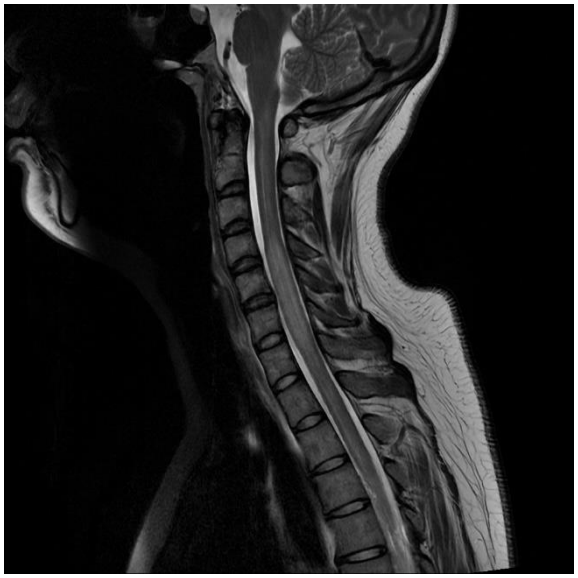
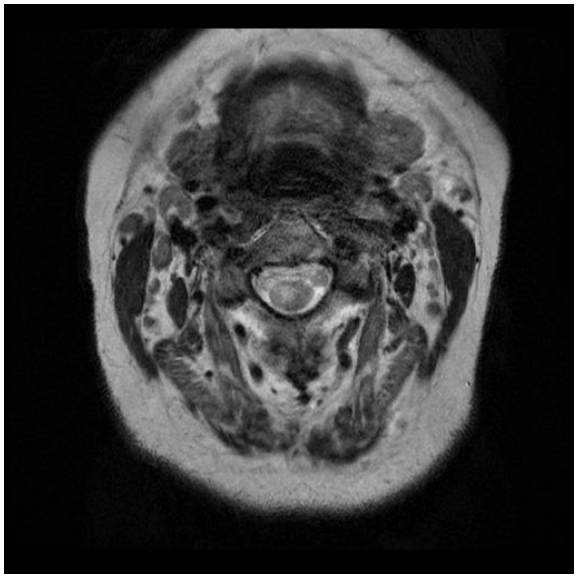
A 22-year-old woman presenting with sudden onset quadriparesis





Dong Rak Kwon M.D., Ph.D.

Department of Rehabilitation Medicine

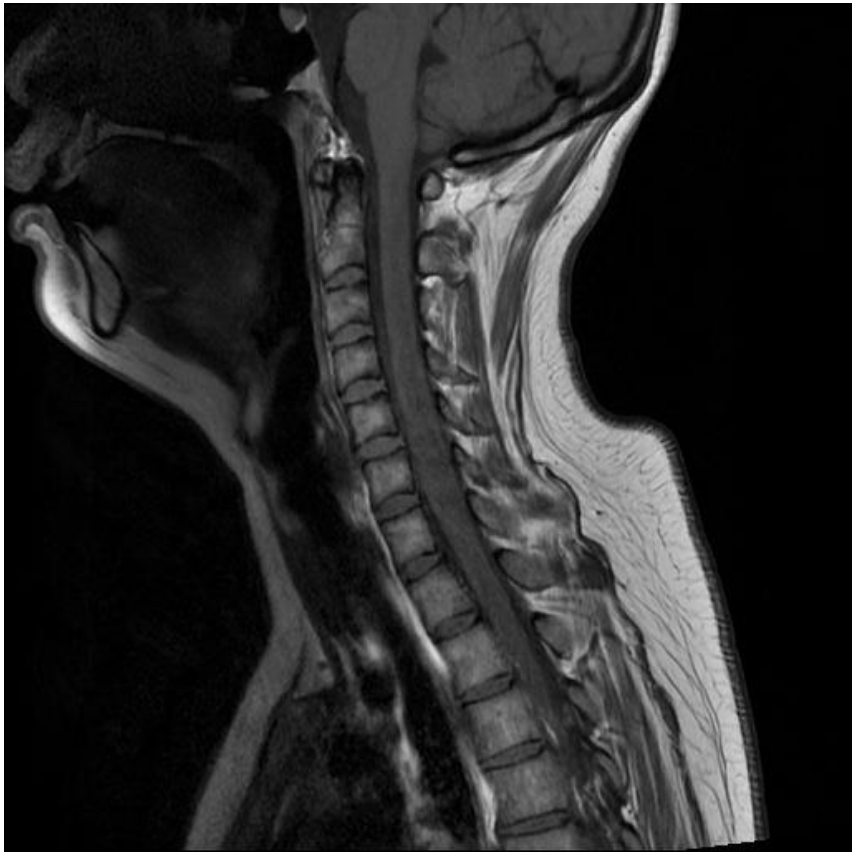
Daegu Catholic University School of Medicine

- F/22, Hyperthyroidism, R/o Graves' disease at 2019
- Fever and headache: 2022년 9월 초, LMC
- Quadriparesis: 2022년 9월 19일, ER
- Bladder and bowel problem
- Lab

Serum (220920)	CSF (220920)	CSF (221114)	Serum (221130)
VDRL (-)	Protein 215 mg/dl	Protein 72 mg/dl	MOG Ab (-)
HIV ab (-)	WBC 40/ul	WBC 0/ul	Aquaporin 4
HSV IgG (-)	Lymph (70%)		IgG Ab (-)
HSV IgM (-)	ADA 11.7 U/l	ADA 7.4 U/l	
Vit B12 (N)	Glucose 27 mg/dl	Glucose 35 mg/dl	
IGRA (-)	M.TB PCR (-)	M. TB PCR (-)	
	AFB stain (-)		

CSF (220920)
HSV IgG,IgM (-)
VZV IgM (-)
EBV PCR (-)
M. pneumonia PCR (-)
Enterovirus PCR (-)
세균성 뇌수막염 PCR 5종 검사 (-)
Fungus culture
Cryptococcus Ag(-)

C-spine MRI (220920)



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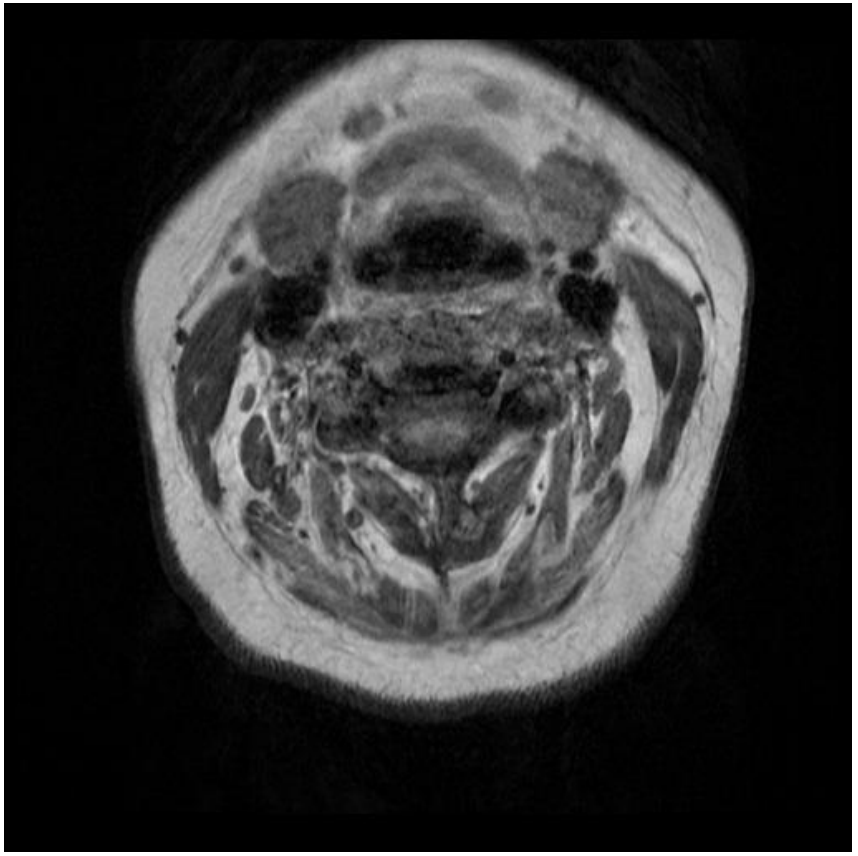
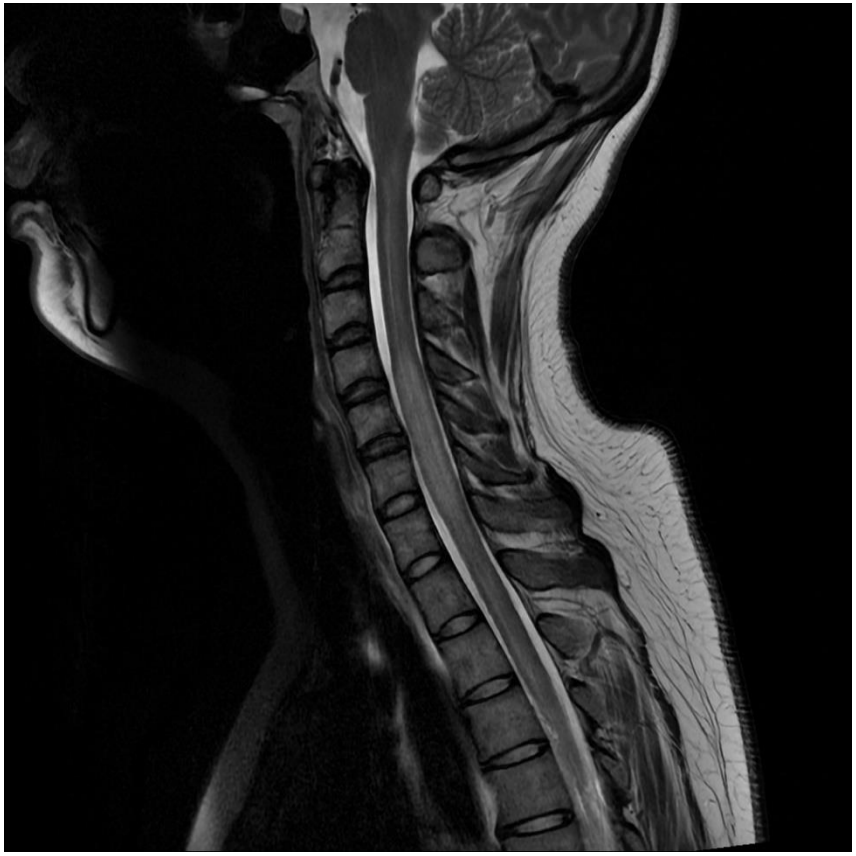


T1 AXL FES

C-spine MRI (220920)



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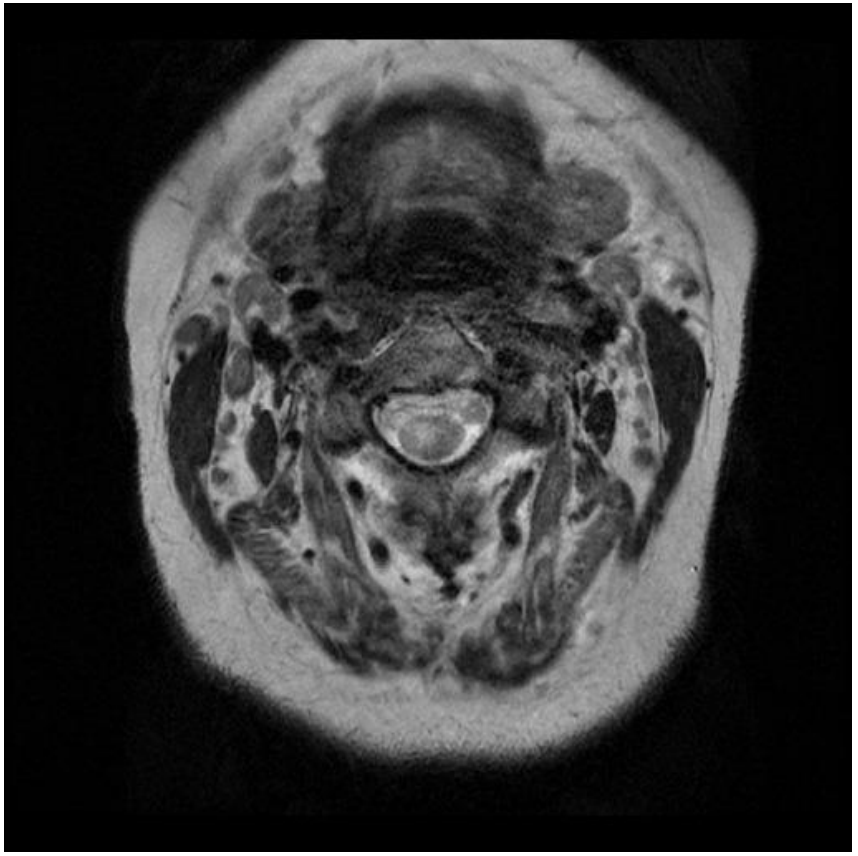
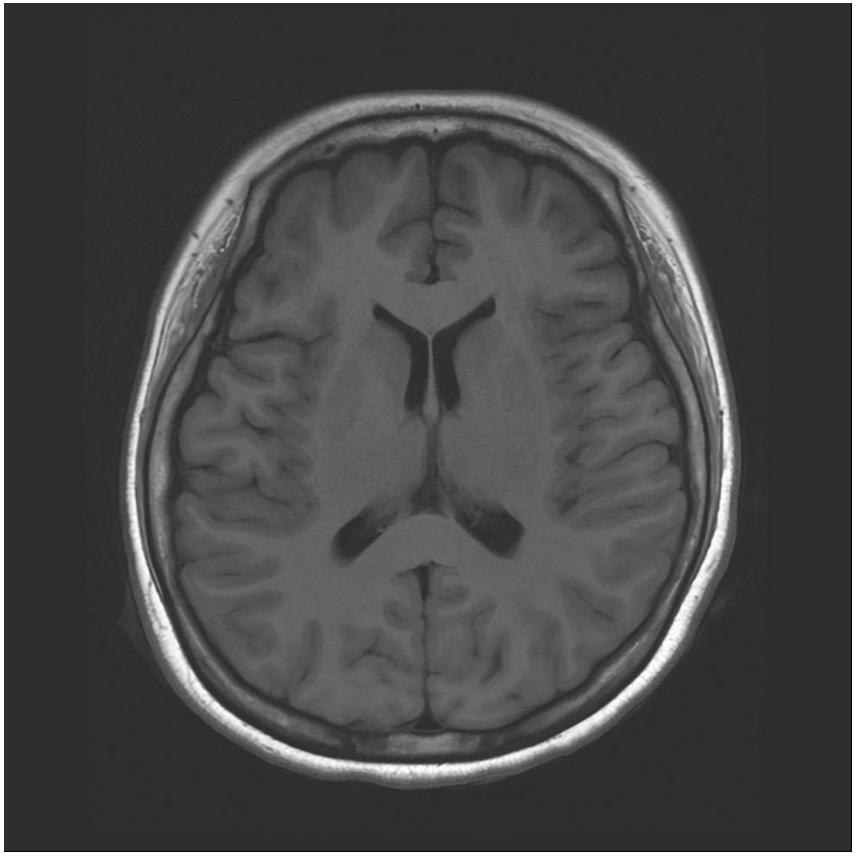


T2 AXL FES

Brain MRI (220927)



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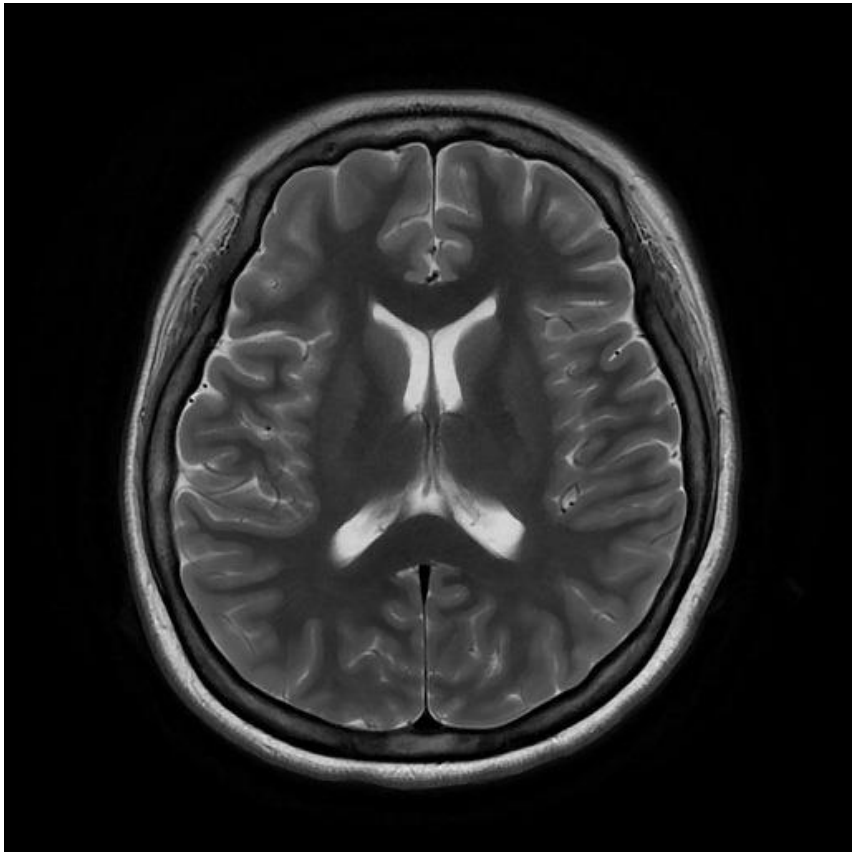


T2 AXL FES

Manual Muscle Test (221010)

Upper limb	Shoulder	Flexor	4-	Extensor	4-
	Elbow	Flexor	4-	Extensor	4-
	Wrist	Flexor	4-	Extensor	4-
	Finger	Flexor	4-	Extensor	4-
Lower limb	Hip	Flexor	0	Extensor	0
	Knee	Flexor	0	Extensor	0
	Ankle	Flexor	0	Extensor	0
	Toe	Flexor	0	Extensor	0

Sensory Exam (221010)

Light touch	Intact	C7/C7
	Impaired	C8/C8
	Absence	T3/T3
Pin Prick	Intact	C7/C7
	Impaired	C8/C8
	Absence	T3/T3
Deep anal sense		-
Perianal sense		-/-

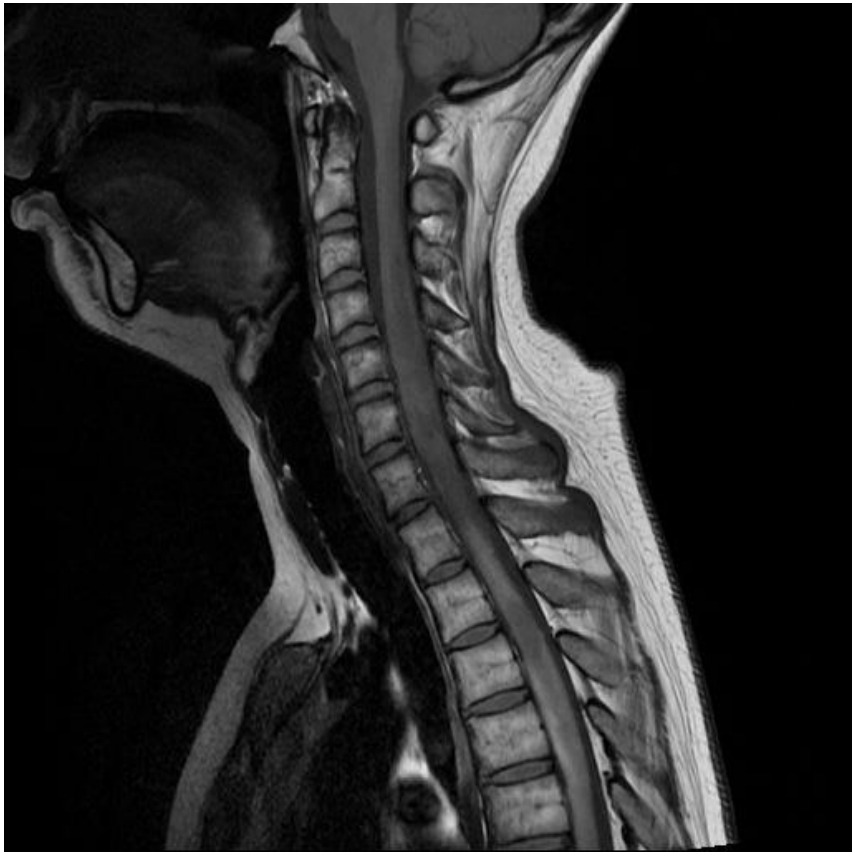
Functional Level (221010)

Functional Level	Roll Over	-
	Come to Sit	-
	Sitting Balance (S/D)	P/Z
SCIM II	Self Care	7/20
	Respiration and sphincter management	10/40
	Mobility	0/40

Medication

- Based on clinical, imaging, and CSF findings
- Isoniazid (INH) 300 mg/D and Rifampin (RMP) 600 mg/D for 10 months
- Pyrazinamide (PZA) 2 g/D and Etambutol (ETB) 1 g/D for 2 months
- Methylprednisolone pulse therapy for 7 days
- Prednisolone 50 mg (10 D) → 40 mg (10 D) → 30 mg (10 D) → 20 mg (10D) → 10 mg (45 D)

C-spine MRI (221011)



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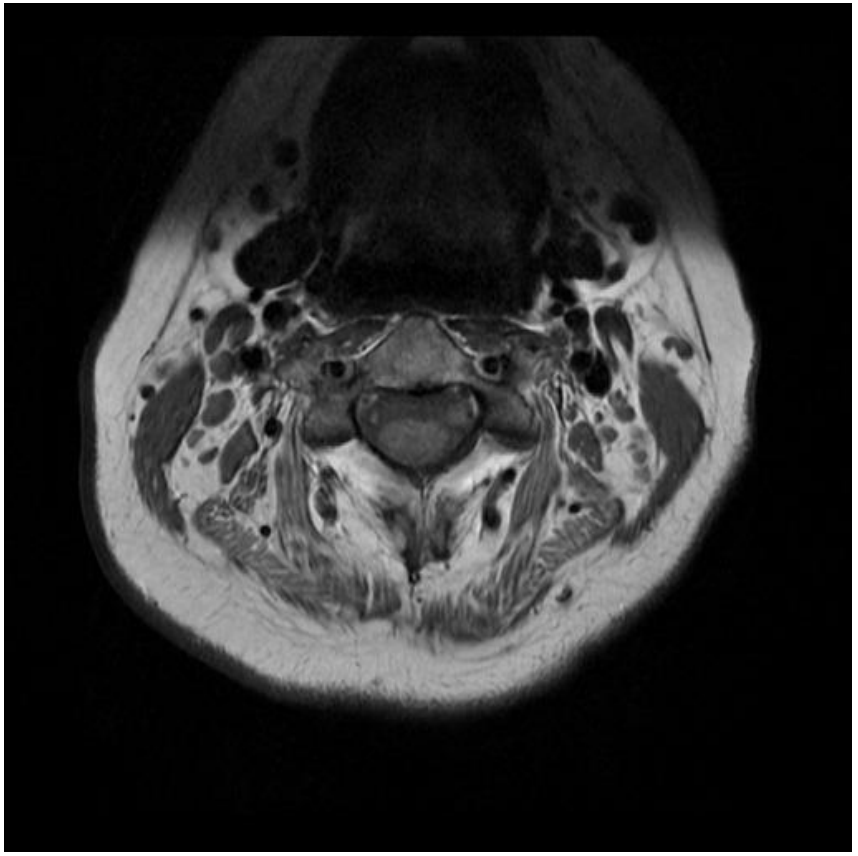
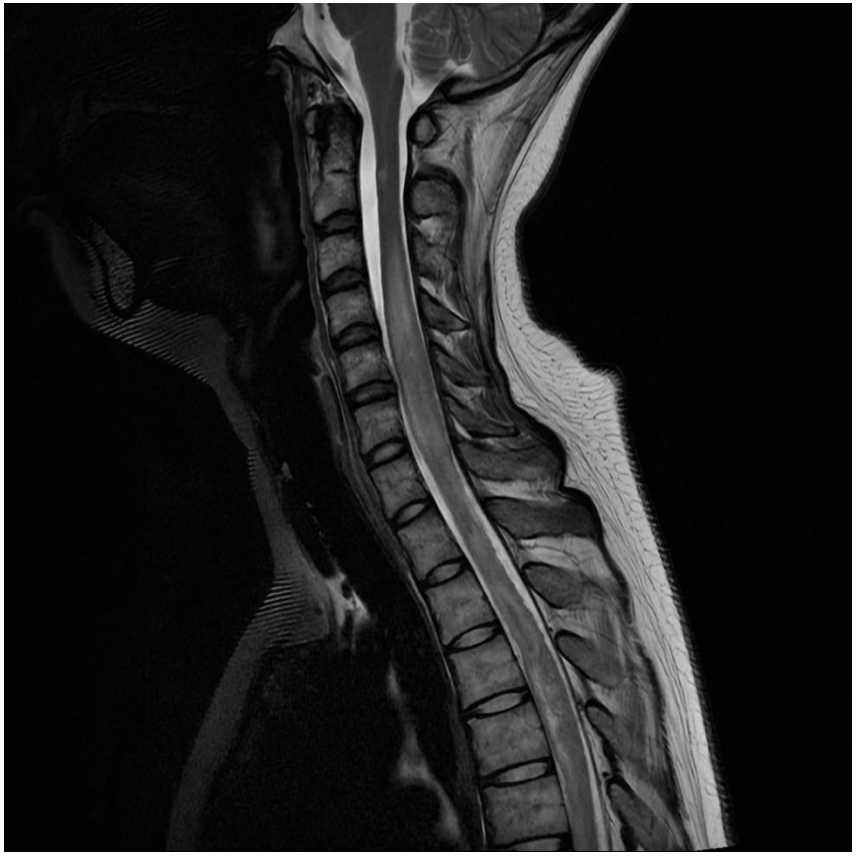


T1 AXL FES

C-spine MRI (221011)



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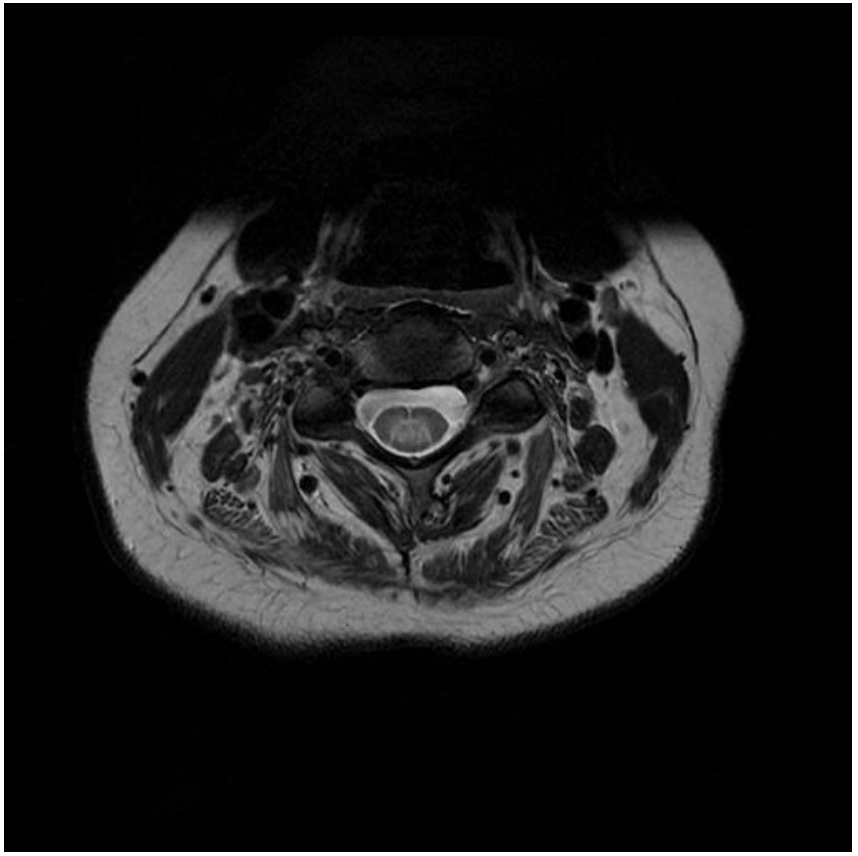
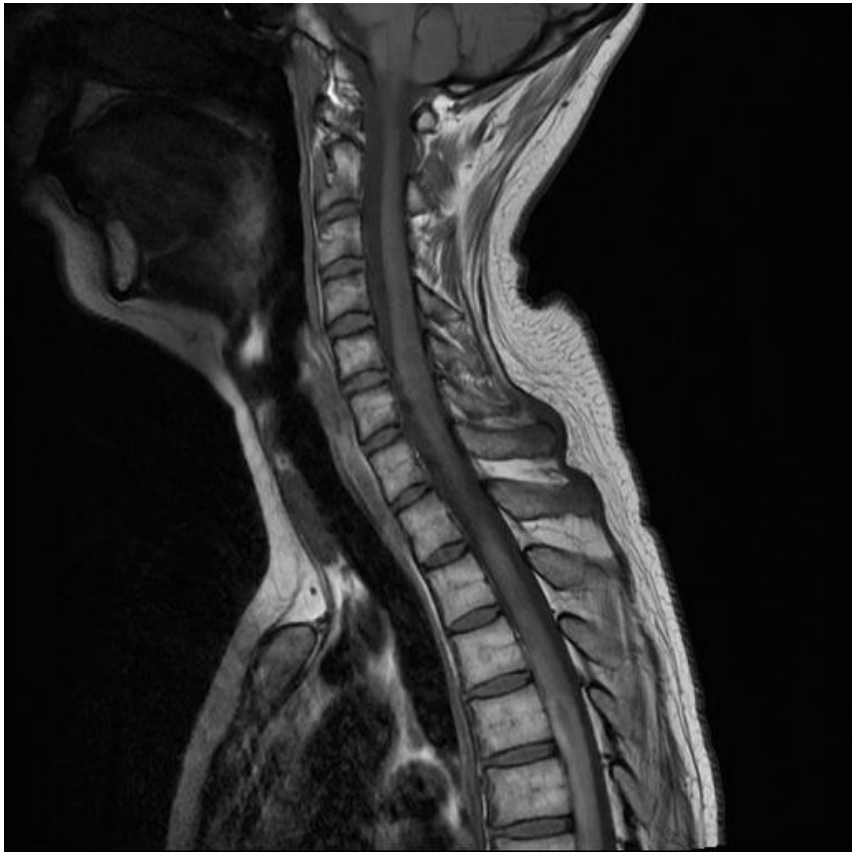


T2 AXL FES

C-spine MRI (221114)



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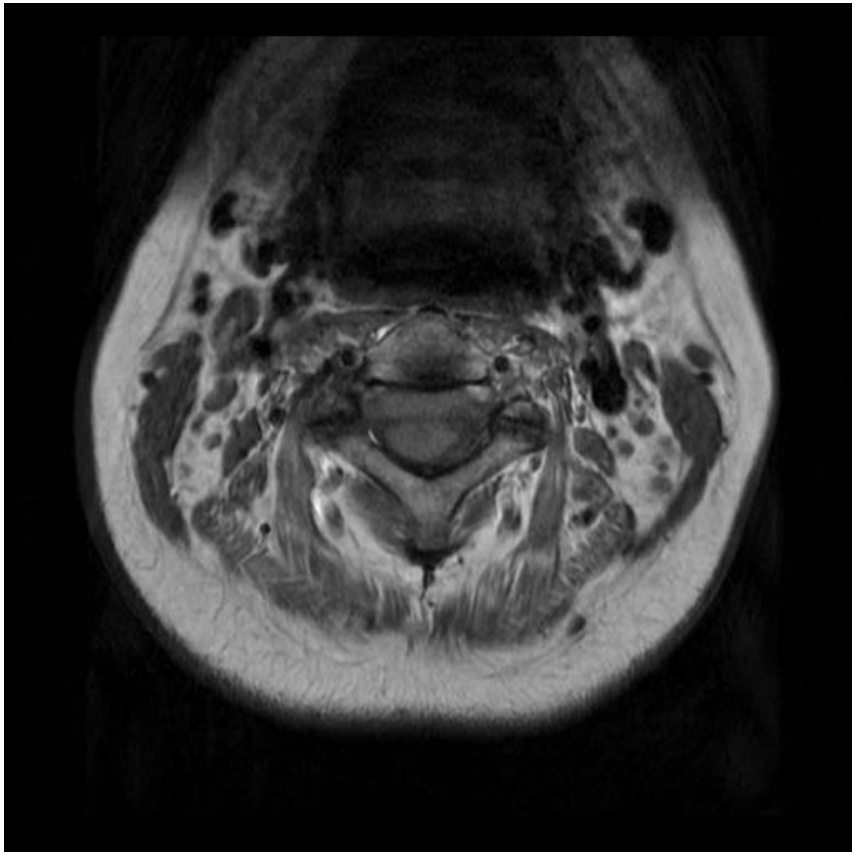

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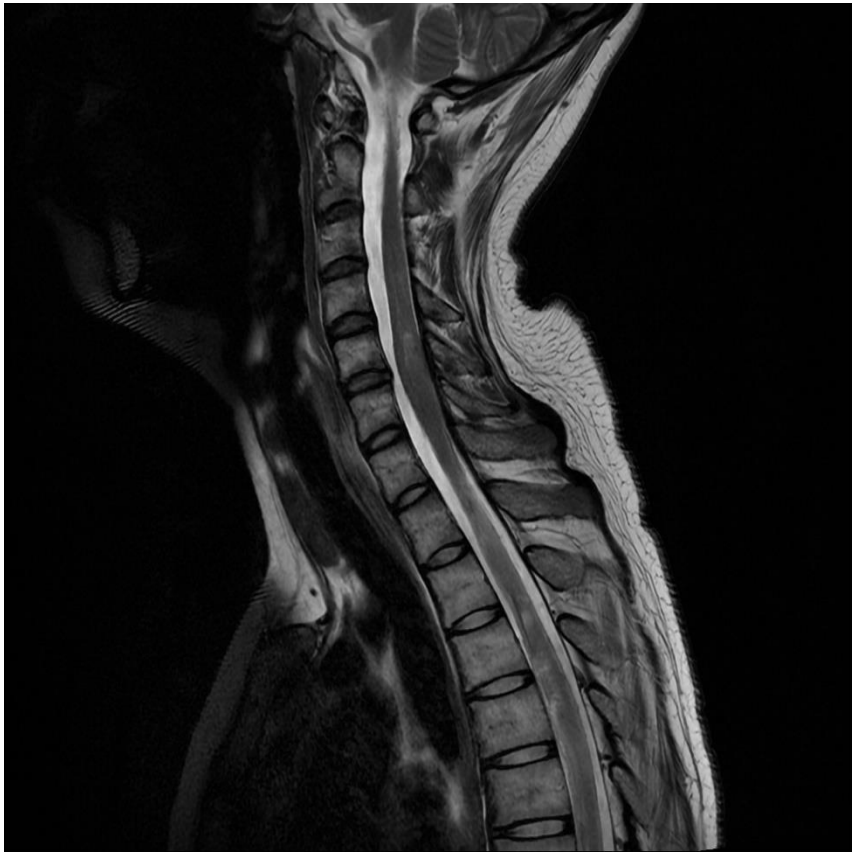
C-spine MRI (221114)



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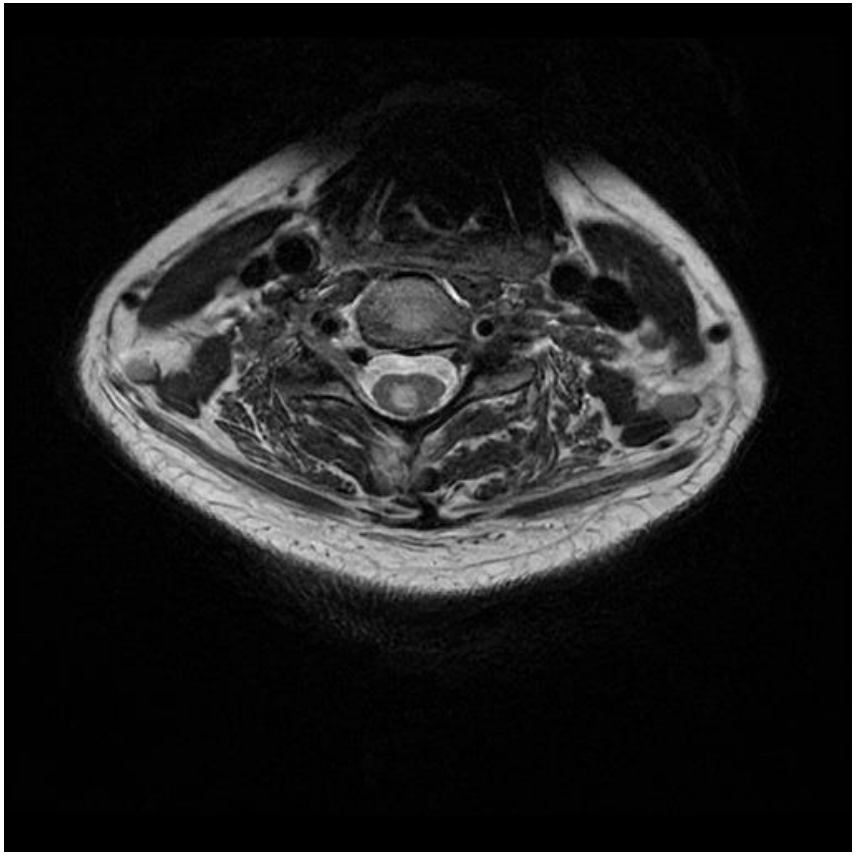


T2 AXL FES

Pancytopenia

- R/O Catheter related blood stream infection, Candidemia : high fever (12.4)

  PICC lumen blood culture: yeast growth (12.7)

  Rt. PICC → Lt. PICC (12.8)

  Tazocin(13.5g) + Anidulafungin(100mg) (12/7~12/14)

  Tazocin(13.5g) + Amphoterisin(300mg) (12/14~12/31)

  Tazocin(13.5g) + Micafungin(100mg) (1/1~)

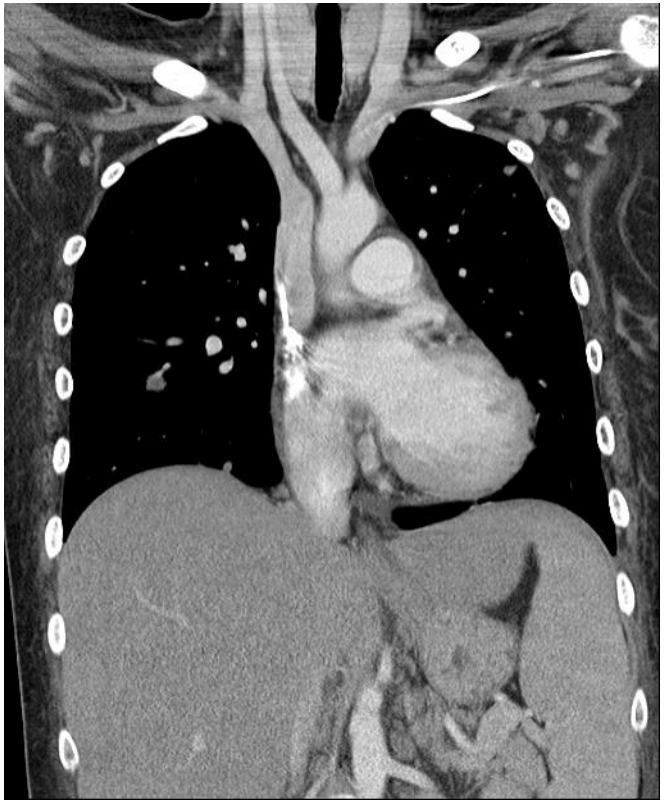
  CRP 7.3(H): 1.6

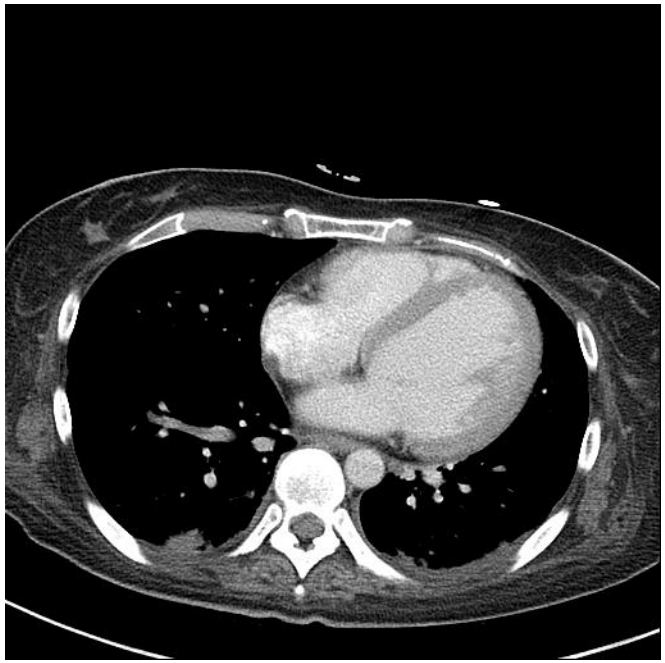
- R/O Invasive pulmonary aspergillosis : dyspnea, sputum (12/14, Chest CT: r/o fungal pneumonia: antifungal agents)

  → improved pneumonia (12/27, Chest CT)

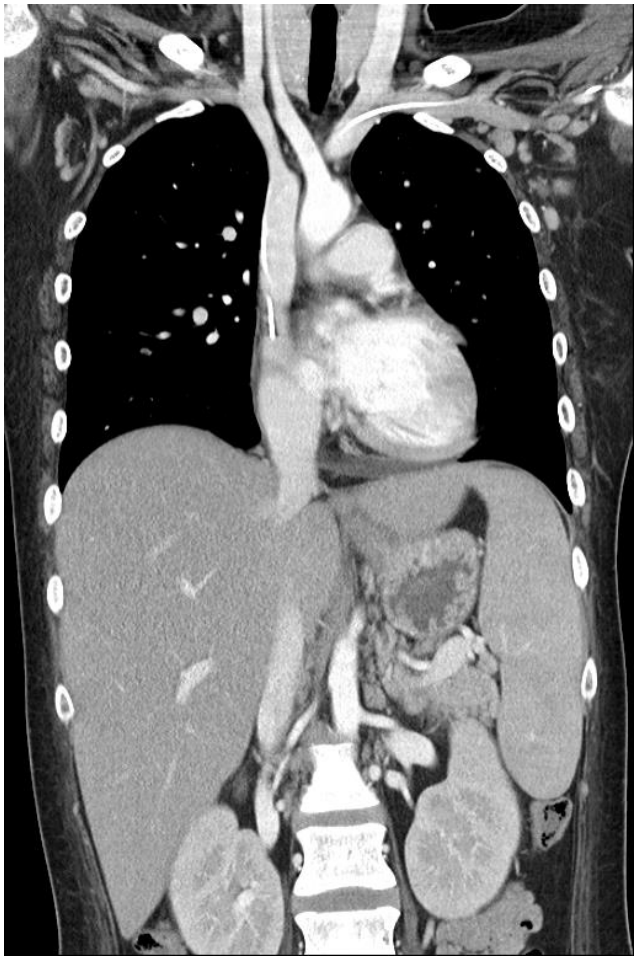
- Bone marrow biopsy and NGS: negative

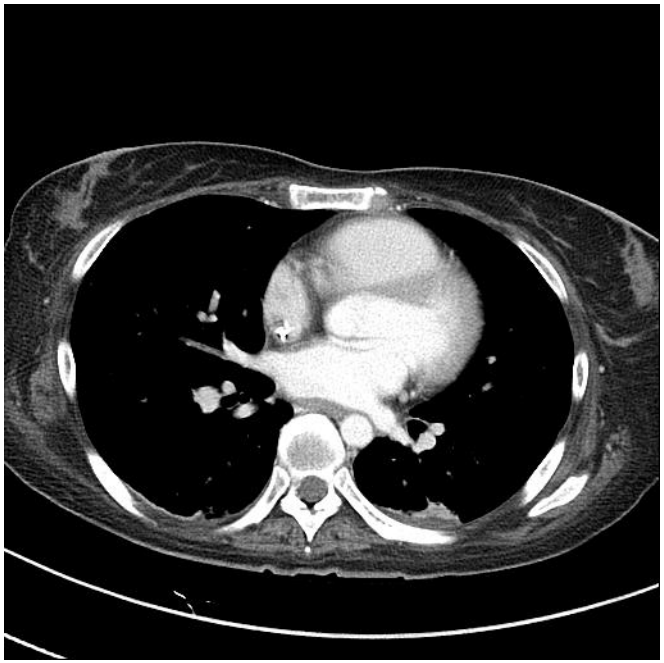
Chest CT (221214)





Chest CT (221227)





Immunologic Test

FANA	1:40
Anti RNP Ab	-
Anti Sm Ab	+
Anti RO Ab	-
Anti La Ab	-
Anti Cardiolipin Ab - IgG	-
Anti Cardiolipin Ab - IgM	-
Anti β2-GPI Ab IgG	-
Anti β2-GPI Ab IgM	-
Anti ds DNA Ab IgG	+
Lupus Anticoagulant	1.11

Medical Problems

- Stress induced cardiomyopathy (SICMP): chest discomfort, elevated cardiac enzyme → TTE, LVEF 36%
- Orthostatic hypotension
- Pressure sore, coccyx stage 4

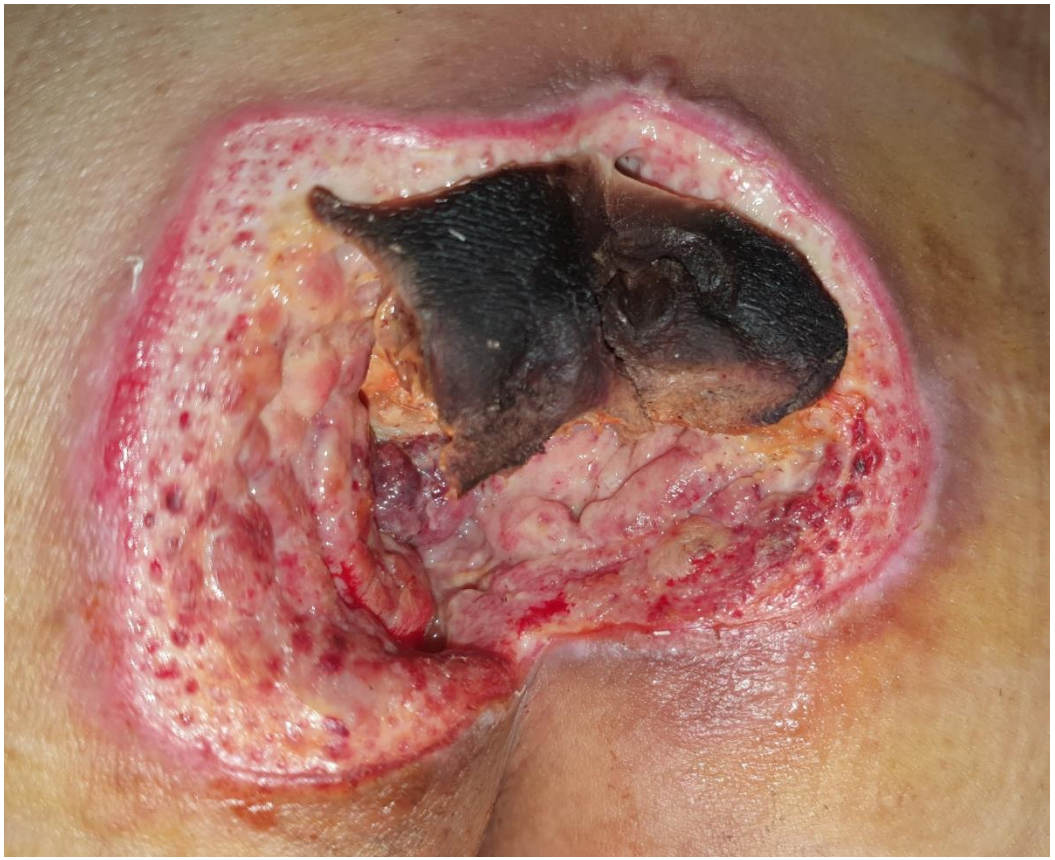
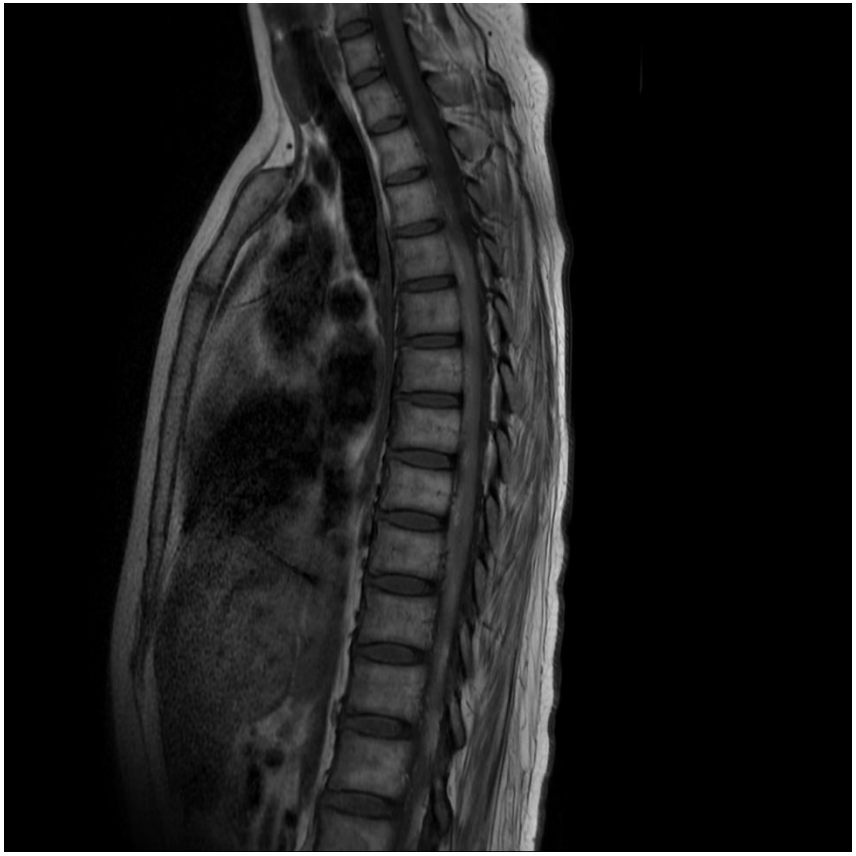


Table 1. Stressor reported to trigger stress-induced cardiomyopathy

Emotional stressors
Death or severe illness of a relative/friend/pet
Bad news
Interpersonal conflict
Financial or job problems
Stressful work
Physical stressors
Surgery, procedure
Acute respiratory failure (e.g. chronic obstructive pulmonary disease, pneumonia)
Sepsis, infection
Malignancy or chemotherapy
Stroke, seizure
No identifiable

J Neurocrit Care 2015;8(1):1-8

T-spine MRI (230114)



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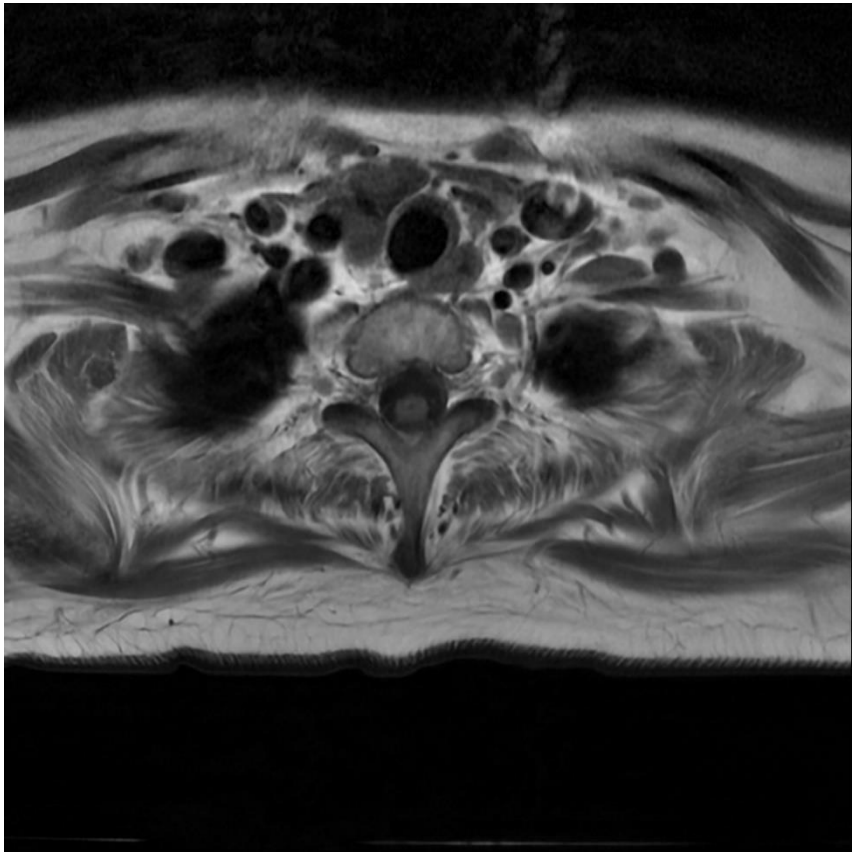
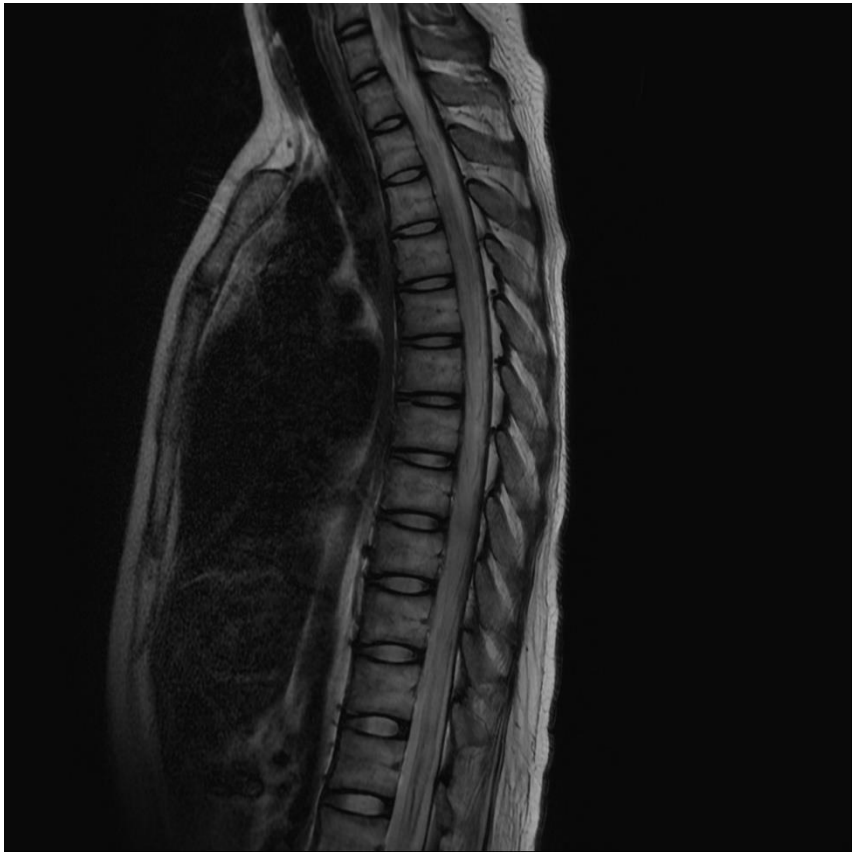


T1 AXL FES

T-spine MRI (230114)



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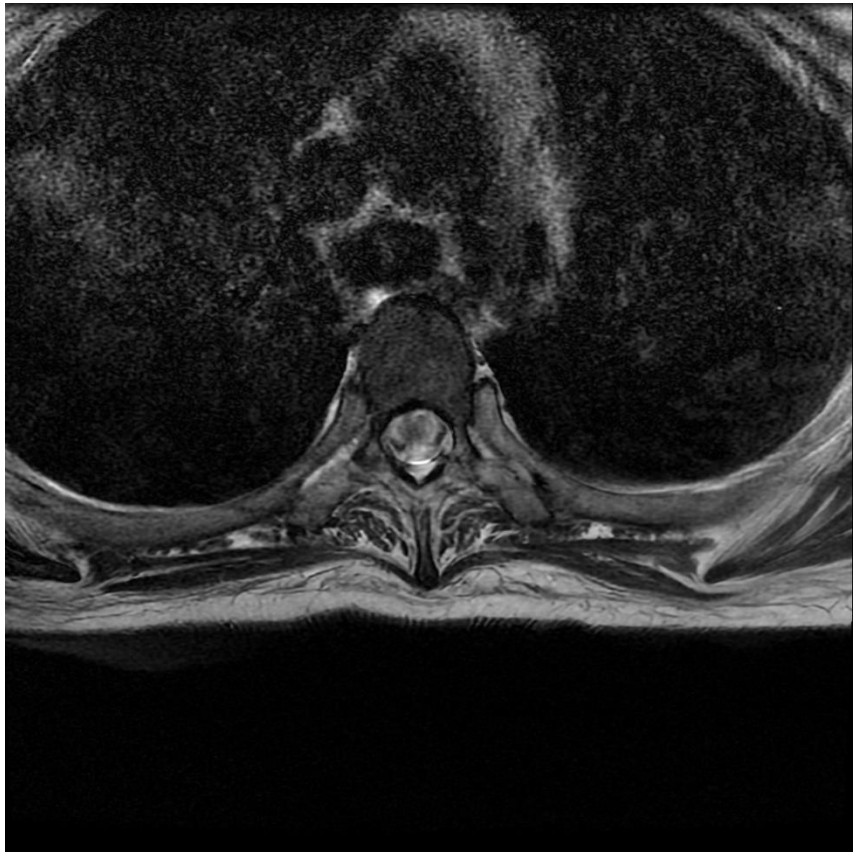

T2 AXL FES

Manual Muscle Test (230208)

Upper limb	Shoulder	Flexor	5	Extensor	5
	Elbow	Flexor	5	Extensor	5
	Wrist	Flexor	5	Extensor	5
	Finger	Flexor	5	Extensor	5
Lower limb	Hip	Flexor	0	Extensor	0
	Knee	Flexor	0	Extensor	0
	Ankle	Flexor	0	Extensor	0
	Toe	Flexor	0	Extensor	0

Sensory Exam (230208)

Light touch	Intact	C7/C7
	Impaired	C8/C8
	Absence	T3/T3
Pin Prick	Intact	C7/C7
	Impaired	C8/C8
	Absence	T3/T3
Deep anal sense		-
Perianal sense		-/-

Functional Level (230208)

Functional Level	Roll Over	-
	Come to Sit	-
	Sitting Balance (S/D)	P/Z
SCIM II	Self Care	7/20
	Respiration and sphincter management	10/40
	Mobility	0/40

Transverse Myelitis

- The term “myelitis” refers to inflammation of the spinal cord, which can involve its cross-sectional area entirely or partially, hence being defined transverse or partial
- Acute progressive inflammatory demyelination or necrosis of the spinal cord caused by various autoimmune reactions
- A rare neurological disease, poor prognosis
- Main pathological changes: myelin swelling, demyelination, significant proliferation of peripheral lymphocytes, axonal degeneration, and infiltration of perivascular inflammatory cells
- Acute transverse myelitis (ATM) refers to lesions that span one to two vertebral segments

Clinical Manifestations

- Symmetric or asymmetric weakness in the limbs (upper and/or lower, depending on the level of involvement)
- Sensory abnormalities (hypoesthesias, paresthesias, allodynia)
- Bladder or bowel dysfunction (urgency or retention) or sexual dysfunction.
- Lower-motor neuron signs (fasciculations, hypo/areflexia and flaccid tone): gray matter involvement

Classifications

- Cell-mediated in the context of autoimmune disorders confined to the CNS (e.g., MS)
- Associated with specific antibodies primarily targeting CNS antigens
- Associated with systemic autoimmune disorders having secondary CNS involvement (e.g., systemic lupus erythematosus, Sjogren’s syndrome, and sarcoidosis)

Longitudinally Extensive Transverse Myelitis (LETM)

- Contiguous inflammatory lesions of spinal cord extending to ≥3 vertebral segments
- The causes of LETM including various infections, neoplastic reason, and autoimmune disease

Table 2. Clinical features to aid in the differential diagnosis of longitudinally extensive transverse myelitis (LETM)

Cause of LETM	Clinical clues to diagnosis
Autoimmune	
• Neuromyelitis optica	Preceding nausea, vomiting, hiccups; endocrine disturbance; history of optic neuritis; previous LETM; history of autoimmune disease; poor clinical recovery
• Systemic lupus erythematosus	Neuropsychiatric symptoms; history of rash, mouth ulcers, arthralgia
• Sjögren's syndrome	Sicca syndrome
• Antiphospholipid syndrome	History of thrombosis or recurrent miscarriage
Inflammatory	
• Multiple sclerosis	Previous symptoms suggestive of central nervous system demyelination
• Acute disseminated encephalomyelitis	Preceding infection or immunization; encephalopathy; good clinical recovery; usually occurs in children or young adults
• Neuro-Behçet's	Mediterranean origin; history of oral or genital ulceration
• Neurosarcoidosis	Evidence of systemic involvement, particularly respiratory symptoms
Infectious	
• Parainfectious: Epstein Barr virus, cytomegalovirus, herpes simplex virus, varicella zoster virus, mycoplasma	Systemic features of infection 1 to 3 weeks prior to onset of neurological symptoms
• Syphilis	
• Tuberculosis	
• HIV	
• Human T cell lymphotropic virus 1	African origin; myelopathy usually slowly progressive
• Schistosomiasis	Recent foreign travel or resident in endemic area
• *Toxocara canis*	Systemic features such as cough, wheezing, urticaria
• *Ascaris suum*	Travel to endemic area. Abdominal pain, weight loss.
Neoplastic	
• Paraneoplastic, particularly CRMP5	Myelopathy usually slowly progressive; may be evidence for systemic malignancy, particularly lung and breast
• Intramedullary tumour: ependymoma, lymphoma	Slowly progressive myelopathy
• Intramedullary metastases	
Metabolic	
• Vitamin B12 deficiency	History of other autoimmune diseases or poor dietary intake or malabsorption
• Copper deficiency	History of gastric band surgery, malabsorption or zinc supplementation
Vascular	
• Spinal cord infarction/ischaemia	Abrupt onset of symptoms; vascular risk factors (although may be absent); usually occurs in patients >55 years; classically dorsal columns spared
• Dural fistula	Limb weakness worse on ambulation or with valsalva manoeuvre; typically occurs in elderly men; usually slowly progressive but acute exacerbations can occur
Other	
• Radiotherapy	History of radiotherapy to neck, mediastinum or thorax; radiotherapy tattoos

Table 3. Radiological and laboratory features to help with the differential diagnosis of longitudinally extensive transverse myelitis (LETM)

Causes of LETM	Investigations to aid in diagnosis	Comment
Autoimmune		
• Neuromyelitis optica (AQP4-mediated)	AQP4 antibody	Highly specific and sensitive for NMO
	ANA	Co-existing autoimmune disease common in NMO
	MRI spine	Lesion involves cervical or thoracic cord; central or holocord location on axial imaging; T1 hypointensity; cord expansion and post-contrast enhancement common
	MRI brain	Periventricular lesions uncommon at presentation. Periaquaductal, hypothalamic, thalamic lesions characteristic
	CSF	Pleocytosis common and may be marked. Mild to moderately elevated CSF protein often seen. OCBs rare.
• Systemic lupus erythematosus	ANA	May coexist with NMO or can cause myelitis per se
• Sjögren's syndrome	ENA	May coexist with NMO; anti-Ro antibodies can be seen in NMO in the absence of Sjögren's syndrome
• Antiphospholipid syndrome	Antiphospholipid antibodies	May coexist with NMO
Inflammatory		
• Multiple sclerosis	MRI brain	Barkhof criteria often met at presentation; Dawson's fingers and u-shaped juxta-cortical lesions characteristic.
	MRI spine	Cord expansion and post contrast enhancement less common than in NMO. May be additional smaller lesions present
	VEPs	Often subclinical abnormalities present
	CSF	OCBs usually positive but otherwise CSF constituents normal
• Acute disseminated encephalomyelitis	MRI brain	Lesions often large and confluent. Basal ganglia commonly affected
	MOG antibodies	May be positive during acute illness
• Neuro-Behcet's	MRI brain	If present, lesions often extend from brainstem up into diencephalic structures
	Pathergy reaction	Often positive
• Neurosarcoidosis	MRI brain	May show MS-like lesions or leptomeningeal enhancement, particularly of basal meninges
	MRI spine	Leptomeningeal enhancement may be present
	Serum ACE	May be elevated, particularly if systemic involvement
Infectious		
• Parainfectious	Serology for Epstein Barr virus, cytomegalovirus, herpes simplex virus, varicella zoster virus, mycoplasma, hepatitis, Lyme	NMO can be triggered by infection so important to test for AQP4 antibodies even if clear systemic infection
• Syphilis	VDRL	Rare but treatable
• Tuberculosis	CSF	AFBs or mycobacterium seen on culture. Often markedly elevated CSF protein
	CXR	Evidence of pulmonary tuberculosis

Tuberculosis

- Tuberculosis (TB) is a devastating disease caused by Mycobacterium tuberculosis (MTB) with over eight million deaths reported annually worldwide.
- Extrapulmonary involvement occurs in 15% of affected individuals.
- TB of the central nervous system comprises 1% of all TB infections
- 95% of these in the form of TB meningitis, and 1% is spinal involvement.
- Diagnosis is usually based on clinical, laboratory and radiological findings.
- TB is a rare cause of TM, and cervicodorsal regions are the most common site of involvement in tuberculosis myelitis

Normal Values of CSF Components

Component	Adults and children	Neonates
Color	Clear	Clear
CSF:serum glucose ratio	0.44 to 0.90	0.42 to 1.10
Differential	70% lymphocytes, 30% monocytes, rare PMNs or eosinophils	PMN count may be normal
Gram stain	Negative for organisms	Negative for organisms
Lactate level*	11.7 to 21.6 mg per dL (1.3 to 2.4 mmol per L)	8.1 to 22.5 mg per dL (0.9 to 2.5 mmol per L)
Opening pressure	Adults and children 8 years and older: 60 to 250 mm H_2O Children younger than 8 years: 10 to 100 mm H_2O	10 to 100 mm H_2O
Protein level*	< 50 mg per dL (500 mg per L)	≤ 150 mg per dL (1,500 mg per L)
White blood cell count*	< 5 per µL	< 20 per µL

CSF Characteristics by Infection Type

Infection type	Differential	Glucose level	Opening pressure
Bacterial (typical)*	Usually 80% to 90% PMNs; > 50% lymphocytes possible	< 40 mg per dL (2.22 mmol per L) in 50% to 60% of cases; CSF:serum glucose ratio < 0.4 is 80% sensitive and 98% specific	Adults and children 8 years and older: 200 to 500 mm H_2O Children younger than 8 years can have lower pressures
Cryptococcal	Lymphocyte predominance	Usually > 40 mg per dL	> 250 mm H_2O in severe cases; serial lumbar punctures or ventriculoperitoneal shunt required to drain CSF if pressure persistently > 250 mm H_2O
Fungal (excluding cryptococcal)	Possible early PMNs progressing to lymphocyte predominance; eosinophils possible	Significant decrease possible	Variable
Neurosyphilis	Variable	Possibly decreased	Usually elevated in immunocompetent patients; may not be elevated in immunocompromised patients
Parasitic	Eosinophilia (> 10 eosinophils per µL or > 10% of total cells)	Usually low normal or normal	Variable but can be persistently elevated, requiring CSF draining
Tuberculosis	Early lymphocyte and PMN predominance progressing to lymphocyte predominance	Median: 40 mg per dL; lower in advanced stages	Variable depending on stage
Viral	Lymphocyte predominance; possible PMN predominance in early infection	Usually normal; decreased in 25% of patients with mumps; mild decrease possible in patients with HIV infection	Usually normal

Table 2. Detailed CSF and relevant biochemical/laboratory and other imaging findings pertinent to the 10 reported cases

CSF findings	Results		
	Present/positive/elevated	Absent/negative/reduced	Not documented
Pleocytosis	70 (7)	20 (2)	10 (1)
Protein	100 (10)	-	-
Glucose	Normal in 60 (6)	Reduced in 20 (2)	20 (2)
PCR for TB / AFB stain	60 (6)	40 (4)	-
AQP4-IgG	-	60 (6)	40 (4)
OCBs	-	40 (4)	60 (6)
ELISA for HIV	10 (1)	30 (3)	60 (6)
Relevant biochemical/laboratory and other imaging findings (for the 4 patients with negative CSF TB findings)	**Present/positive/elevated**	**Absent/negative/reduced**	**Not documented**
Tuberculin skin test	20 (2)	-	-
Biopsy	Positive in 2 patients (1 paratracheal lymph node, 1 right lung nodule)	-	-
Chest CT	2 patients with positive chest CT findings (1 right-lung nodule, 1 left upper lobe cavitating lesion)	-	-
Brain MRI	Tuberculomas on brain MRI (1 patient)	-	-

Longitudinal Extensive Transverse Myelitis due to Tuberculosis

- The MRI findings in patients with intraspinal TB have both diagnostic and prognostic significance
- Poor outcome: cord atrophy or cavitation and the presence of syrinx
- Since the neurologic deficits are mainly secondary to the inflammatory process
- These lesions usually respond to medical therapy alone, and with early diagnosis

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Table 1: Summary of CSF and radiological findings in the four cases

Investigations	Case 1	Case 2	Case 3	Case 4
CSF biochemistry				
Protein	75 mg/dl	40 mg/dl	200 mg/dl	440 mg/dl
Sugar	48 mg/dl	152 mg/dl	35 mg/dl	40 mg/dl
AFB	Negative	Negative	Negative	Positive
CSF cytology				
Total cells	5/ml	5/ml	20/ml	140/ml
Neutrophils (%)	0	0	50	40
Lymphocytes (%)	100	100	50	60
CSF ADA (IU/L)	15	6	10	6
CSF TB-PCR	Positive	Positive	Positive	Positive
MRI brain	Tuberculomas	Tuberculomas	Normal	Normal
MRI spine	Case 1: Continuous centrally located intramedullary cord signal alteration from C7 spinal segment to the conus, appearing hyperintense on T2W images without cord expansion Case 2: Continuous intramedullary T2 hyperintensity extending from D1 to conus Case 3: Involving the dorsal cord from D6 to D10 levels, appearing hypointense on T1W and hyperintense on T2W images Case 4: Patchy ill-defined T2 hyperintensities in the dorsal cord from D2 to D9 levels			

Systemic Lupus Erythematosus (SLE)

- A life-threatening autoimmune connective tissue disease with poor prognosis, affecting multiple organs
- Prevalent in women of childbearing age
- Clinical manifestations: ranging from cutaneous and mucous dysfunction to multisystemic involvement (disorders of immune function, kidney function, and nervous system function)
- Transverse myelitis has been reported in approximately 1–2% of people with SLE while its prevalence in the general population is estimated at 1–4 cases per million per year.
- TM tends to occur within the first 5 years from the diagnosis of SLE and at least one recurrence occurs in 21–55% of patients.

Diagnostic criteria over the years.

	1971 ACR	1982 ACR	1997 ACR	2012 SLICC
Cutaneous manifestation	**6 items** • Facial erythema (butterfly rash) • Discoid rash • Raynaud's phenomenon • Alopecia • Photosensitivity • Oral or nasopharyngeal ulceration	**4 items** • Malar rash • DLE lesion • Photosensitivity • Oral ulcers	**4 items** • Malar rash • Discoid rash • Photosensitivity • Oral ulcers	**4 items** • ACLE/SCLE • CCLE • Oral ulcers • Nonscarring alopecia
Joints	**1 item** Arthritis without deformity ≥ one peripheral joint, characterized by pain, tenderness or swelling	**1 item** Nonerosive arthritis ≥2 peripheral joints, characterized by pain, tenderness or swelling	**1 item** Nonerosive arthritis ≥ 2 peripheral joints, characterized by pain, tenderness or swelling	**1 item** Synovitis ≥ 2 peripheral joints, characterized by pain, tenderness, swelling or morning stiffness ≥30min
Serositis	**1 item** Serositis (any of the following): pleuritis, rub, history, evidence of both pleural thickening and fluid, pericarditis, EKG	**1 item** Serositis (any of the following): pleuritis, pericarditis	**1 item** Serositis (any of the following): pleuritis rub, evidence of pleural effusion, pericarditis, EKG	**1 item** Serositis (any of the following): pleuritis, typical pleurisy > 1day, history, rub, evidence of pleural effusion, pericarditis, typical pericardial pain >1day, EKG evidence of pericardial fusion
Renal disorder	**2 items** •Proteinuria ≥ 3.5g/day • Cellular casts	**1 item** Renal disorder (any of the following): proteinuria > 0.5g/day, cellular casts	**1 item** Renal disorder (any of the following): proteinuria > 0.5g/day, cellular casts	**1 item** Renal disorder (Any of the following): urine protein/creatinine ratio or urinary protein concentration of 0.5 g of protein/24 h, Red blood cell casts
Hematologic disorder	**1 item** Hematologic disorder (any of the following): hemolytic anemia, leukopenia (<4000/mm^3) ≥ 2 occasions, thrombocytopenia (<100,000/mm^3)	**1 item** Hematologic disorder (any of the following): hemolytic anemia, leukopenia (<4000/mm^3), thrombocytopenia (<100,000/mm^3)	**1 item** Hematologic disorder (any of the following): hemolytic anemia with elevated reticulocytes, leukopenia <4000/mm^3 on ≥ 2 occasions, lymphopenia < 1500/mm^3 or ≥ 2 occasions, thrombocytopenia <100,000/mm^3	**3 items** •Hemolytic anemia •Leukopenia or lymphopenia (<4000/mm^3, <1000/mm^3 separately at least once) •Thrombocytopenia (<100,000/mm^3) at least once
Immunologic abnormal	**2 items** • LE cells • Chronic false-positive serological test for syphilis	**2 items** • Positive lupus erythematosus preparation, anti-dsDNA and anti-Sm and false-positive for syphilis serological test • Positive ANA	**2 items** • Positive anti-dsDNA, anti-Sm or antiphospholipid antibodies • Positive ANA (by immunofluorescence or an equivalent assay)	**6 items** • Positive ANA • Positive anti-dsDNA (except ELISA) on ≥2 occasions • Anti-Sm • Antiphospholipid antibody (including lupus anticoagulant, false-positive RPR, anticardiolipin, anti-β2 glycoprotein 1) • Low complement (C3, C4 or CH50) • Direct Coombs test in the absence of hemolytic anemia
Diagnosis	**Satisfy 4 or more items**	**Satisfy 4 or more items**	**Satisfy 4 or more items**	**Satisfy 4 items** (with one having to be a clinical item and one having to be an immunologic item), e.g. lupus nephritis, in the presence of at least one of the immunologic variables

Longitudinal Extensive Transverse Myelitis due to SLE

- Patients were subdivided into two clinical forms: partial (non-paralyzing: independence in activities of daily living and mobility were preserved); or complete (paralyzing or incapacitating).
- Partial forms; almost half of our patients had non-paralyzing TM at onset and about a third of these patients had suspected or documented peripheral neuropathy
- Fever has been observed preceding SLE-related TM before.
- Fever was significantly more common in patients with complete TM than in those with partial TM.
- Fever is probably due to a combination of factors including severe inflammation, and also infections related to immobility or bladder catheterization, common in patients with complete TM.

Review

Longitudinally extensive transverse myelitis in systemic lupus erythematosus: Case report and review of the literature

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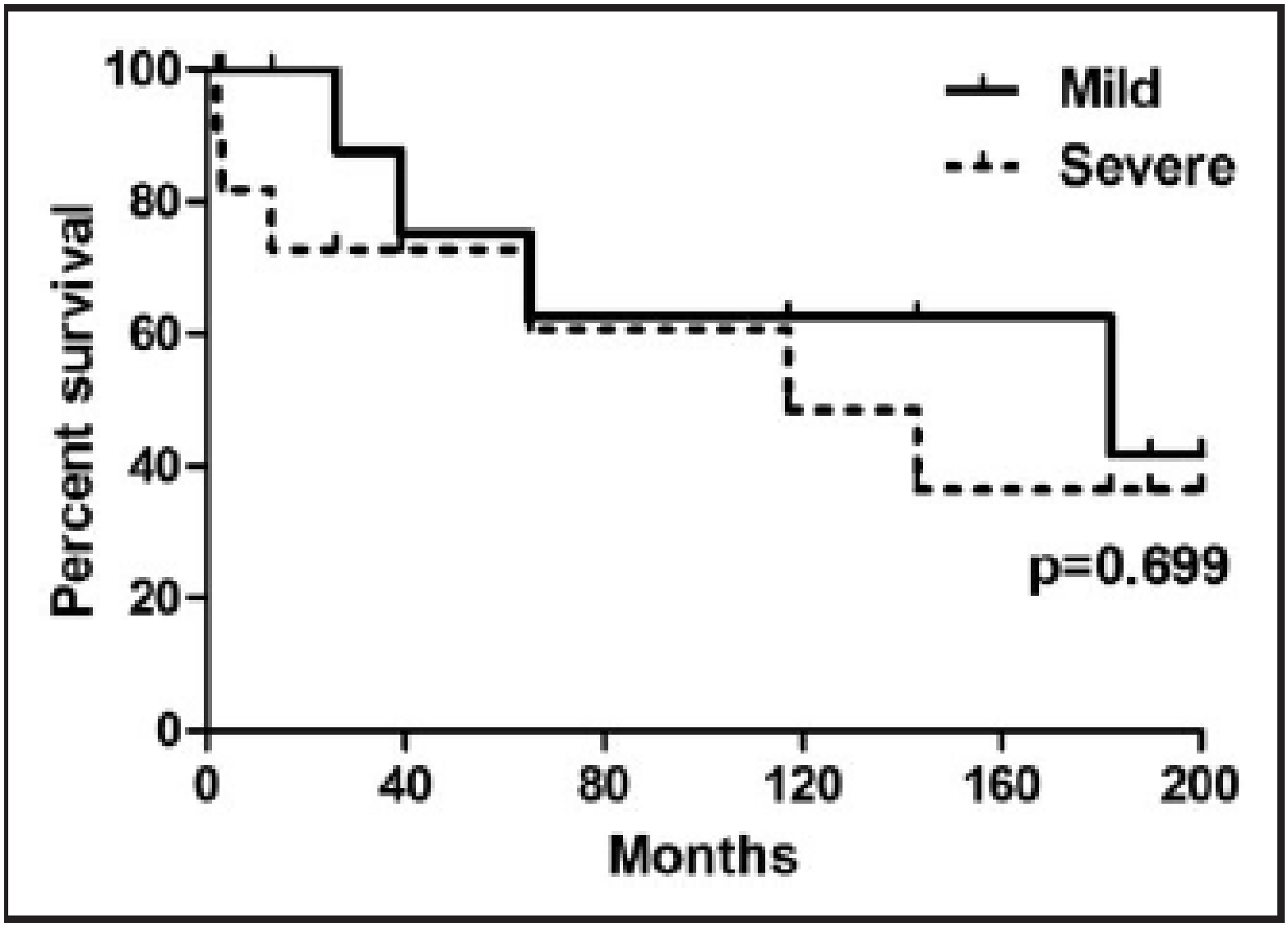
ABSTRACT

Objective: To report a case of longitudinally extensive transverse myelitis (LETM), a rare but disabling condition defined as a lesion of the spinal cord that extends over four or more vertebrae on MRI, in association with systemic lupus erythematosus (SLE).
Methods: We present a rare case of LETM involving the cervical and thoracic spinal cord in a patient with SLE and review the existing literature on the association of lupus-associated myelitis.
Results: LETM is included within the diagnostic criteria for Neuromyelitis Optica (NMO), but is also known to be associated with a wide range of auto-immune diseases. Only 37 cases of LETM in patients with SLE have been previously described. We performed an updated review on epidemiology, pathophysiology, clinical features, diagnosis, management, and prognosis of LETM in the setting of SLE.
Conclusion: Due to the generally poor prognosis of LETM in SLE patients, prompt diagnosis and treatment is of critical importance for a positive clinical outcome. We provide a comprehensive perspective of past and current literature in order to aid diagnosis and management of this rare phenomenon.

Variable	*All (n = 35)*	*Partial (n = 15)*	*Complete (n = 20)*	p *value (partial versus complete)*
Age	31 (24–38)	27 (20–34)	36.5 (27–42)	0.014
Female (%)	33 (94)	13 (86)	20 (100)	0.176
Interval from SLE to myelitis (months)	52 (7–78)	52 (26–78)	15 (0–70)	0.202
SLE-defining criteria	6 (5–8)	6 (5–8)	6 (6–7)	0.438
Malar rash (%)	18 (51)	10 (66)	8 (40)	0.118
Discoid rash (%)	8 (23)	3 (20)	5 (25)	1.000
Photosensitivity (%)	12 (34)	3 (33)	7 (35)	0.918
Oral ulcers (%)	20 (57)	8 (53)	12 (60)	0.693
Non-erosive arthritis (%)	32 (91)	13 (86)	19 (95)	1.000
Pleuritis/pericarditis (%)	19 (54)	9 (60)	10 (50)	0.557
Renal disorder (%)	19 (54)	8 (53)	11 (55)	0.922
Neurological involvement other than myelitis (%)	11 (31)	2 (13)	9 (45)	0.049
Hematological disorder (%)	31 (88)	12 (80)	19 (95)	0.292
Immunological disorder (%)	30 (85)	12 (80)	18 (90)	0.631
Positive antinuclear antibodies (%)	28 (80)	11 (73)	17 (85)	0.430
History suggestive of ON at any time (%)[a]	8 (23)	5 (33)	3 (15)	0.242
SLEDAI	4 (1–11)	5 (0–9)	4 (1–13)	0.626
APS (%) ($n = 34$)	10 (29)	7 (46)	3 (15)	0.382
Positive aPL (aCL, LA, aβ2-GPI) (%)	28 (80)	13 (87)	15 (75)	0.572
aCL	22 (63)	9 (60)	13 (65)	0.596
LA	7 (20)	3 (20)	4 (20)	0.678
aβ2-GPI	16 (45)	8 (53)	8 (40)	0.596
Systemic manifestations (%)				
Fever	16 (45)	4 (26)	12 (60)	0.05
Unintended weight loss	17 (48)	4 (26)	13 (65)	0.025
Raynaud's phenomenon	8 (23)	6 (40)	2 (10)	0.054
Lymph node enlargement	7 (31)	2 (13)	5 (25)	0.672
Cutaneous vasculitis	11 (31)	5 (33)	6 (30)	1.000
Alopecia	18 (51)	5 (33)	13 (65)	0.064
Treatment at onset of myelitis (%)				
Prednisone	25 (71)	10 (67)	15 (75)	0.712
Azathioprine	12 (34)	7 (46)	5 (25)	0.181
Cyclophosphamide	2 (5)	2 (13)	0	0.176
Oral methotrexate	1 (3)	0	1 (5)	1.000
CLQ/HCQ	12 (34)	5 (33)	7 (35)	0.918
Aspirin	5 (14)	3 (20)	2 (10)	0.631
Oral anticoagulants	4 (11)	3 (20)	1 (5)	0.292

Table 2 Myelitis-related findings

	All (n = 35)	*Partial (n = 15)*	*Complete (n = 20)*	p *value (partial versus complete)*
Interval from first TM-related neurological manifestation to diagnosis of TM (days)	10 (3–23)	9 (1–25)	10 (6–23)	0.502
Clinical presentation of myelitis (%)				
Motor	33 (94)	13 (86)	20 (100)	0.176
Sensory	34 (97)	15 (100)	19 (95)	1.000
Muscle stretch reflexes	29 (83)	14 (93)	15 (75)	0.207
Sphincter	24 (68)	11 (73)	13 (65)	0.721
Spine MRI: involved region (%)				
Brainstem	9 (26)	2 (13)	7 (35)	0.224
Cervical	25 (71)	13 (86)	12 (60)	0.142
Thoracic	28 (80)	12 (80)	16 (80)	0.350
Lumbar-cone	15 (43)	6 (40)	9 (45)	0.754
Spine MRI: other findings (%)				
Enlarged	15 (43)	4 (26)	11 (55)	0.094
Contrast enhancement	9 (25)	3 (20)	6 (30)	0.729
Myelitis-specific treatment (%)				
Methylprednisolone (IV)	21 (60)	7 (46)	14 (70)	0.163
Cyclophosphamide (IV)	19 (54)	5 (33)	14 (70)	0.048
Oral corticosteroids	16 (45)	8 (53)	8 (40)	0.433
Azathioprine	5 (14)	5 (33)	0	0.004
Myelitis-related hospital stay (days)	9 (6.5–27)	7 (0–10)	23 (8–43)	0.003
30-day mortality (%)	0	0	0	–
Long-term mortality (%)	11 (31)	4 (26)	7 (35)	0.721


Mild
Severe
p=0.699
Percent survival
100
80
60
40
20
0
0
40
80
120
160
200
Months

Meta-analysis results for the relationship between risk factors and poor neurological outcome in SLE-related TM patients.

Variables	No. of studies	Number of enrolled patients	Effects model	I^2 (%)	OR/SMD (95%CI)	P values	Publication bias (*P* value of Begg's test)
Demographic and clinical characteristics							
Age of onset of myelitis	5	272	Random	75.10%	-0.08 (−0.69, 0.53)	0.80	None (0.81)
SLEDAI score	4	111	Random	59.50%	0.39 (−0.31, 1.09)	0.28	None (0.73)
An AIS grade of A	4	119	Fixed	20.10%	11.22 (4.00, 31.47)	<0.001	None (0.09)
An AIS grade of A or B	4	119	Fixed	46.30%	21.78 (7.29, 65.06)	<0.001	None (1.00)
An AIS grade of A, B or C	4	119	Random	50.60%	56.05 (6.29, 499.25)	<0.001	None (0.31)
Laboratory and image data							
aPLs	5	225	Fixed	0.00%	0.92 (0.52, 1.62)	0.78	None (0.81)
aCL	5	206	Fixed	0.00%	0.64 (0.33, 1.21)	0.17	None (0.22)
Anti-β2GPI	4	114	Fixed	46.20%	1.00 (0.39, 2.52)	0.99	None (1.00)
LA	5	171	Fixed	0.00%	0.83 (0.40, 1.71)	0.61	None (0.46)
Drop of complement	3	99	Fixed	0.00%	1.42 (0.58, 3.43)	0.44	None (1.00)
Anti-dsDNA antibody	5	199	Fixed	43.00%	1.06 (0.57, 1.97)	0.86	Yes (0.027)
Hypoglycorrhachia	3	95	Fixed	38.20%	10.78 (3.74, 31.07)	<0.001	None (0.30)
LETM	5	238	Fixed	0.00%	1.62 (0.87, 3.01)	0.13	None (0.46)
Treatment							
Methylprednisolone pulse	4	256	Fixed	12.90%	1.54 (0.74, 3.24)	0.25	None (0.73)
CTX treatment	5	270	Fixed	0.00%	1.14 (0.67, 1.96)	0.63	None (0.46)

Thank You for Your Attention