SOME SPINAL SYNDROMES: COMMON AND UNCOMMON

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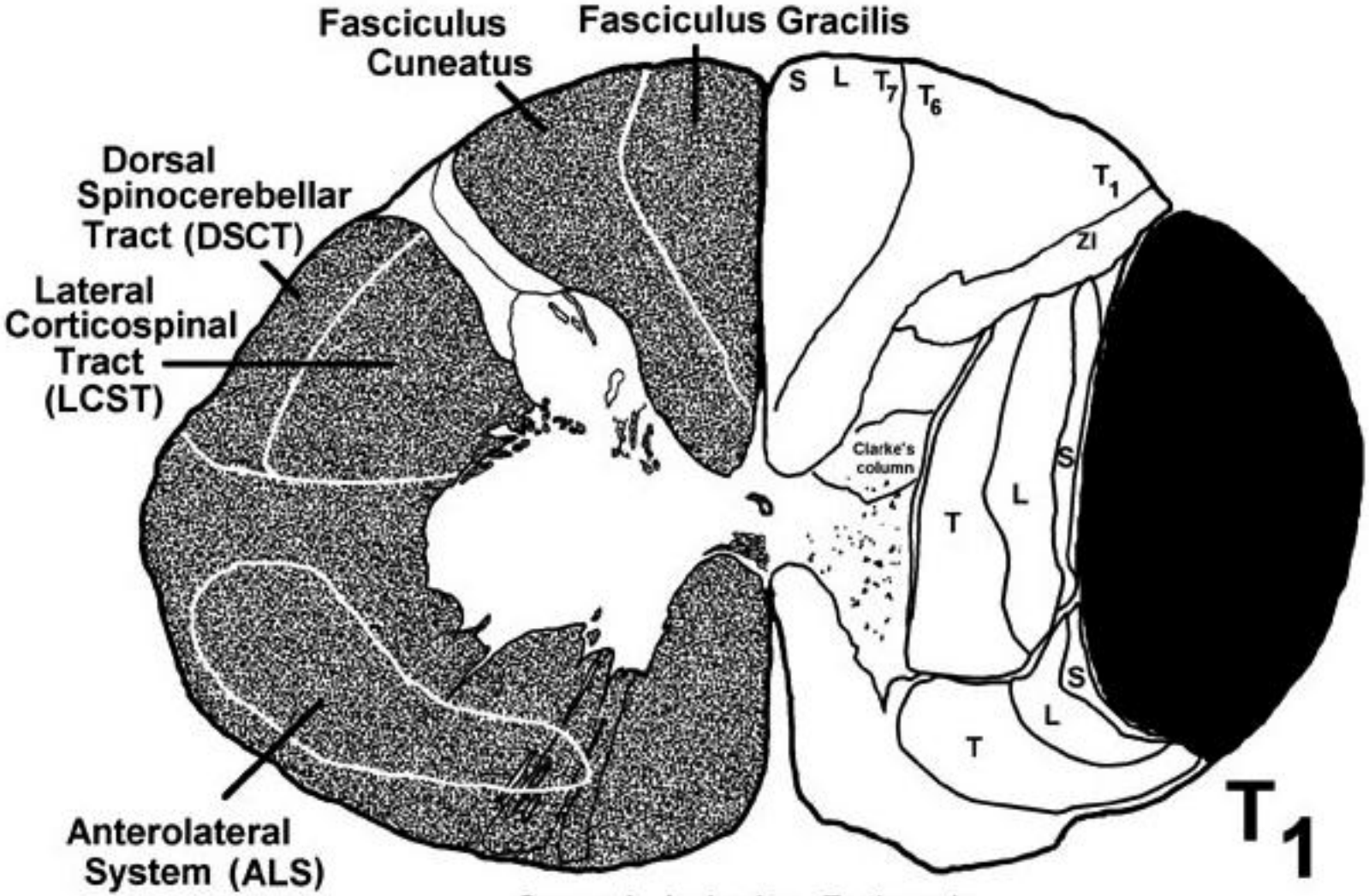
Outline

- Spinal syndromes
  - Classical
  - Less common
- Clinical aspects of
  - disc disease / spondylosis
  - primary and secondary neoplasia
  - vascular disease
  - Infection

Clinical approach to spinal cord disease

Classical cord lesions

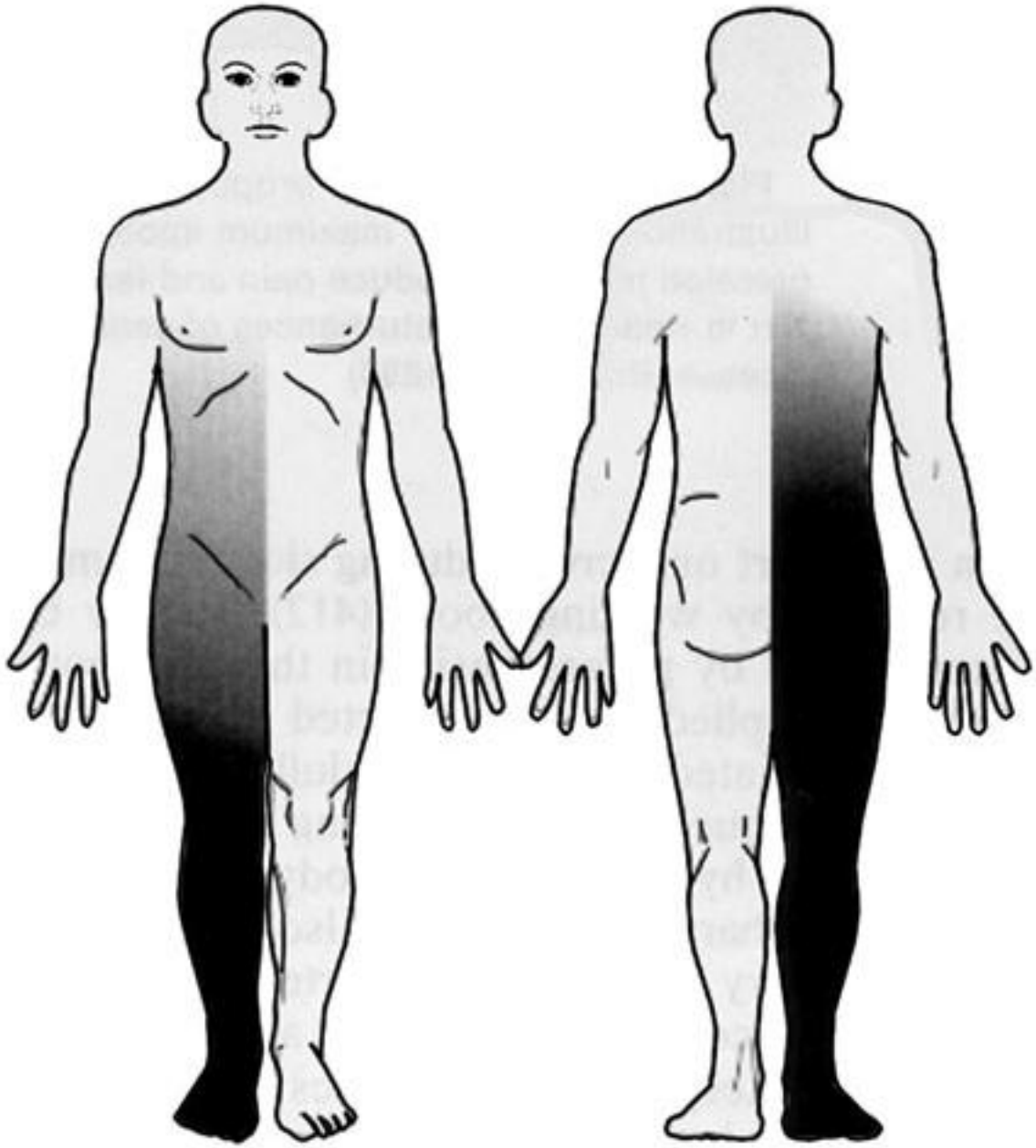
- Extrinsic compression
- Intrinsic cord lesion with sacral sparing
- Central cord lesion with suspended sensory level (syrinx syndrome)
- Brown-Séquard lesion
- Anterior spinal artery syndrome

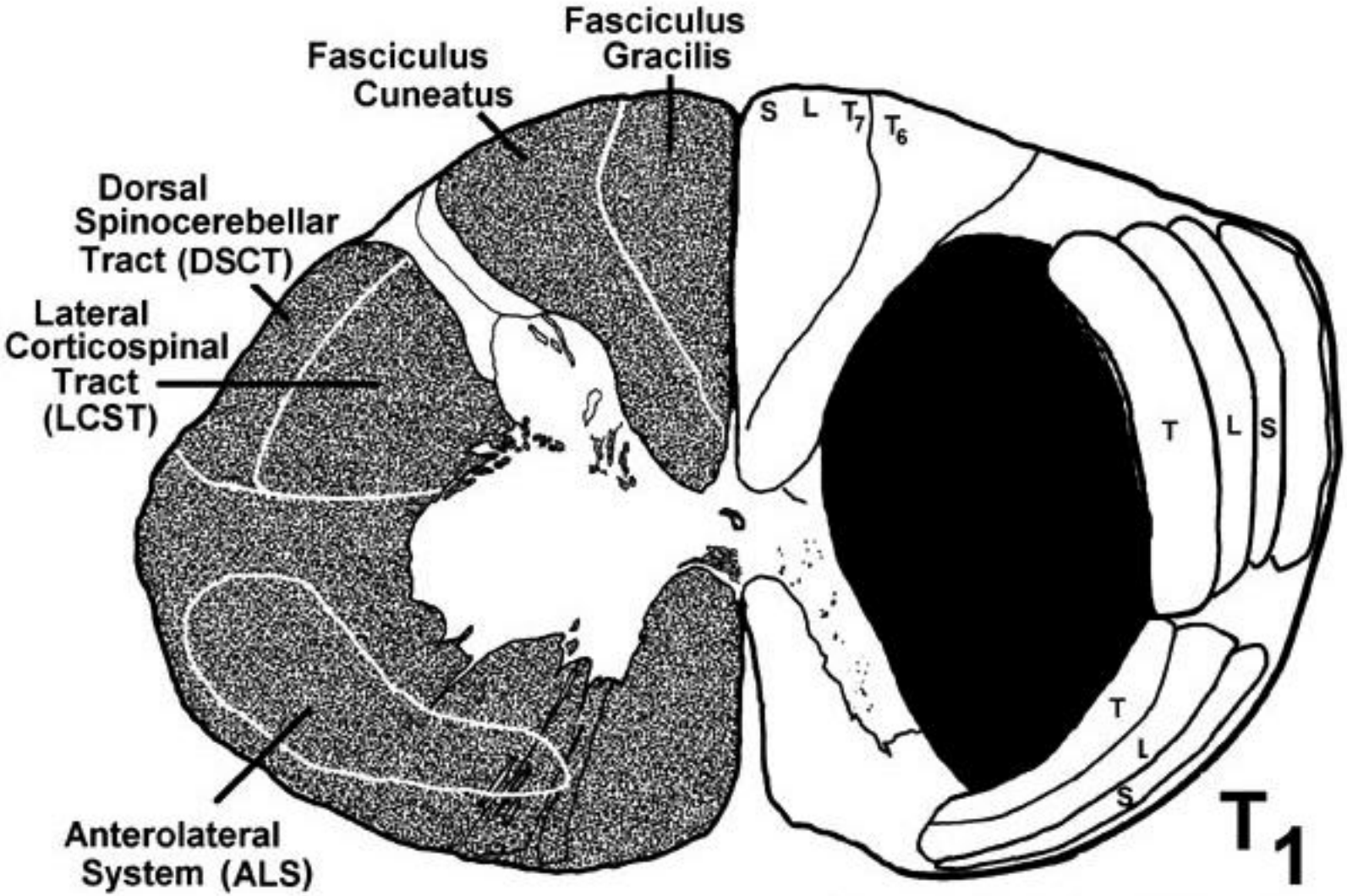


S- sacral L - lumbar T - thoracic

ZL - Zone of Lissauer
I - Marginal zone of the dorsal horn
II - Substantia Gelatinosa

VII - Intermediate zone (Includes neurons of Clarke's column C_8 - L_2)
IX - Motor Nuclei





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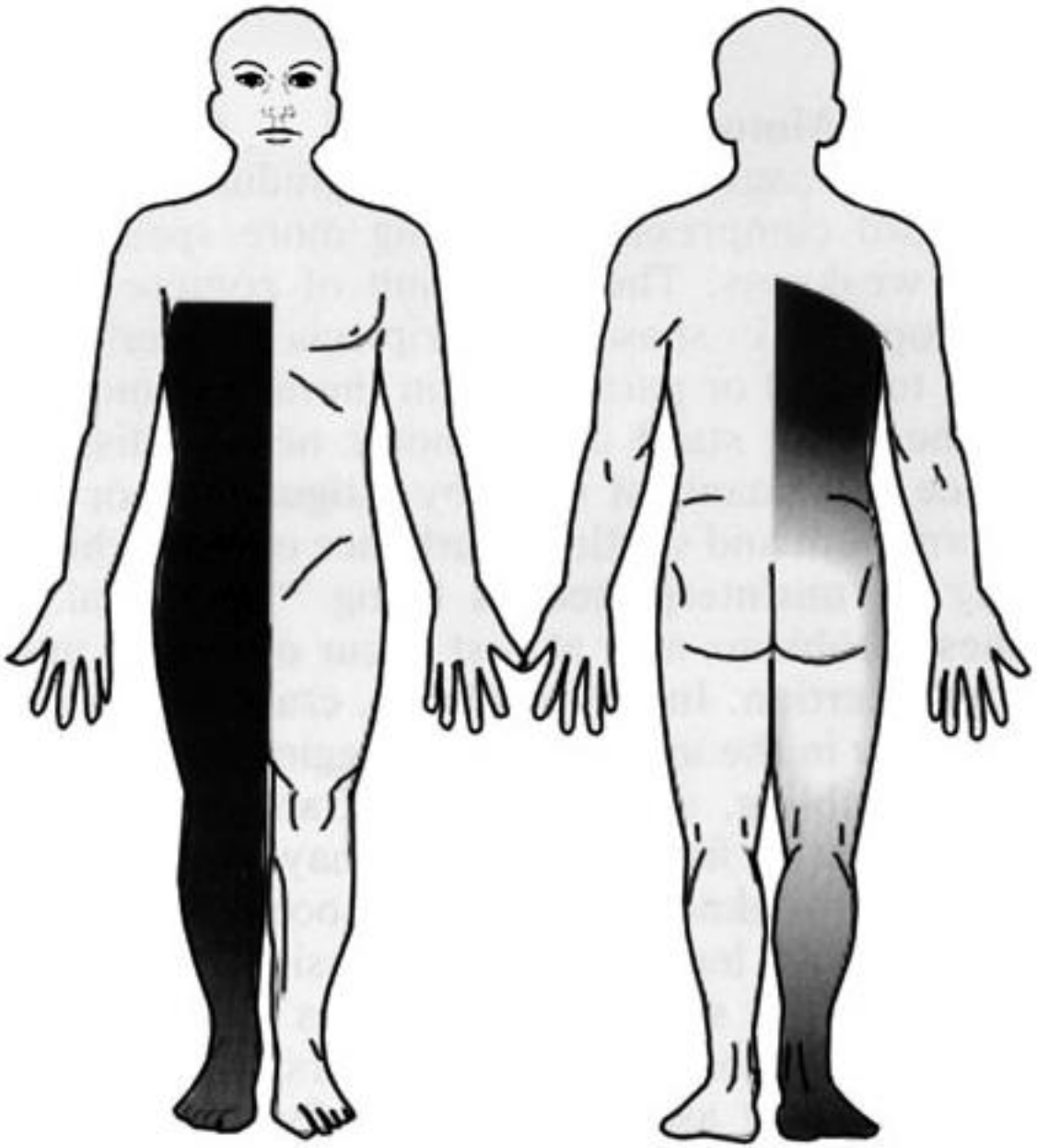

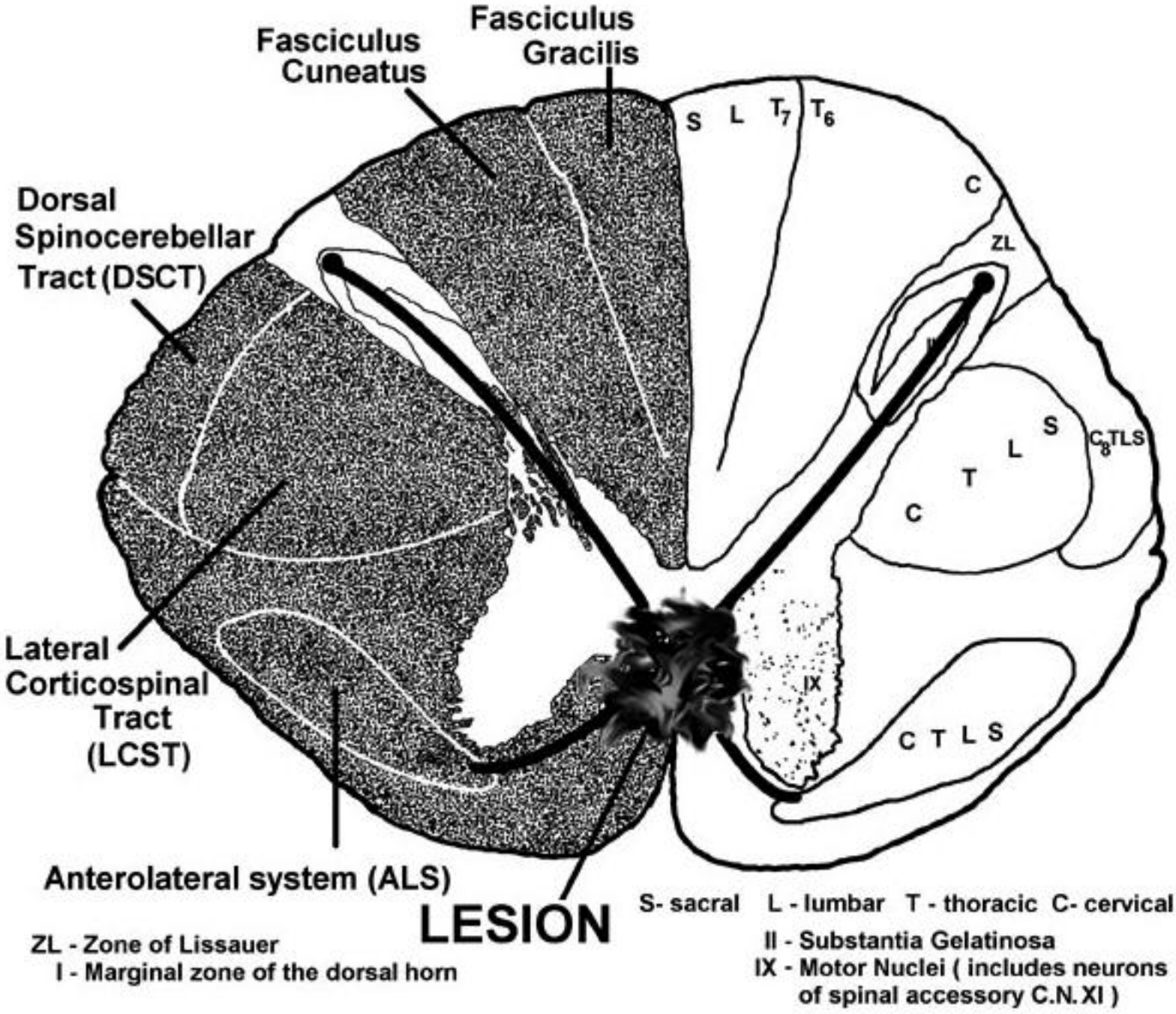
Fasciculus Cuneatus
Fasciculus Gracilis
Dorsal Spinocerebellar Tract (DSCT)
Lateral Corticospinal Tract (LCST)
Anterolateral system (ALS)
LESION
S L T7 T6
C
ZL
S
L
T
C
C8TLS
IX
C T L S
ZL - Zone of Lissauer
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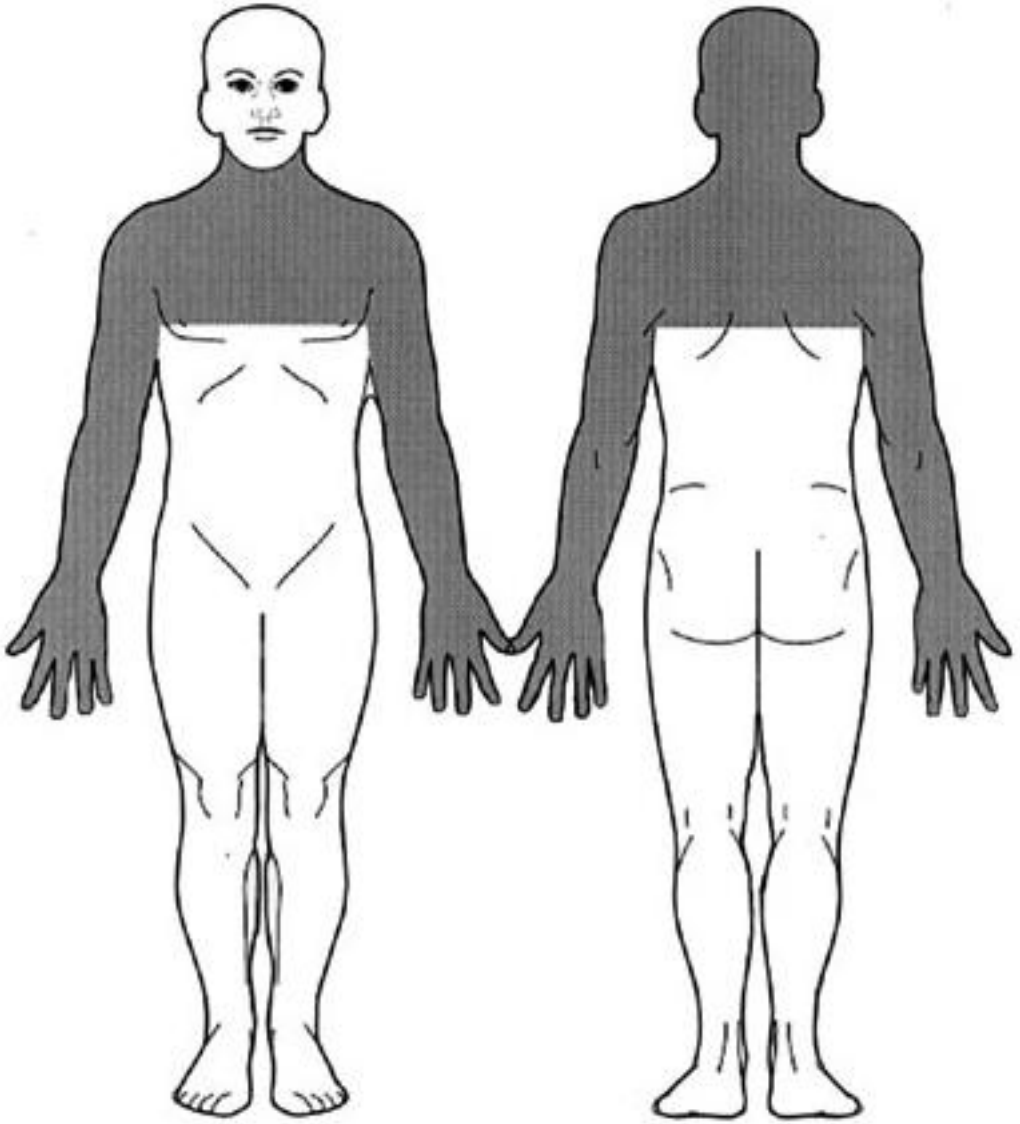
II - Substantia Gelatinosa
IX - Motor Nuclei (includes neurons of spinal accessory C.N. XI)



The lesion in this case involves the decussating fibers of the ALS, or spinothalamic tract, for the entire cervical spinal cord

A 30 yo woman develops weakness of the right leg over a few days

There is moderate weakness in all R leg muscles. Tone is increased, ankle clonus present.

There is also subtle weakness of R biceps & triceps and reduced dexterity R fingers

Where is the lesion?
Try to guess sensory findings

Reflexes:

BJ	±	+
BRJ	±	+
TJ	+++	+
FJ	+++	+
KJ	+++	+
AJ	+++	+
Pl	?x	?x

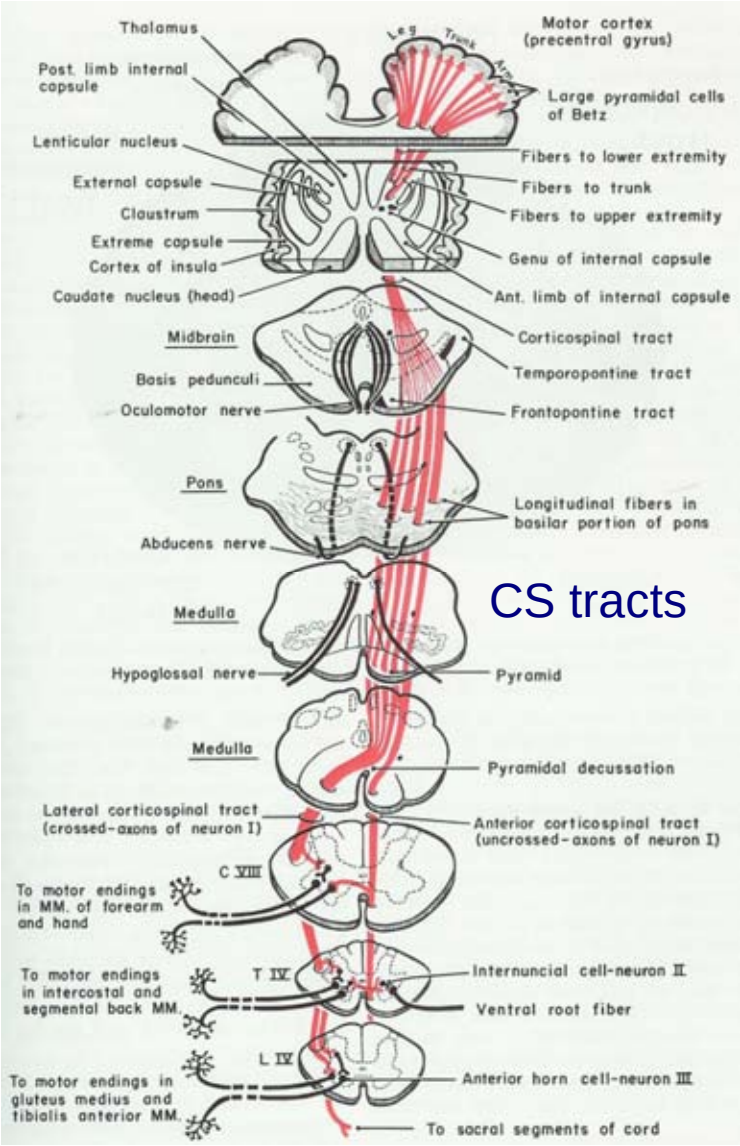
Sensation:

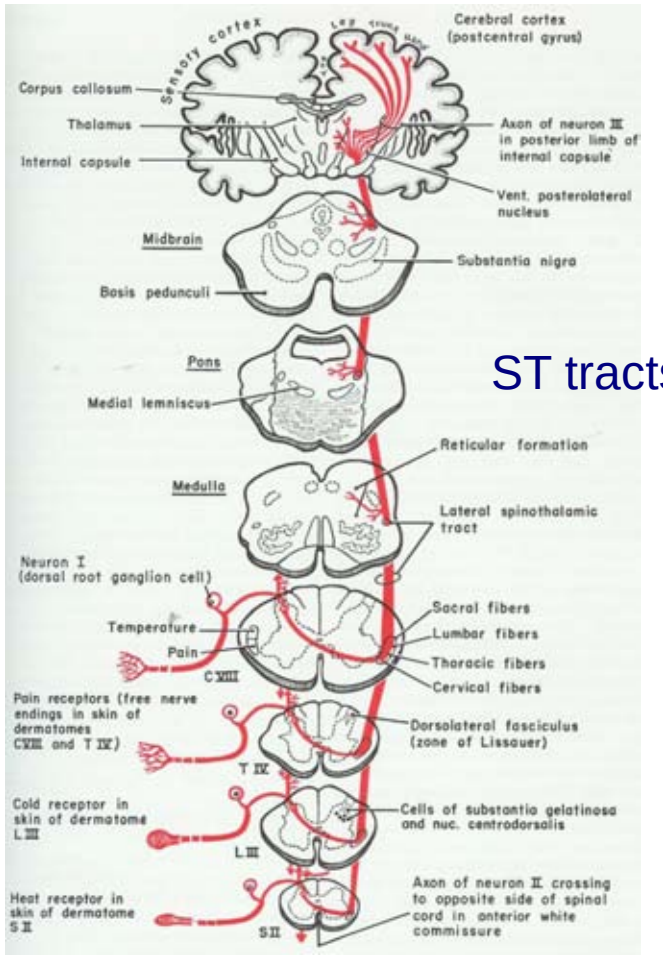
JPS and VS slightly reduced in R foot

PP & TM reduced on ***left*** side below nipple line (i.e. about T4)

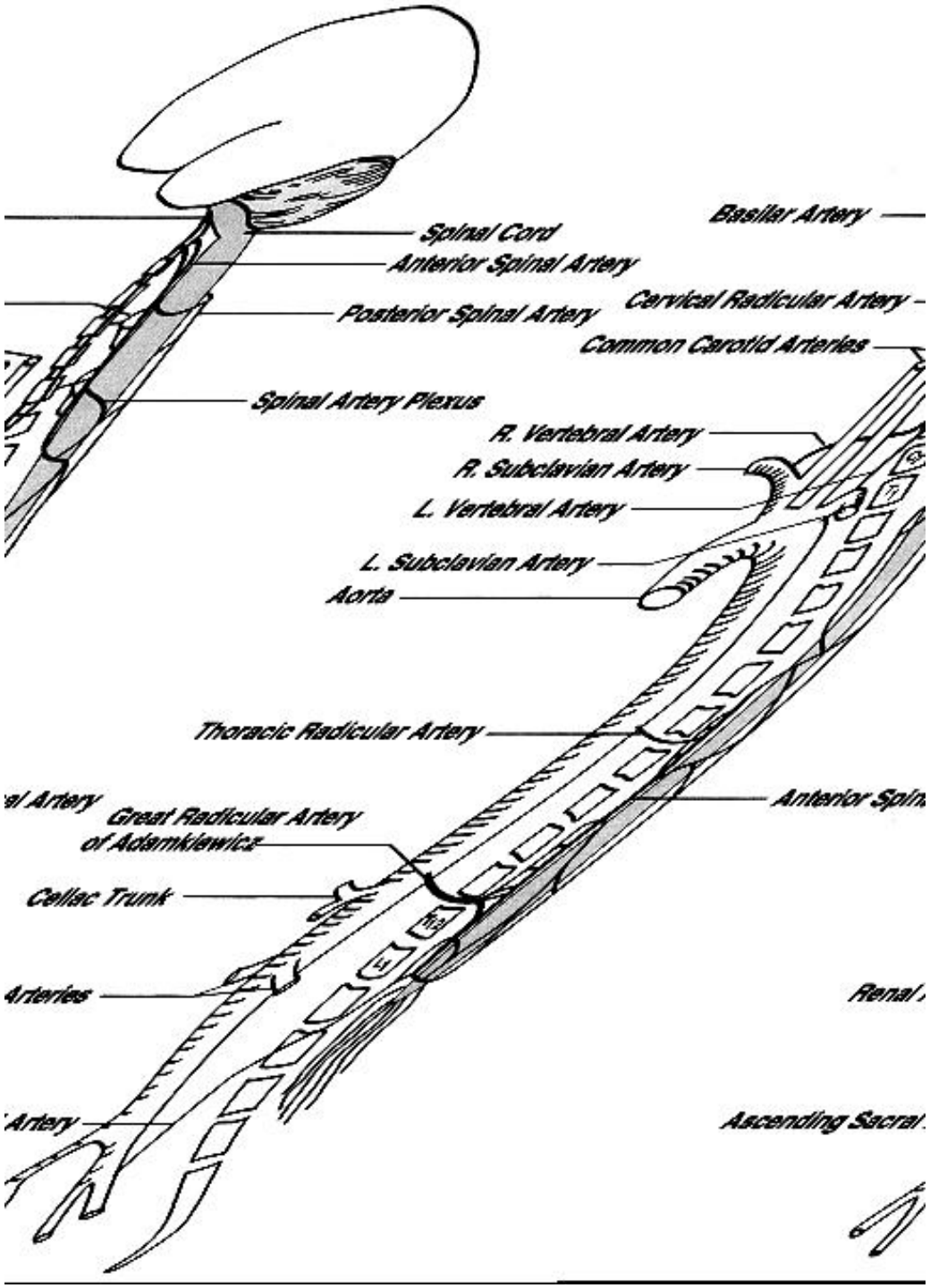
PP & TM reduced in right thumb & index finger

Remember corticospinal tracts cross in the medulla (above lesion in this case) and spinothalamic tracts cross just above the site of entry of sensory roots into the cord

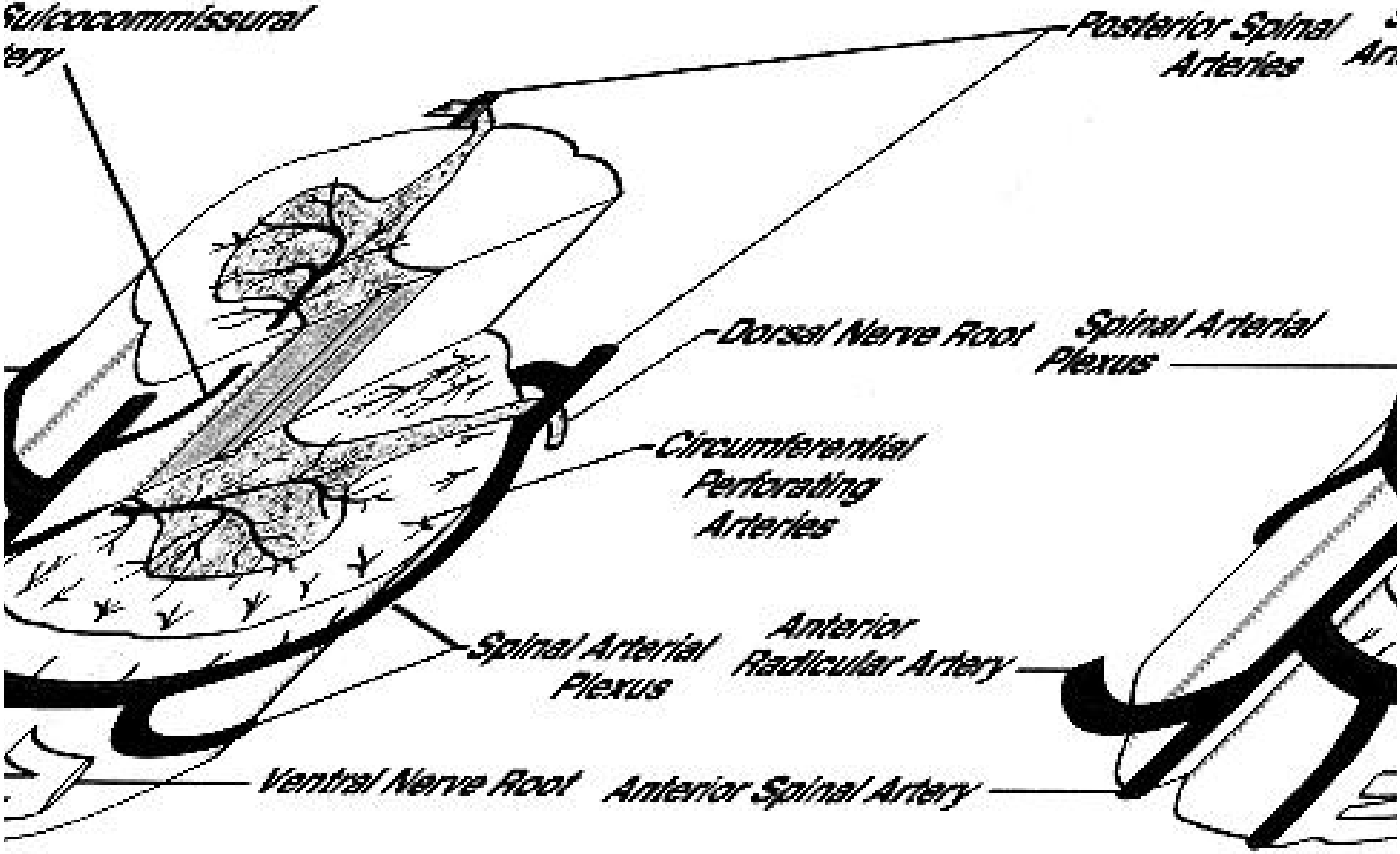




Spinal cord circulation



Spinal cord circulation



Some other spinal syndromes

- Traumatic Central cord syndrome
- Foramen magnum / High cervical cord compression
- Posterior column syndromes
- Excessive cord mobility
- Spinal canal stenosis

Central Cord Syndrome: confused terminology

From Harrison's: "The central cord syndrome results from damage to the gray matter nerve cells and crossing spinothalamic tracts near the central canal. In the cervical cord, the central cord syndrome produces arm weakness out of proportion to leg weakness and a "dissociated" sensory loss signifying a loss of pain and temperature sense in a cape distribution over the shoulders, lower neck, and upper trunk in contrast to intact light touch, joint position, and vibration sense in these regions. Trauma, syringomyelia, tumors, and anterior spinal artery ischemia are main causes."

Confuses traumatic "central cord syndrome" with "syrinx syndrome" – in fact they are totally different

Traumatic Central Cord Syndrome

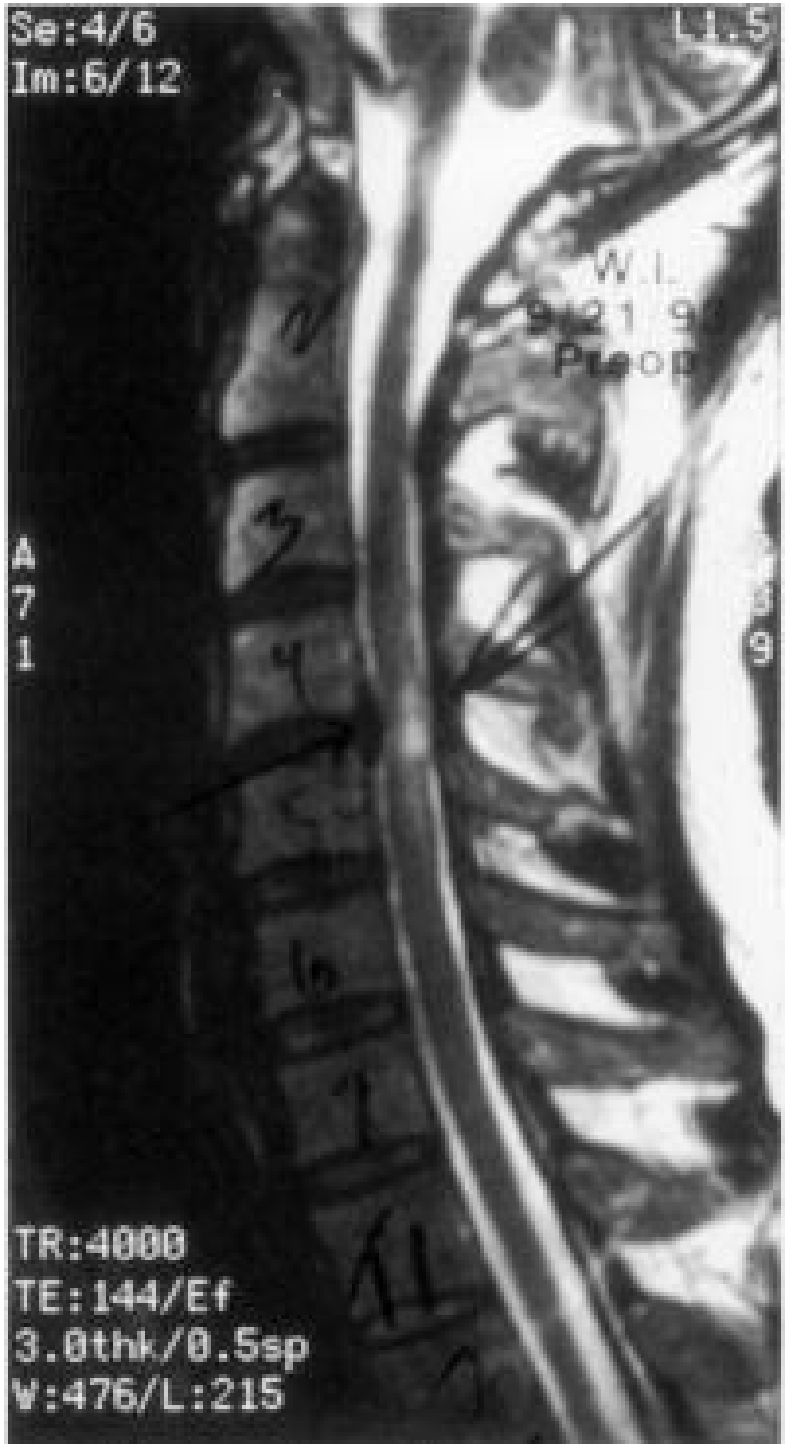
- Typically cause by acute extension injury in a patient with pre-existing cervical spondylosis
- Cord is contused against a spondylotic bar
- Typically motor loss arms>legs:
  - “man in a barrel” (whole of upper limb)
  - “flipper hands” (hands predominantly affected)
- Sensory, lower limb motor and sphincter dysfunction are usually relatively spared
  - (sometimes dorsal column loss in hands)

Traumatic Central Cord Syndrome

- Usually no or mild **ongoing** cord compression, so surgery is not helpful
- Debate about pathogenesis, but a histopathological analysis in 5 cases suggests:
  - In acute cases & chronic high cervical cases, the most obvious pathology was in lateral cortico-spinal tracts
  - Chronic cases with low cervical injuries did lose some LMNs at C7-T1
- Signs vary, but often brisk finger jerks and disproportionate loss of dexterity

Traumatic Central Cord Syndrome: acute radiology.

Note congenitally narrow canal, moderately severe, but not critical, stenosis at the spondylotic level and cord signal change



Lesions of the Foramen Magnum

Pyramidal tract fibres are decussating.

Fibres to legs cross below those to the arms

Patterns of weakness and progression may be unusual

E.g. weakness of one arm, then the ipsilateral leg, then the other leg, and finally the other arm (an **"around the clock" pattern** that may begin in any of the four limbs).

There is typically suboccipital pain spreading to the neck and shoulders.

Atypical / “false localizing” signs from high cord compression

- Predominant involvement of the hands, with proprioceptive loss, paraesthesias and atrophy, can occur with disorders afflicting the upper cervical spinal cord.
- The signs might thus suggest lower cord pathology

Atypical / “false localizing” signs from high cord compression

One review: 11 patients with an extradural lesion above C4 (most commonly C3-C4).

All had a syndrome of **finger and hand dysaesthesias**, **hand atrophy**, and occipital or cervical pain.

These complaints usually preceded the development of spasticity and gait disturbance.

Pathophysiology: theories include

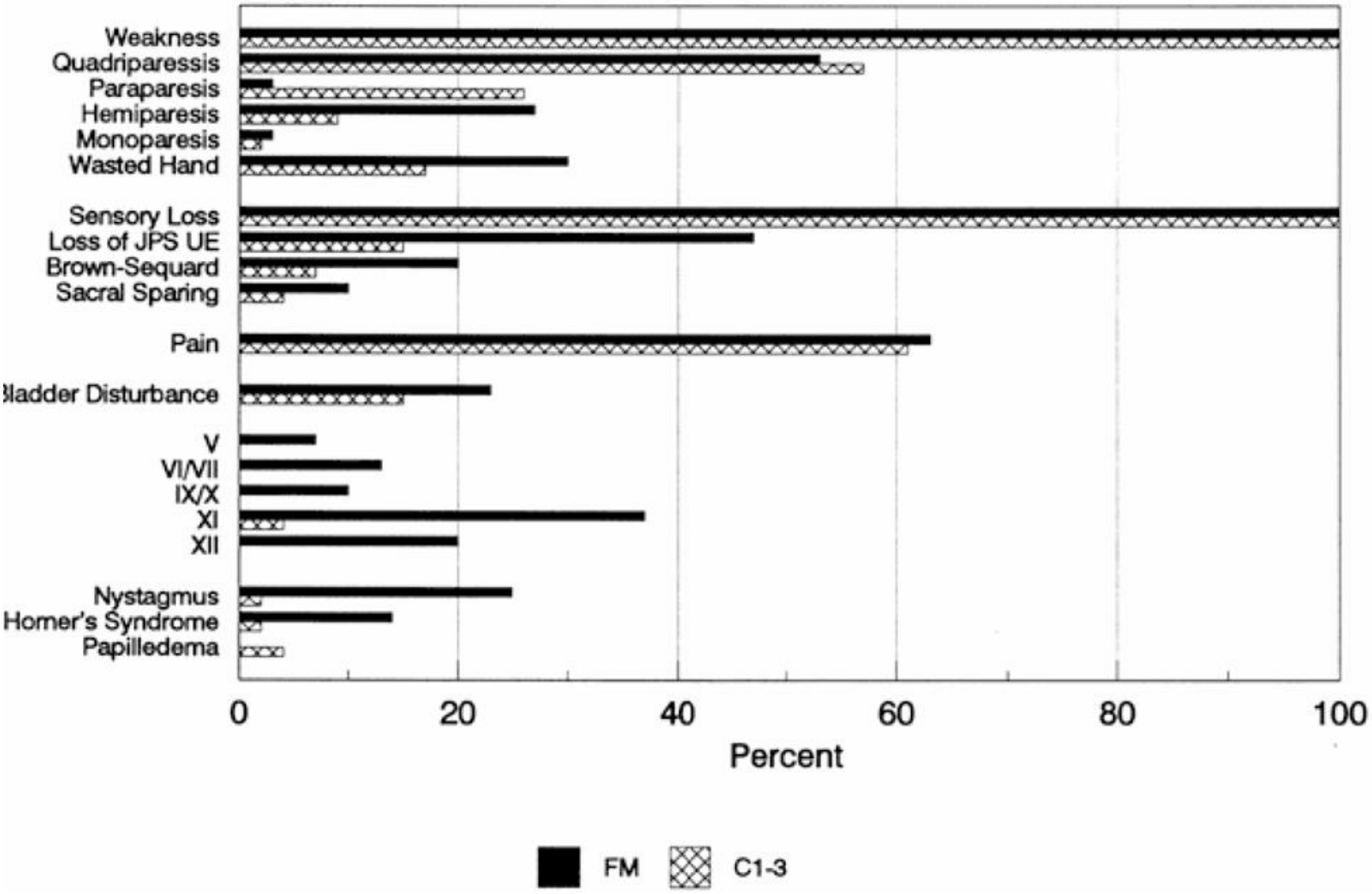
- anterior spinal artery ischemia,
- **venous obstruction**, and
- differential decussation of the forelimb and hindlimb fibers of the corticospinal tract.

Clinical features in 11 patients with extradural cord compression above C4

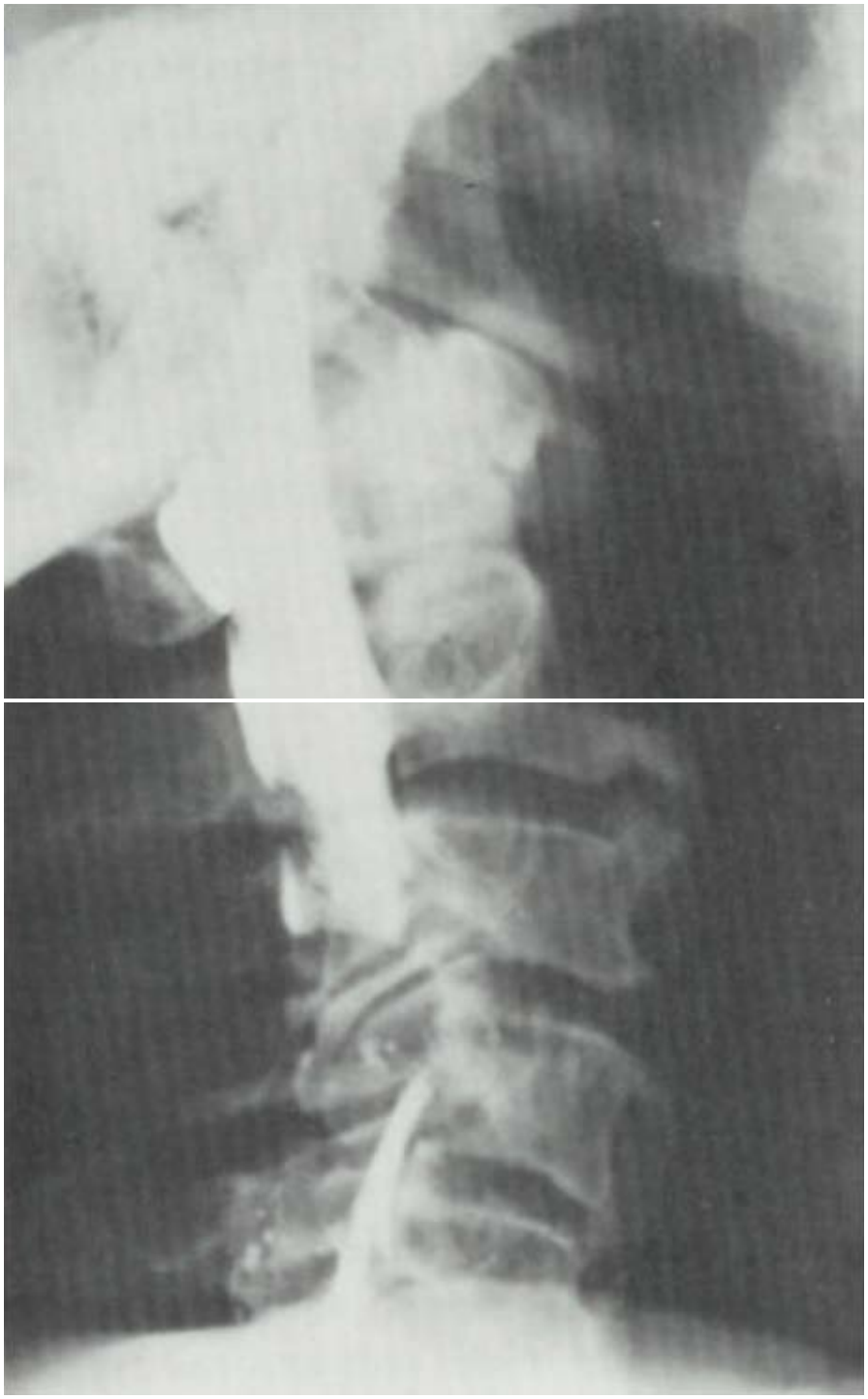
(Sonstein, Neurosurgery 1996)

Symptom	No. of Patients	%
Dysesthesias		
Hands	5	45
Arm	2	18
Lower extremity	1	9
Numbness in hands	5	45
Weakness		
Hands	6	55
Arm	5	45
Lower extremity	6	55
Atrophy		
Hands	9	82
Arm	1	9

Clinical features of foramen magnum and high cervical (C1-C3) tumours

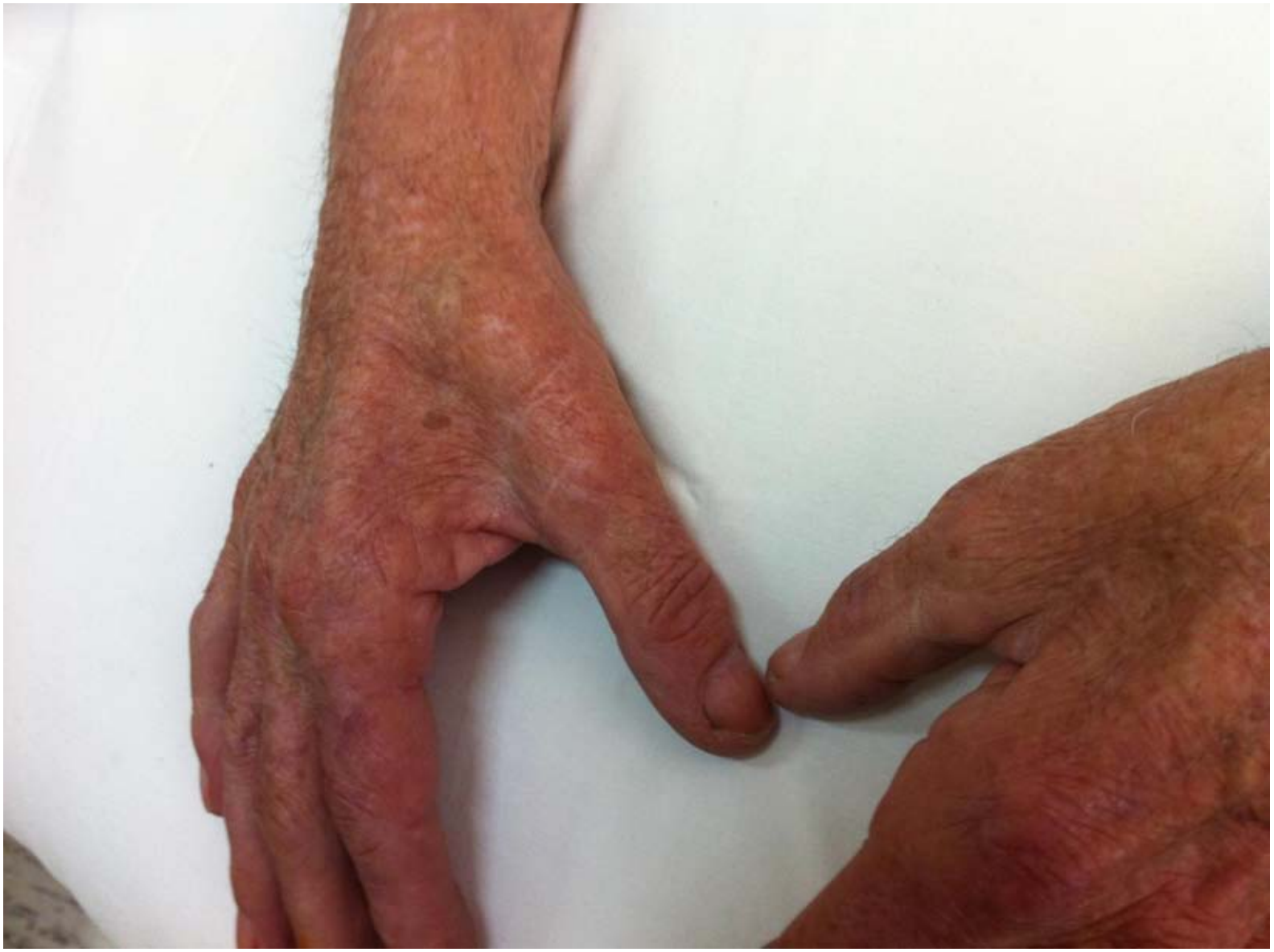


Patient presented with wasted and weak small muscles of hand

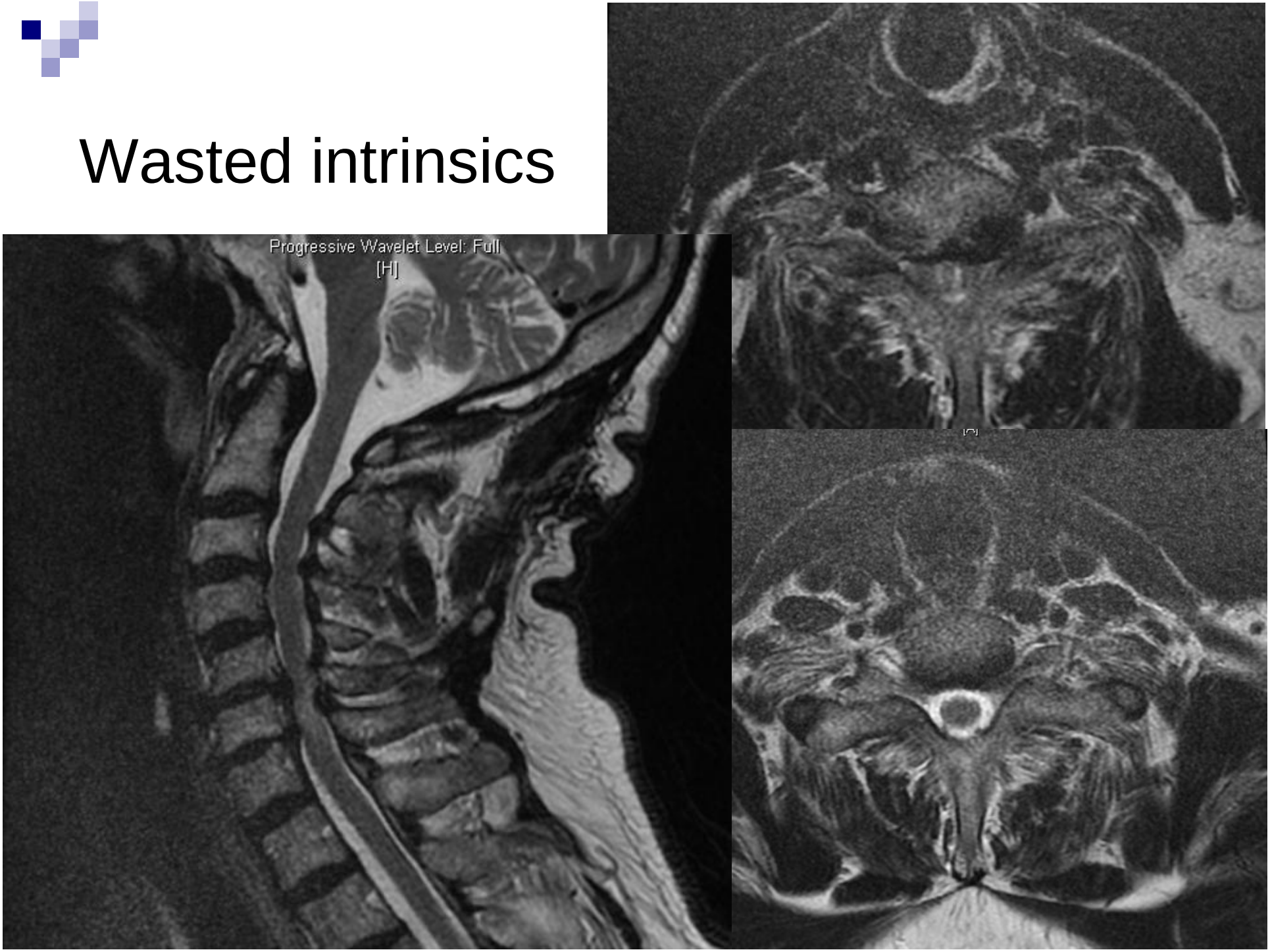


Stark RJ**,** Kennard C, Swash M. Hand wasting in spondylotic high cord compression; an electromyographic study. *Annals of Neurology* 1981; 9:58-62.

Cervical cord compression C5-6-7



Wasted intrinsics



Dural defects in the spine

Cause CSF leak and may result in:

- Low CSF pressure syndrome

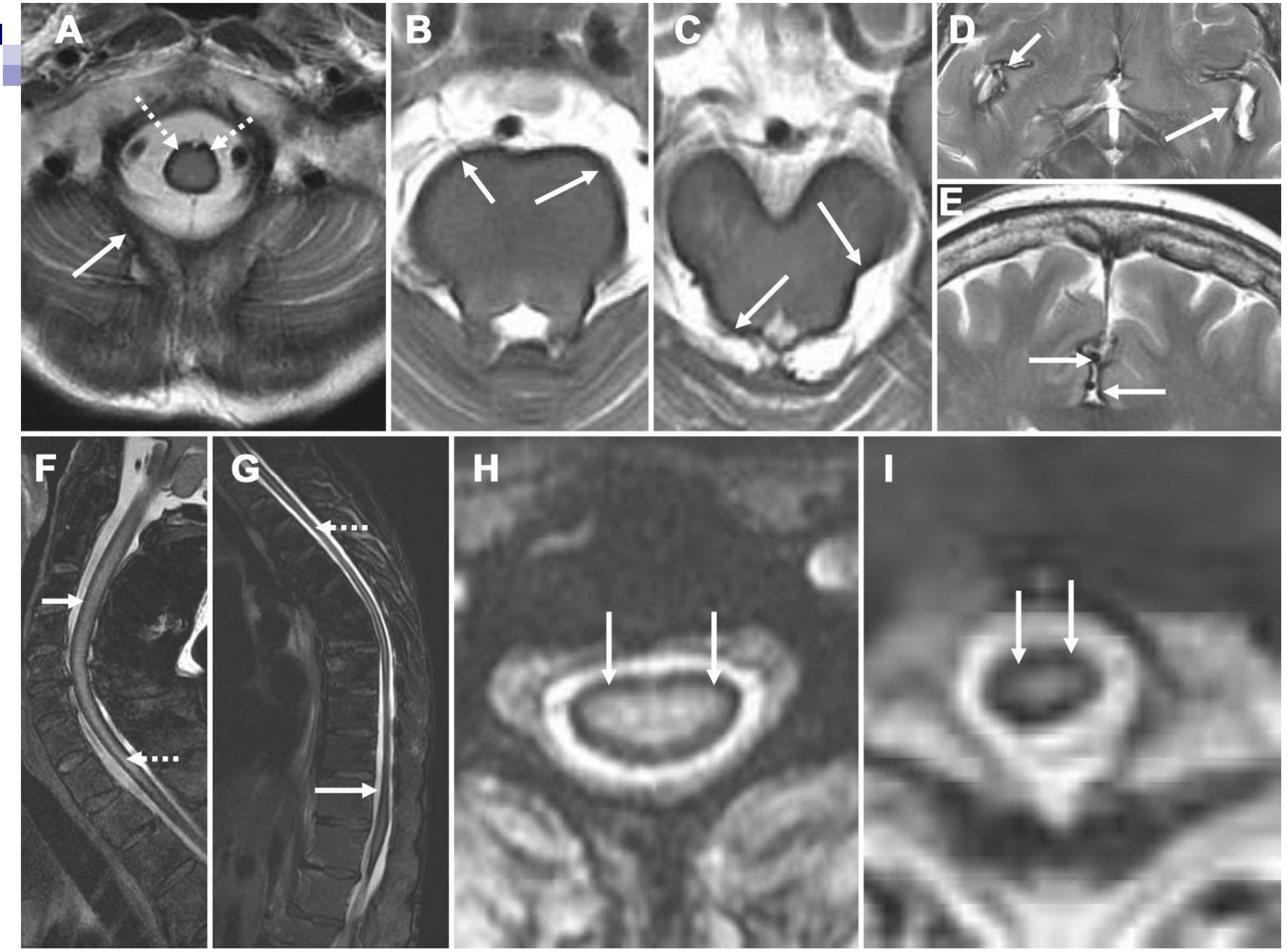
  Postural headache, pachymeningeal enhancement on Brain MRI etc

- Superficial siderosis on brain or spine MRI
- Epidural CSF collections in the spine

  (which can be a cause of upper limb amyotrophy)

Deluca GC, Boes CJ, Krueger BR, Mokri B, Kumar N. Ventral intraspinal fluid-filled collection secondary to CSF leak presenting as bibrachial amyotrophy. Neurology 2011;76:1439 –1440.

Kumar N. Beyond superficial siderosis: Introducing “duropathies”. Neurology 2012;78:1992-1999.



MRI in superficial siderosis

Upper limb amyotrophy in VLISFC

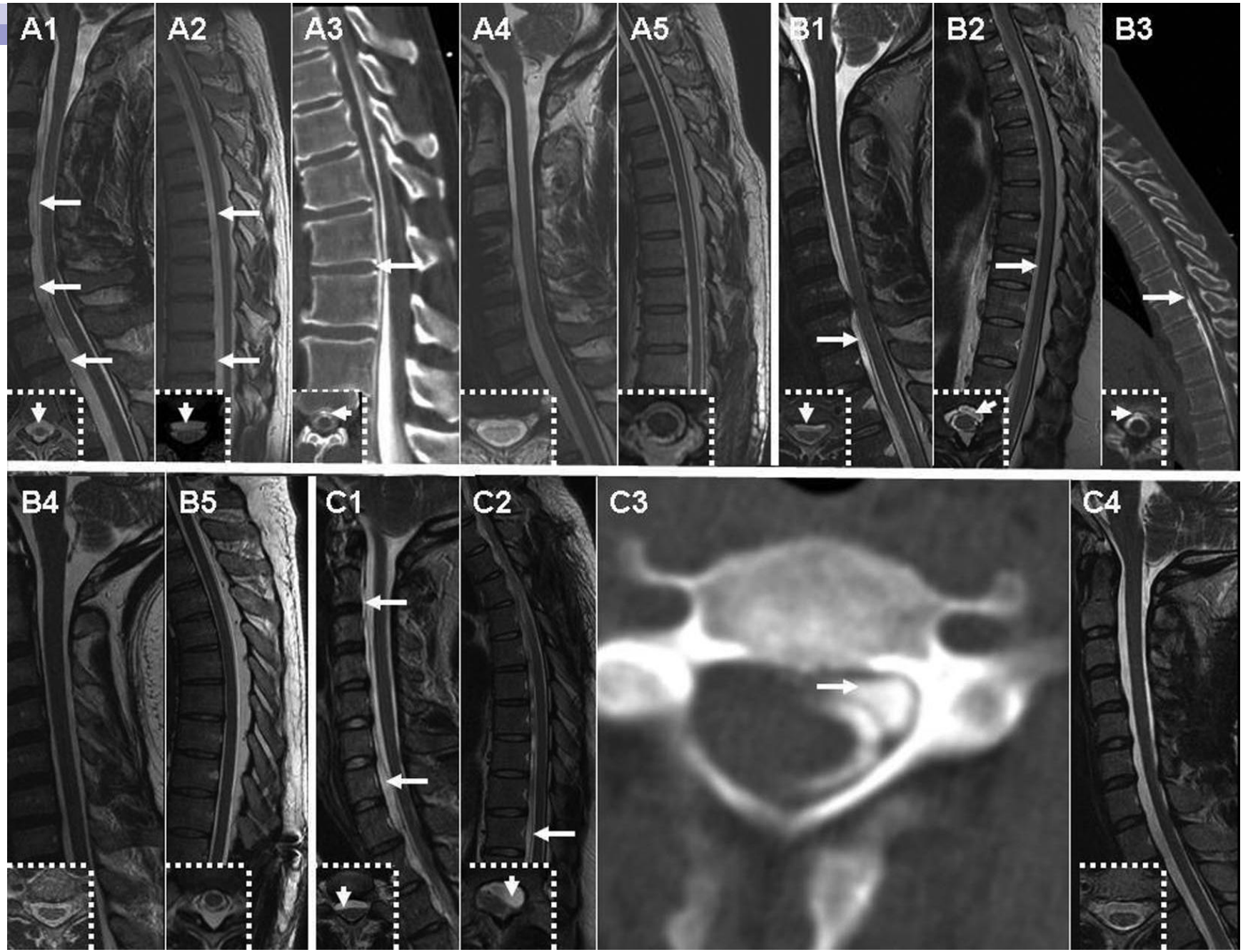
A 48-year-old man presented with a 5-year history of progressive, bilateral, asymmetric, proximal greater than distal, upper limb atrophy and weakness. EMG demonstrated chronic neurogenic changes in the C4–C8 myotomes.

A 40-year-old man presented with a 2-year history of progressive, asymmetric, upper limb atrophy and weakness. EMG demonstrated chronic neurogenic changes in the cervical and upper thoracic myotomes.

A 32-year-old man presented with a 10-year history of progressive, proximal greater than distal, right upper limb weakness and atrophy and a 4-month history of progressive distal left upper limb weakness and atrophy. MRI brain revealed brain sag suspicious for intracranial hypotension. EMG showed chronic neurogenic changes in the C5–T1myotomes.

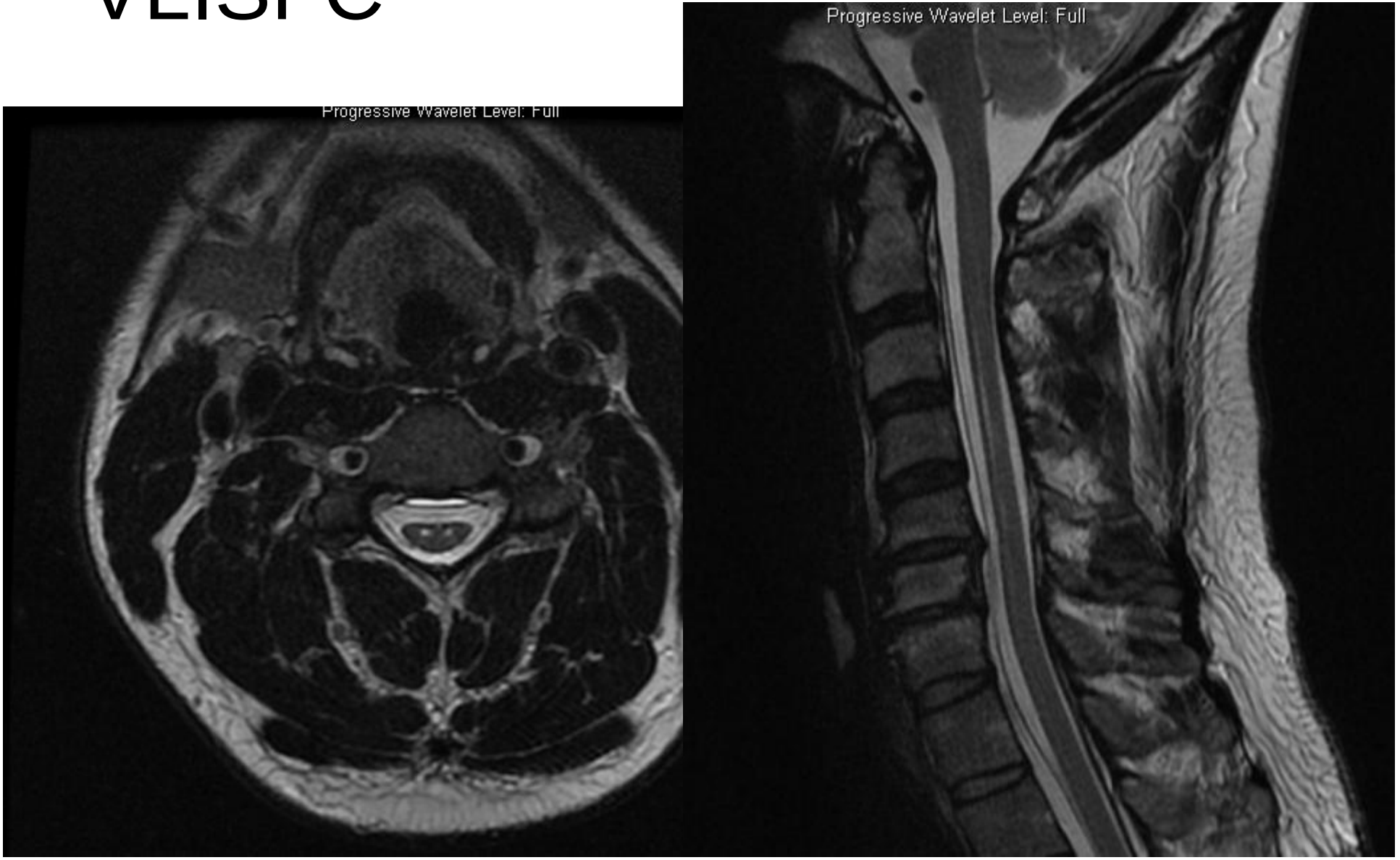
All had VLISFC on spinal imaging

Deluca GC, et al Neurology 2011;76:1439 –1440.



Ventral longitudinal intraspinal fluid collection

VLISFC


Progressive Wavelet Level: Full
Progressive Wavelet Level: Full

3 different cases: same problem

- 60 year old artist ?neuropathy 2 years numbness in hands and feet without weakness
- Ataxic gait; hypertonicity of lower limbs
- Weak L iliopsoas and R triceps and APB
- Brisk reflexes to the pectoral jerks but no trapezius jerks. Plantars extensor
- Decreased VS and JPS left hand

58 year old woman numb hands; sister with MS

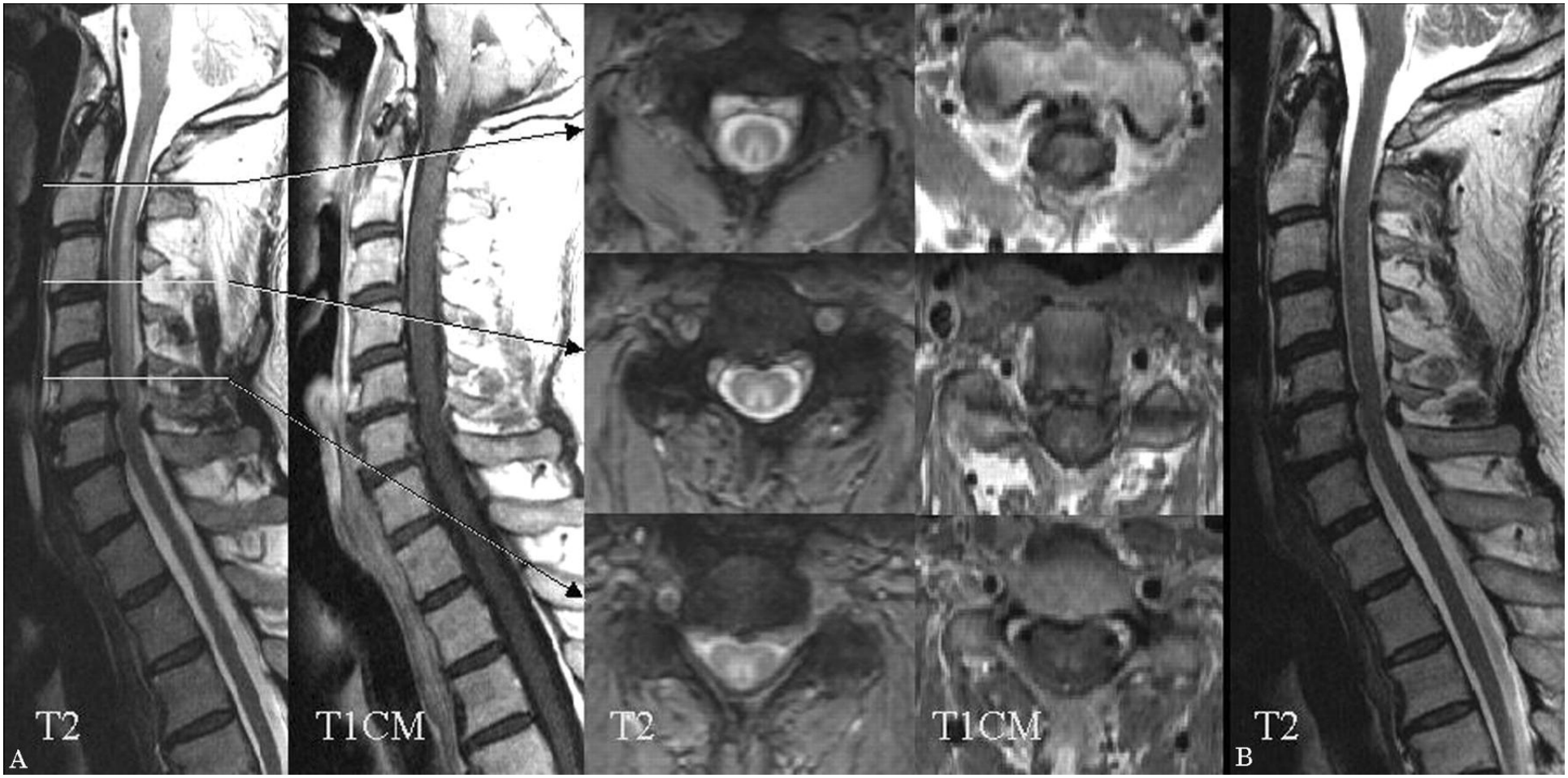
- 6 months hand numbness dropping things
- Also slightly unsteady on feet
- Romberg -ve
- JPS down in fingers
- Tinel’s -ve
- Reflexes brisk
- Plantars flexor

28 yo woman: post-op

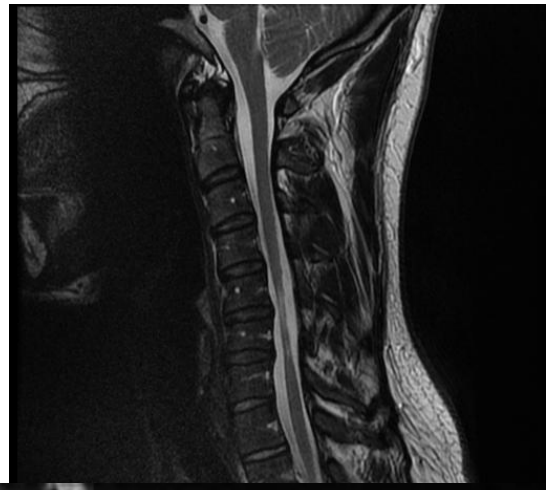
Woke after routine gynae surgery (ovarian cyst) with severe sensory ataxia.

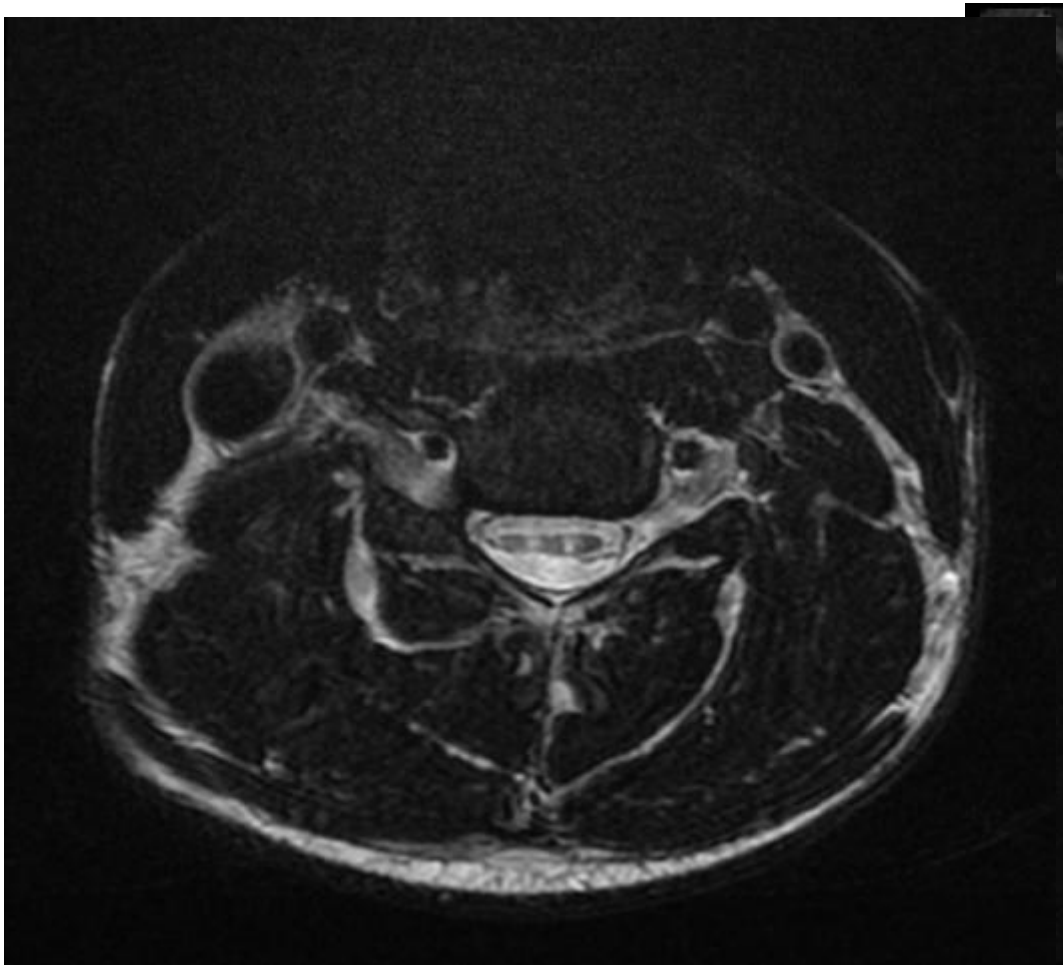
Sluggish reflexes, extensor plantars

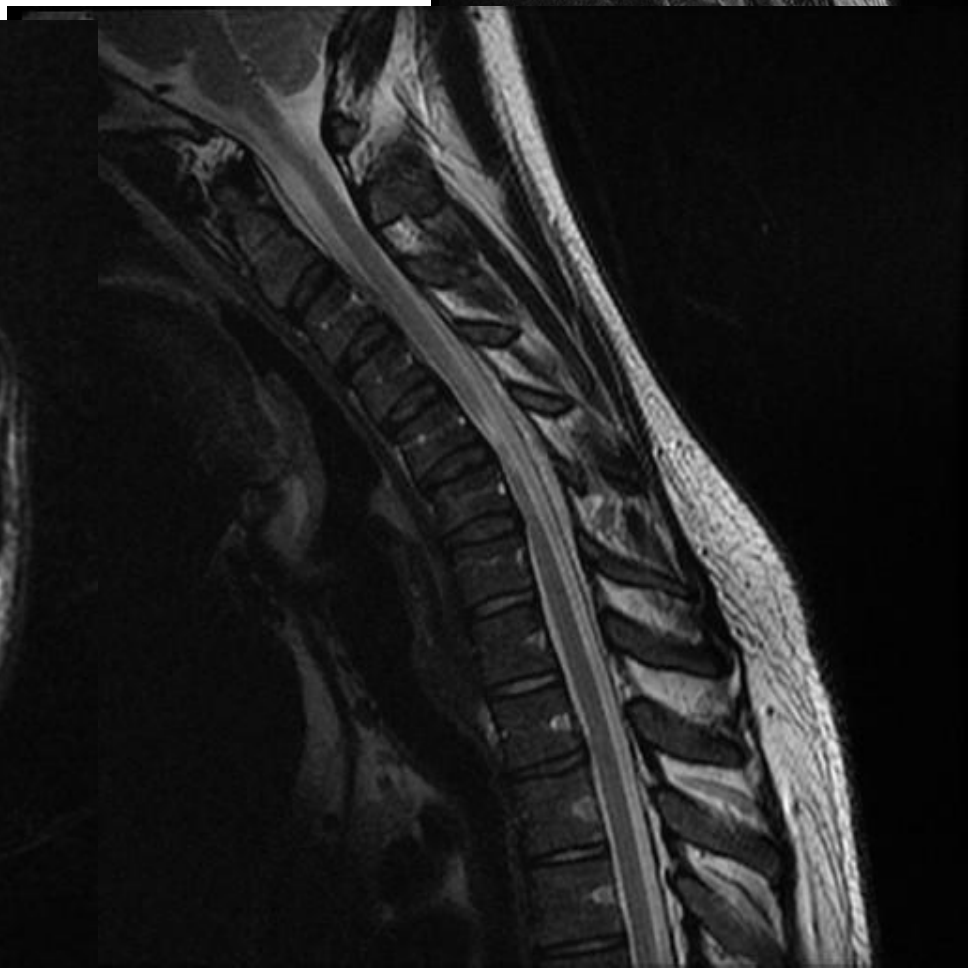
Similar MRI appearances & clinical features for SACD (B12), Nitrous oxide toxicity & Cu deficiency.



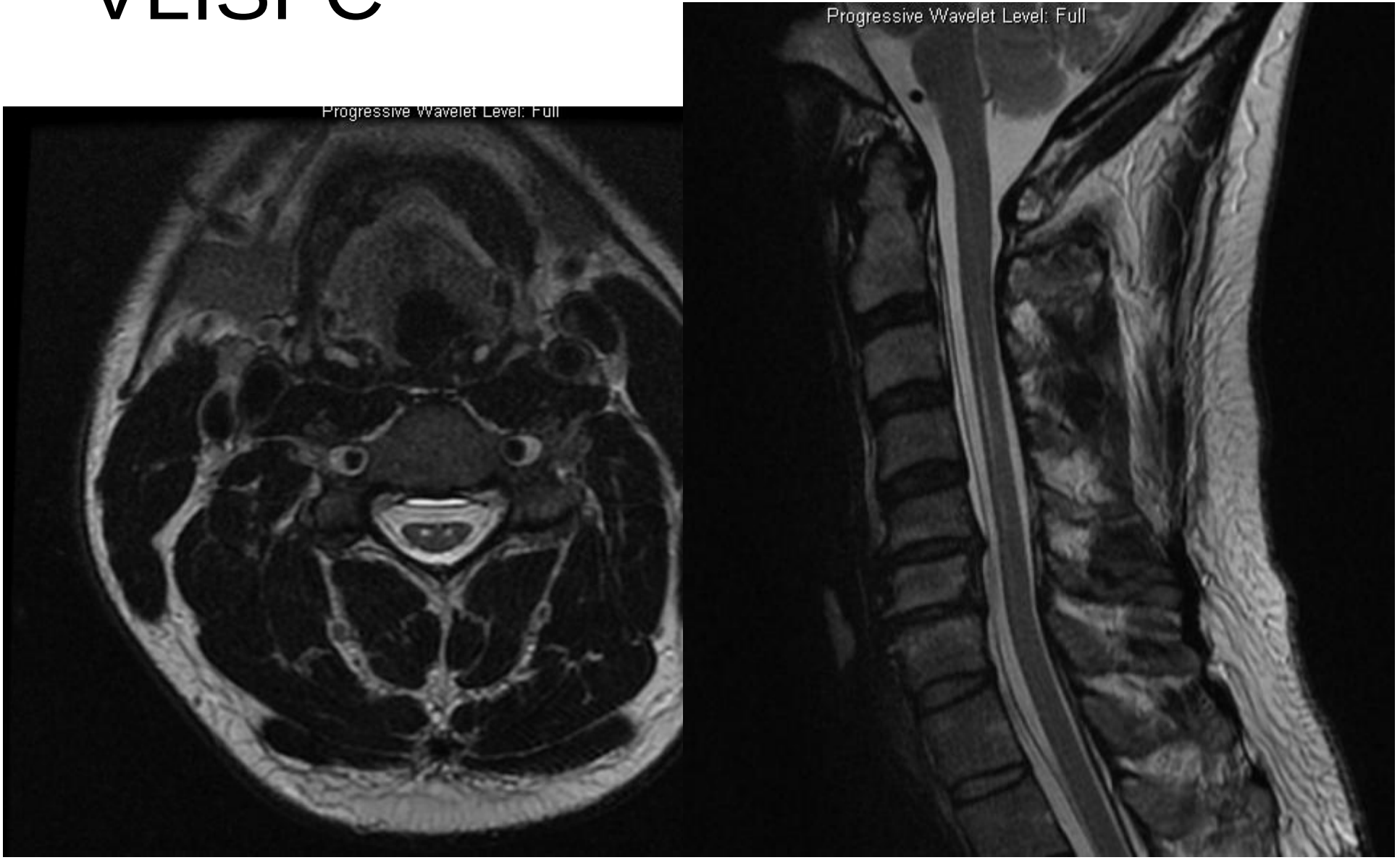
Hirayama







VLISFC


Progressive Wavelet Level: Full
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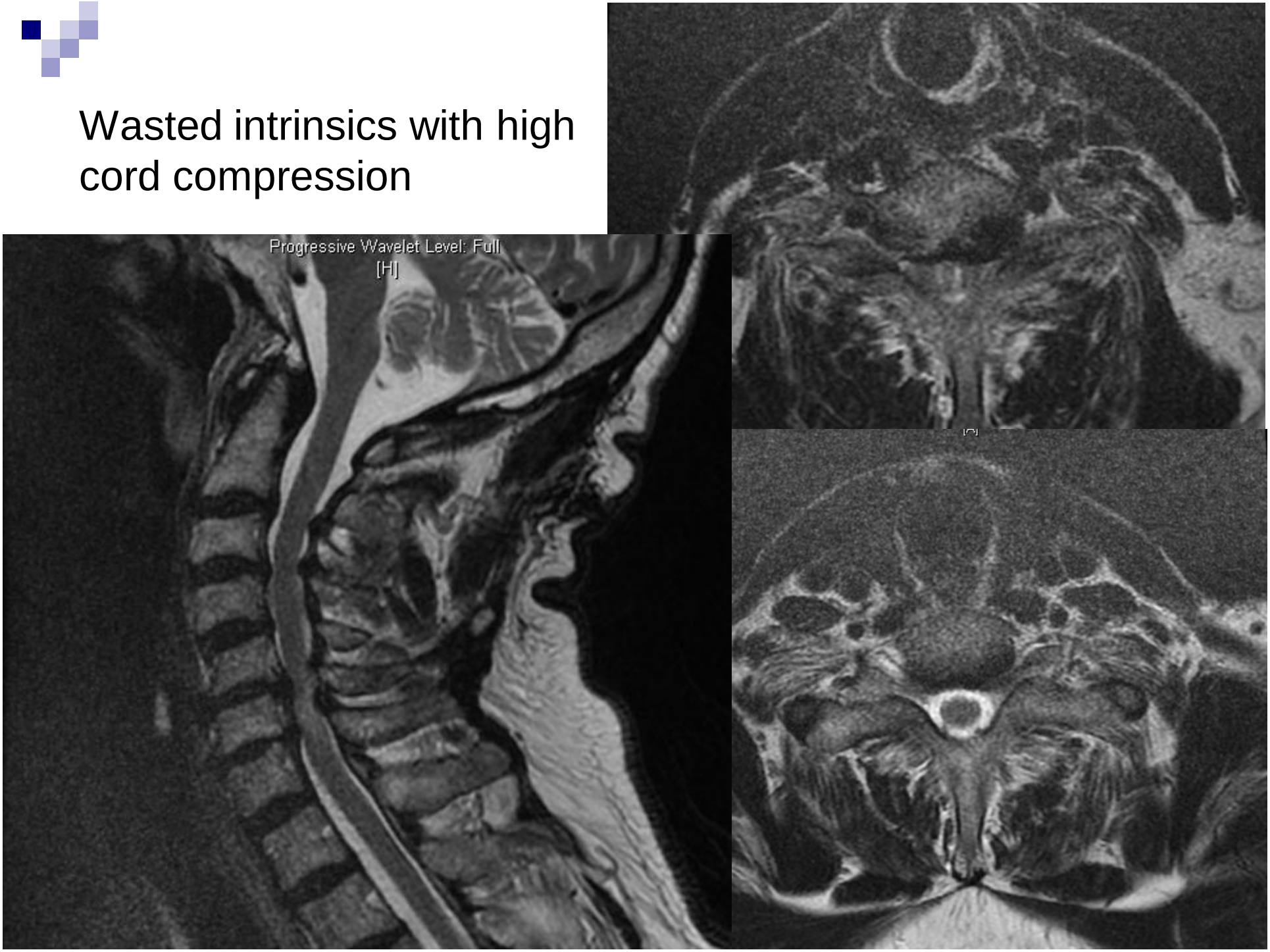
Wasted intrinsics with high cord compression



Lumbar spinal stenosis: symptoms

The typical clinical feature of severe lumbar spinal stenosis is **neurogenic claudication**

This consists of back and buttock or leg pain induced by walking or standing and **relieved by sitting** (or flexing the spine in some other way).

Symptoms in the legs are usually bilateral.

Unlike vascular claudication, symptoms are often provoked by standing without walking.

Lumbar spinal stenosis: signs

Focal weakness, sensory loss, or reflex changes may occur when spinal stenosis is associated with radiculopathy. Severe neurologic deficits, including paralysis and urinary incontinence, occur rarely.

Congenital **lumbar** stenosis often co-exists with congenital **cervical stenosis** resulting in mixed UMN & LMN signs in the legs

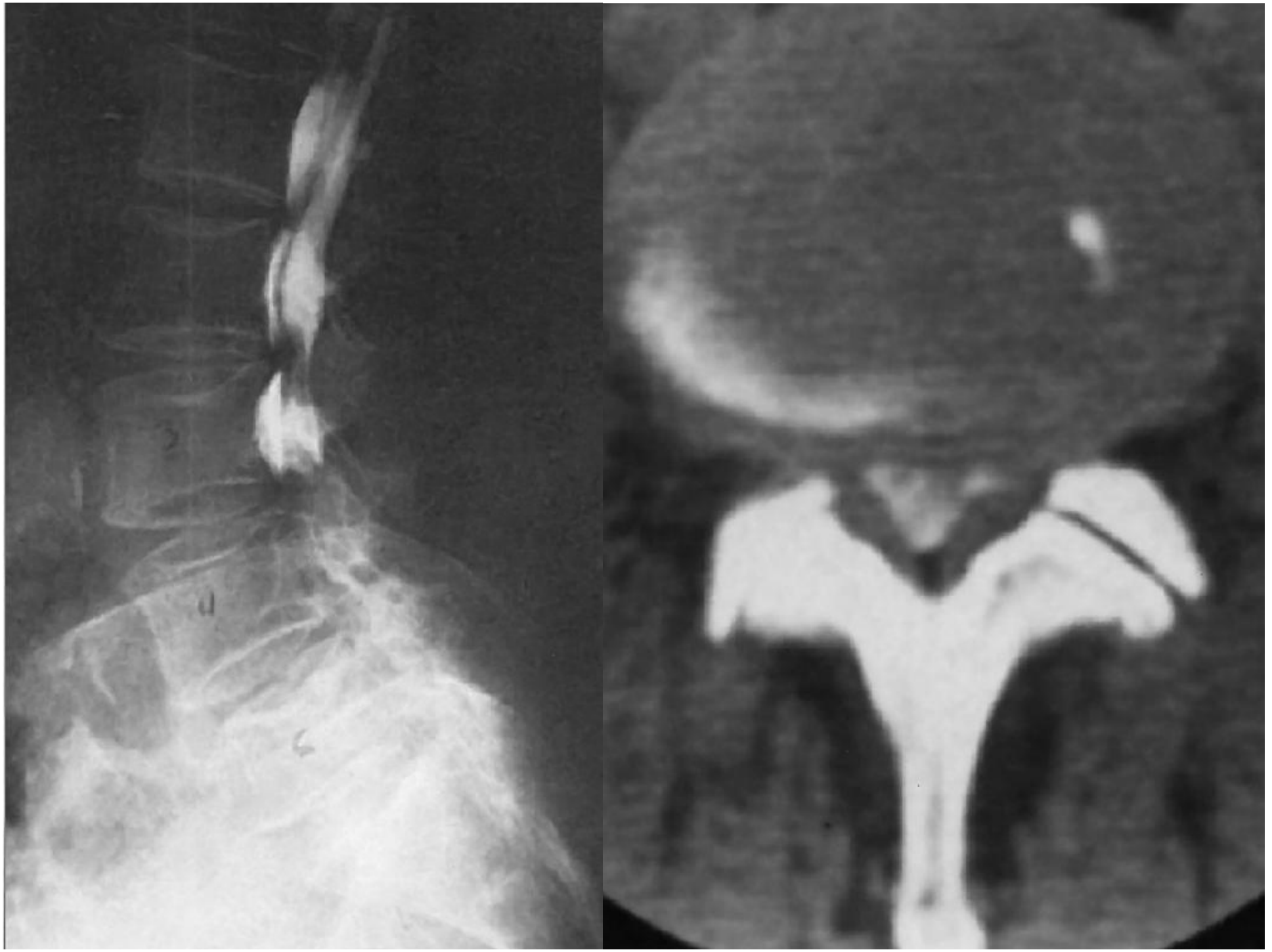
Lumbar spinal stenosis

Spinal stenosis can be acquired, congenital, or **due to a combination of the two causes**.

Congenital (achondroplasia, idiopathic): short, thick pedicles that produce both spinal canal and lateral recess stenosis.

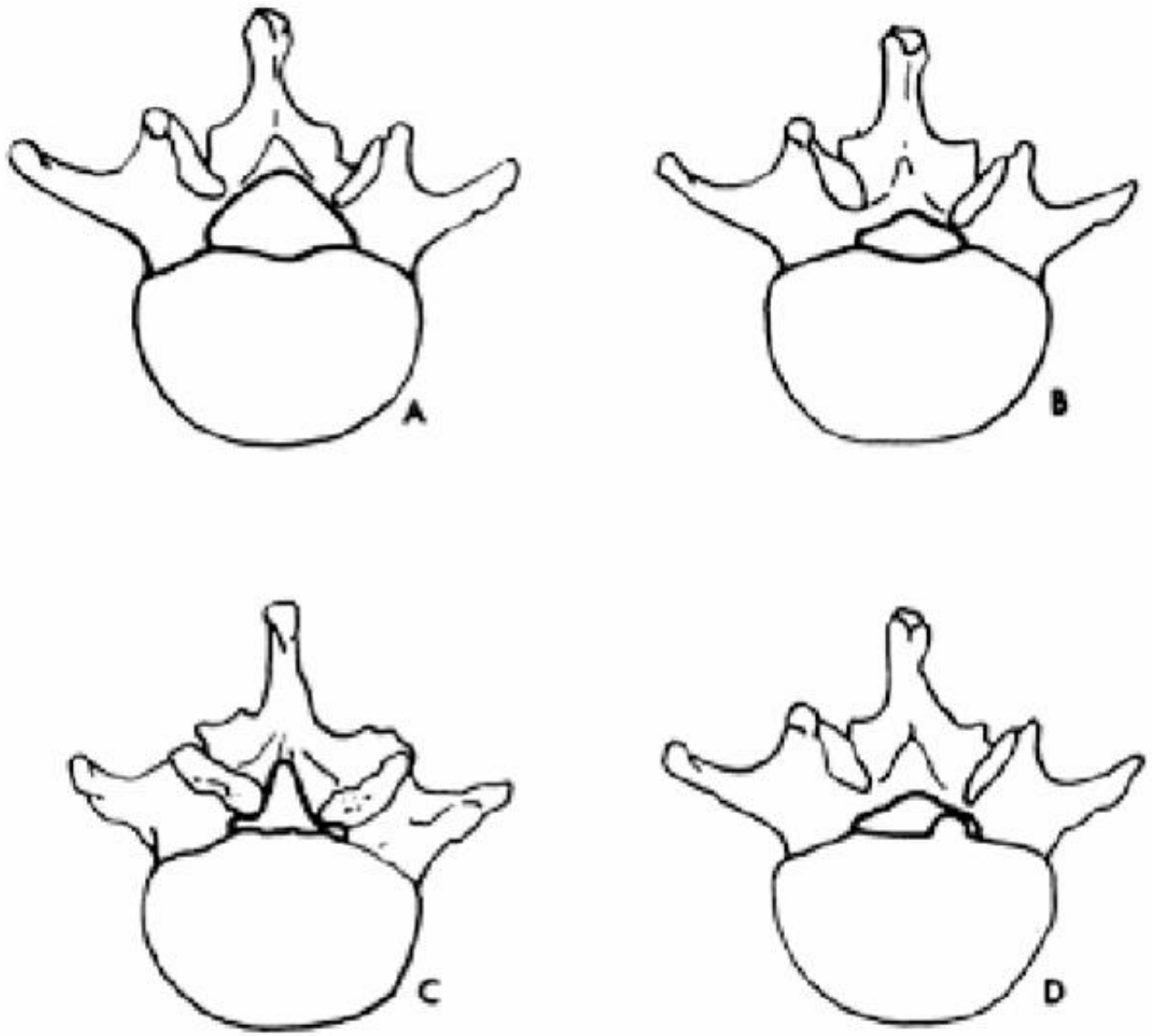
Acquired factors that may contribute to spinal stenosis include:

- **degenerative** diseases (spondylosis, spondylolisthesis, scoliosis)
- trauma, spine surgery (postlaminectomy, fusion),
- metabolic or endocrine disorders (epidural lipomatosis, osteoporosis, acromegaly, renal osteodystrophy, hypoparathyroidism), and
- Paget's disease.



Types of spinal stenosis

A. Normal
B. Congenital
C. Acquired
D. Congenital with disc prolapse



Neoplastic disease

Extradural / Spinal metastases

Intradural / extramedullary

- Meningeal metastases (often tiny)
- Neurofibroma / meningioma

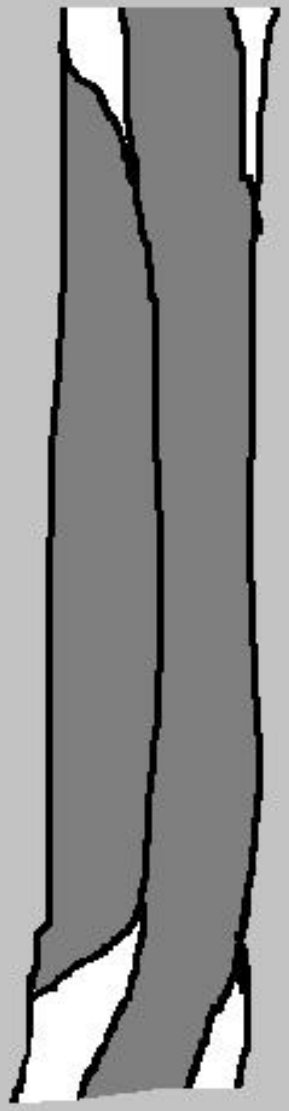
Intramedullary neoplasia

Radiology

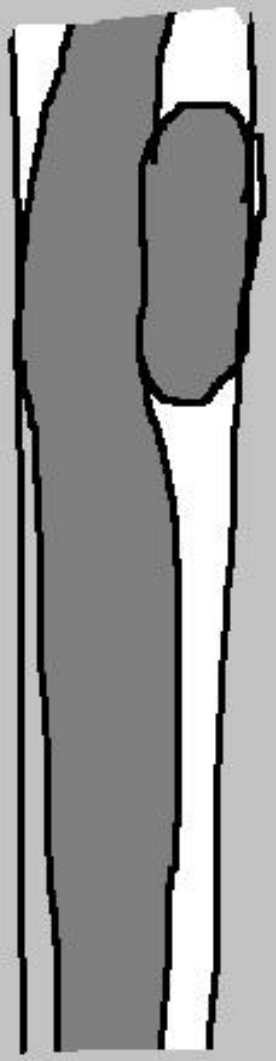
Radiology of lesions by site:

- Intramedullary
- Intradural / extramedullary
- Extradural

Intramedullary

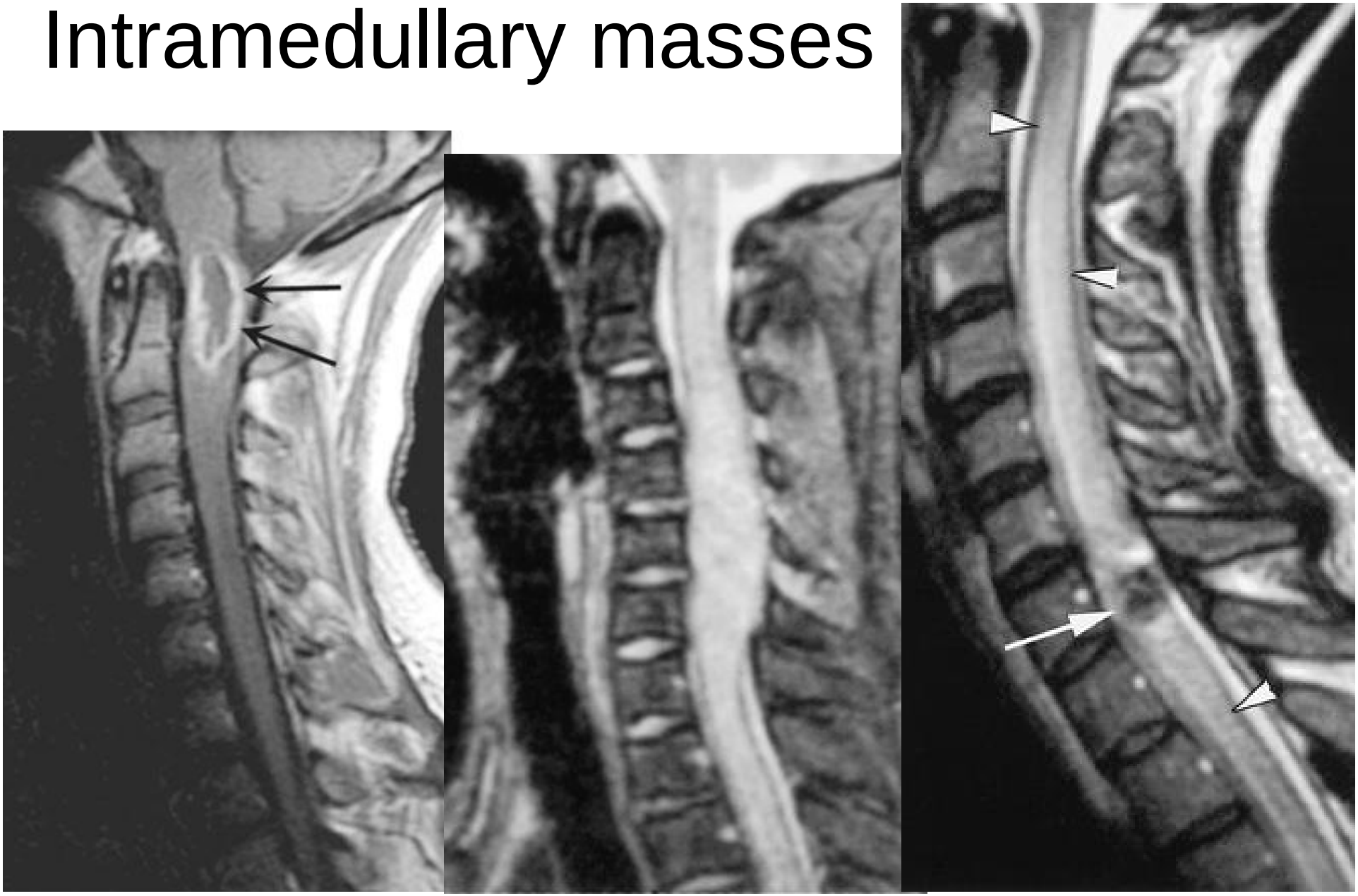


Extradural

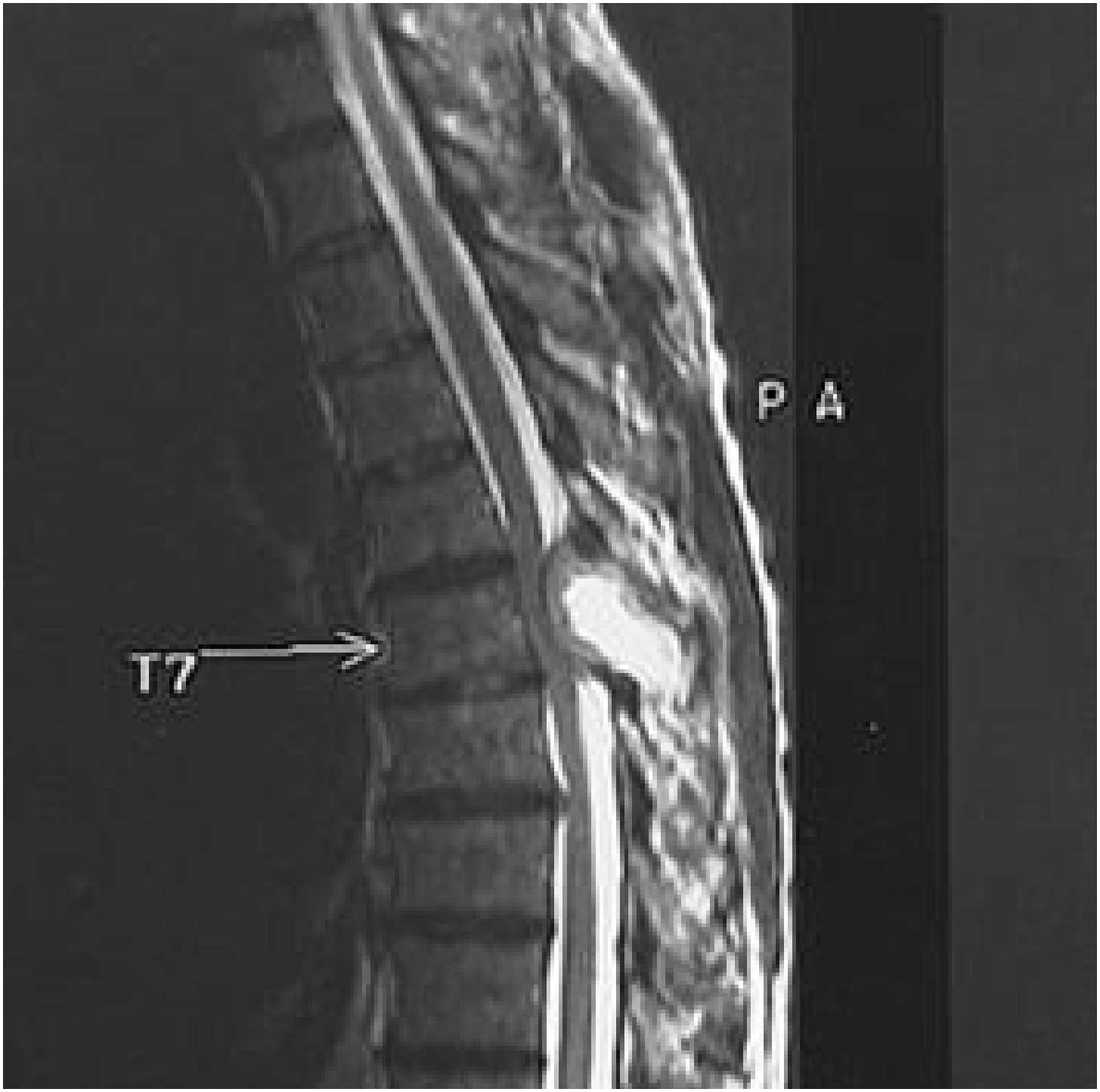


Intradural /
extramedullary

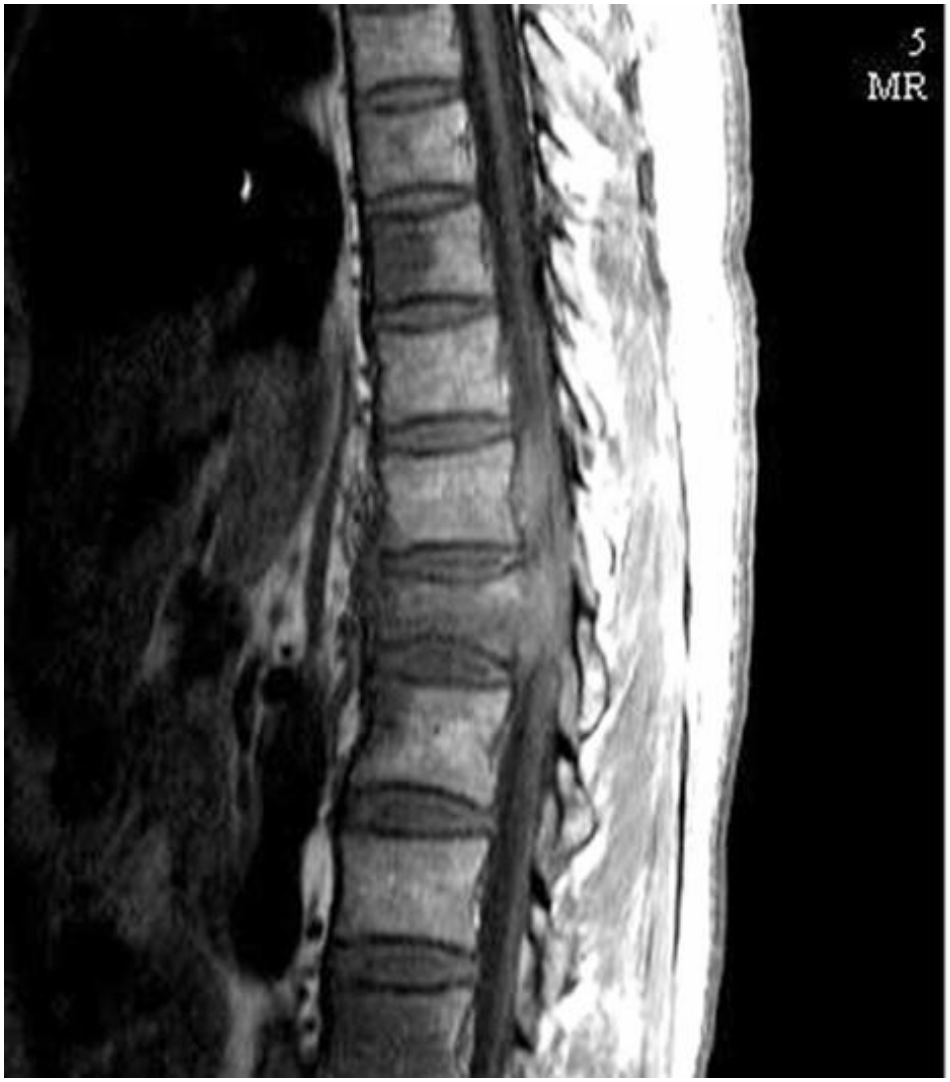
Intramedullary masses

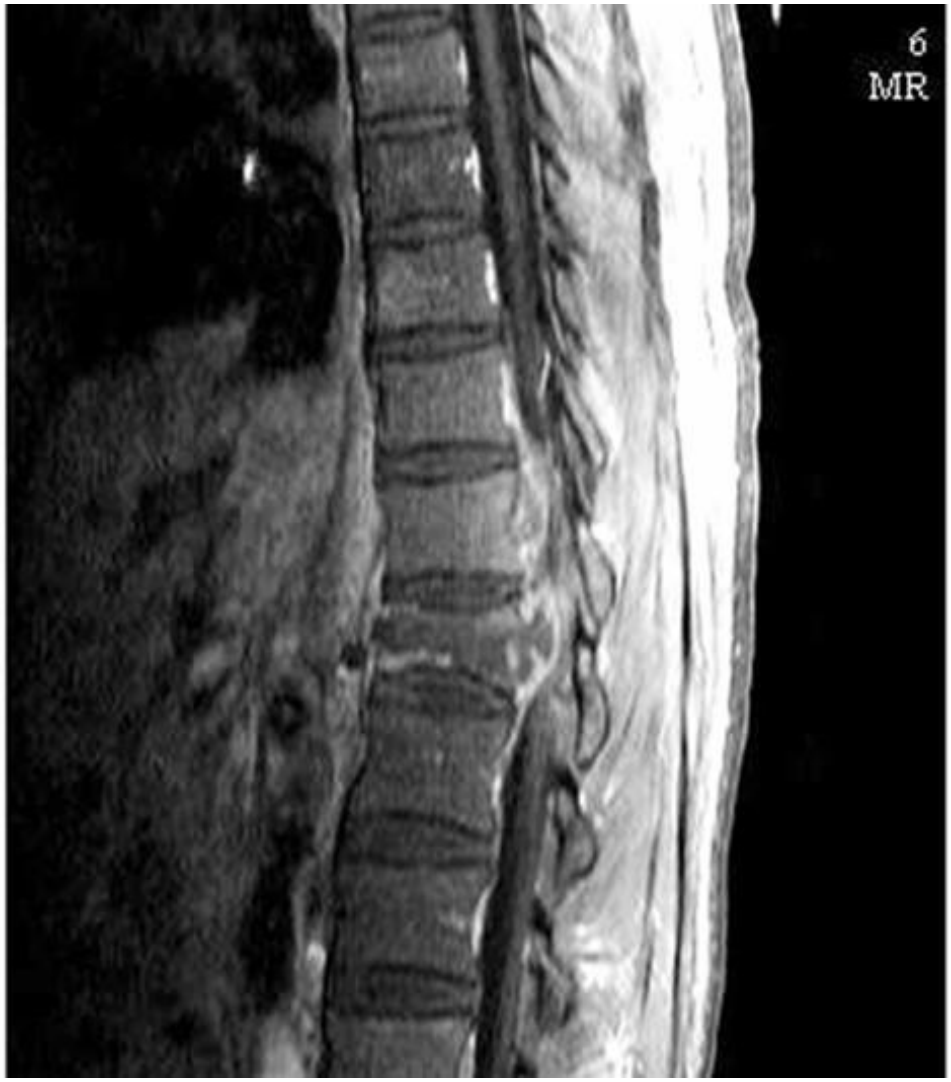


Extradural mass

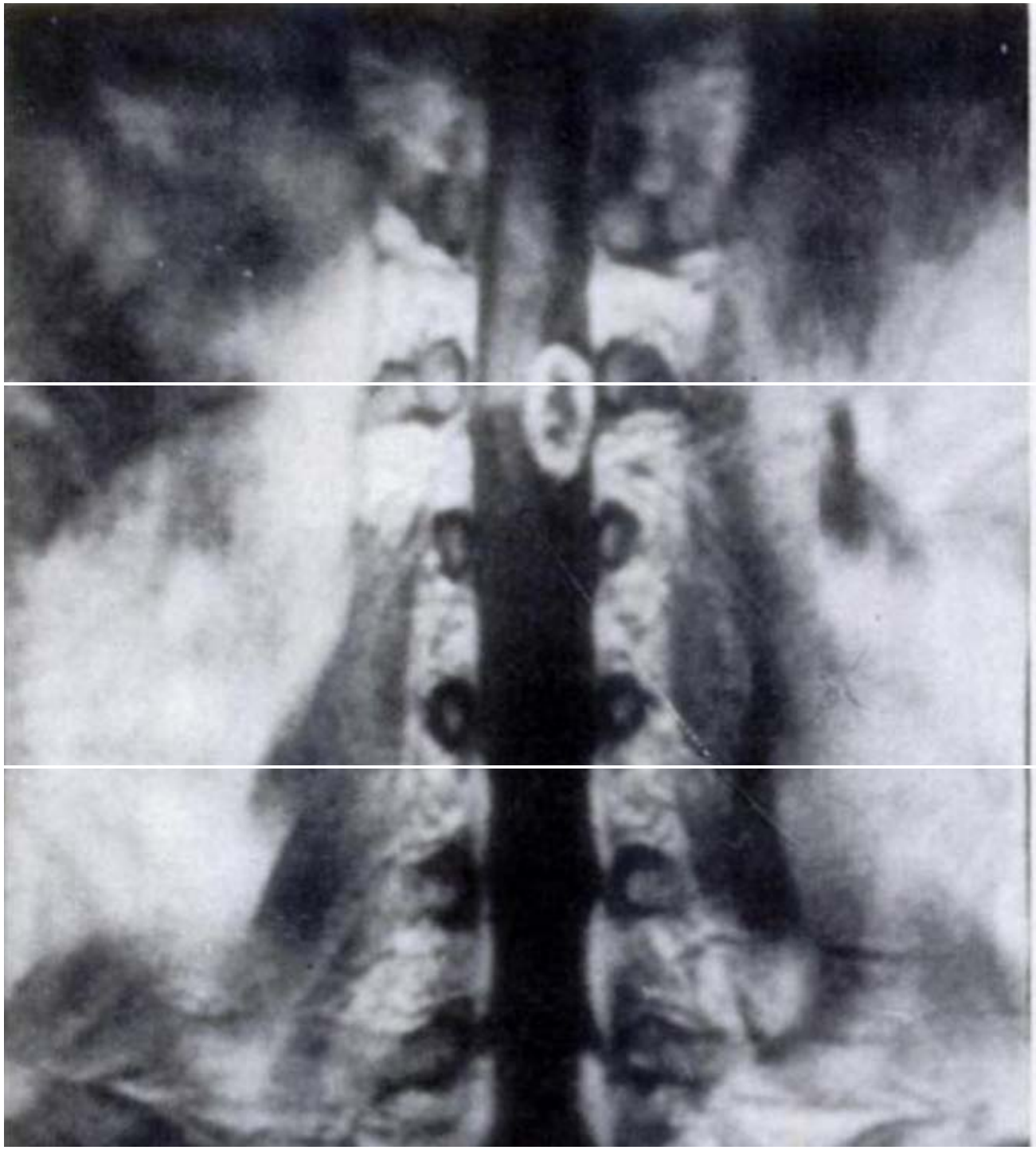


Extradural mass

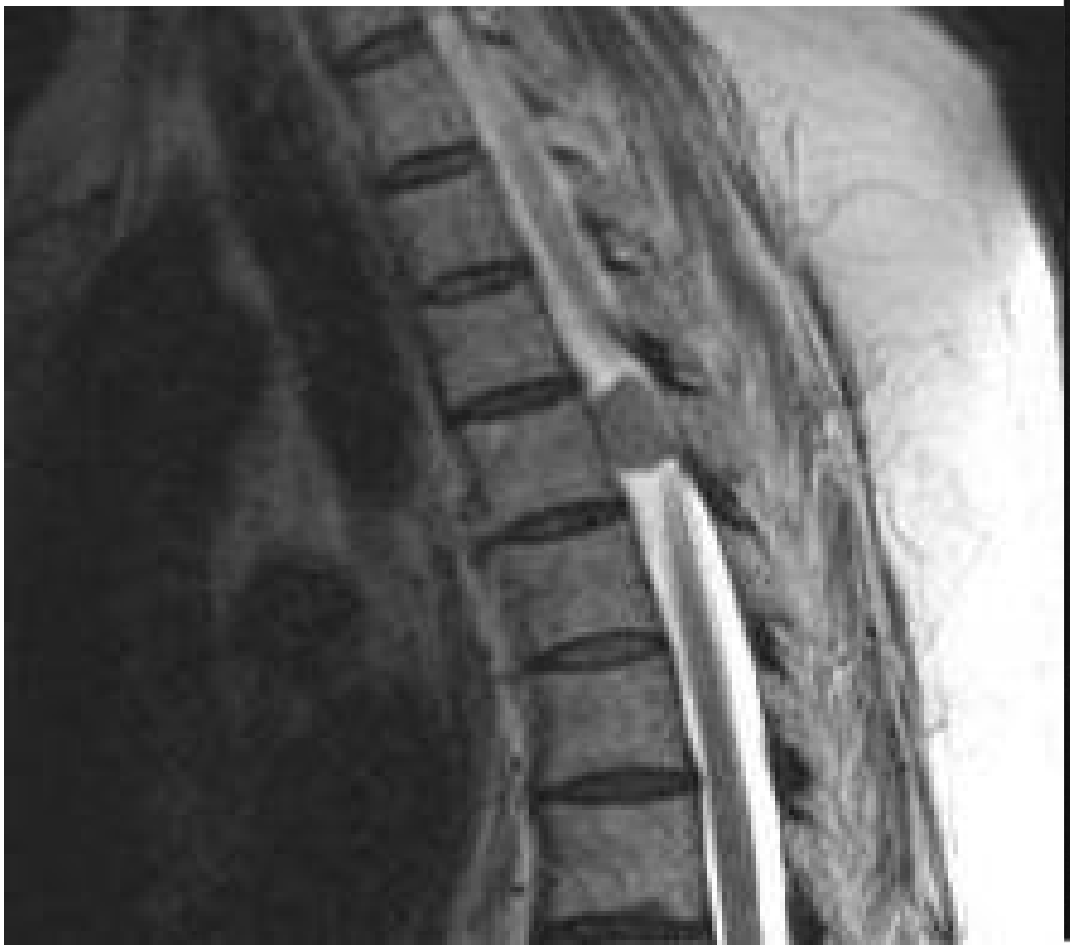


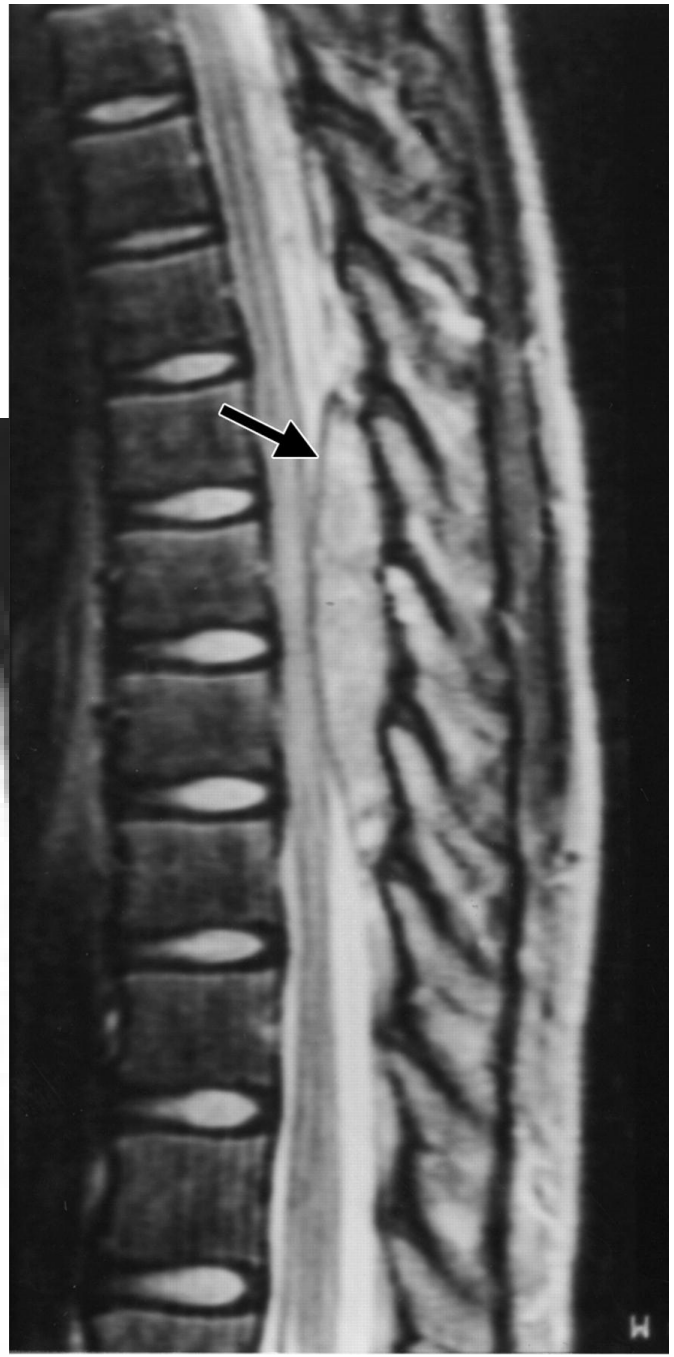


Extra-medullary intradural mass

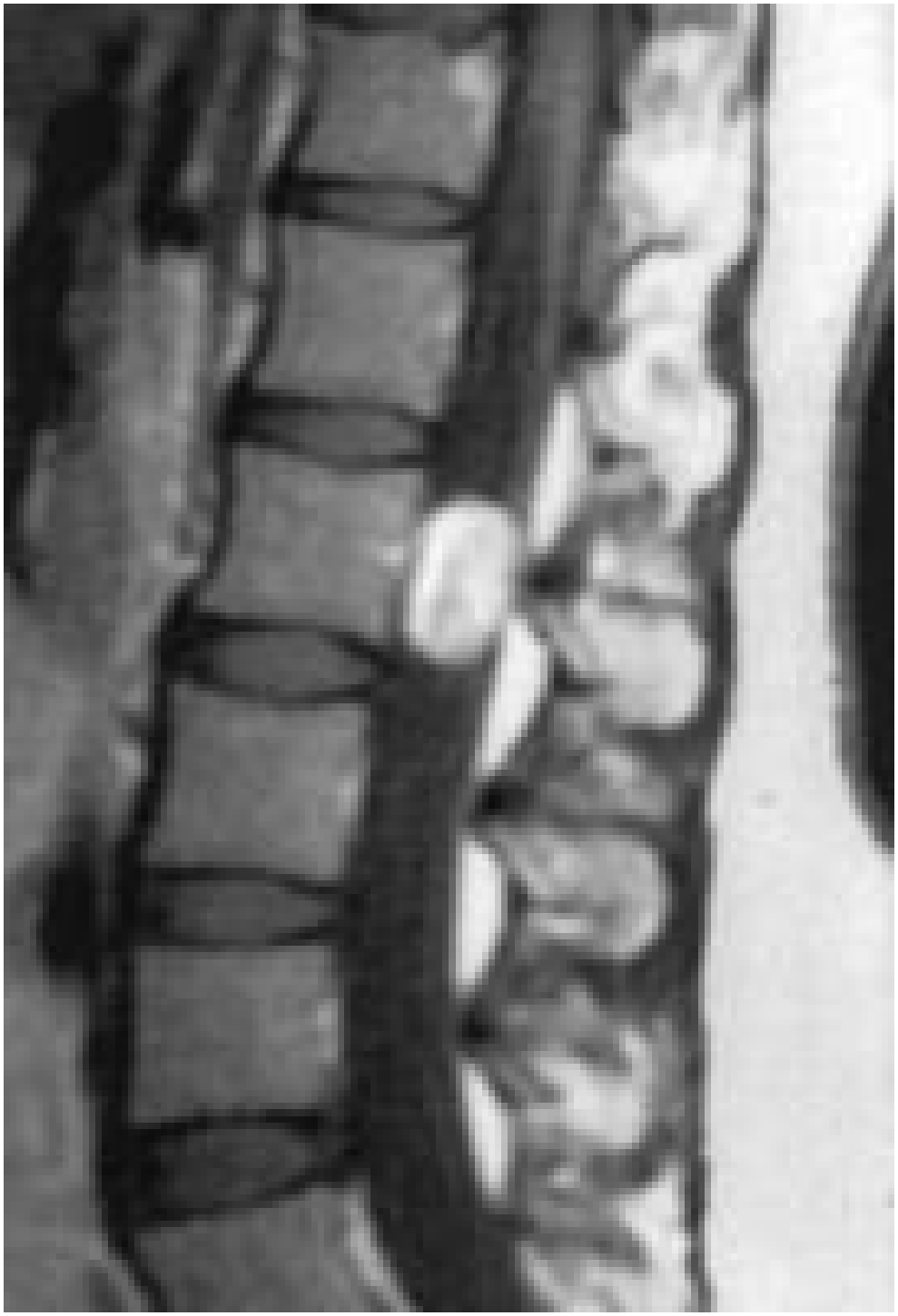


Extra-medullary intradural mass





Extra-medullary intradural mass (at cauda equina level)



Brain (1982), **105**, 189–213

SPINAL METASTASES

A RETROSPECTIVE SURVEY FROM A GENERAL HOSPITAL

by R. J. STARK,[1] R. A. HENSON *and* S. J. W. EVANS

(*From the Neurological Department, Section of Neurological Sciences, The London Hospital, and the Department of Clinical Epidemiology, The London Hospital Medical College, London E1*)

INTRODUCTION

Spinal Metastases: Primary Site

SITE	M/F	Total
Lung	35/8	43
Breast	0/37	37
Miscellaneous rapid	10/6	16
Miscellaneous slow	12/5	17
Undetermined	9/9	18
TOTAL	**66/65**	**131**

Spinal metastases
Patients not known to have cancer

Lung	30/43
Breast	3/37
Miscellaneous	11/33
Undetermined	18/18
TOTAL	**62/131**

Symptoms of Spinal Metastases

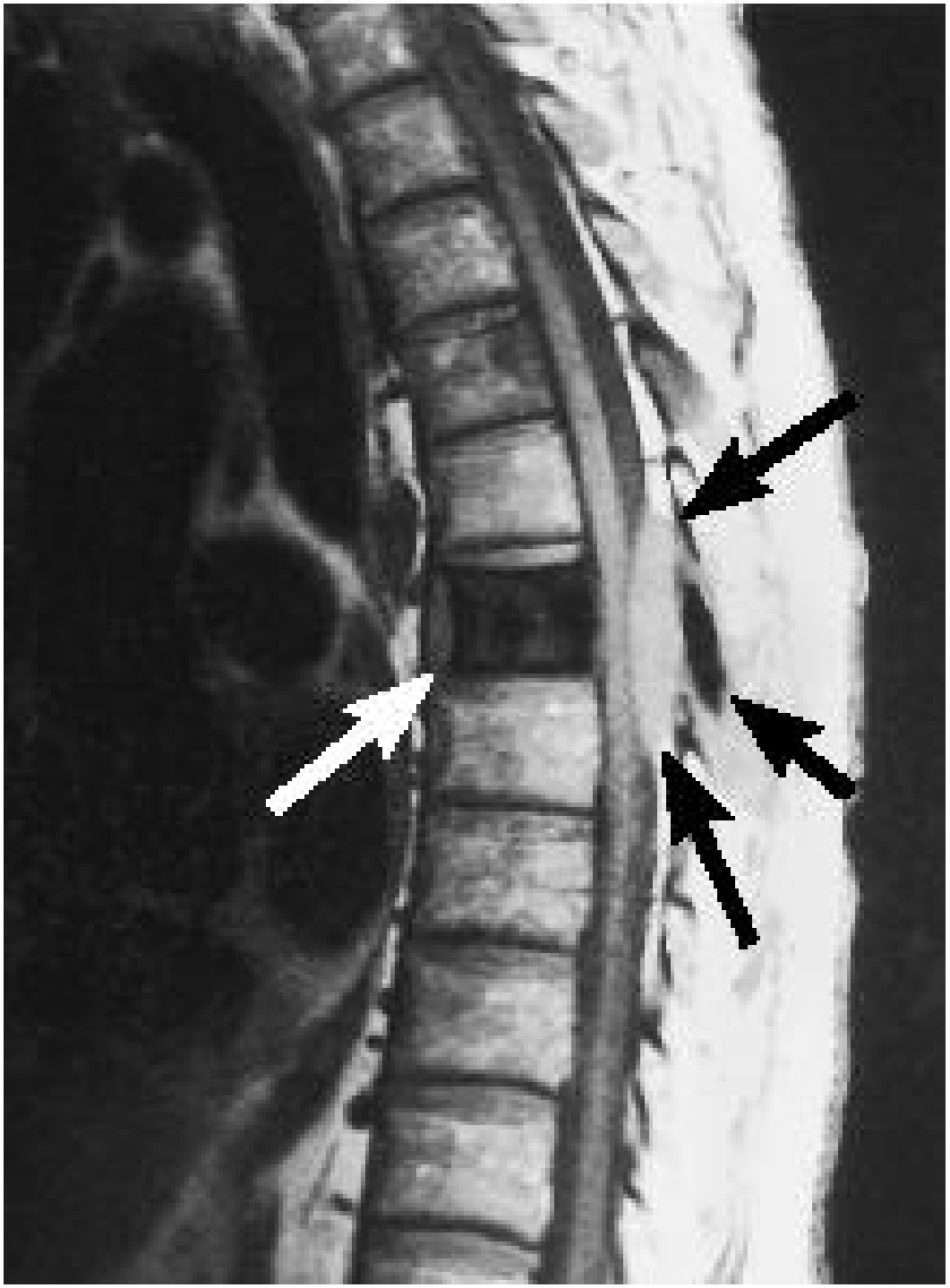
Axial pain	72%
Radicular pain	41%
No pain	14%
Pain precedes deficit	69%
Leg weakness	82%
Arm weakness	14%
Sphincter disturbance	63%

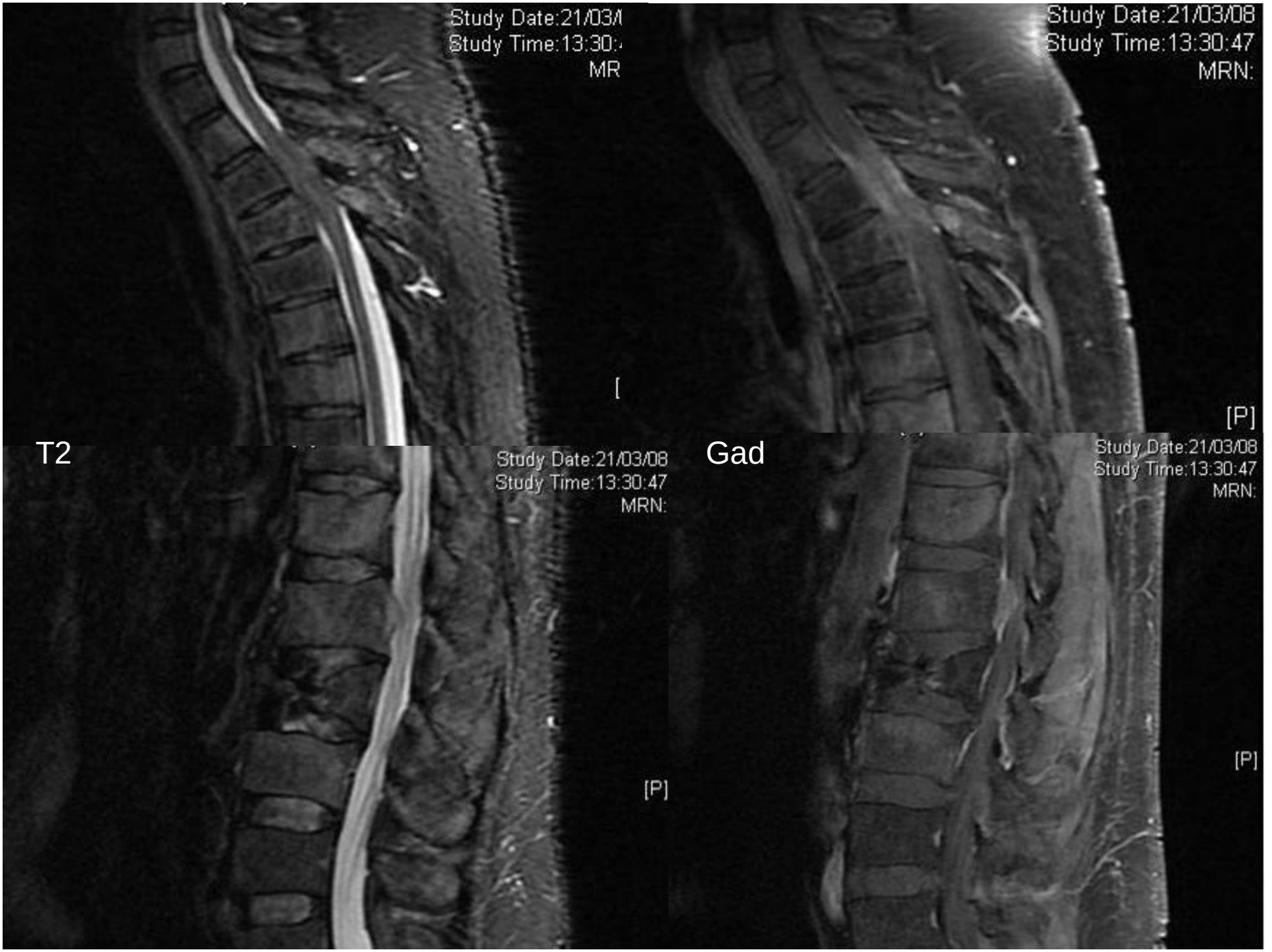
Signs of spinal metastases

Spinal tenderness	74%
Signs in arms	16%
Sensory level (PP)	72%
Power in legs: Normal	11%
Mild-moderate weakness	65%
Both Grade 0 or 1	24%
“Clinical transection”	11%
Decreased anal tone	35%

Spinal metastasis

Note involvement of vertebral bone, extradural location of metastasis

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MRN:
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Gad
Study Date:21/03/08
Study Time:13:30:47
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Meningeal cancer: primary site

Primary	My cases	Henson (1982)
N-H lymphoma	31	+++
Breast	30	+++
Lung	17	+++
Melanoma	6	+
Prostate	3	-
Leukaemias	3	++
Stomach	1	(+++)
Other sites	10	AdenoCa
TOTAL	101	

Meningeal cancer: Clinical patterns

1. Encephalopathy
   - Drowsiness, confusion, headache, papilloedema

2. Multifocal lesions, randomly scattered
   - **Nerve roots**
   - Cranial nerves
   - **Spinal cord**

Meningeal cancer: Presentation

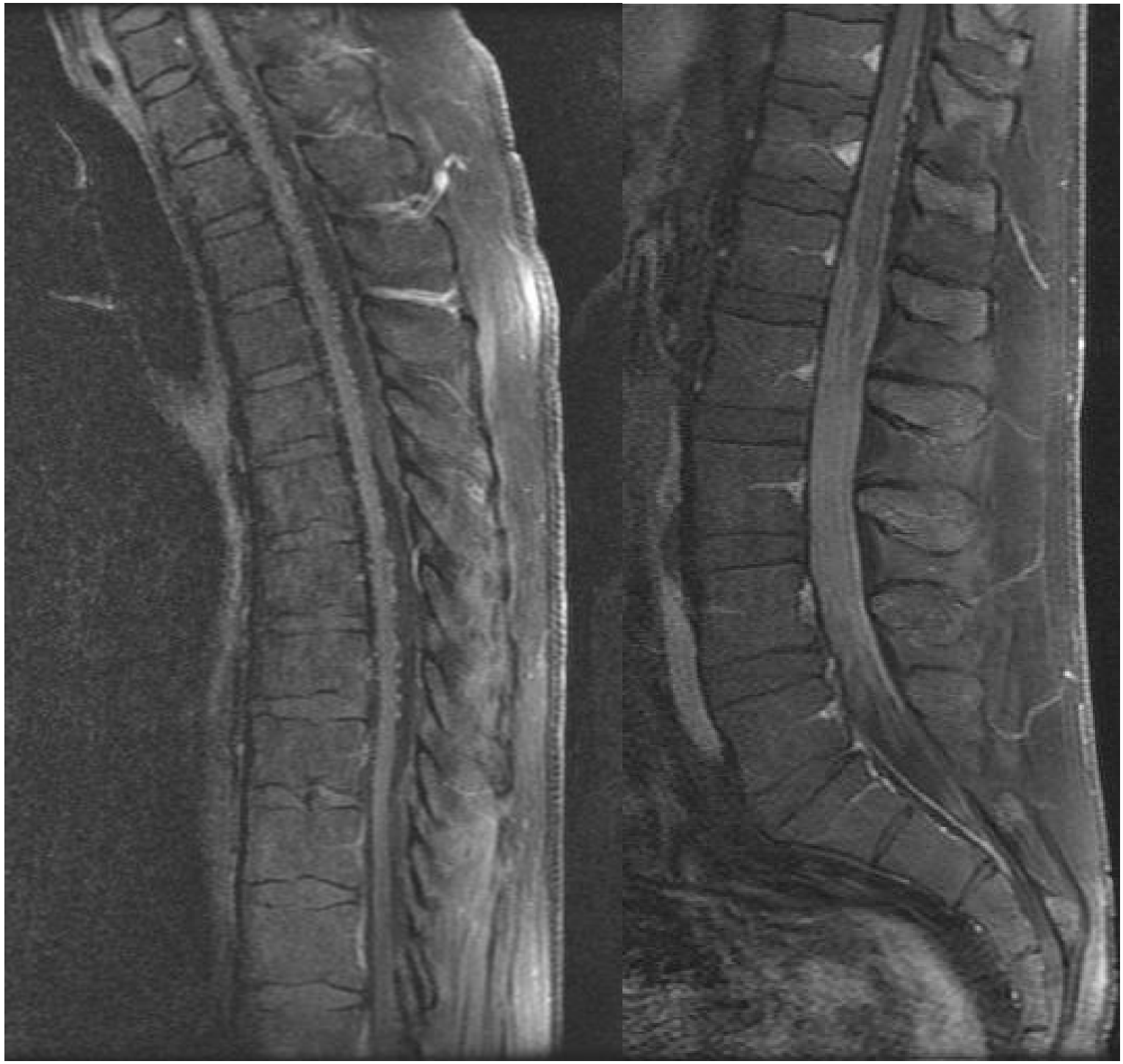
(my series)

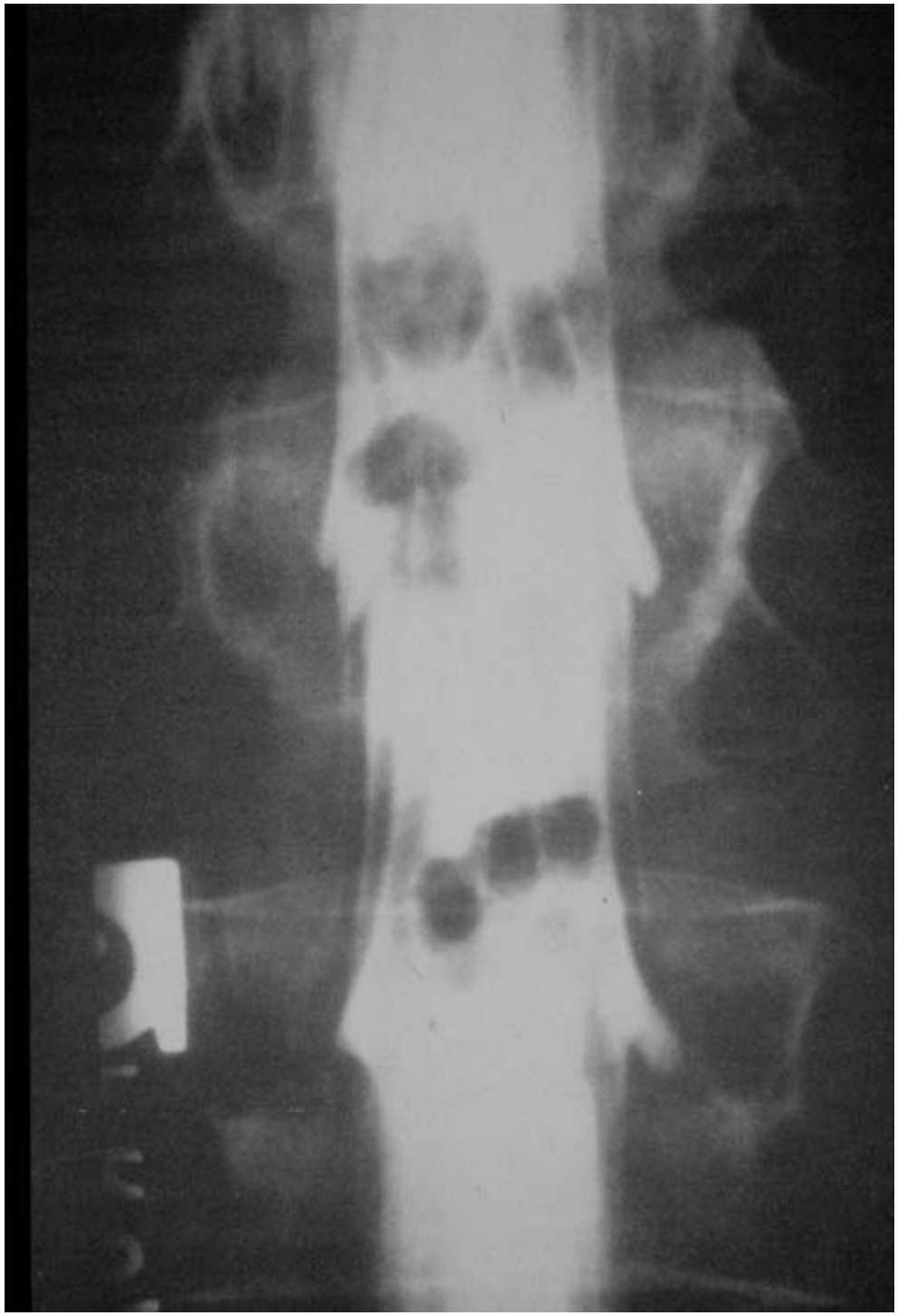
Radiculopathy &/or cranial nerves (scattered)	46	67%
Encephalopathy	12	17%
Both radiculopathy & encephalopathy	4	6%
Myelopathy	**7**	**10%**
TOTAL	69	100%

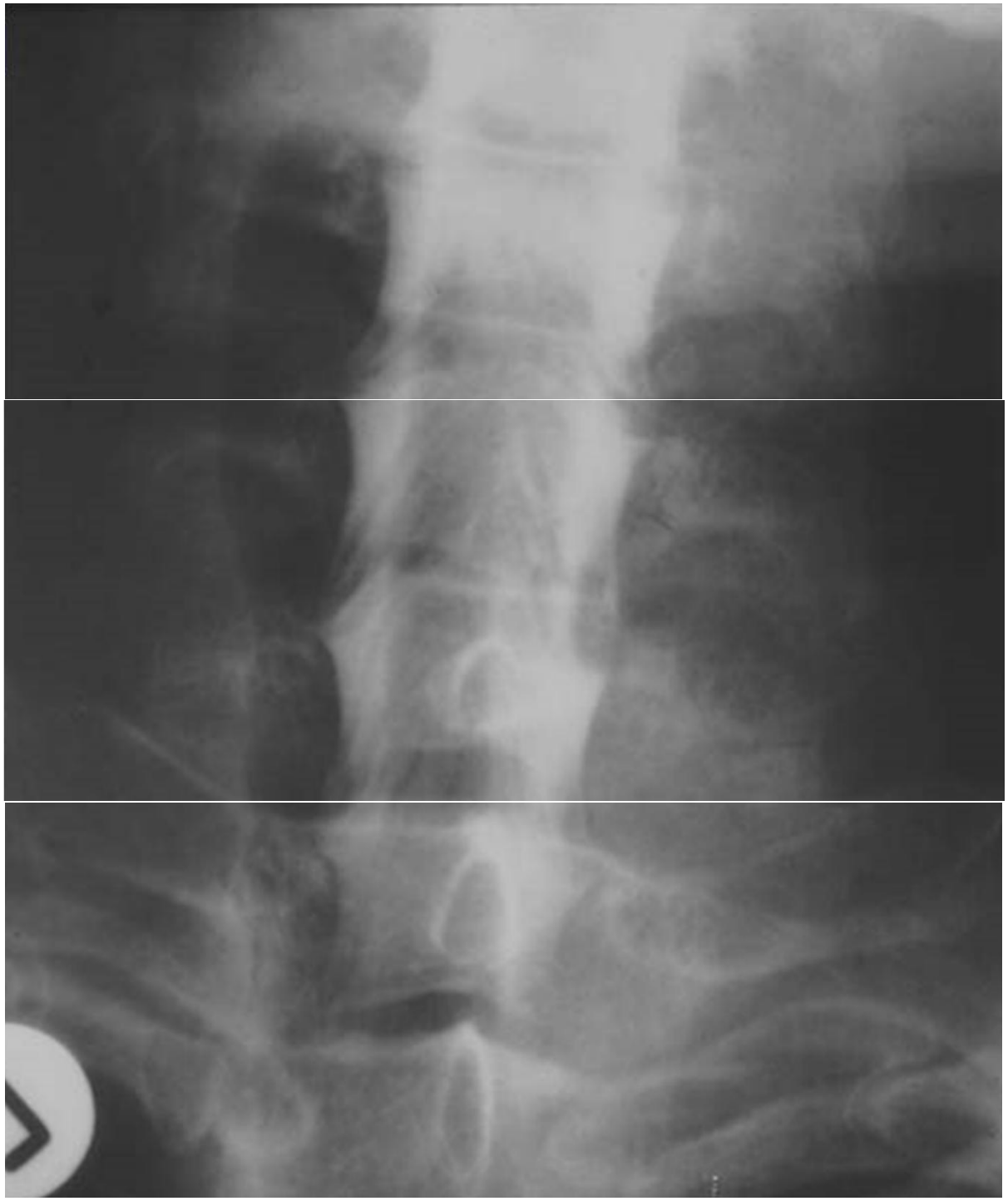
Meningeal cancer: Diagnosis

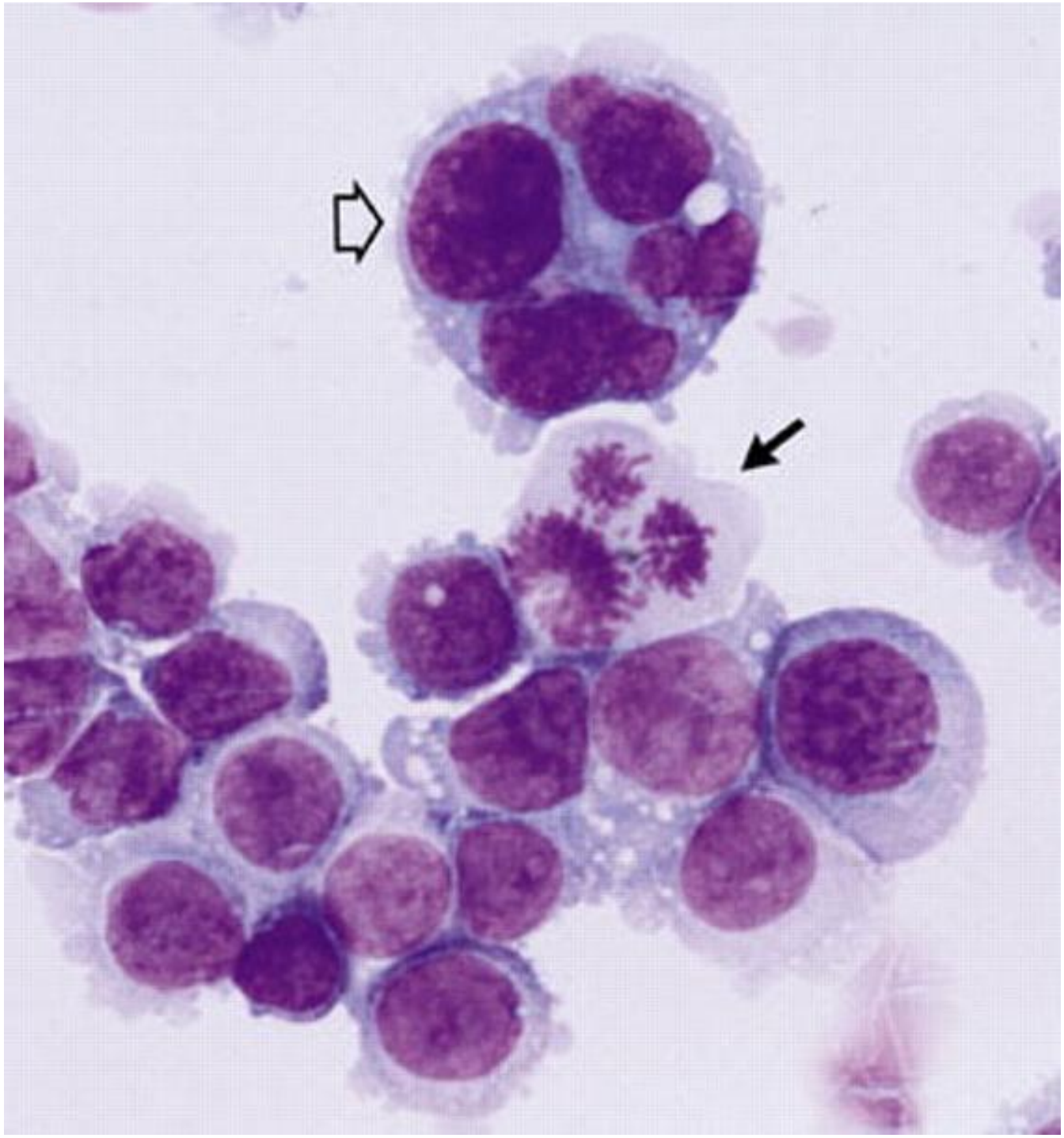
- MRI - Gadolinium greatly increases sensitivity
- Lumbar puncture (multiple if necessary)
  - Cytology may be specific (Gold standard)
  - Even with negative cytology, CSF leucocytosis with typical clinical picture is very suggestive
  - Interest in new CSF markers (research)
- Myelogram & post-myelo CT - big nerve roots
- CT Brain - rarely helps

Multiple tiny meningeal metastases post contrast









Non-compressive Myelopathy

(my series: 13 cases referred as “NCM”)

Intramedullary metastases	0
Meningeal cancer	4
Radiation Myelopathy	2
Paraneoplastic (subacute necrotising myelopathy)	1
Intrathecal chemotherapy	1
Spinal Infarction	1
Unexplained	2
(Inadequate imaging: actually compressive)	*
(Pseudo-myelopathy: cerebral parasagittal)	2

A clinical scenario

GP desperate for you to see patient who is complaining bitterly of back pain and requiring narcotics.

CT lumbar spine reported as normal.

GP accedes to patient request for MRI lumbar spine – also reported normal.

Patient reports severe low back pain with radiation to buttocks.

A clinical scenario

GP desperate for you to see patient who is complaining bitterly of back pain and requiring narcotics.

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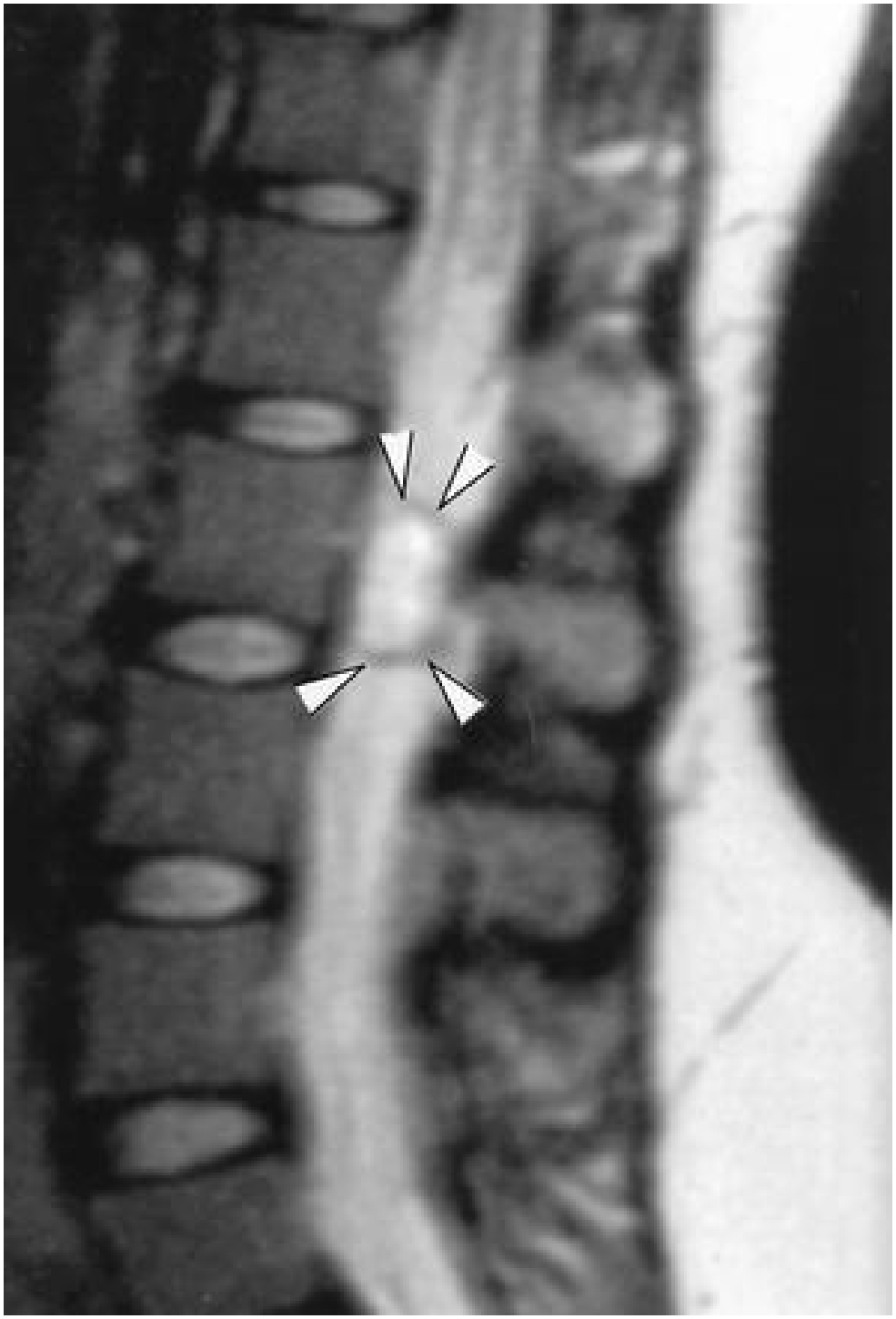
Restless and avoids certain postures – won't straighten legs on couch.

Examination difficult but perhaps reflexes a little sluggish. Nothing definitive.

Next step?



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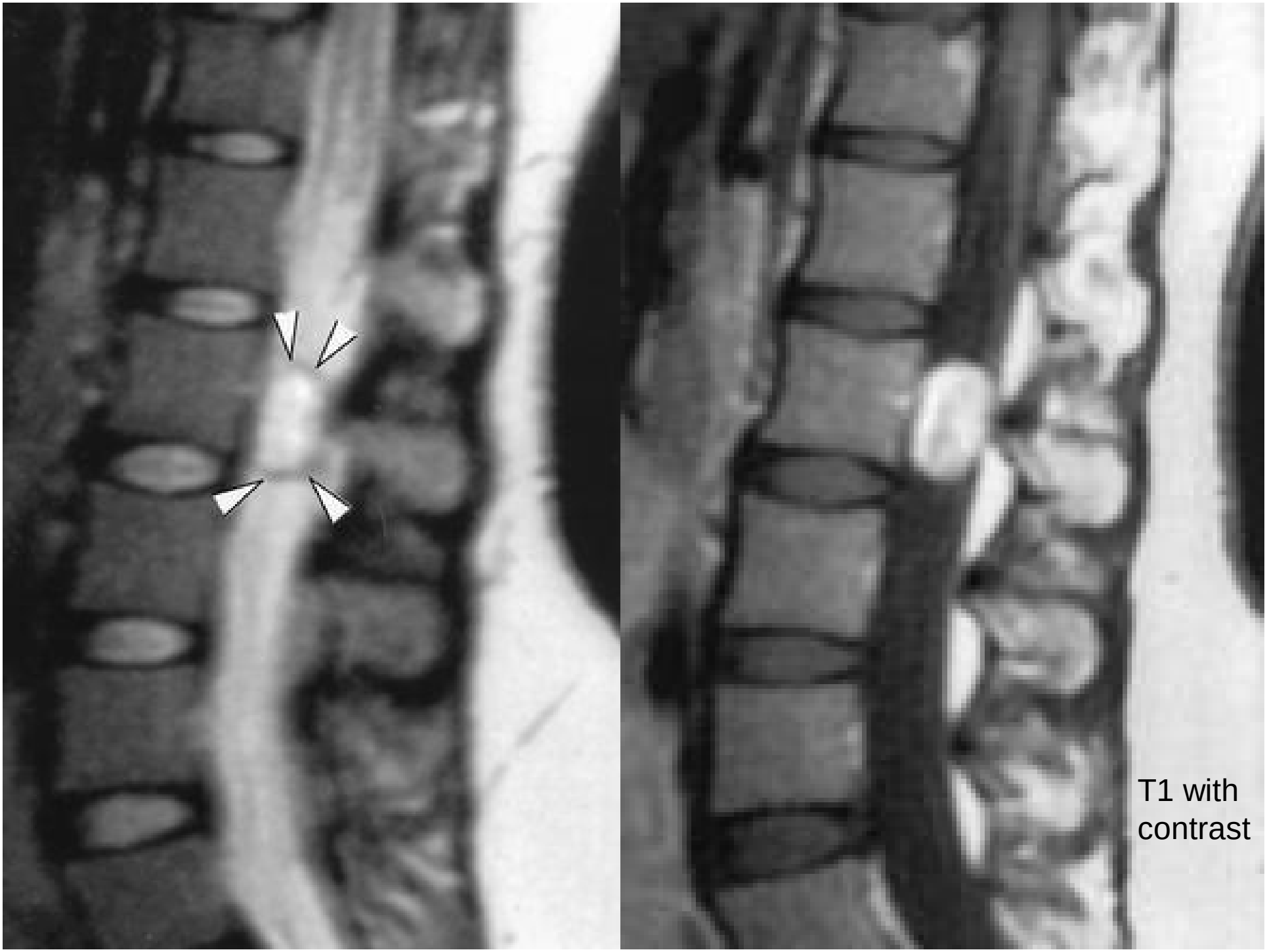
Next Step: Review the films (T2 image shown with helpful arrows added)

Next Step?


T1 with
contrast

Moral of the tale

Neurofibromas of cauda equina can be very painful and show few signs.

Radiological diagnosis is easy if the correct tests are requested, but may be missed with routine studies

Meningiomas and neurofibromas

- Signs are usually pretty obvious at cord level (UMN signs as expected)
- Meningiomas typically in thoracic region
- Neurofibromas typically in cervical and lumbar regions

Intramedullary spinal tumours

Table from

Koeller K et al, AFIP ARCHIVES Neoplasms of the Spinal Cord and Filum Terminale: Radiologic-Pathologic Correlation.
Radiographics. 2000;20:1721-1749.

Neoplasms of the Spinal Cord and Filum Terminale

- Common
  - Ependymoma
    - Myxopapillary ependymoma
  - Astrocytoma
    - Pilocytic astrocytoma
    - Anaplastic astrocytoma
  - Hemangioblastoma
- Less common
  - Subependymoma
  - Ganglioglioma
  - Paraganglioma
  - Metastasis
  - Lymphoma
  - PNET
  - Neurocytoma
  - Oligodendroglioma
  - Mixed glioma
  - Glioblastoma multiforme

Vascular disease of the spinal cord

Infarct

Haemorrhage

AVM

Dural venous fistula

Mechanisms of spinal cord infarction

- Hypoperfusion
- Atheroembolism
- Arterial thrombosis or dissection
- Arteritis
- Fibrocartilaginous embolism
- Compression of vessels by cervical spondylosis or tumours
- Decompression sickness
- Surgery, to aorta or spine, angiography
- Spinal anaesthetic or other injection

Selective Vulnerability of the Lumbosacral Spinal Cord After Cardiac Arrest and Hypotension

N. Duggal, MD; B. Lach, MD, PhD

Background and Purpose—It is generally accepted that the gray matter in the watershed area of the midthoracic level of the spinal cord is the ischemic watershed zone of the spinal cord. We performed a retrospective study to reevaluate the frequency and distribution of spinal cord injury after a global ischemic event.

Methods—Clinical files and neuropathology specimens of all adult patients with either a well-documented cardiac arrest or a severe hypotensive episode, as well as pathologically confirmed ischemic encephalopathy and/or myelopathy, were reviewed by an independent reviewer.

Results—Among 145 cases satisfying selection criteria, ischemic myelopathy was found in 46% of patients dying after either a cardiac arrest or a severe hypotensive episode. Among the patients with myelopathy, predominant involvement of the lumbosacral level with relative sparing of thoracic levels was observed in >95% of cardiac arrest and hypotensive patients. None of the examined patients developed neuronal necrosis limited to the thoracic level only.

Conclusions—Our findings indicate a greater vulnerability of neurons in the lumbar or lumbosacral spinal cord to ischemia than other levels of the spinal cord. (***Stroke*****. 2002;33:116-121.)**

Operation

AVM (Anson & Spetzler 1992)

Type 1 Dural AVF

Type 2 glomus, intramedullary AVM

Type 3 juvenile, extensive

Type 4 middle age or later, extramedullary

Miscellaneous Spinal disorders

Infection

After anaesthesia

Radicular syndromes

Spinal epidural abscess stages

1. Back pain
2. Radicular radiation
3. Motor, sensory, sphincter decline
4. Paralysis

Risk factors

- Constitutional (diabetes, alcoholic, IV drug use…..)
- Local spinal problem (arthritis, trauma, surgery, percutaneous procedure, implanted device….)
- Infective source elsewhere (skin, UTI, IV lines…..)

Investigations

FBE

ESR

CRP

MRI spine + Gd

Treatment

Surgery

Antibiotics

Outcome relates to neurological function at the time of intervention

Pitfalls (NEJM 2006;355:2012-2020)

- Systemically unwell, obtunded, difficult to examine patient
- Staph septicaemia treatment without finding cause
- Accepting vertebral osteomyelitis as sufficient diagnosis
- Imaging wrong section of spine
- Misssing second site of epidural abscess
- Delaying investigations

Spinal cord injury and anaesthesia

- Anaesthetists quote 0.45 per 10,000 for spinal anaesthetics
- Epidural haematoma
- Abscess
- Needle trauma
- Intracord injection
- Spinal cord infarction

Spinal cord infarction with regional anaesthesia

- Hypotension
- Prothrombotic states
- Hyperlordotic operative position
- Pre-existing structural disease or vascular disease

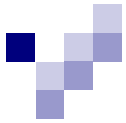
Summary

Spinal syndromes

- Classical
- Less common

Clinical aspects of

- disc disease / spondylosis
- primary and secondary neoplasia
- vascular disease
- Infection



End

Progressive amytrophy of hand

19 years old male previously well

Right handed

Started to notice right hand twitching 6 months ago

2-3 months ago noted increasing weakness and wasting in the right hand

In the last 1-2 month felt that left hand also became weak

History

- Currently spending the year overseas (Israel) in the army
- Weakness worse in cold weather
- No sensory symptoms
- No neck pain
- No bulbar symptoms
- No diplopia or ptosis

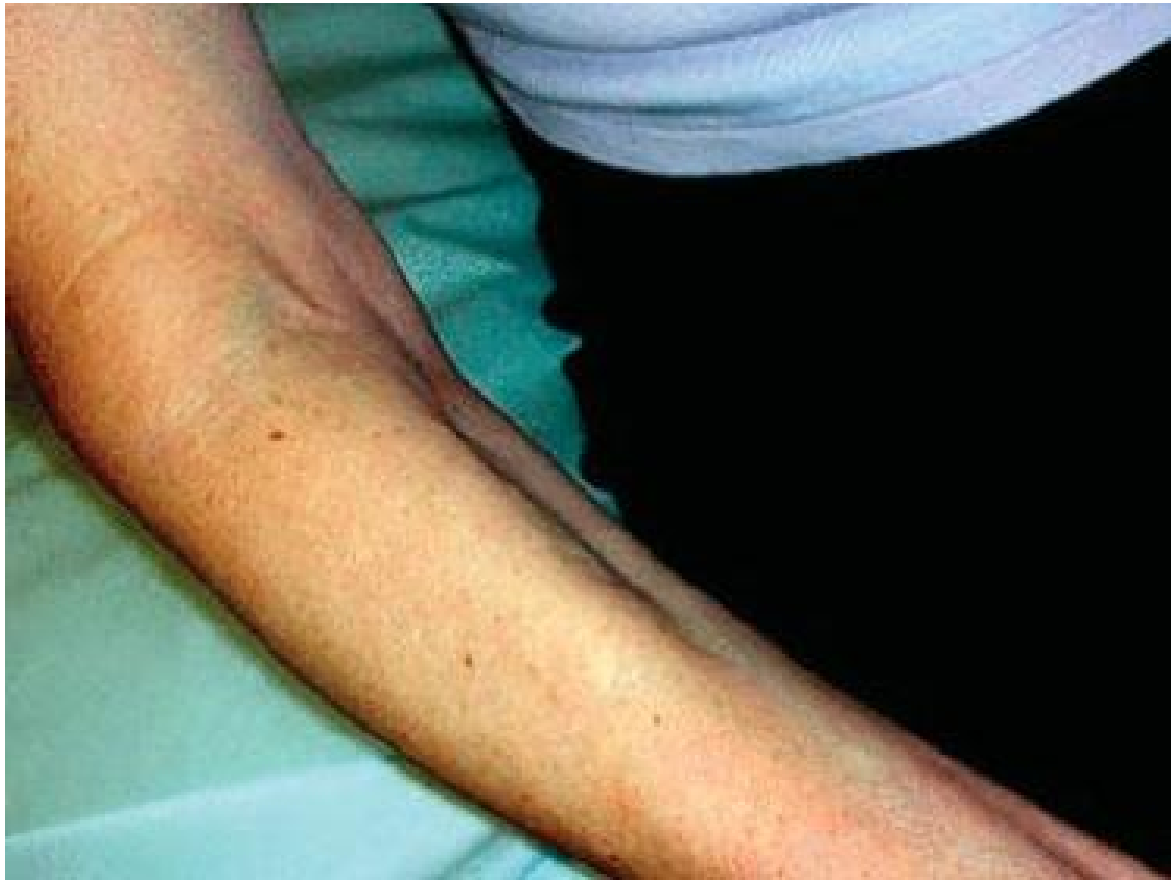
Examination

Wasting, weakness and fasciculations of all intrinsic muscles of right hand and some wasting of flexor compartment of forearm

Left arm normal, legs normal

Reflexes normal

Oblique Atrophy



Sensory NCS

Nerve / Sites	Rec. Site	Latency ms	Pk Amp µV	Duration ms	Distance cm	LatDiff ms	Vel m/s
R MEDIAN - Dig II Ortho							
1. Dig II-Wrist	Wrist	2.95	33.0	1.30	17	2.95	57.6
R ULNAR - Digit V Ortho							
1. Dig V-Wrist	Wrist	2.45	21.0	1.25	14	2.45	57.1

Motor NCS

Nerve / Sites	Latency ms	Amp mV	Dur. ms	Area mVms	Dist. cm	Lat Diff ms	Vel m/s
R MEDIAN - APB							
1. Wrist	4.25	6.8	6.65	28.7		4.25	
2. Elbow	8.90	6.8	6.65	28.7	24	4.65	51.6
R ULNAR - ADM							
1. Wrist	**5.45**	**2.0**	5.80	5.8		5.45	
2. B.Elbow	10.25	**2.2**	6.35	6.0	22	4.80	45.8
3. A.Elbow	12.20	**2.1**	6.40	6.3	10.5	1.95	53.8

EMG

	Spontaneous					MUAP			Recruitment
	IA	**Fib**	**PSW**	**Fasc**	**H.F.**	**Amp**	**Dur.**	**PPP**	**Pattern**
R. FCU	2+	3+	2+	None	None	1+	2+	3+	Reduced
R. APB	1+	1+	1+	None	None	1+	1+	1+	Reduced
R. BICEPS	N	None	None	None	None	N	N	N/+	N
R. BR	N	None	None	None	None	N	N	N	N
R. FCR	N	None	None	None	None	N	N	N/+	N
R. FDIO	2+	3+	2+	None	None	1+	2+	3+	Discrete
R. EDC	N	None	None	None	None	N	N	N	Reduced
R. PT	N	1+	1+	None	None	N	N	1+	N
R. FDS	N	None	None	None	None	N	N	1+	N

Differential Diagnoses?

Monomelic Amyotrophy

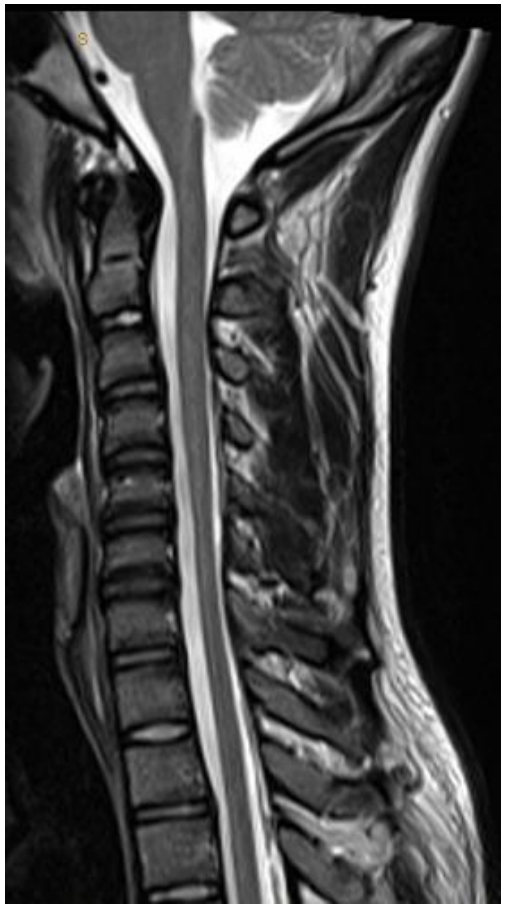
Focal SMA

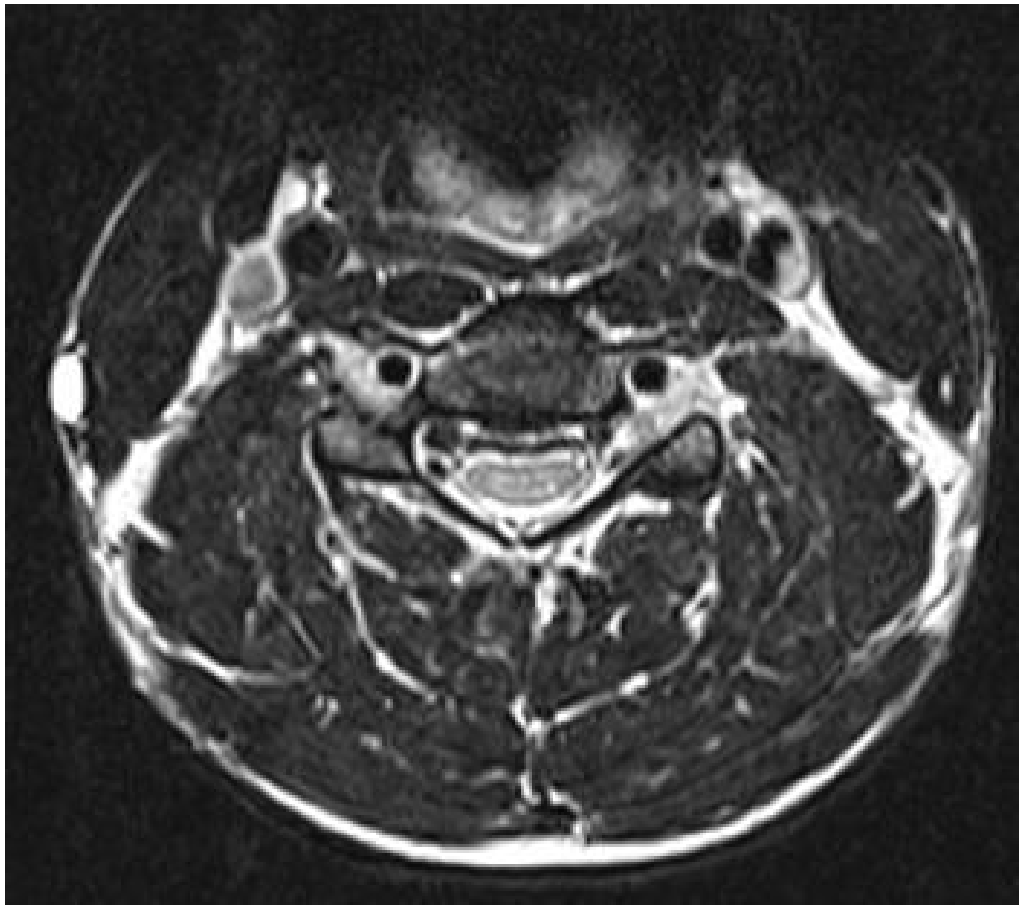
MMN

Toxic motor neuronopathy

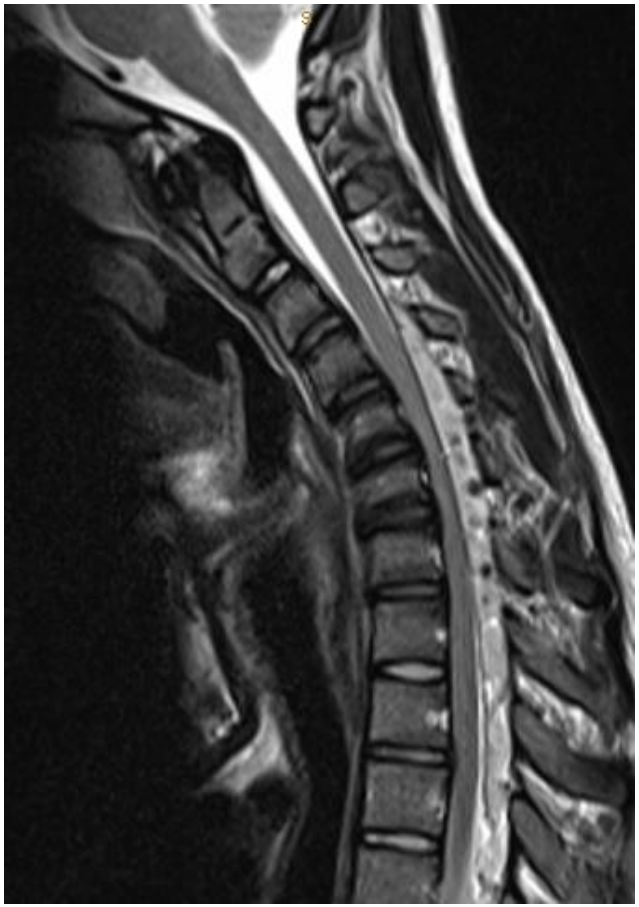
Variant ALS???

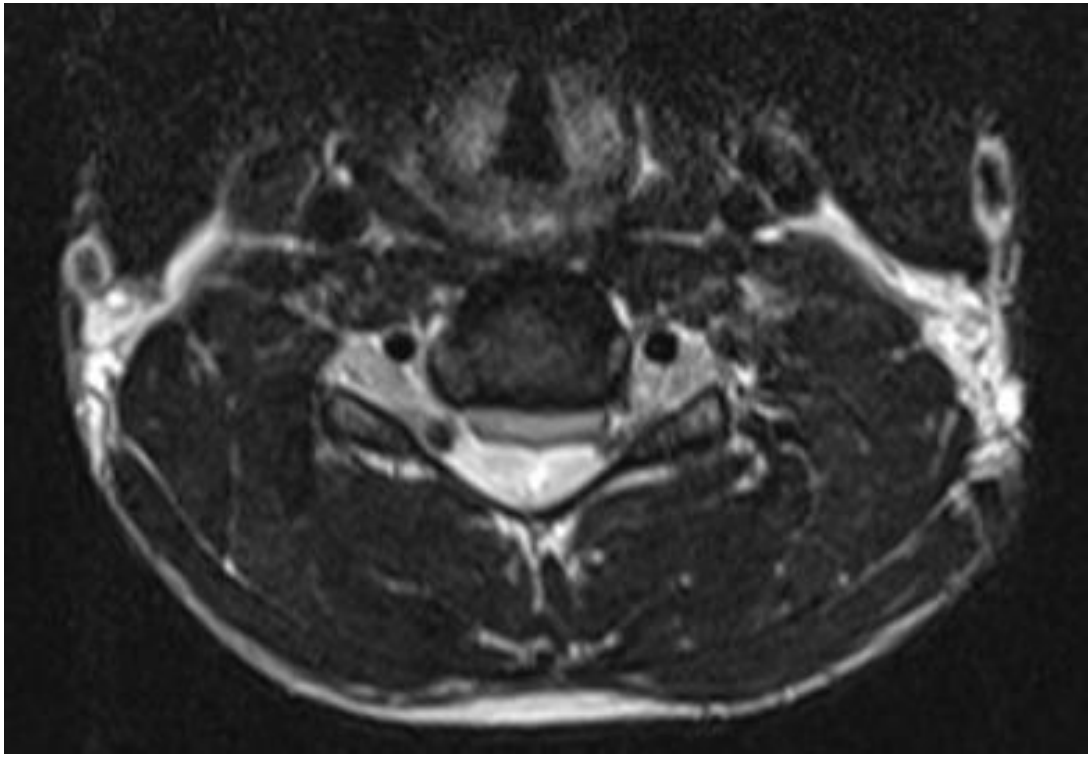
MRI



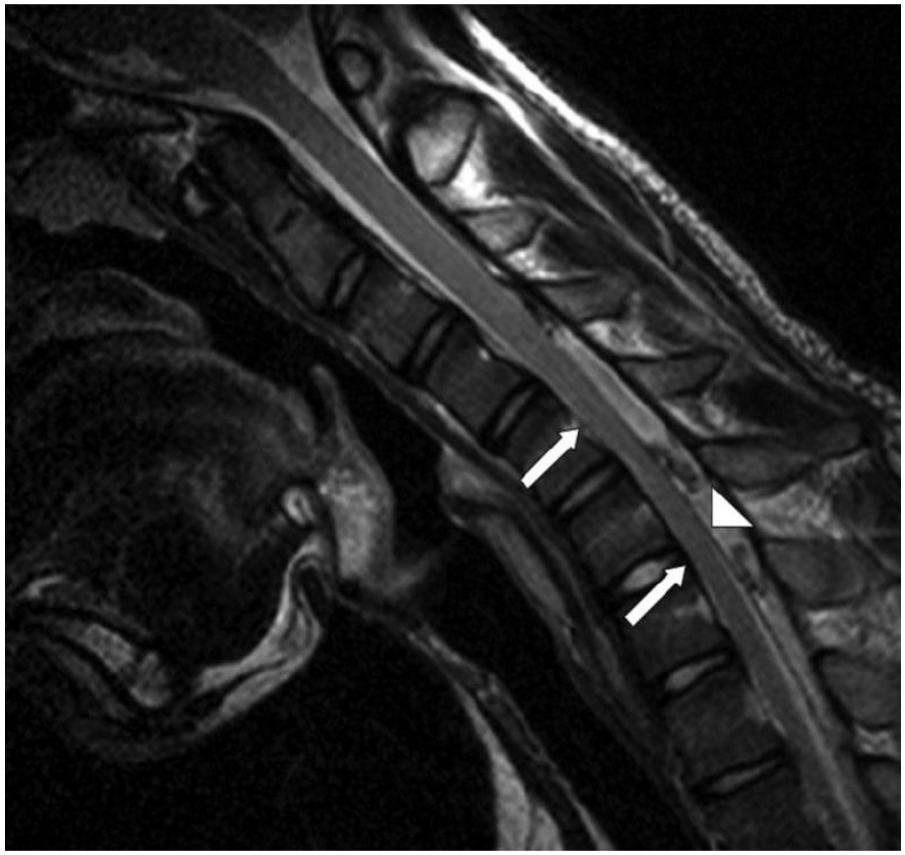


MRI





MRI from another patient



Synonyms

Monomelic atrophy
Benign focal atrophy
Benign focal amyotrophy
Benign monomelic amyotrophy
Distal amyotrophy of predominantly the upper limbs
Juvenile segmental muscular atrophy
Juvenile type of distal and segmental atrophy of upper extremities
Juvenile muscular atrophy of the upper extremity
Juvenile nonprogressive muscular atrophy localized in hand and forearm
Juvenile distal spinal muscular atrophy of upper extremities
Juvenile amyotrophy of distal upper extremity
Non-familial spinal segmental muscular atrophy in juvenile and young subjects
Non-familial juvenile distal spinal muscular atrophy of upper extremity
Non-familial juvenile central neurogenic muscular atrophy
Focal cervical poliopathy causing juvenile muscular atrophy of distal upper extremity
Monomelic spinal muscular atrophy
Benign juvenile focal muscular atrophy of upper extremities
Spinal monomelic amyotrophy
Unilateral juvenile muscular atrophy of upper limbs

Pathophysiology

Anterior displacement of posterior wall of lower cervical dural sac with widening of epidural space and obliteration of subarachnoid space

- Anterior displacement of cervical cord on neck flexion occurs in normal individuals and in disease control with cervical cord atrophy
- Disproportional length between vertebrae and dural canal in adolescents leading to overstretching of cord on flexion
  - Long cervical spine but short spinal cord
- Repeated or sustained flexion causes compression of cord leading microcirculatory compromise in ASA territory