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INTRODUCTION

Neuromyelitis optica (NMO) is a chronic demyelinating inflammatory disease of the central nervous system, mediated by autoantibodies against astrocyte podocyte-based AQ4 receptors, which typically involves the optic nerve and transverse myelitis spanning more than three myelotomies. In the spectrum of NMO, other autoimmune diseases also coexist, though its association with systemic lupus erythematosus (SLE) is rare. SLE myelitis, as a neuropsychiatric manifestation, refers most commonly to acute transverse myelitis of one to two myelotomies, with more extensive involvement occurring in only 1-2% of cases.

We present two cases of patients in their 70’s who were diagnosed with NMO in the context of SLE.

Case Series Presentation

	Case 1	Case 1 – follow up	Case 2	Case 2 – follow up
Age	78	79	71	72
Duration of Symptoms	Three weeks		Two months	
Level	T3-T8	T3-T8	C3-C7	C4-C7, T10-T12
Neurological Manifestation	Incomplete paraparesis T4 level	Indifferent	Incomplete paraparesis C4 level, worse on the left side	Improved – slight residual weakness on the left lower extremity peripherally
LP:	0 cells, Glucose: - 62mg/dl, Protein: 69mg/dl, LDH: 51, culture (-) cytology (-) for malignancy		0 cells, Glucose: 51mg/dl, Protein: 32mg/dl, LDH: 46, culture (-) cytology (-) for malignancy	-
Oligoclonal Bands	Type 3	-	Type 3	-
Anti-AQP4	320	100	3200	100
Significant Workup	PCR HHV-6 (+) ANA (+) & dsDNA (+)	-	↓C3: 71, ↓C4: 4	-
Treatment	Iv methyl-prednisolone 1gr x 5 days -> stop due to pancreatic reaction 2 cycles CYC	RTX	Iv methyl-prednisolone 1gr x 5 days, with gradual per os tapering up to 16mg. 2 cycles CYC	RTX
EDSS	7.5	7.5	7	4
Outcome	Stagnant – bedridden		Improved - able to stand and walk with support and self-service capability	

Table. Overview of the two cases. Abbreviations: LP= Lumbar Puncture; AQP4= Aquaporin – 4; CYC= Cyclophosphamide; RTX= Rituximab; EDSS= Expanded Disability Status Scale

Case Series Presentation

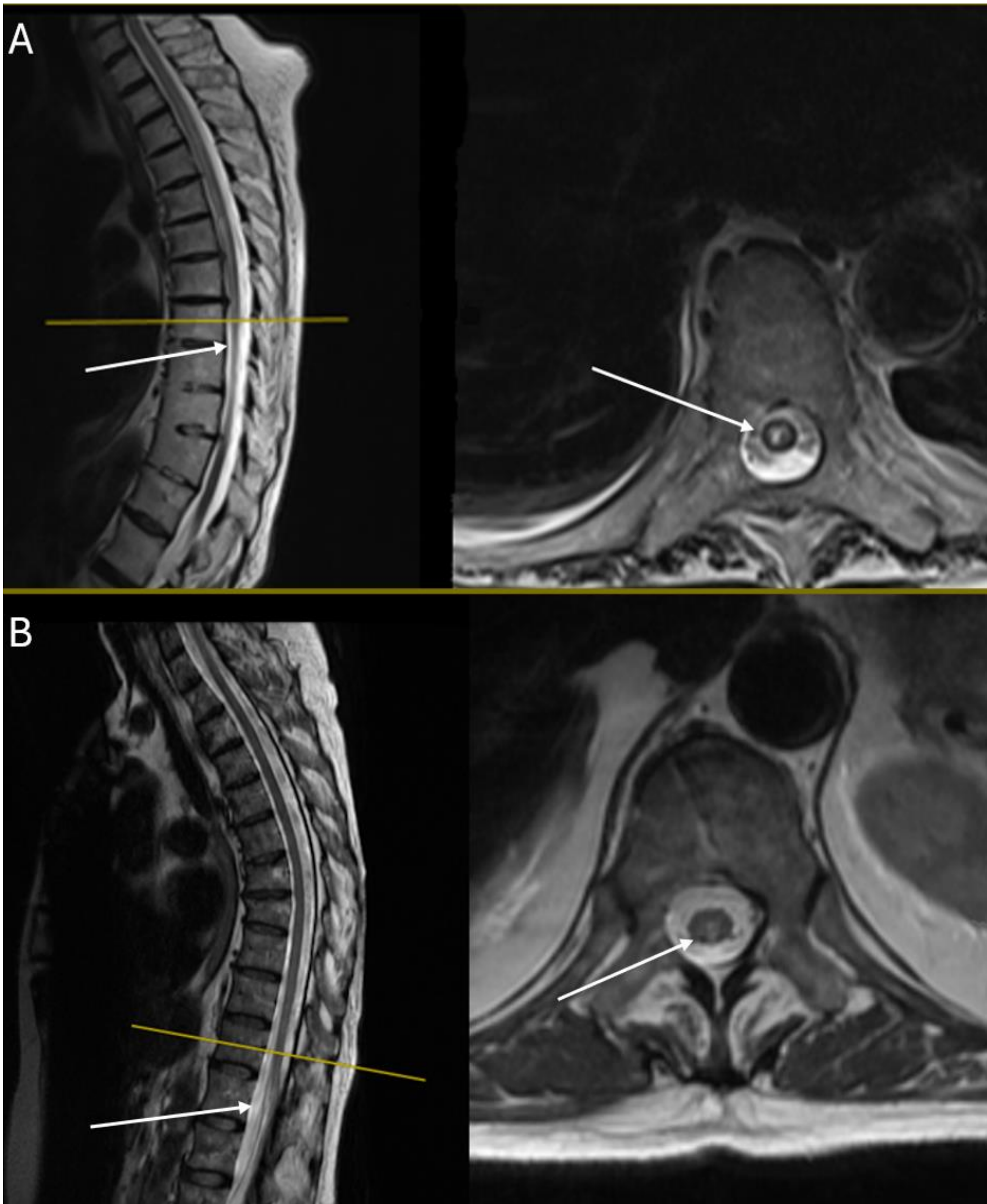


Figure. MRI imaging of the first (A) and second (B) cases demonstrating a long segment well-defined lesion extending within the thoracic spine; appearing low on T1 with no gadolinium enhancement and high on T2. A: axial and sagittal T2 STIR – weighted MRI images of the thoracic spine at admission of the first case. White arrows show the lesions of hyperintensities involving the central and the right half of the cord, sparing the dorsolateral part. B: axial and sagittal T2 STIR – weighted MRI images of the thoracic spine at admission of the second case. White arrows show the lesions of hyperintensities involving the dorsolateral part of the cord.

DISCUSSION

We present two rare cases of elderly women with SLE and NMO, illustrating heterogeneity in the context of the serological and clinical manifestation of the lupus spectrum, as well as of the NMO severity and progression.

CONCLUSION

The coexistence of NMO with SLE is rare, but the occurrence of myelitis in patients with connective tissue diseases should raise the suspicion of NMO, especially in elderly women and several years after the diagnosis of SLE. Time to treatment is critical, as delays in treating NMO can result in cumulative and disabling damage.

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