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Neuromyelitis Optica With Systemic Sclerosis: Case Report

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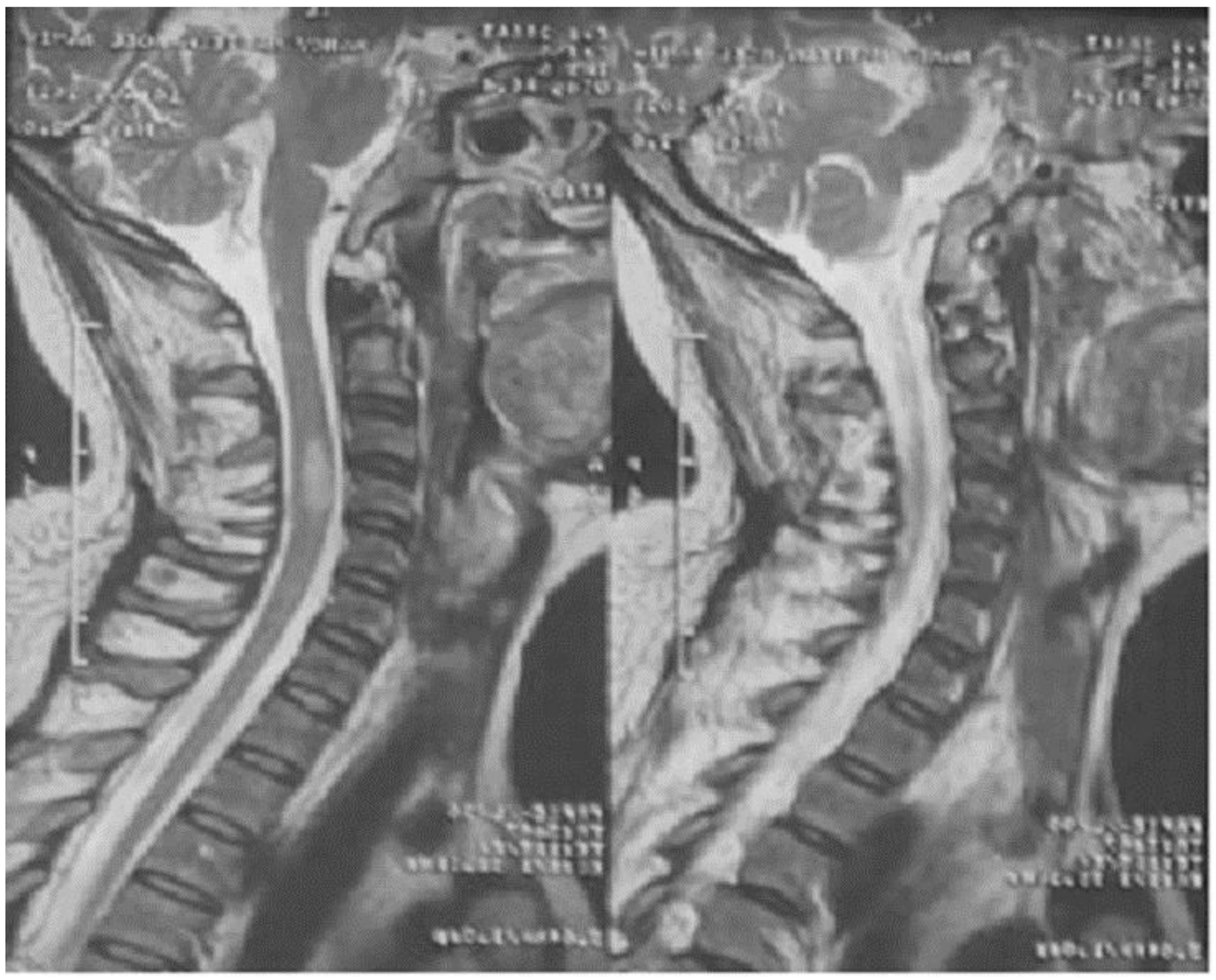
Objectives Neuromyelitis optica spectrum disorders (NMOSD) are an immune-mediated neurological pathology that affects the central nervous system (CNS), especially the optic nerves, and myelitis that affects a long segment of the spinal cord. Considering NMOSD as an autoimmune disease, several reports address the association with other systemic autoimmune diseases such as organ-specific autoimmune disorders, neoplasia, and infectious diseases. Interestingly, the strong association between NMO and autoimmune disorders, such as systemic lupus erythematosus (SLE) or Sjogren syndrome (SS), in addition to non-specific autoantibodies, such as antinuclear antibodies against antigens extracted from the nucleus, has also been detected.

Methods A 39-year-old woman with a 10-year history of systemic sclerosis, consulted with headache, loss of muscle strength, and bladder tenesmus. Physical examination reveals lethargy, quadriplegia, sphincter abnormalities, skin with loss of turgor on the hands, forearms, anterior chest and abdomen, Raynaud's phenomenon on the hands and pitting scars. Brain MRI images in T2 FLAIR showed hyperintensities in the dentate nucleus, thalamus, left and right cerebral peduncle, posterior limb of the left internal capsule, temporal periventricular subcortical hyperintensities, dorsal column in T2 hyperintensities in C3-C4, multiple hyperintensities in mesencephalon and medulla oblongata (Figure 1). The aquaporin-4 (AQP4) antibody profile was negative, ANA reactive (1:40 Speckled). Treatment with methylprednisolone was given for 5 days, followed by immunoglobulins and cyclophosphamide. She was discharged with good clinical parameters and stable immune response.

Results The association of NMOSC with other disorders of the immune system is known, amongst them, the diseases with the greatest relationship to NMOSC are SLE and SS. It has also been reported in sarcoidosis, rheumatoid arthritis, ankylosing spondylitis, antiphospholipid syndrome, and systemic sclerosis.



Conclusion The association of NMOSC, transverse myelitis with systemic sclerosis is documented in case reports.