

A Rare Central Nervous System Finding of Sjögren's Syndrome: Unilateral Fronto-Parietal Cortical Atrophy

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Abstract: Sjögren's Syndrome is a common, chronic autoimmune disease that typically affects salivary and lacrimal glands. The immune response damages the exocrine glands and results in the typical symptoms of dry eye and dry mouth. There are other various systemic and focal symptoms, as well. Vasculitis is one of the most common extraglandular manifestations of the disease and skin vasculitis, and leukocytoclastic vasculitis is the most frequent ones. However, some studies revealed an acute necrotizing arteritis of medium-sized vessels similar to polyarteritis nodosa may be seen in Sjögren's Syndrome. MRI is the best diagnostic tool for detecting the findings of cerebral vasculitis and nonspecific, T2-weighted hyperintensities affecting subcortical and periventricular white matter are the most common finding. In the current report, we present MRI findings of unilateral fronto-parietal cortical atrophy, a rare finding in a case with long-standing Sjögren's syndrome.

Keywords: Sjögren's Syndrome, magnetic resonance imaging, cortical atrophy, central nervous system.

INTRODUCTION

Sjögren's syndrome (SS) is a chronic disorder that is characterized by immune attack on the exocrine glands with resultant dry eyes and dry mouth.

Patients may usually have problems in other organs and systems, and one of the most common extraglandular manifestations is vasculitis. The most common vasculitic manifestations are skin vasculitis with palpable purpura and leukocytoclastic vasculitis. Additionally, similar to polyarteritis nodosa (PAN), necrotizing vasculitis of medium-sized vessels may be seen in SS [1, 2]. Central nervous system (CNS) involvement may be seen and commonly mimics multiple sclerosis [3]. The cerebral vasculitis of SS mimics PAN and is characterized by vessel stenoses and dilatations [4].

Magnetic resonance imaging (MRI) is more sensitive than computed tomography in revealing the anatomic anomalies associated with the CNS findings of SS. The most common MRI findings is multiple areas of increased signal intensity on T2-weighted images predominantly affecting subcortical and periventricular white matter [5-7].

In the current report, we present MRI findings of unilateral fronto-parietal cortical atrophy, a rare finding in a case with Sjögren's syndrome.

CASE REPORT

A 64-year-old female was admitted to our department for brain MRI examination for the further evaluation of her headache. She had a 30-year history of SS and was on medication. She had no history of another systemic disease, stroke or cerebral disease. Her physical examination, including the neurologic exam and laboratory findings were within the normal limits.

Brain MRI revealed cortical atrophy on the right frontal and parietal lobes. Multiple, nonspecific T2-weighted and Fluid Attenuated Inversion Recovery (FLAIR) hyperintensities were also seen in the subcortical and deep white matter of both frontal and parietal lobes, more pronounced on the right side (Figure 1 and 2). The imaging of the other brain parts was normal.

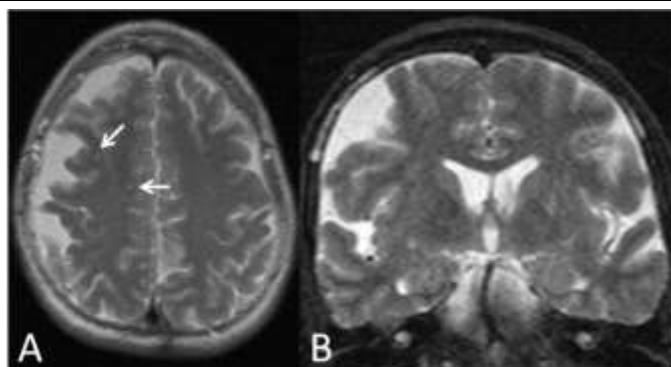


Fig-1: Axial (A) and coronal (B) T2-weighted MR images demonstrate prominent cortical atrophy in the right frontal and parietal lobes. A few hyperintense foci are also seen in the white matter of the right hemisphere (arrows)

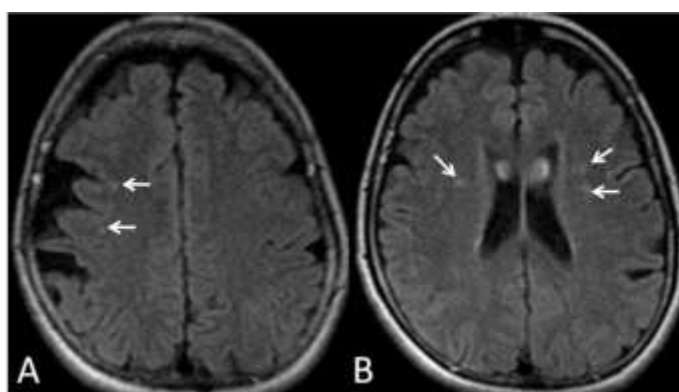


Fig-2: Axial Fluid Attenuated Inversion Recovery (FLAIR) images from two different levels (A and B) show nonspecific hyperintense foci affecting bilateral fronto-parietal white matter (arrows). On A, right fronto-parietal cortical atrophy is seen, as well

DISCUSSION

SS is a relatively common autoimmune disease, affecting 2-3% of the adult population. The main pathology contains lymphocytic infiltration and destruction of the exocrine glands. The salivary and lacrimal glands are the most commonly affected glands and dry mouth and dry eyes are the leading symptoms. Other exocrine structures such as pancreas, gastrointestinal tract, and bronchial tree may be affected. SS may be primary or in association with other connective tissue diseases (secondary). Clinical spectrum is wide and ranging from mucosal dryness to more systemic findings mainly affecting renal, pulmonary, musculoskeletal, and vascular [7, 8]. Involvement of CNS in SS is a serious controversy in medical literature and there is no consensus on even the definition of this entity. Additionally, diagnostic criteria of SS are not uniform, and there is no consensus on the inclusion of the mild symptoms such as headache or mood problems, as well [7]. The etiology of the CNS involvement in SS is not clear. However, this may be immunologically mediated [7]. Histopathological examination of brain tissue from some patients showed a small vessel mononuclear inflammatory and ischaemic/haemorrhagic vasculopathy [9].

MRI is the most useful diagnostic tool in showing the CNS involvement of SS and increased signal intensity predominantly affecting subcortical and periventricular white matter on T2-weighted images is the most common finding [5-7]. Those may be caused by edema, infarction, ischemia, or demyelination. However, MRI abnormalities may also be associated with arteriosclerosis, myelin pallor, dilated perivascular spaces, and periventricular gliosis [5]. Belin *et al.* [10] showed no abnormal brain MRI findings in their study group including 19 patients with SS, all of whom had neuropsychological abnormalities mostly frontal lobe syndrome and memory problems. On the other hand, Alexander *et al.* [11] demonstrated focal CNS findings on brain MRI in their 5 of 8 patients with psychiatric or cognitive dysfunction. The situation in patients with SS without clinical CNS disease is also not clear. Alexander *et al.* [9] reported such patients have a very low frequency of abnormal MRI examinations. Pierot *et al.* [12] showed abnormalities on brain MRI in their 9 of 15 patients, involving mainly punctate areas of high signal in the basal ganglia and the white matter of the basal ganglia. However, high signal intensities on brain MRIs are very common in normal population and especially increase with age. Herein, it should be underlined that evaluating the findings mentioned above

in SS population and linking all those findings to the disease itself is rather difficult [13].

In SS, cerebral angiography may be performed for excluding the other CNS diseases such as arteriovenous malformations, congenital aneurysms, and other vascular abnormalities and cerebrovascular disease. Alexander *et al.* [9] found angiographic findings suggesting small vessel vasculitis, such as stenosis, dilatation, or occlusion of small cerebral blood vessels up to 45% in their study group consisting highly selected SS patients with active CNS disease.

CONCLUSION

We found T2-weighted and FLAIR hyperintensities in the subcortical and deep white matter of both frontal and parietal lobes, more pronounced on the right side and prominent cortical atrophy on the right frontal and parietal lobes in our case with 30 year-history of SS. There was no history of another systemic disease and stroke or any other cerebral disease can affect brain tissue in our case. To the best of our knowledge, this is the first paper reporting unilateral focal fronto-parietal cortical atrophy in a case with SS.

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