NEUROSJOGREN'S SYNDROME – A REPORT OF THREE CASES

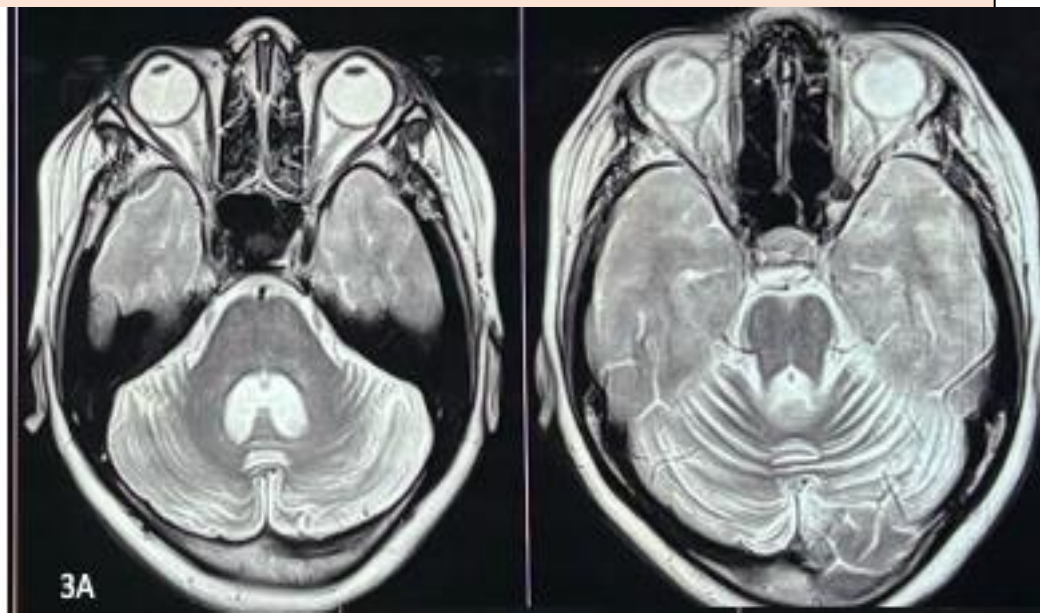
BACKGROUND:

Sjogren's syndrome(SS) is a chronic autoimmune disease characterized by lymphocytic infiltration and inflammation of exocrine glands. SS is notorious for its multifaceted systemic involvement including the nervous system. We report three cases of Neuro-Sjogren's syndrome(NSS) highlighting the diverse clinical manifestations.

CASE I

A young female presented with progressive quadriparesis and dysarthria for 4 weeks. She had episodes of dryness of mouth for past few years.

Examination revealed scanning speech, spastic quadriperesis and cerebellar ataxia. MRI brain showed diffuse cerebellar atrophy (3A). CSF analysis, autoimmune and para-neoplastic panel were unremarkable. ANA immunoblot revealed strong positive anti-Ro antibodies with focus score>1 on labial biopsy. A diagnosis of NSS with spastic ataxia was considered and she received IV pulse followed by Rituximab.



CASE II

A 40-year-old female presented with one episode of seizure, right hemiparesis followed by progressive cognitive decline for 3 months. On examination MMSE-8/30 with right spastic hemiparesis. Serum potassium was low (2.8mEq/L), and ABG showed non-anion gap metabolic acidosis. MRI brain revealed multiple chronic infarcts (1A,1B) and diffuse narrowing of bilateral MCA, ACA PCA and VA (1C) with enhancement along ICA (1D) suggestive of vasculitis. ANA immunoblot showed strong positive anti-Ro and anti–La antibodies suggestive of NSS. She received IV pulse MPS and Rituximab.

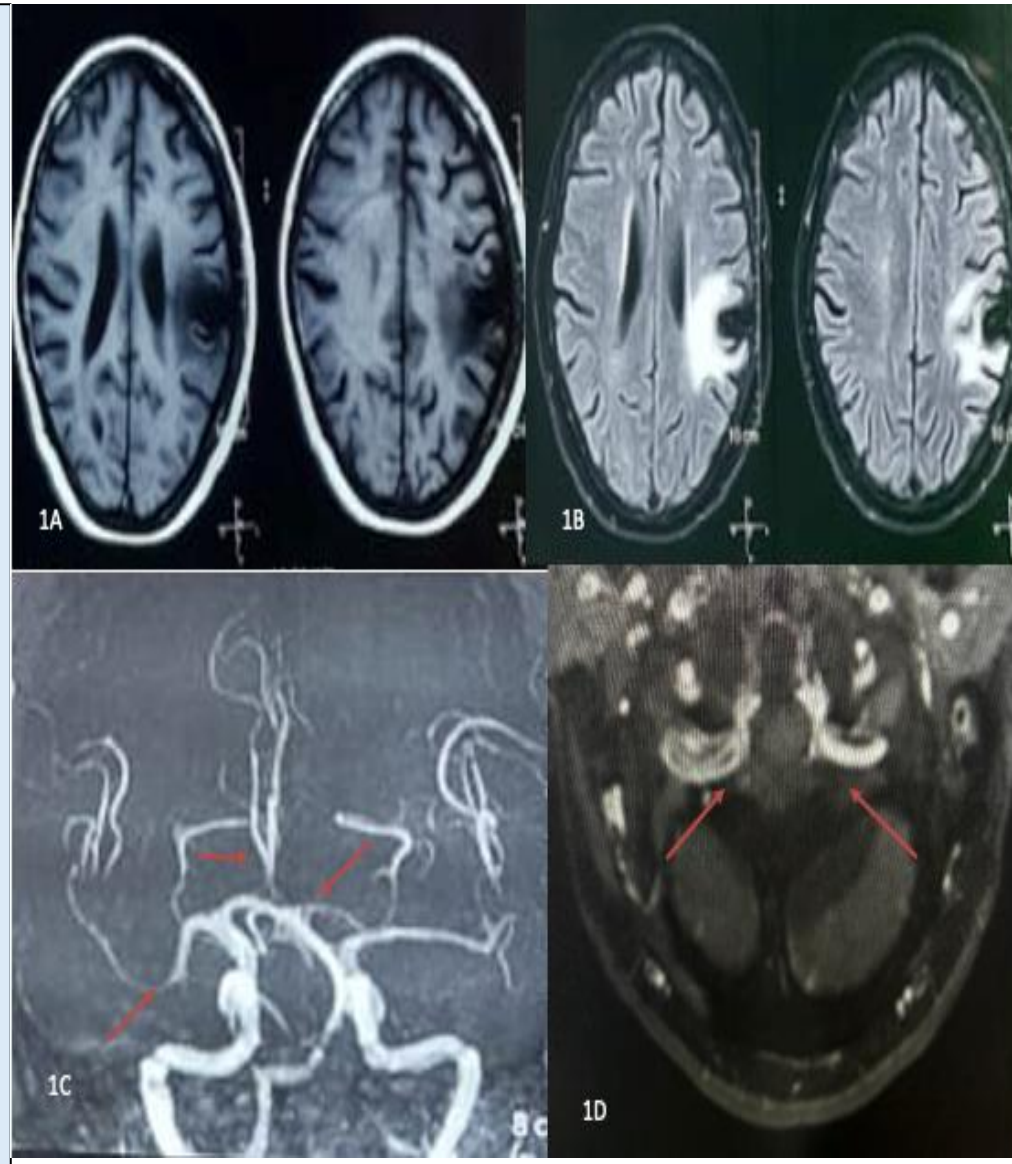


FIGURE 1:MRI Brain showing chronic infarct in left frontal region with diffuse narrowing of MCA, ACA and PCA and contrast enhancement along bilateral ICA

CASE III

A young female presented with progressive quadriparesis and bulbar weakness, recurrent episodes of vomiting, loose stools and syncopal episodes for 2 months. Examination showed bilateral facial and bulbar weakness, flaccid quadriparesis, orthostatic hypotension. NCS revealed demyelinating sensorimotor polyradiculoneuropathy. MRI brain and spine was unremarkable. CSF examination showed albumin-cytological dissociation, ANA immunoblot revealed strong positive SS-A and SS-B antibodies with a focus core > 1 in lip biopsy (2A,B). With a diagnosis of NSS with acute onset CIDP she received fludricortisone, pulse IV MPS and Rituximab.

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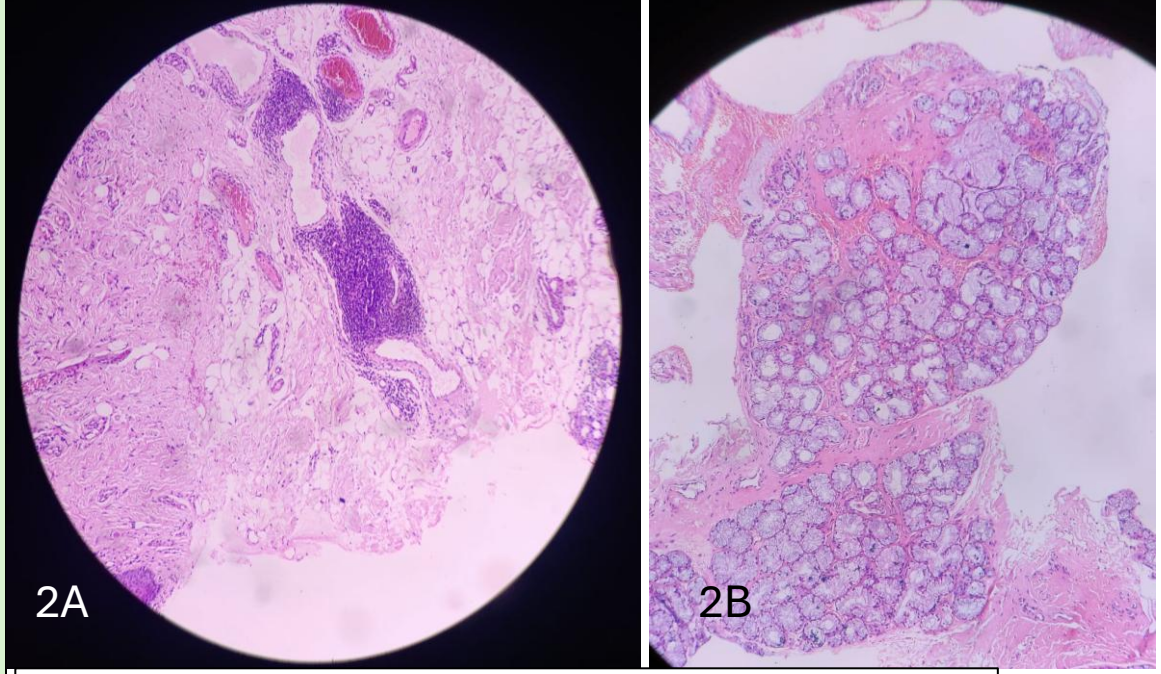

FIGURE 2: Salivary gland biopsy showing peri Ductal lymphocyte infiltration and aggregates

CONCLUSION

A high index of suspicion along with clinical history, serological tests, lip biopsy and parotid gland enlargement establishes diagnosis of NSS.