Case report

Sjögren's syndrome presenting with recurrent strokes

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Introduction

Sjögren's syndrome (SS) is an autoimmune rheumatic disease characterized by lymphocytic infiltration and destruction of exocrine glands. The spectrum of clinical manifestations of SS is wide, ranging from mucosal dryness to extra glandular systemic manifestations. Involvement of the central nervous system (CNS) in patients with SS is variable and the peripheral nervous system involvement is well recognized. We report a patient with SS presenting with strokes as the first clinical manifestation.

Case report

A 49-year-old female was admitted with acute onset vomiting, vertigo and difficulty in walking for three days. She also had difficulty in swallowing and worsening weakness of her right upper limb and lower limb. One year ago she had presented with sudden onset right hemiparesis and vertigo. MRI brain at that time had revealed multiple lacunar infarctions in left corona radiata, right cerebellar hemisphere and brainstem. She was given physiotherapy and was started on aspirin, dipyridamole and atorvastatin. She made a good recovery with mild residual weakness of right side but had defaulted follow up after three months. Her medical history was otherwise unremarkable.

On examination she had right hemiparesis with bilateral cerebellar signs, left Horner's syndrome and left seventh (lower motor neurone), ninth and tenth cranial nerve palsies. Cardiovascular system examination was normal with a blood pressure of 120/80 and regular rhythm. There were no neck bruits.

MRI brain revealed acute and old multiple lacunar infarctions in basal ganglia bilaterally, left corona radiata, pons (Figure 1) and cerebellum. MRA of neck and intracranial arteries was normal. MRI further revealed multiple cystic lesions in both parotid glands possibly due to sialectasis (Figure 2). Subsequent inquiry revealed the patient to have dry mouth (xerostomia) and dry eyes for one year.

Routine stroke work up including lipid profile, fasting blood glucose, transthoracic and transoesophageal echocardiogram, carotid and vertebral duplex studies were normal.

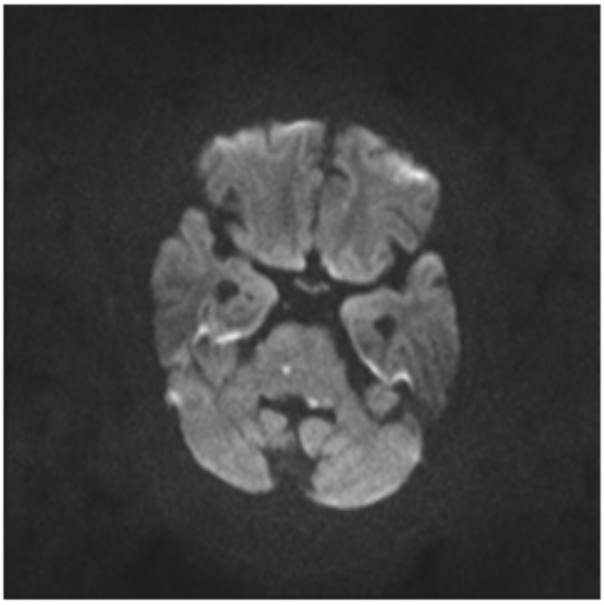

Figure 1. MRI brain indicating acute infarction in pons.

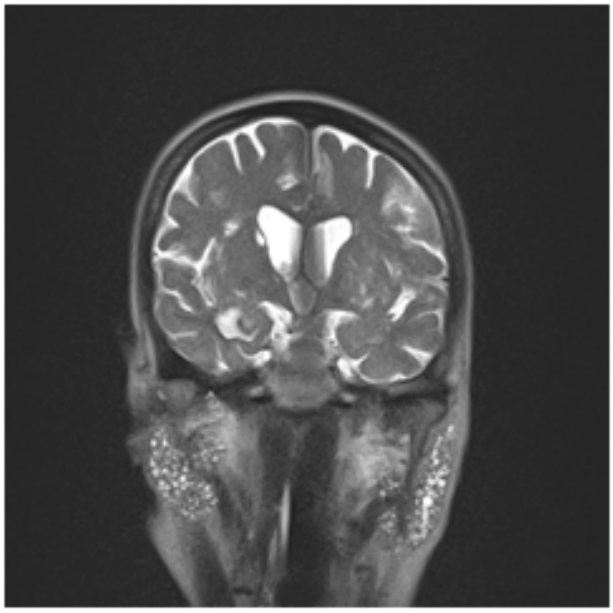

Figure 2. MRI brain indicating parotid gland sialectasis.

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Basic laboratory testing was normal except elevated ESR of 95mm. Vasculitic screen revealed positive rheumatoid factor and anti SS-A antibodies. ANA, anti DS DNA, anti SS-B, anti cardiolipin, VDRL and HIV antibodies and C-ANCA, P-ANCA were negative and C3, C4 and LDH levels were normal.

Lip biopsy showed marked lymphocytic infiltrate, acinar atrophy and lymphoepithelial lesion formation compatible with SS (Figure 3). Cerebrospinal fluid (CSF) had a high protein content of 125mg/dl but was acellular.

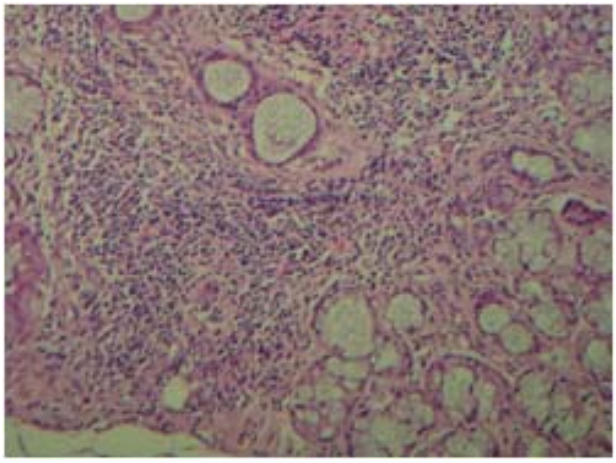

Figure 3. Marked lymphocytic infiltration in lip biopsy.

Diagnosis of SS was made and she was given intravenous immunoglobulin (IVIG) 0.4 mg/kg for 5 days, intravenous methyl prednisolone 1g daily for 3 days followed by oral prednisolone and azathioprine together with physiotherapy. During the hospital stay of three weeks she made a good recovery and barthel index improved from 40 to 80.

Discussion

SS is the second commonest autoimmune rheumatic disease after rheumatoid arthritis, affecting 3-4% of the adult population. Spectrum of CNS SS include focal lesions presenting as motor and/or sensory deficit, aphasia, dysarthria, seizures, cerebellar syndromes and non-focal lesions presenting as encephalopathy, seizures, aseptic meningitis, dementia, and psychiatric disorders. Spinal cord lesions can give rise to transverse myelitis, chronic progressive myelopathy, neurogenic bladder and Brown Sequard syndrome. CNS SS may mimic multiple sclerosis and present as a relapsing-remitting or primary progressive syndrome with increased IgG synthesis and oligoclonal bands in the CSF[1]. Immunologically mediated mechanisms, mainly vasculitis is thought to play a role in CNS manifestations of SS. It has also been found that anti SS-A positivity to be associated with severe disease with frank necrotizing angiitis[2]. Histopathologically there is small vessel mononuclear inflammatory and ischaemic/ haemorrhagic vasculopathy.

Recurrence of stroke in a patient at forty nine years of age in the presence of uncontrolled multiple vascular risk factors may not be uncommon. However, in situations where vascular risk factors are absent one need to be vigilant and should search for uncommon causes. While raised ESR directed towards a systemic pathology, positive Rheumatoid factor and anti SS-A antibodies along with oral and ocular symptoms and lip biopsy findings confirmed Sjögren's syndrome in the reported patient[3]. For diagnosis of CNS SS, MRI brain is nonspecific and CSF commonly shows lymphocytosis, a raised IgG index and oligoclonal bands. Treatment of CNS SS remains largely empirical. There are no randomized clinical trials but intravenous pulses of corticosteroids, cyclophosphamide, plasmapheresis and IVIG have been reported to be beneficial[4,5].

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