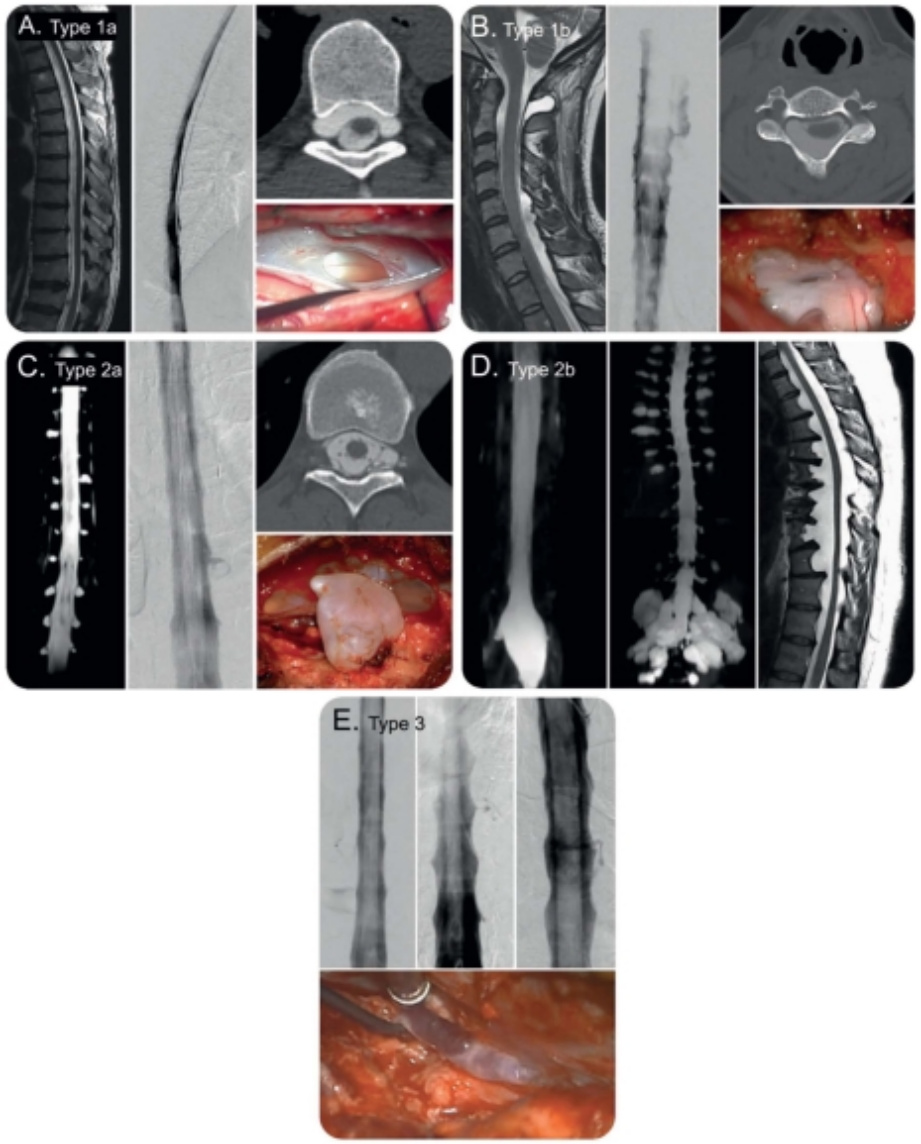
Dural ectasia and orthostatic headache

Marcus Stoodley

MACQUARIE University

Disclosures: Nil

Dural ectasia



Type 1 – dural tears ~ 25%

a: ventral ~ 95%

b: postero-lateral

Type 2 – meningeal diverticula ~ 40%

a: simple ~ 90%

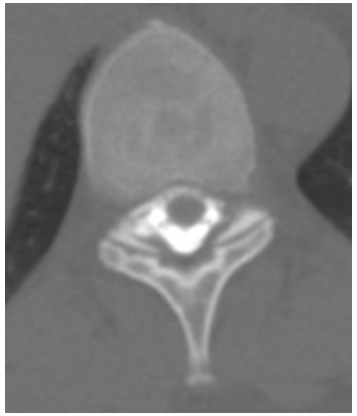
b: dural ectasia

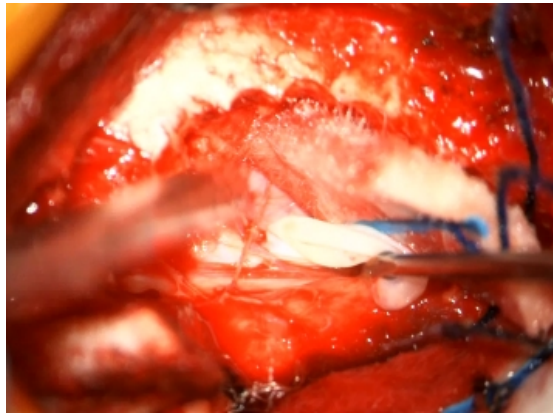
Type 3 – CSF-venous fistulae ~2.5% (in 2016)

Schievink et al, Neurology 2016

Type 2: Meningeal diverticula

a) Simple





Usually paraspinal CSF

a) Dural ectasia

Meningeal cysts

Tarlov (1938): ‘perineural cysts’

TABLE 1

Classification of spinal meningeal cysts

Type	Description
I	extradural meningeal cysts without spinal nerve root fibers
IA	"extradural meningeal cyst" ("extradural arachnoid cyst")
IB	"sacral meningocele" ("occult sacral meningocele")
II	extradural meningeal cysts with spinal nerve root fibers ("Tarlov's perineurial cyst," "spinal nerve root diverticulum")
III	spinal intradural meningeal cysts ("intradural arachnoid cyst")

J Neurosurg 68:366–377, 1988

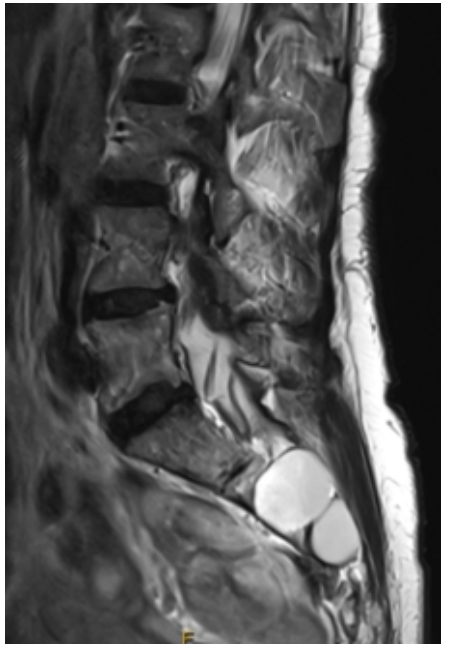
Updated assessment and current classification of spinal meningeal cysts

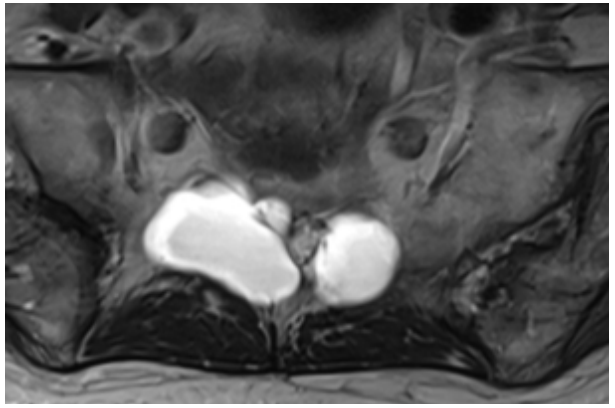
Michael W. Nabors, M.D., T. Glenn Pait, M.D., Edward B. Byrd, M.D., Najmaldin O. Karim, M.D., David O. Davis, M.D., Arthur I. Kobrine, M.D, Ph.D., and Hugo V. Rizzoli, M.D.

Meningeal cysts

Tarlov cysts

- Junction of dorsal root and ganglion
- Between perineurium (arachnoid) and endoneurium (pia)
- Sacral common
- Pathogenesis unclear
  - ? Ball-valve
  - ? Disruption of CSF-venous drainage
  - ? Congenital – dural ectasia





67 y.o.woman, SIH, no connective tissue disorder

Murphy et al, Neuroradiology (2024) 66:1–30

Meningeal cysts

Tarlov cysts

- ~ 5 – 10% of population
- Women > men
- Increase with age (rare in children)
- Associated with IIH
- ~4% of SIH patients Schievink et al Neurology 87(7):673–679

Meningeal cysts

Tarlov cysts

- Can be symptomatic
  - Nerve pain
  - Periosteal pain
  - Fractures
  - Pre-sacral meningoceles
- Never spontaneously regress
- Can increase in size
  - *Associated with SIH* Yang et al Neurosurgery 86(1):88–92

Meningeal cysts

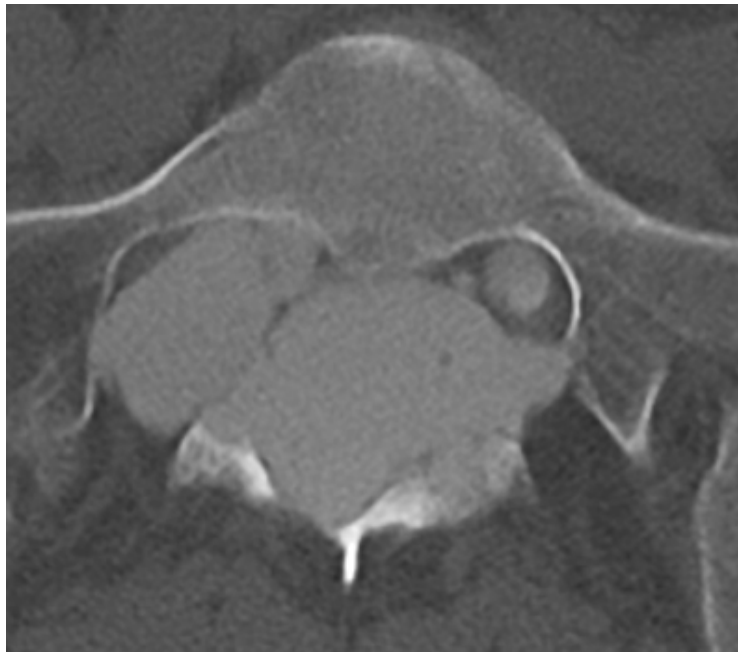
Tarlov cysts

- Treatment
  - Percutaneous fibrin glue (*note narrow vs wide-neck*)
  - Surgery: varied techniques

Meningeal cysts

Tarlov cysts *vs* simple meningeal diverticula?

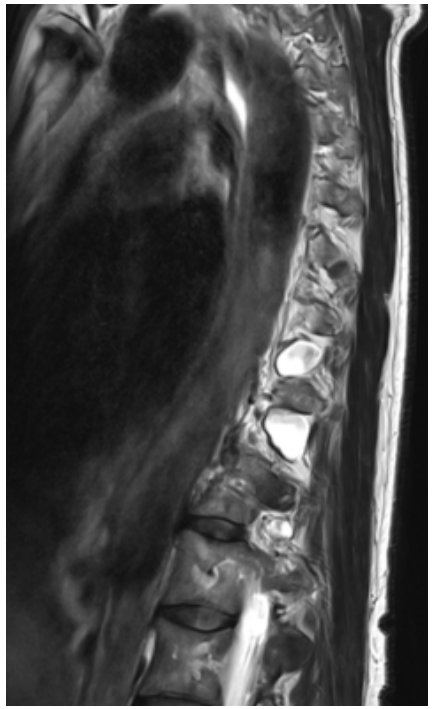
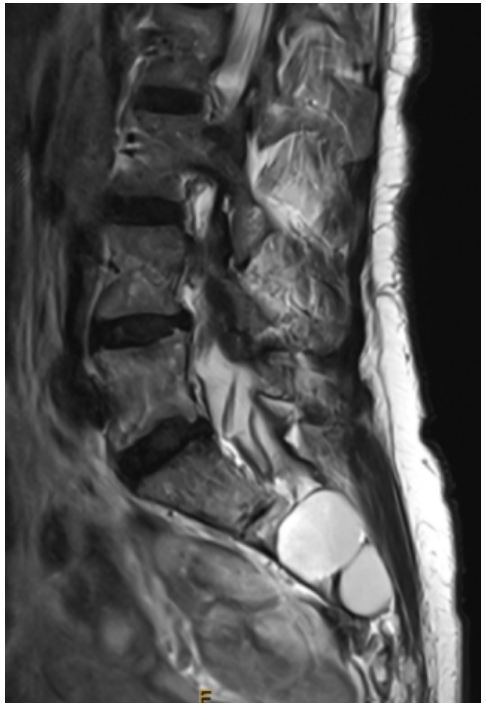
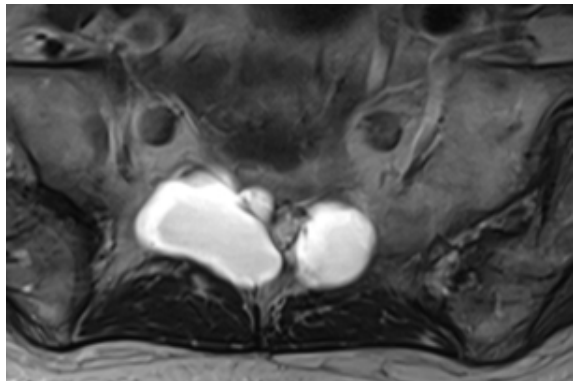
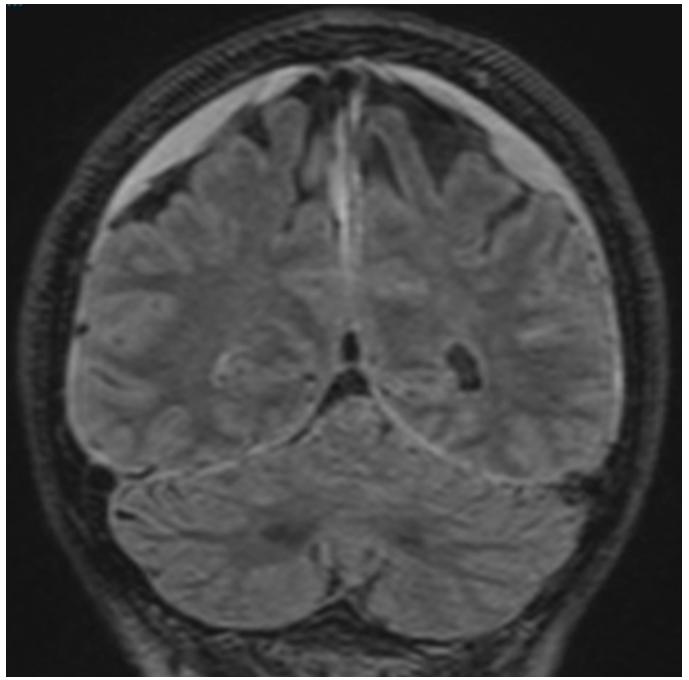
- Tarlov cyts



Meningeal cysts

Tarlov cysts *vs* simple meningeal diverticula?

- Tarlov cyts

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Meningeal cysts

Tarlov cysts *vs* simple meningeal diverticula?

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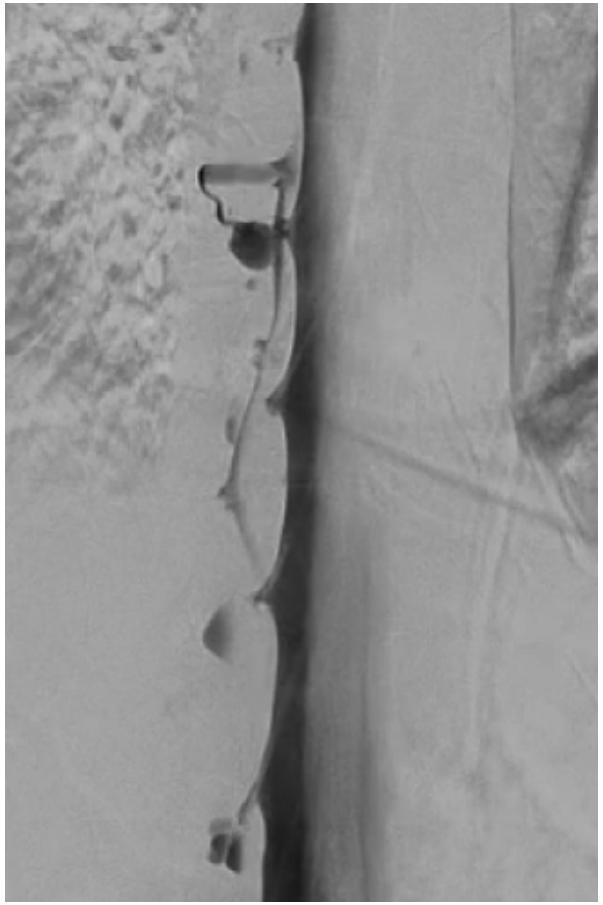
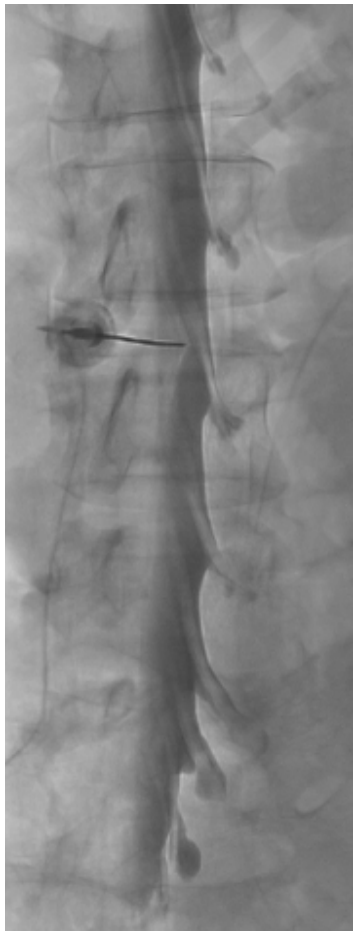
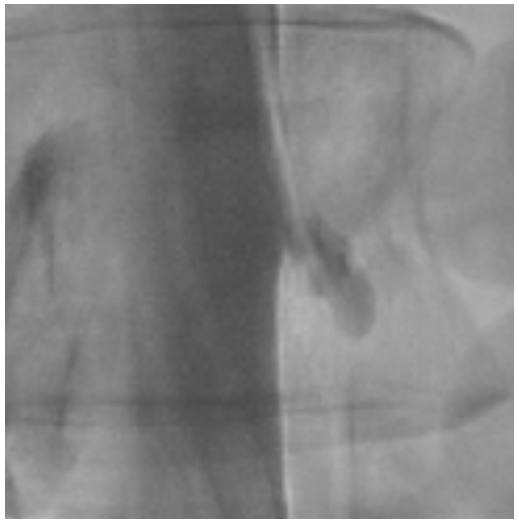


Meningeal cysts

Tarlov cysts *vs* simple meningeal diverticula?

- Diverticulum

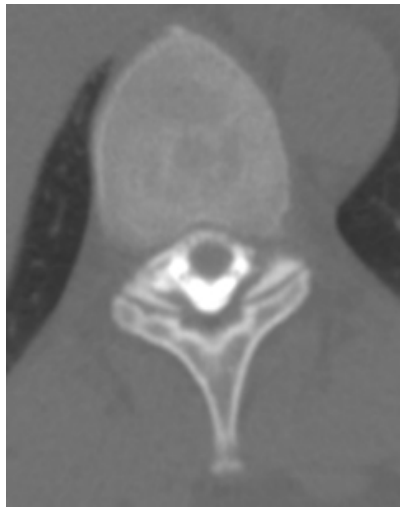




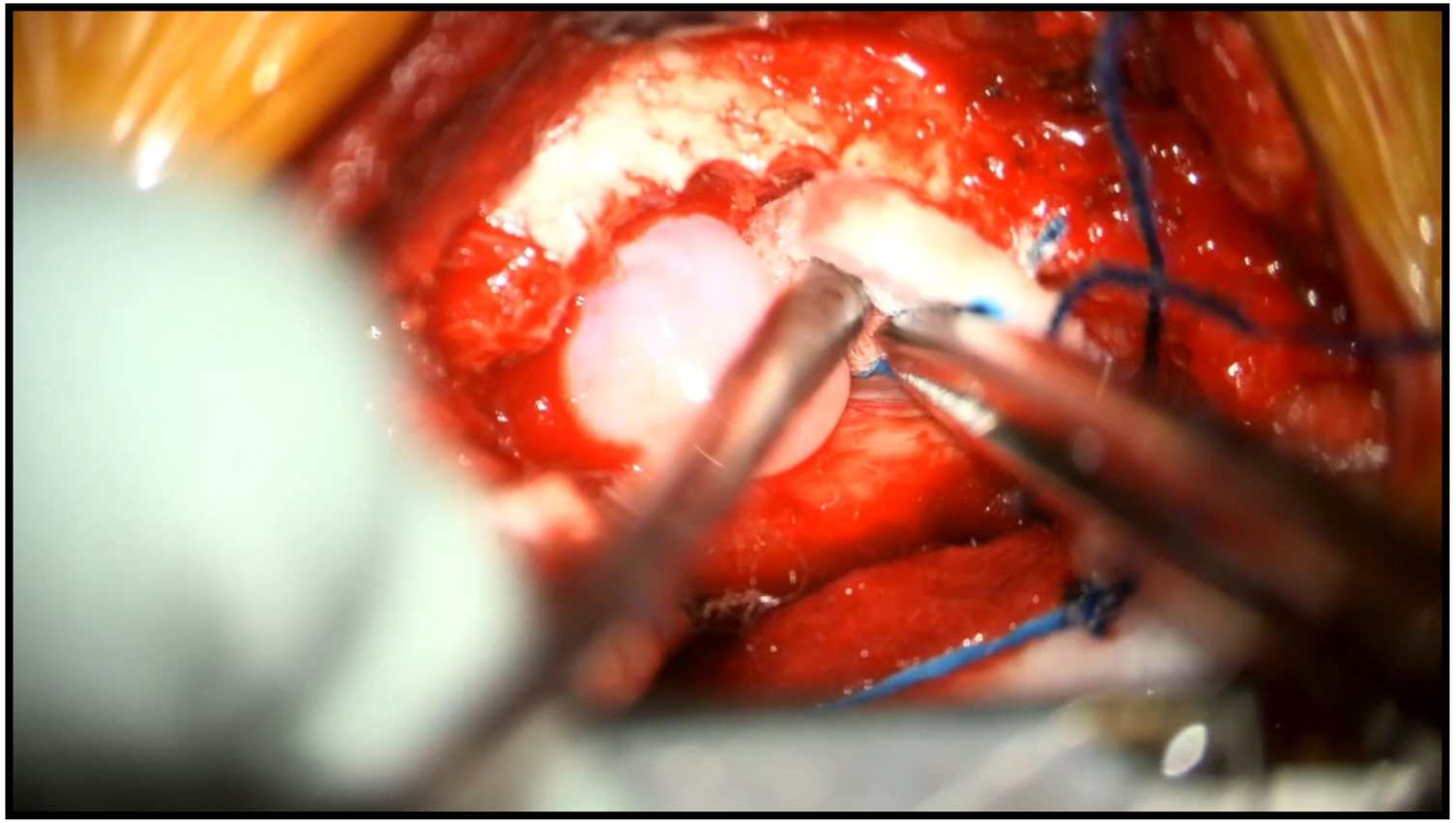
Meningeal cysts

Tarlov cysts *vs* simple meningeal diverticula?

- Simple diverticula



Usually paraspinal CSF



Dural ectasia

- Widening or enlargement of dural sac
- Most commonly lumbosacral
- Weak dura or high CSF pressure

Dural ectasia

- Marfan (⅔ of cases)
- Loeys-Dietz
- Ehlers-Danlos
- Neurofibromatosis type 1
- Ankylosing spondylitis
- Idiopathic

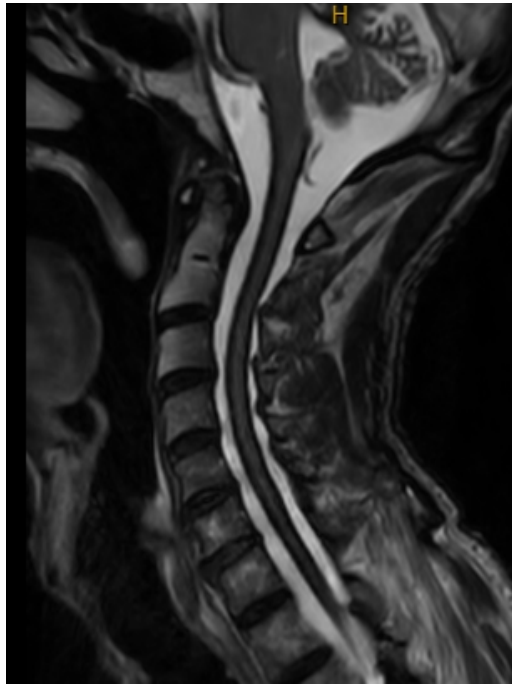
- ✂prevalence of Tarlov cysts with ectasia

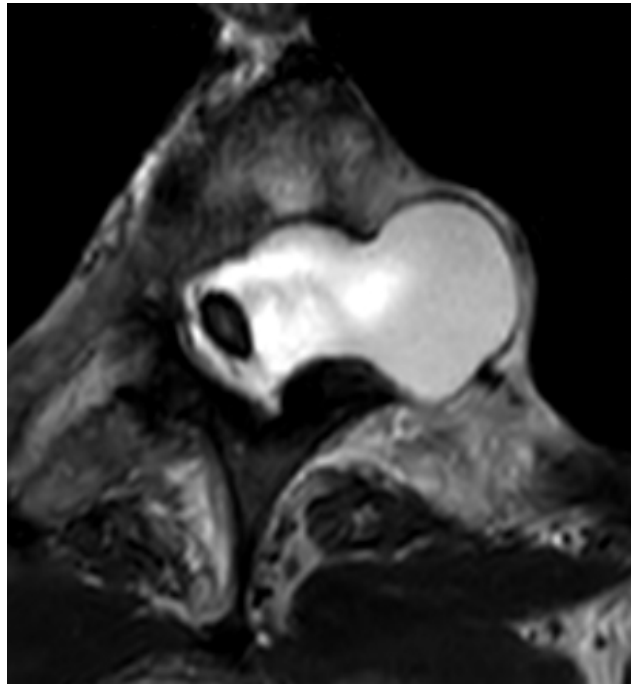
Dural ectasia

- Cause of SIH?
  - Increased compliance
  - Dural tears (Type I)
  - Meningeal diverticula (Type II)
  - Tarlov cysts → CSF-venous fistula

Dural ectasia

- Cause of superficial siderosis?
  - Bleeding from low pressure
  - Blood entering through dural tear
  - Vascular fragility





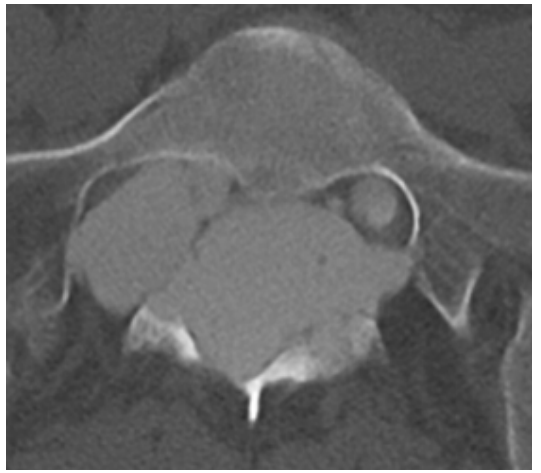
Dural ectasia

- Marfan syndrome

  - 1:5,000 – 1:10,000
  - Mutation in FBN1 → abnormal fibrillin (extracellular matrix glycoprotein)
    - Formation and maintenance of elastic fibres
    - Normal fibrillin sequesters TGF-β
  - Autosomal dominant
  - 25% spontaneous
  - Multisystem connective tissue disorder
    - Heart, aorta, lens, dura
  - Abnormal joint flexibility

Dural ectasia

- Marfan



Dural ectasia

- Loeys-Dietz syndrome
  - Described in 2005
  - Incidence unknown
  - Mutation of TGF genes (5 types)
    - Abnormal collagen
  - Autosomal dominant
  - 75% spontaneous
  - Clinical similarities to Marfan syndrome
    - Plus hypertelorism, abnormal skin

Dural ectasia

- Ehlers-Danlos

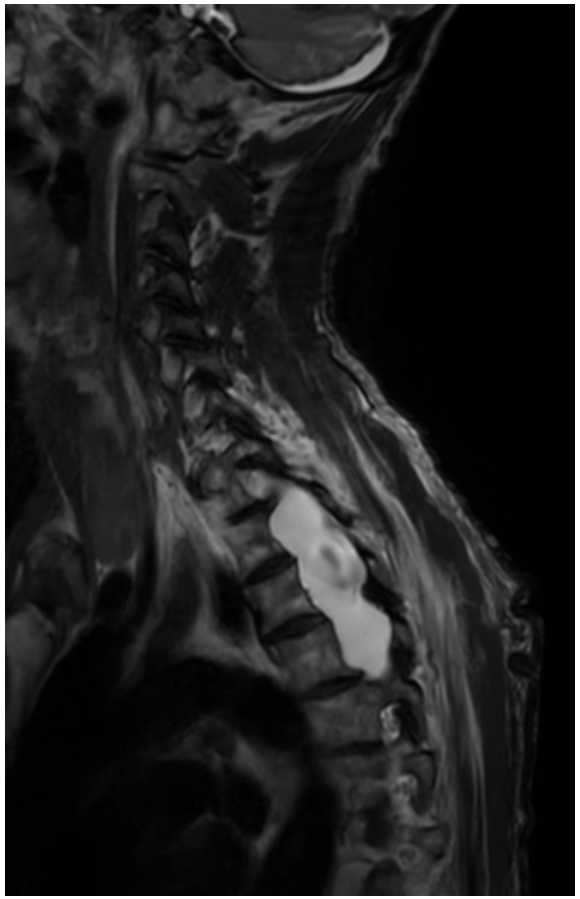
  - Multiple types (19 genes)
  - Hypermobile type 1:5,000
  - Abnormality of collagen or tenascin
  - Autosomal dominant or recessive
  - Hypermobile:
    - Cranio-cervical instability
    - Mast cell activation

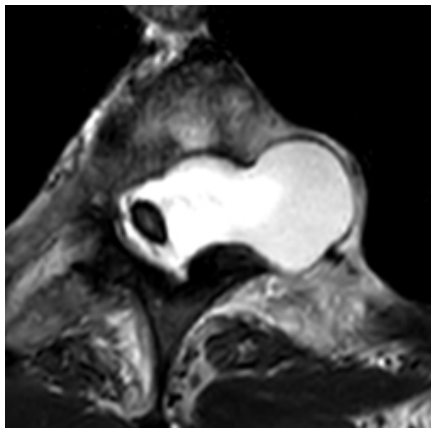
Dural ectasia

- Neurofibromatosis type 1
  - Mutation of Neurofibromin
    - GTPase-activating protein, regulates RAS/MAPK
  - Autosomal dominant
  - ? Infiltration of dura by plexiform neurofibroma
  - Dura thin and fragile
  - ? Sacral less common

Dural ectasia

- Neurofibromatosis type 1





Dural ectasia

- Ankylosing spondylitis

  - Aetiology unknown
    - ? autoimmune
  - Associated with HLA-B27
  - Chronic inflammation of spinal joints
  - Sacral nerve root sheath ectasia

Dural ectasia

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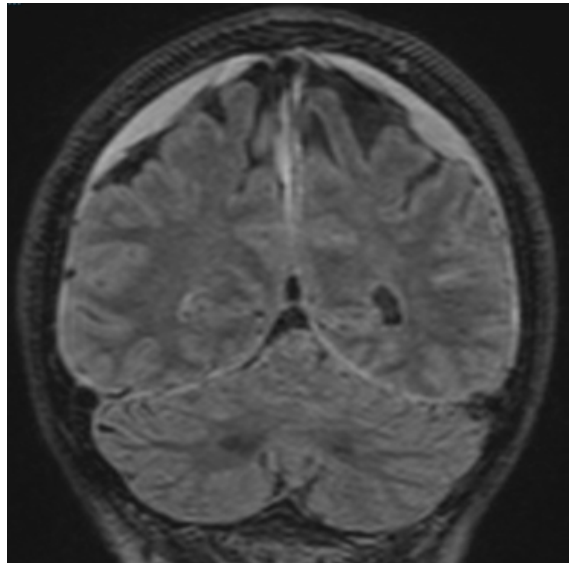
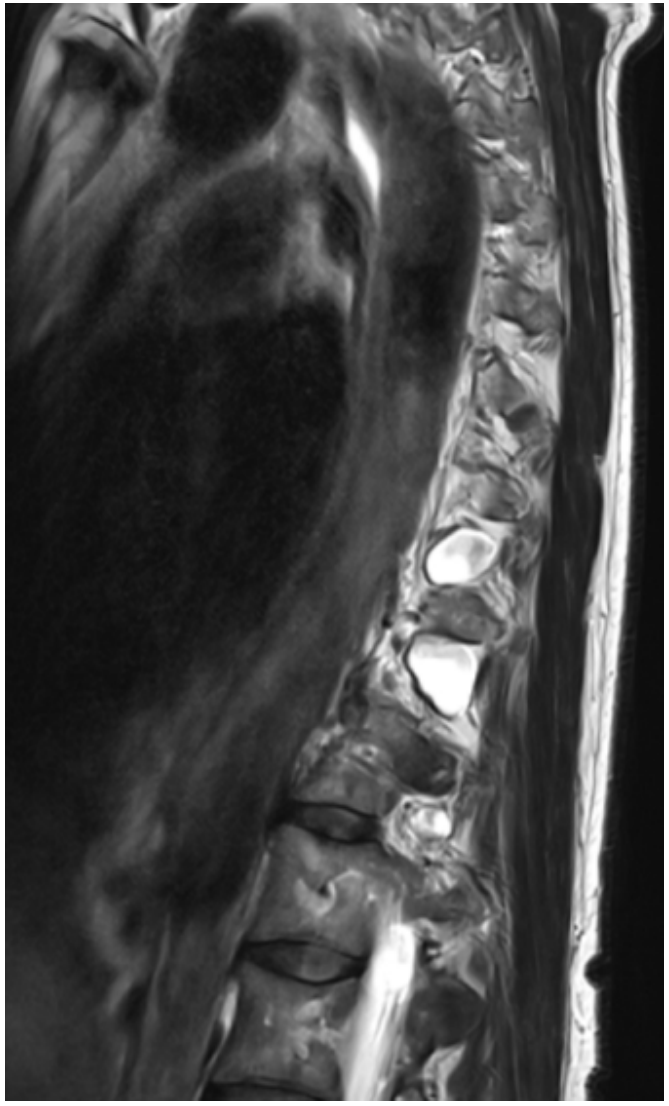
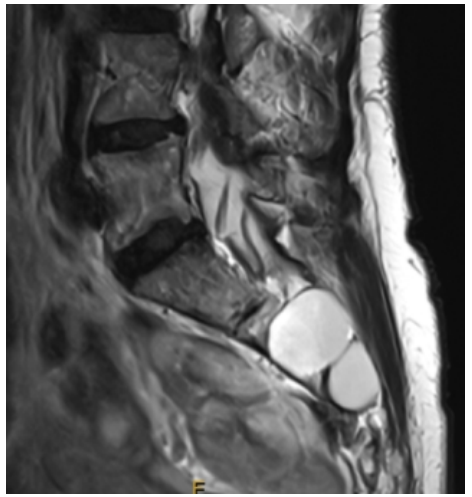
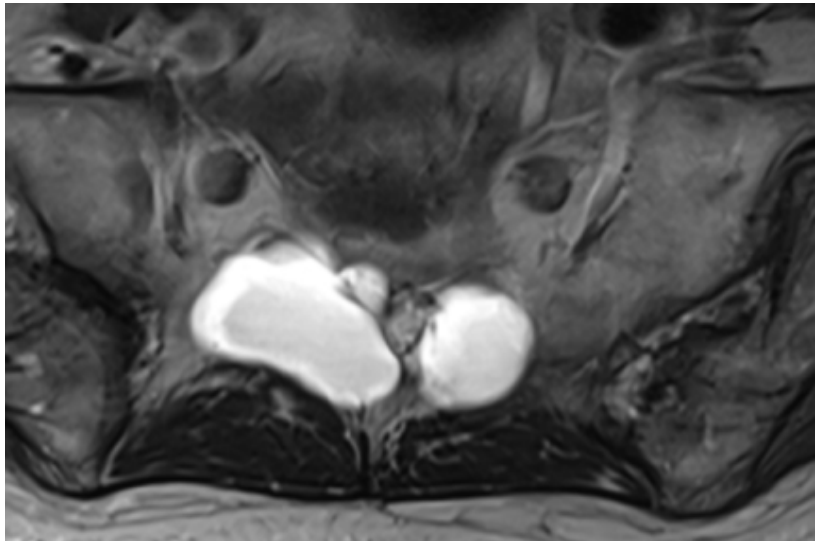
Investigation: MRI









67 y.o.woman, SIH, no connective tissue disorder

Dural ectasia

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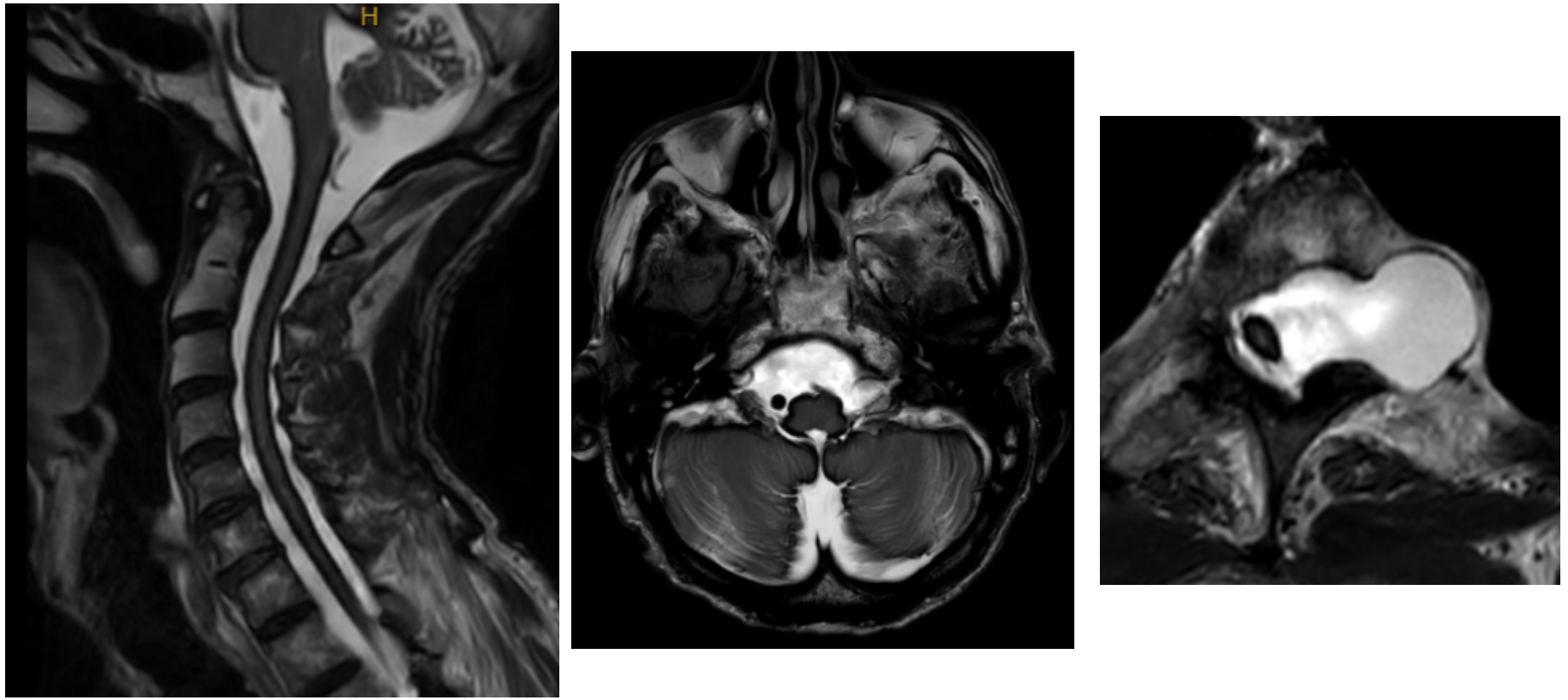
Investigation: MRI



Dural ectasia

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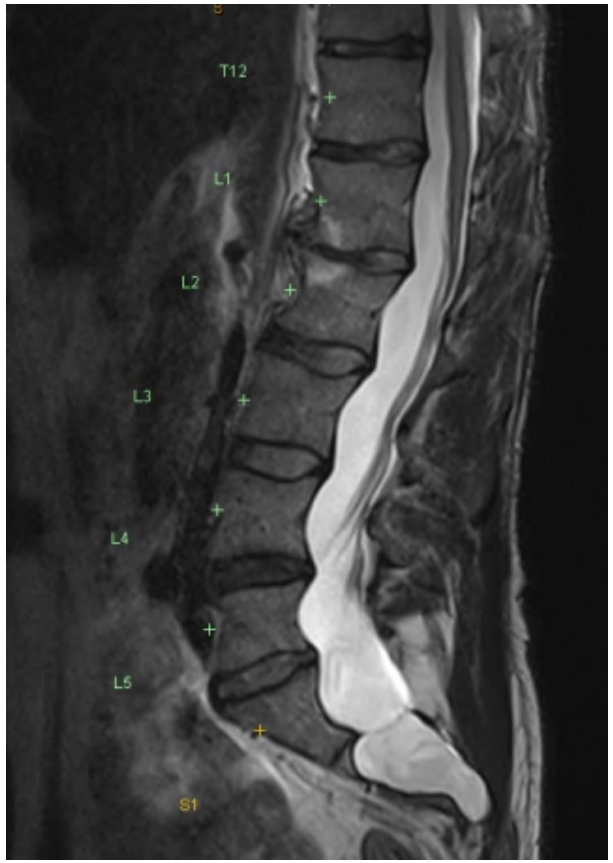

Dural ectasia

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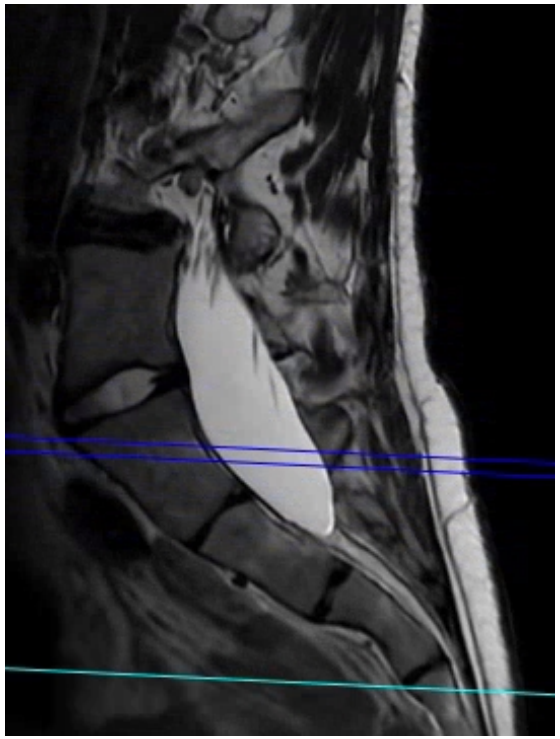

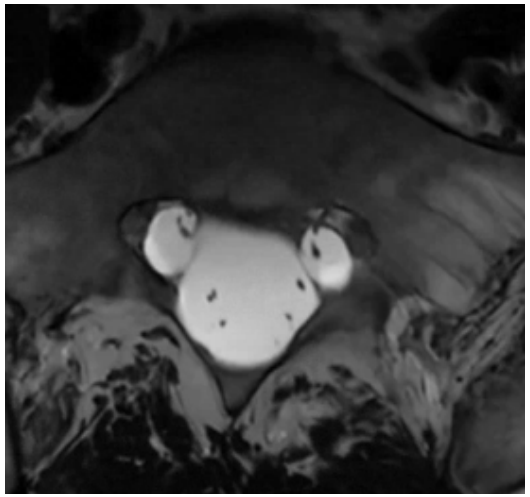

Dural ectasia

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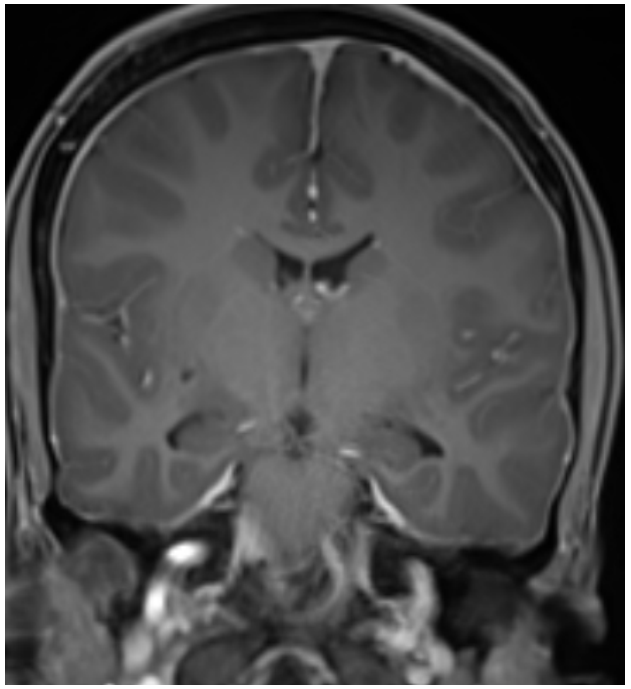

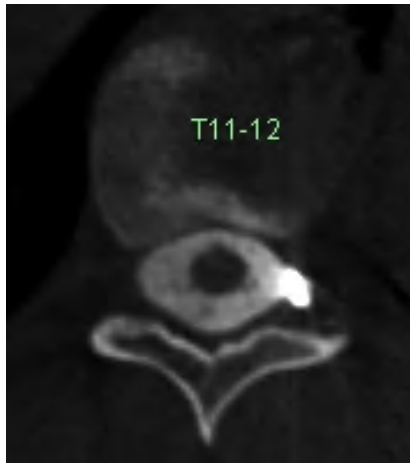

Dural ectasia

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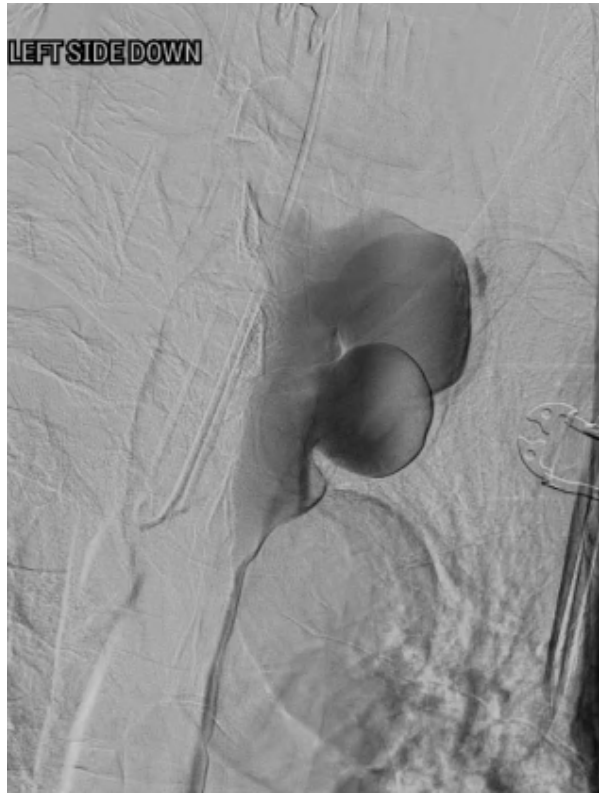
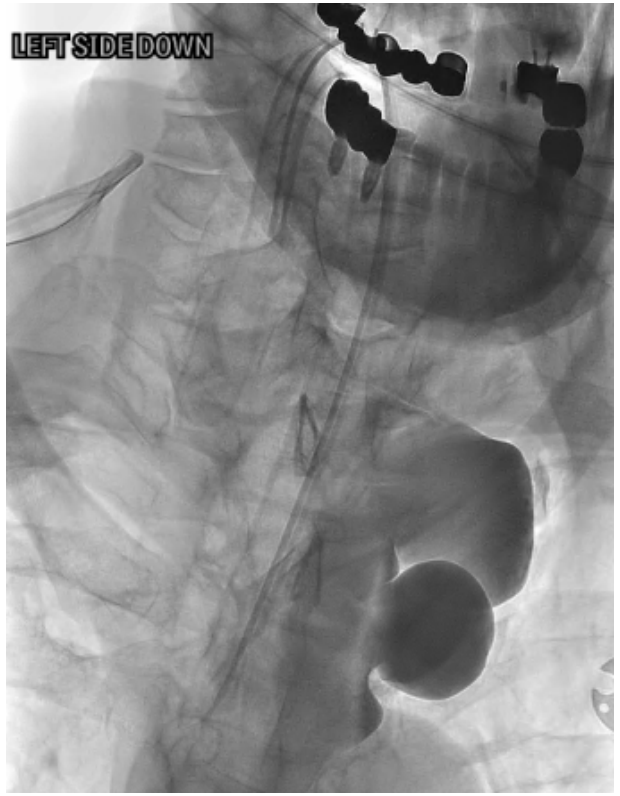
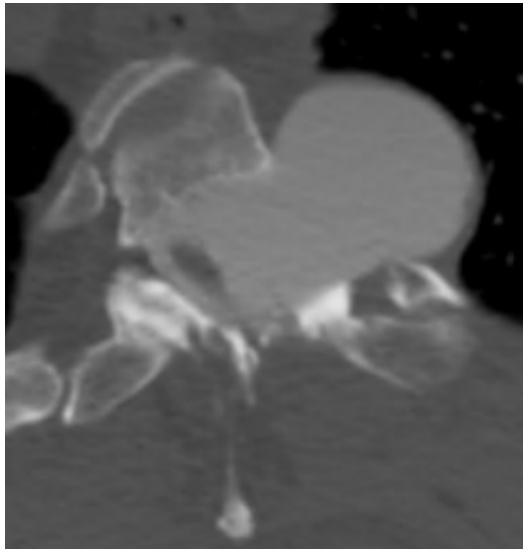
Investigation: Myelography







Dural ectasia

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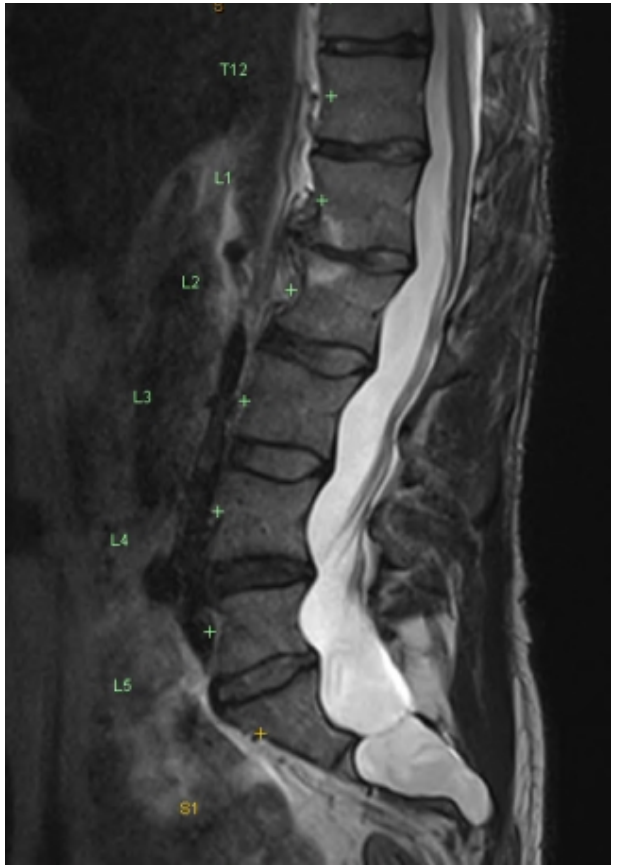
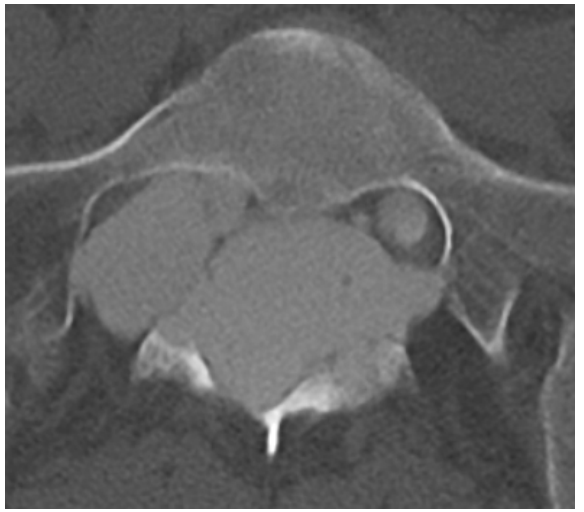
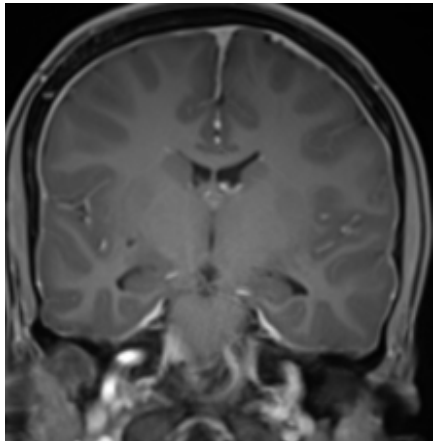
Investigation: CT myelogram

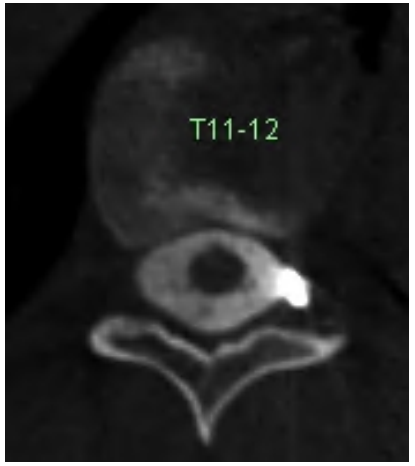




Dural ectasia

Investigation: CTM





Dural ectasia

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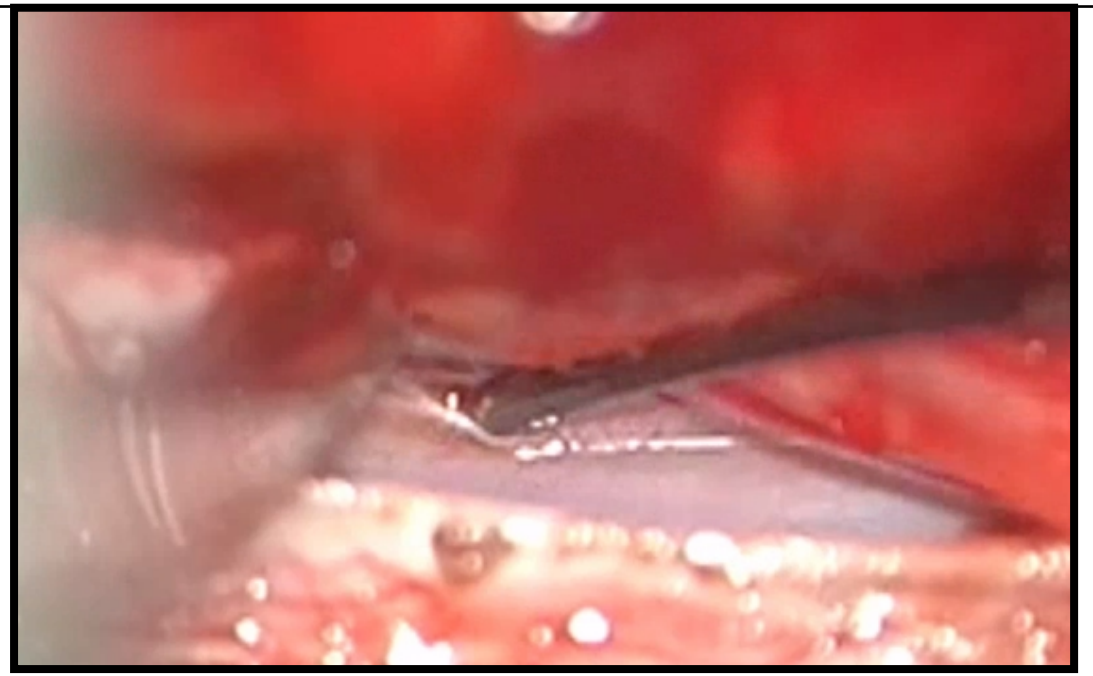

Dural ectasia

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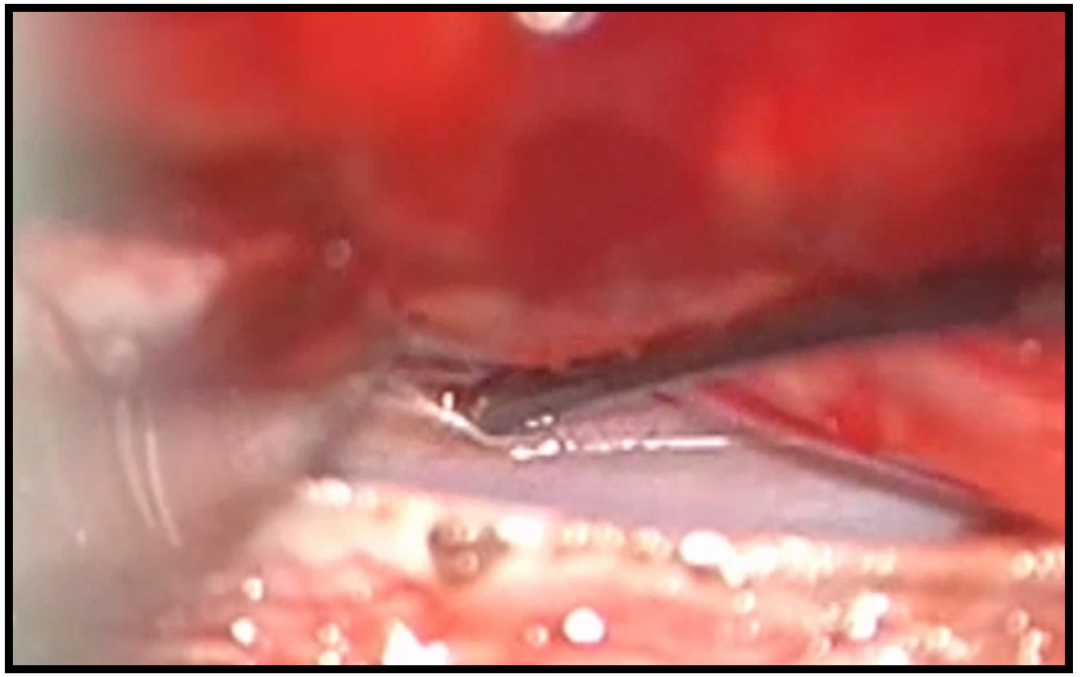
Treatment

- Blood patch

Dural ectasia

Treatment

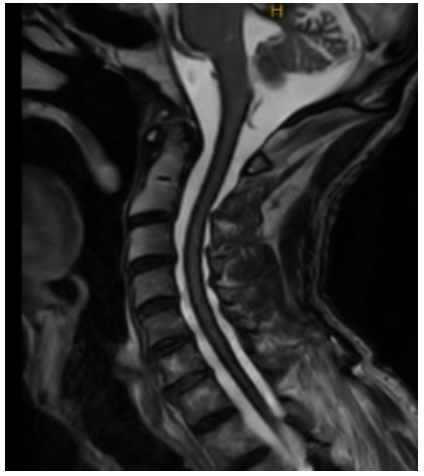
- Surgery – repair ventral leak

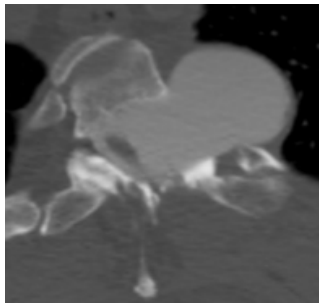


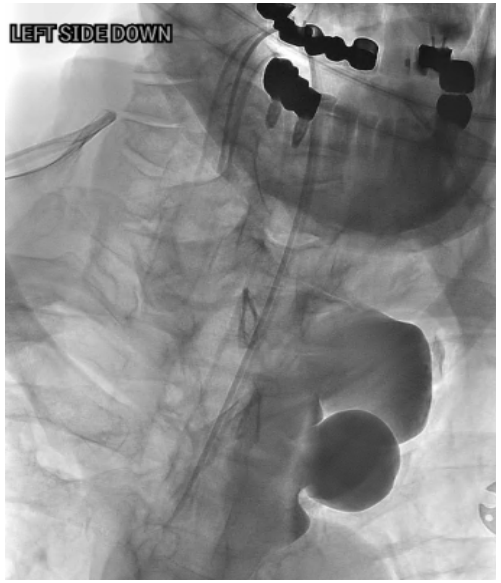
Dural ectasia

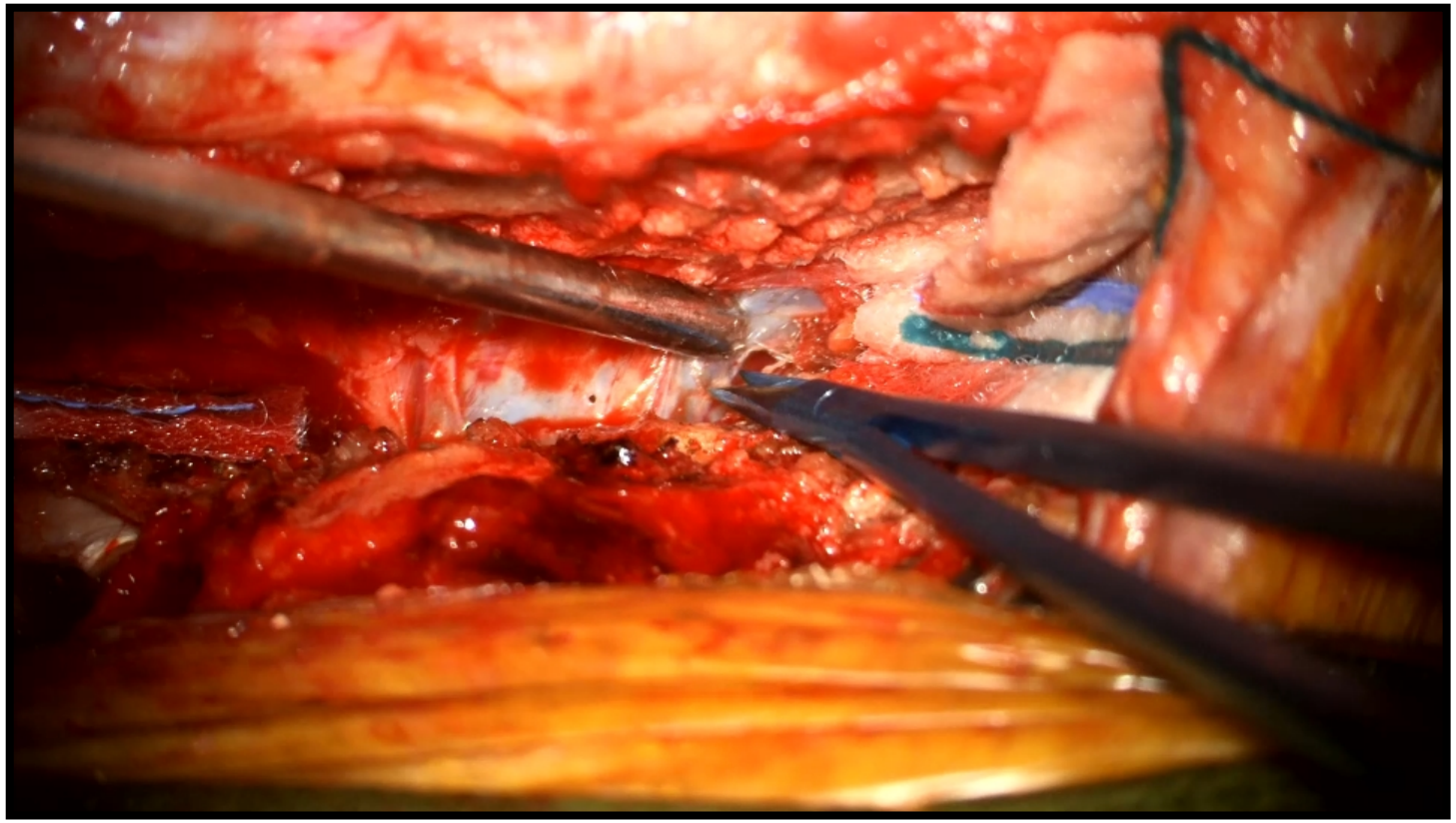
Treatment

- Surgery – resect nerve root cyst









Dural ectasia

Treatment

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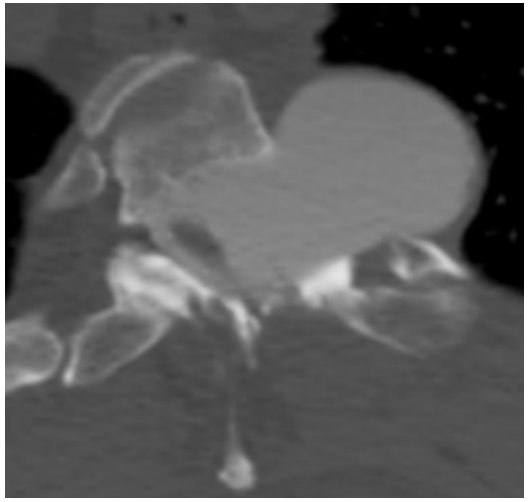

Dural ectasia

Treatment

- Surgery – dural reduction
  - Plication
  - Imbrication
  - Resection

Dural ectasia

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Conclusions

- SIH with dural ectasia is especially difficult!
  - Identifying site of leak
  - Blood patch seems to work well
  - Surgery results unclear