Taking further history after hospitalization, she had been suffering from dry eyes and had been using artificial tears for several years. Schirmer test and tear breakup time test were abnormal. Salivary gland scans showed a severe decrease of uptake (Figure 2A). Serologic studies revealed positive anti-Ro/SSA and La/SSB antibodies. A diagnosis of Sjögren syndrome was made. Lip biopsy showed lymphocytic and plasmacytic infiltrates in the minor salivary gland (Figure 2B). Intravenous methylprednisolone (1000 mg/day) was administered for seven days, followed by oral prednisolone (1 mg/kg) as treatment for the diagnosis of SS-related HP. Subsequently, the steroid was gradually reduced, and after several months, the patient was treated with prednisolone 2.5 mg/day, tacrolimus 1 mg/day, and hydroxychloroquine 200 mg/day. After the steroid pulse therapy, her headache moderately improved. Proximal weakness and distal limb paresthesia improved gradually over the next few months. Nerve conduction study of the upper and lower limb nerves showed mild improvement at three months.

DISCUSSION

HP is a rare disease characterized by localized or diffuse thickening of cranial or spinal dura mater.[1,2] Various conditions have been associated with HP, including malignancy, infection, autoimmune

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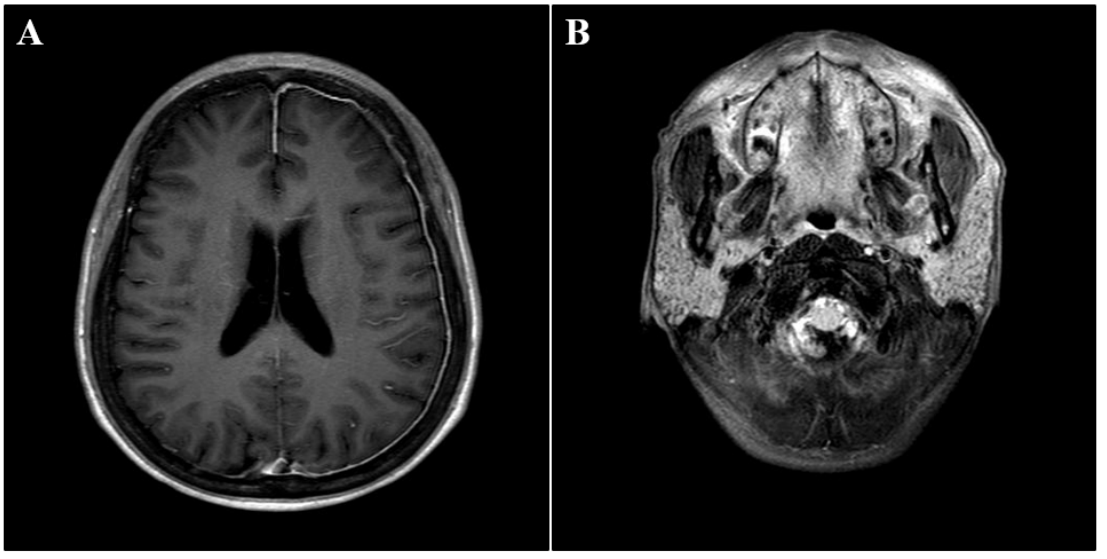

Figure 1. Patient's Magnetic Resonance Imaging (MRI). (A) MRI with gadolinium enhancement reveal hypertrophic enhancement of the pachymeninges in the left cerebral convex. (B) T2-weighted image showing diffuse enlargement of the lacrimal and parotid glands.

disease including sarcoidosis, relapsing polychondritis, Sjögren syndrome, IgG4-related disease, and intracranial hypotension; with some cases remaining unknown.[1,2] HP in autoimmune disease mainly appears diffusely on bilateral dura mater, it is rare to manifest only on one side.[3,4] Furthermore, the mechanism of its occurrence has not yet been sufficiently understood.

Sjögren's syndrome is a systemic chronic autoimmune disorder of the exocrine glands associated with lymphocytic infiltration.[3] It mainly involves the parotid and lacrimal glands that result in distinct symptoms of dry eyes, dry mouth, and swelling of the major salivary glands. Patients with Sjögren syndrome can develop extraglandular manifestations, such as joint, muscle, pulmonary, kidney, skin, nervous system involvement. Neurologic manifestations of Sjögren syndrome are reported to range from 8% to 49%.[3] Among the neurological manifestation, sensory polyneuropathies are most common. Other manifestations include cranial neuropathy, radiculopathy, mononeuritis muliplex, and other central nervous system involvement.[3] In the central nervous system, HP occurs infrequently[4], especially as unilateral manifestation.

In summary, we report here a patient who has sicca symptoms, enlargement of the lacrimal

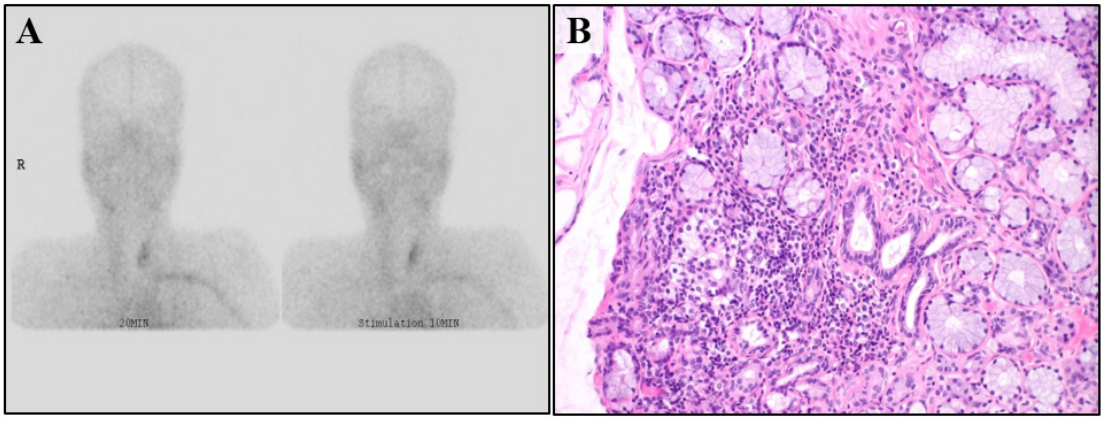

Figure 2. Findings of salivary scans and lip biopsy. (A) Salivary gland scans show severely decreased uptake in both the parotid and submandibular glands. (B) Lip biopsy show moderate to severe lymphocytic and plasmacytic infiltrates in the minor salivary gland.

gland, severely impaired salivary function, peripheral neuropathy confirmed by NCS, and positive Anti-Ro, La antibody, and histology are supportive of Sjögren syndrome. Our patient also has an unusual unilateral HP.

DISCLOSURE

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Conflicts of interest: None

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