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Case Report

Beyond sicca: A rare case of neuro- sjogren's presenting as recurrent cerebral infarctions and syringomyelia

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Abstract

Background: Neurological involvement in primary Sjögren's syndrome (pSS) is rare but can include serious central nervous system (CNS) manifestations such as recurrent strokes and myelopathy.
Case Presentation: A 27-year-old woman presented with progressive quadriparesis and dysarthria. Six months earlier, she had recovered from a similar episode of limb weakness. Neurological examination revealed spastic quadriparesis and right 12th cranial nerve palsy. Brain MRI showed chronic infarcts in the bilateral fronto-parietal cortex, right occipital lobe, and basal ganglia, with spinal MRI revealing diffuse cord thinning and a holocord syrinx. Carotid artery Doppler was normal. Autoimmune workup revealed ANA (1:320, speckled) and anti-Ro/SSA positivity. Labial gland biopsy confirmed pSS. CNS vasculitis secondary to pSS was diagnosed, and treatment included IV methylprednisolone followed by weekly rituximab.
Conclusion: This case underscores a rare but severe neurological manifestation of pSS with recurrent infarctions and myelopathy, highlighting the importance of early recognition and immunosuppressive treatment in young patients with unexplained CNS disease

Keywords: Fronto-temporo, ENA, Syringomyelia

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1. Introduction

Primary Sjögren's syndrome is an autoimmune connective tissue disorder that primarily affects the exocrine glands, characterised by lymphocytic infiltration of the glands thus leading to their structural damage and functional impairment. Patients commonly present with complaints of dry mouth and eyes. Diagnosis is established using a combination of subjective and objective measures as determined by 2016 ACR/EULAR classification criteria. Beyond glandular involvement, it can also affect various other internal organs. Neurological complications are among the most frequent extra glandular manifestations. The prevalence of neurological involvement varies widely between reports, ranging from 0% to 67.5%.[1] The most common neurological manifestation is peripheral neuropathy, particularly sensory polyneuropathy, while central nervous system involvement is relatively uncommon. Additionally, there have been reports of cranial nerve involvement and autonomic nervous system dysfunction.

2. Case Discussion

A 27-year-old married woman, with uneventful obstetric history and no prior comorbidities initially presented with right-sided hemiparesis. MRI of the brain revealed a subacute infarct in the left fronto-temporo-parietal region and chronic infarcts in the right fronto-temporo-parietal region. She was treated symptomatically and showed complete neurological recovery within a month.

Six months later, she presented again with a five-day history of progressive weakness affecting all four limbs, associated with numbness over upper and lower limbs and history of dysarthria and difficulty in forming food bolus in

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mouth. There was no history suggestive of autonomic, or systemic features such as rash, joint pain, oral ulcers, alopecia, sicca symptoms, or thrombotic tendencies. There was also no family history of recurrent cerebrovascular events.

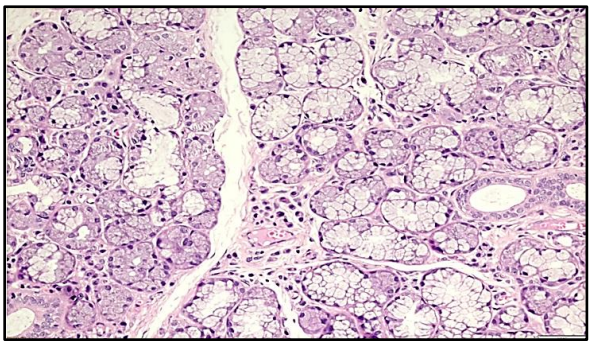

Figure 1: H & E stain 40 X image of lip biopsy shows fragment of minor salivary gland with moderate lymphoplasmacytic infiltrate in interlobular septa around the ducts and acini

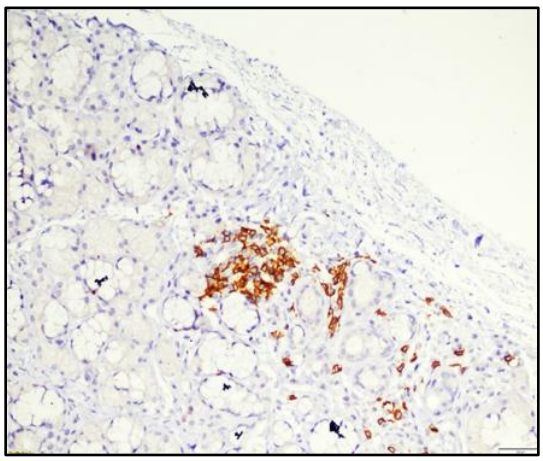

Figure 2: 20 x image showing IHC positivity for CD 20

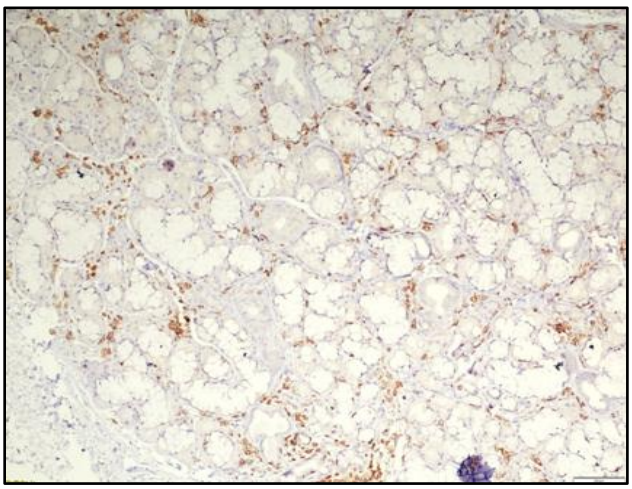

Figure 3: 20 x image showing IHC positivity for CD 45

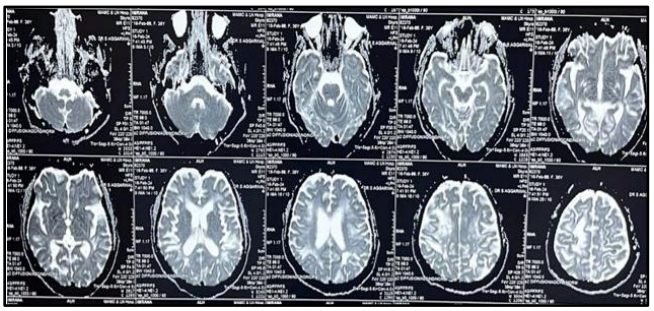

Figure 4: MRI Brain axial T2 weighted imaging showing hyper intense lesions inn peri ventricular and deep white matter

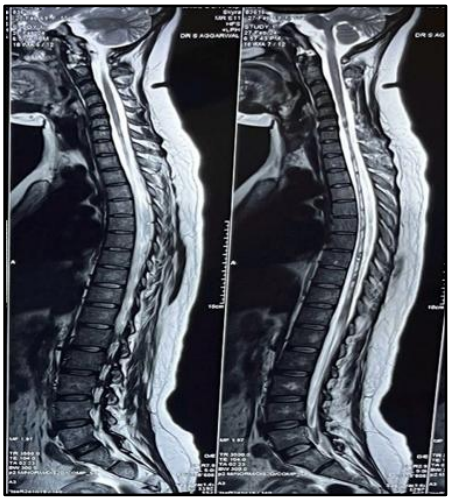

Figure 5: MRI spine: diffuse thinning of the entire spinal cord with holocord syrinx formation in dorsal and lumbar cord.

On examination, she was conscious, oriented, and hemodynamically stable and exhibited emotional incontinence. Hypertonia was present in all limbs, with muscle strength graded at 4/5 (MRC scale) in the upper limbs and 3/5 in the lower limbs. Deep tendon reflexes were brisk and plantar reflexes were bilaterally extensor. Decreased perception of superficial touch, pain, temperature was present over upper and lower limbs. Cranial nerve examination revealed a right lower motor neuron palsy of the twelfth nerve, while cerebellar functions remained intact. There were no signs of meningeal irritation. Cardiovascular, Respiratory, Gastrointestinal system examination were normal.

Complete blood picture, liver and renal function tests were normal. ESR and CRP were elevated: 60 mm/hr and 78 mg/L respectively. Montoux test was negative. HIV, hepatitis B,C were non-reactive. Initial non-contrast CT of the head showed chronic infarcts in the bilateral fronto-parietal lobes, right occipital lobe, and right basal ganglia. Given the young age and presence of multifocal infarcts, an extensive evaluation for hypercoagulable states and vasculitis was undertaken. Tests including lipid profile, serum homocysteine, protein C and S, antithrombin III, Factor V

Leiden mutation were done which did not suggest of any abnormality .The antiphospholipid antibody panel was also negative. S. Calcium, S. ACE levels as well as chest X-ray which were done to look for evidence of sarcoidosis were normal.

Autoimmune workup revealed ANA speckled pattern at 1:320 titers, and ENA profiling showed anti-Ro/SSA positivity. MRI Brain demonstrated white matter changes involving the periventricular and subcortical U-fibers with FLAIR suppression and surrounding gliosis (**Figure 4**). MRI spine showed diffuse thinning and signal alteration of the spinal cord with a holocord syrinx, predominantly in the dorsal and lumbar segments (**Figure 5**) —likely sequelae of prior insult. Bilateral carotid Doppler studies showed no evidence of atherosclerosis. (**Figure 1**, **Figure 2**, **Figure 3**) was consistent with focal lymphocytic sialadenitis, confirming the diagnosis of Sjögren's syndrome. For histopathological diagnosis, focal lymphocytic sialadenitis is the histological hallmark: 1 or more dense aggregates with 50 or more lymphocytes, usually in perivascular or periductal areas. The differentials are--- focal lymphocytic sialadenitis, Sialosis, nonspecific chronic sialadenitis or chronic sclerosing sialadenitis.[2]

In view of recurrent ischemic insults and imaging features suggestive of vasculitic involvement of cerebral and spinal vessels, the patient was diagnosed with Sjögren's syndrome-associated secondary CNS vasculitis. She was initiated on high-dose pulse corticosteroid therapy, followed by maintenance steroids and weekly rituximab infusions. Although no new cerebrovascular events occurred following immunosuppressive therapy, the patient's motor function showed limited recovery, likely due to irreversible spinal cord damage from prior insults.

3. Discussion

Neurological involvement in pSS is diverse and may affect both the peripheral and central nervous systems. Neurological symptoms may precede the classic sicca symptoms or immunological diagnosis by several years, necessitating high clinical suspicion in patients with unexplained neurological deficits. The spectrum of CNS manifestations includes cognitive impairment, psychiatric symptoms like anxiety, depression, movement disorder like ataxia and tremors, seizures, headache, white matter lesions, transverse myelitis, aseptic meningitis, and multiple sclerosis-like presentations.

The underlying pathogenesis of neurological involvement in pSS remains incompletely understood but is thought to involve autoimmune-mediated vasculitis and perivascular lymphocytic infiltration, particularly around the dorsal root ganglia. T lymphocytes, dendritic cells, and various cytokines such as IL-1β, IL-6, IL-2, and TNF-α are implicated in neural damage.[3] B-cell dysfunction is a key feature of pSS. The excessive B-cell activating factor (BAFF) production is a pathological mechanism in pSS. Nonlymphoid cells, such as astrocytes, can express BAFF and trigger CNS manifestations. In patients with primary Sjögren's syndrome who have central nervous system involvement, brain tissue analysis has shown evidence of inflammation and vascular damage affecting small vessels, including mononuclear cell infiltration and ischemic-hemorrhagic changes. Furthermore, cerebral angiography often reveals abnormalities indicative of small-vessel vasculitis, supporting the theory that vascular injury contributes significantly to the neurological complications seen in this condition.[4-5]

Spinal cord involvement, most notably in the form of acute transverse myelitis, represents one of the more severe CNS complications. The exact mechanism of syringomyelia in Sjögren's syndrome in unclear. It is a consequence of inflammatory demyelinating pathology. Given the limited literature available further research is necessary to elucidate the relationship between syringomyelia and Sjögren's syndrome

Diagnosing SS in patients with neurological manifestations can be challenging, especially in the absence of sicca symptoms. In such cases, ancillary investigations such as labial salivary gland biopsy and serological markers (particularly anti-Ro/SSA antibodies) play a critical role. Imaging modalities like MRI can reveal characteristic glandular changes and CNS lesions that support the diagnosis. The ACR–EULAR classification criteria.[6] provide a structured framework to guide diagnosis, incorporating objective tests such as Schirmer's test, ocular staining, and unstimulated salivary flow, in conjunction with serologic and histopathologic findings.

Management of neurological involvement in SS generally includes immunosuppressive therapy. High-dose corticosteroids remain the cornerstone for acute exacerbations. In cases with CNS manifestations, additional immunosuppressant's such as cyclophosphamide, methotrexate, azathioprine, and biologics like rituximab[7] are often employed. Rituximab, in particular, has shown promise in achieving disease stabilization and preventing further neurological deterioration. Close monitoring and long-term immunomodulatory therapy are essential to prevent relapses and irreversible neurological deficits.

Several studies and case reports have highlighted the clinical heterogeneity and varying outcomes of central nervous system (CNS) involvement in primary Sjögren's syndrome (pSS). Chen et al.[8] described a 33-year-old male with subacute speech and gait disturbances and gadolinium-

enhancing punctate MRI lesions, who achieved full recovery following corticosteroid therapy—demonstrating the potential reversibility of early inflammatory CNS involvement. Afzali et al.[9] reported on 19 patients where myelitis and optic neuritis were predominant. Although corticosteroids were used, most patients experienced incomplete recovery, indicating the chronic and disabling potential of CNS involvement in pSS. Jaskolska et al.[10] reported a male with encephalopathy and infarct-like lesions who responded well to IVIG, cyclophosphamide, and azathioprine. XiangLing Li[11] described recurrent demyelination with syringomyelia in a patient with pSS and NMOSD overlap. Bragoni et al presented a patient with transient ischemic symptoms and MCA territory infarct onMRI who remained relapse-free with steroid and methotrexate therapy. Li, Jia et al presented a 66-year-old female with no atherosclerotic risk factors who presented with recurrent cerebral infarctions and intracranial arterial stenosis. CNS vasculitis was suspected and workup was suggestive of Sjögren's syndrome. Patient was started on immunosupressive therapy, but there was no improvement in symptoms although further progression of disease was prevented.[12]

A retrospective study conducted by Wei Fan et al in a hospital at china found that among the 412 patient with primary sjogrenes syndrome. The prevalence of central nervous system involvement in the studied pSS patients was 10.2%, with 31.3% of patients having neurological manifestations as the initial symptom. The manifestations of hemiparesis (35.7%), paraparesis (28.6%), dysphonia (31.0%), blurred vision (21.4%), and dysfunctional proprioception (23.8%) were more common in the pSS-CNS patients. Cerebral infarction (57.1%), demyelination (31.0%), myelitis (23.8%), and angiostenosis (21.4%) were most often found on MRI or CT scan imaging in the pSS-CNS patients.[13]

These studies and case reports collectively underscore the clinical diversity, diagnostic challenges, and variable outcomes associated with central nervous system involvement in primary Sjögren's syndrome.

Together, these findings emphasize the diverse neurological manifestations in pSS, ranging from reversible inflammatory events to chronic disabling disease, and highlight the importance of early recognition and individualized immunosuppressive therapy.

4. Conclusion

This case highlights the importance of considering autoimmune etiologies such as Sjögren's syndrome in young patients presenting with recurrent stroke-like episodes and spinal cord pathology, even in the absence of classic sicca symptoms. Given the potential for serious neurologicalsequelae, early recognition and aggressive treatment of NeuroSjogren's is vital.

5. Source of Funding

None.

6. Conflict of Interest

None.

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