Idiopathic spinal cord herniation

I have nothing to disclose !

Patient

- Male 59 Y
- Antifosfolipid syndrome
- Otherwise healthy
- Slowly evolving complaints in 2 years

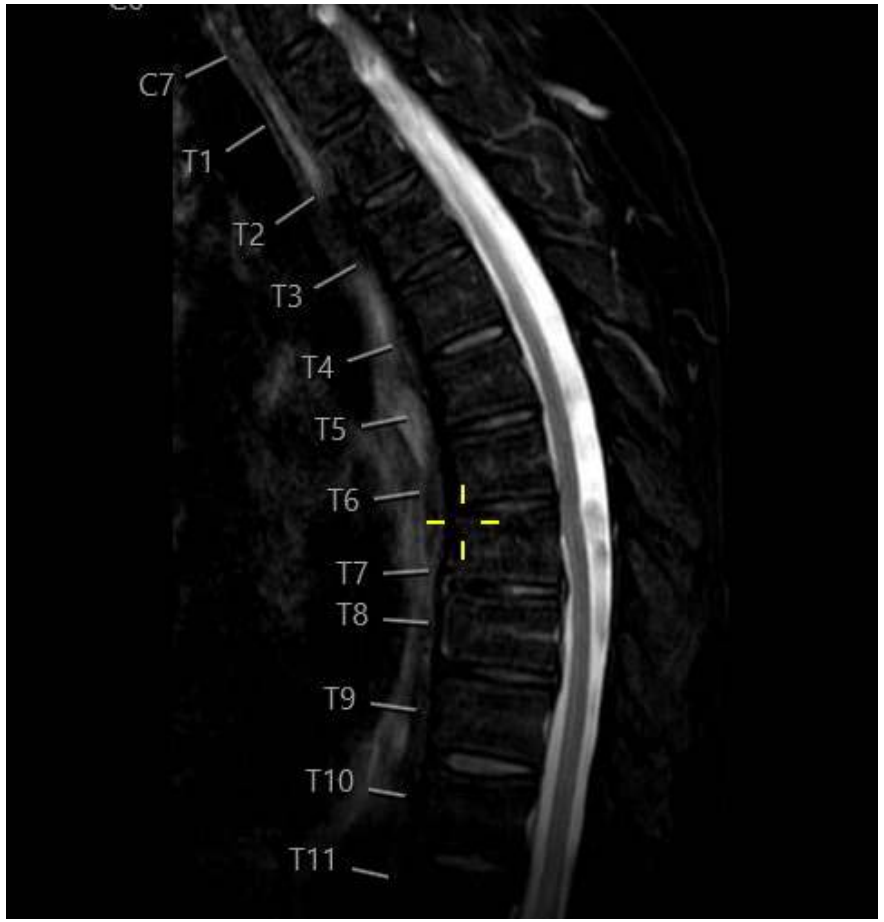
History

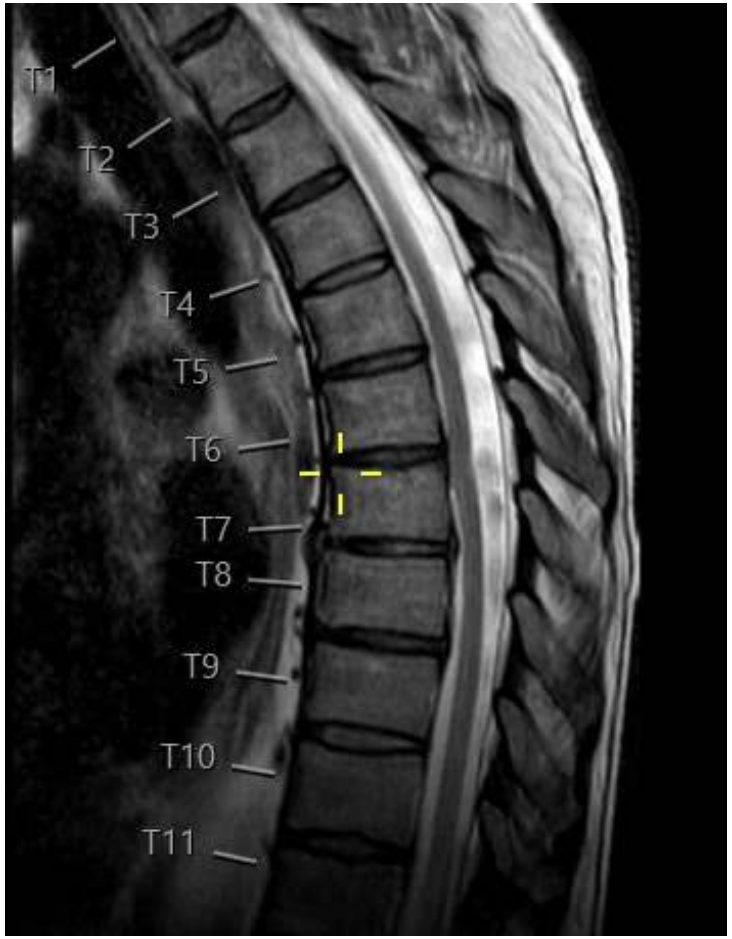
- Sensory disturbances left leg and lower abdomen, trouble discriminating warm cold when taking showers
- Some pain inside left leg and painful sensations left side of the trunk 10 cm below the nipples
- Increased stiffness in legs in the morning and mild difficulty walking

Neurological examination

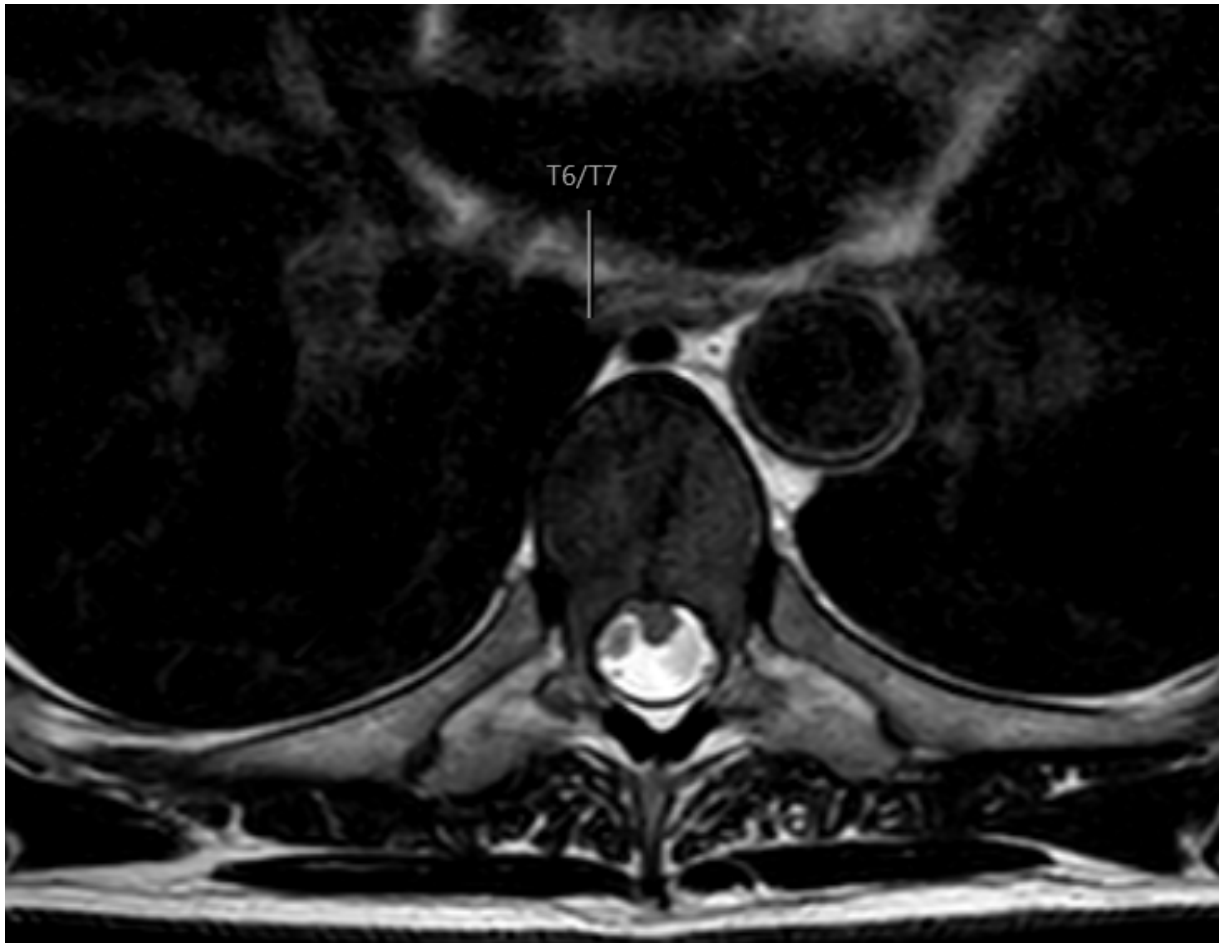
- Diminished sensation for touch and pain left leg and trunk up th Th10
- Inability to discriminate warm /cold (decreased temperature sense) left leg
- Gnostic sensibility intact on left side
- No right sided sensory disturbances (vital and gnostic)
- Subtle piramidal signs legs: slight reflex dissociation legs /arms and discutable left plantar reflex (Babinski ?)
- Otherwise normal

MRI anterior displaced spinal cord Th6-7





Clear anterior spinal cord herniation



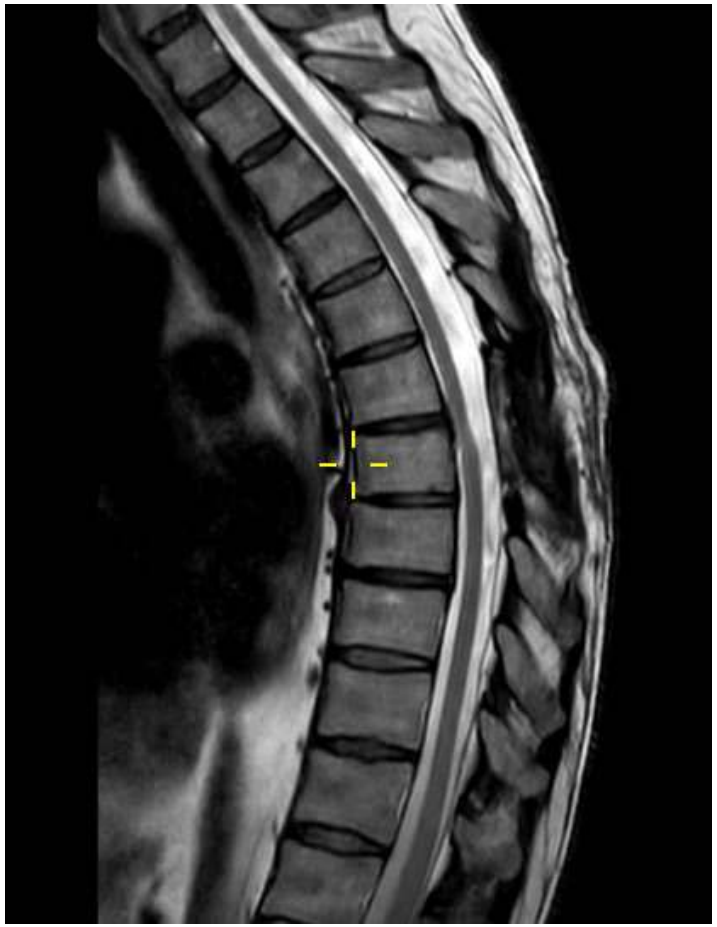
Surgery

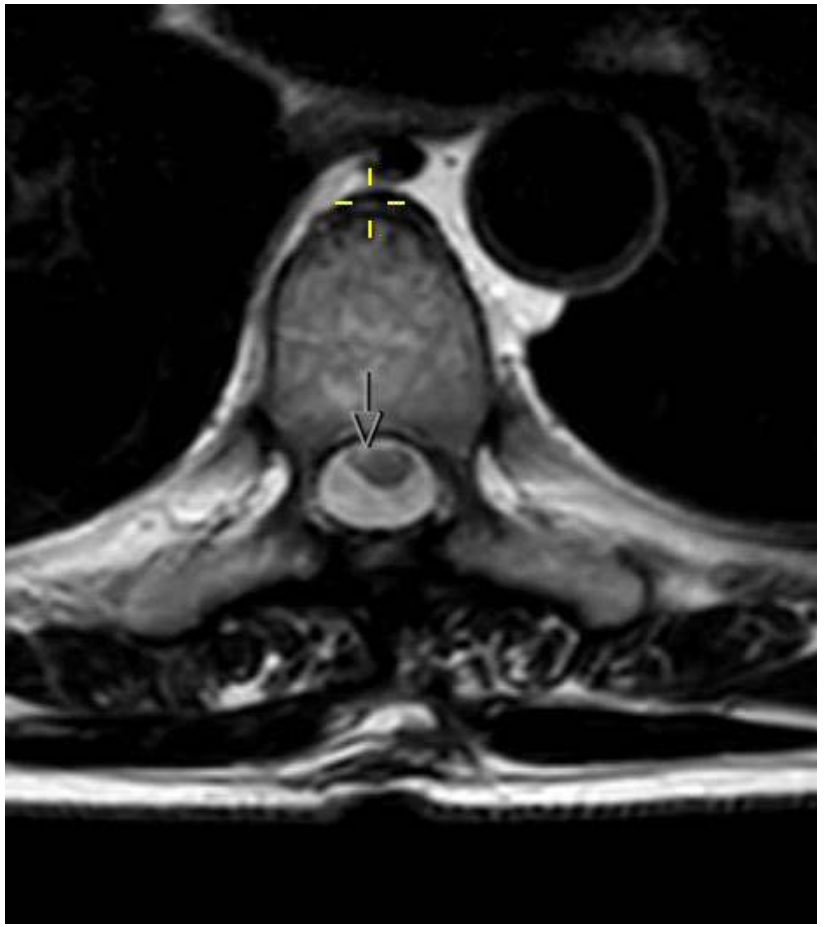
- General anesthesia under IONM
- Posterior approach targeted laminectomy
- Posterior dural opening and spinal cord mobilization
- Reduction of the herniated spinal cord from the anterior dural defect
- Closure of the anterior dura defect with muscle and fibrin glue

Postoperatively

- Some improvement in perceived stiffness of the legs
- Otherwise no changes
- Most disturbing persisting painful zone left side of the trunk

Postoperative MRI





Spinal cord herniation

- Anterior spinal cord is herniation through a dural defect leading to strangulation and tethering of the cord in the defect
- Almost exclusively through anterior defect in thoracic spine.

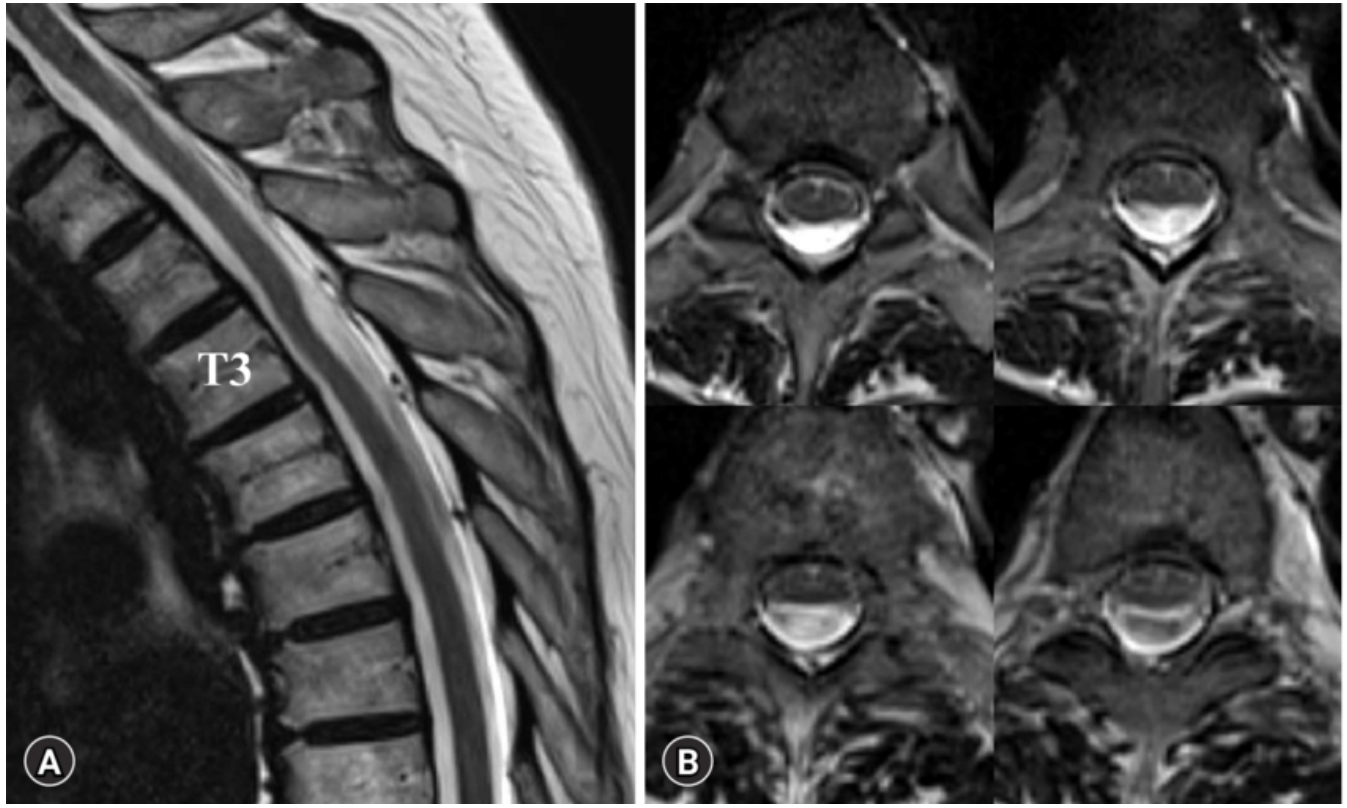
  Case report dorsal herniation (Tri, C et al. The Spine journal 12 (2012) e9-e12)
- Thoracic spine because of the kyfosis and ventral position of the cord
- Age at presentation 31-72 years (mostly 40-70); Female preponderance (1.72 v 1)
- Clinical Presentation: BSS, myelopathy, lower extremity motor/sens deficits, rarely SIH

Spinal cord herniation

- First described 1974 accidental finding during intradural thoracic disc surgery
- Rare condition used to be difficult to diagnose
- (high quality e.g. 3 D T2)), MRI, CT myelography
- Important to diagnose, surgical intervention is often useful

Differential diagnosis

- Mostly posterior intradural archnoid webs or cysts



Pathogenesis & pathophysiology

Mostly seen around the apex of the thoracic kyphosis because of the proximity of the cord to the ventral dura

Ventral dural defect:

- Developmental or congenital e.g dural duplicature/dissection
- Iatrogenic e.g after surgery
- (repetitive) Trauma?
- Degenerative disc changes eroding the ventral dura causing slitlike defects:

Cord herniation, Ventral CSF leakage causing SIH

(Cornips et al. World Neurosurgery 140:e311-e319 august 2020)

Spontaneous intracranial hypotenion

- Low pressure /positional headaches
- Typical cranial MRI abnormalities
- Spinal leak
  - Anterior or posterolateral
  - Dural ectasia/diverticula
  - CSF venous fistula’s
- Often extrathecal/epidural spinal fluid collections
- Long standing complications
  - Bibrachial amyotrophy
  - Superficial siderosis

Another patient

(courtesy of Mathijs Brouwer and Peter Vandertop Amsterdam, University Hospital (VU –AMC))

- Referred from other hospital
- Unexplained myelopathic syndrome, mild bibrachial atrophy ?
- Evidence of anterior epidural fluid collections

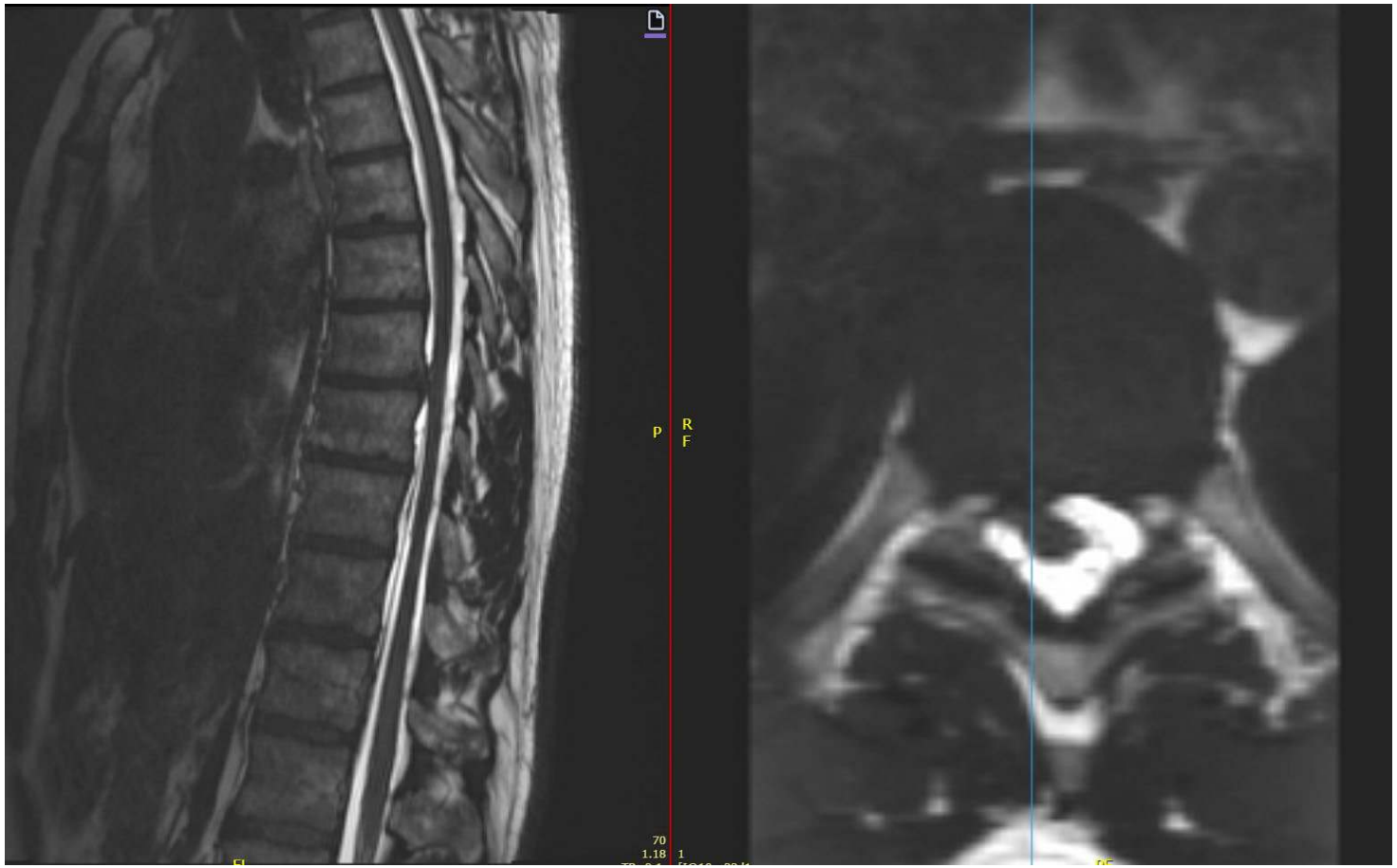
History

- 7 Y awkward sensations upper arms not painfull, related to sports?
- MRI cervical spine disc protrusion C5-6 managed conservatively
- 2 Y later weakness upper arms, mild (increased fatique), fasciculations? No explanation (EMG normal)
- Stable no progressive S & S for years
- 2 M decreased sensory sense left leg (warm/cold discrimination)
- Skin left leg felt different (hyperpathic)
- No walking difficulties or other complaints
- No interference with ADL (GI doctor!)

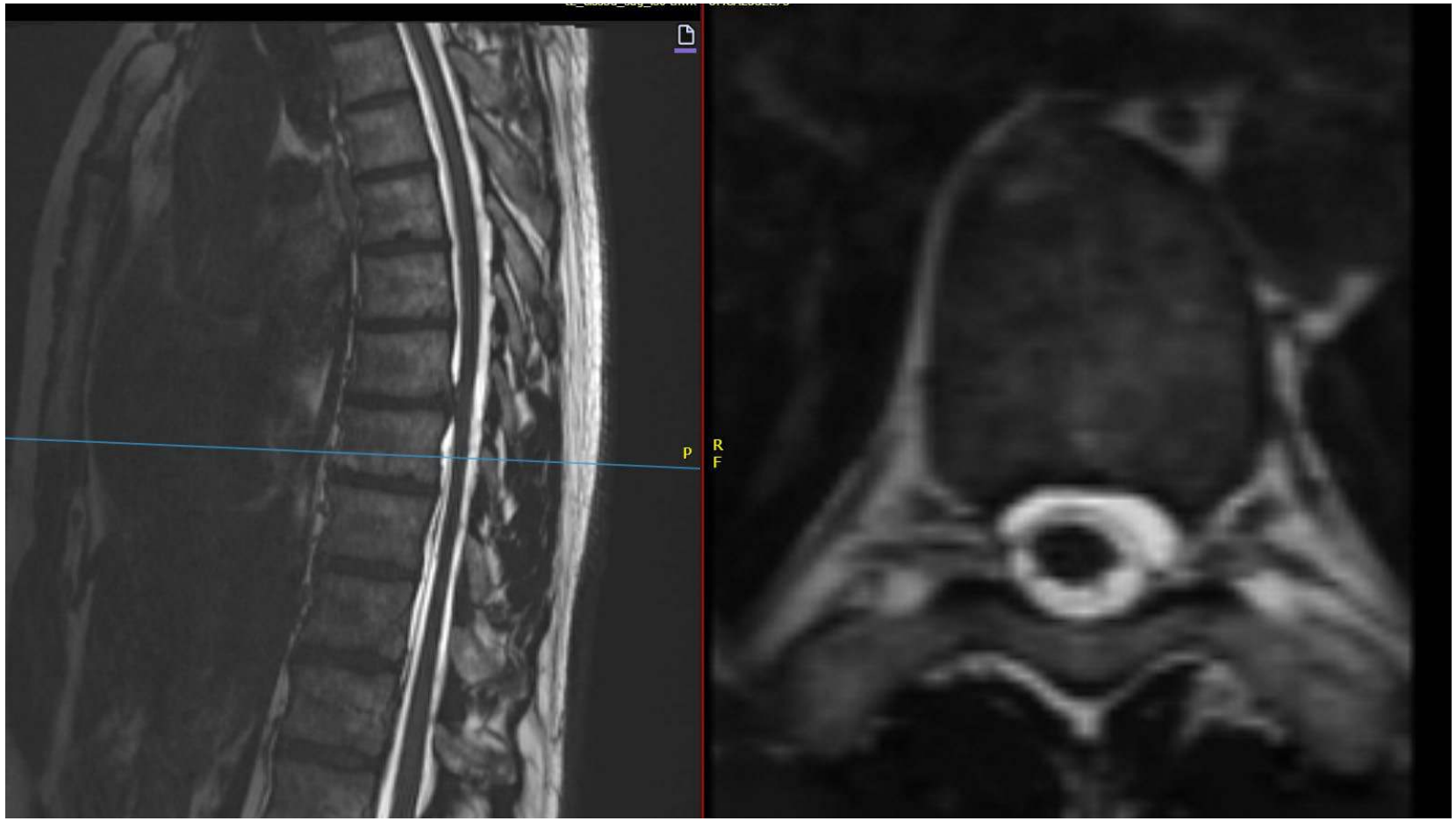
Neurological examination

- Right handed, normal consciousnesss orientation, articulation speech
- Cranial nerves no abnormalities
- Right sided atrophy of the deltoid and biceps and subtle also right quad
- Right sided weakness deltoid and biceps and triceps gr 4
- Normal tendon reflexes arms, brisk patellar reflexes, clonus achilles tendon, plantar reflexes downward
- Coordination no abnormalities
- Diminished sensory sense of the left leg

MRI Th 8-9 anteriorly displaced and herniated spinal cord



MRI epidural anterior epidural fluid collections C3-L1

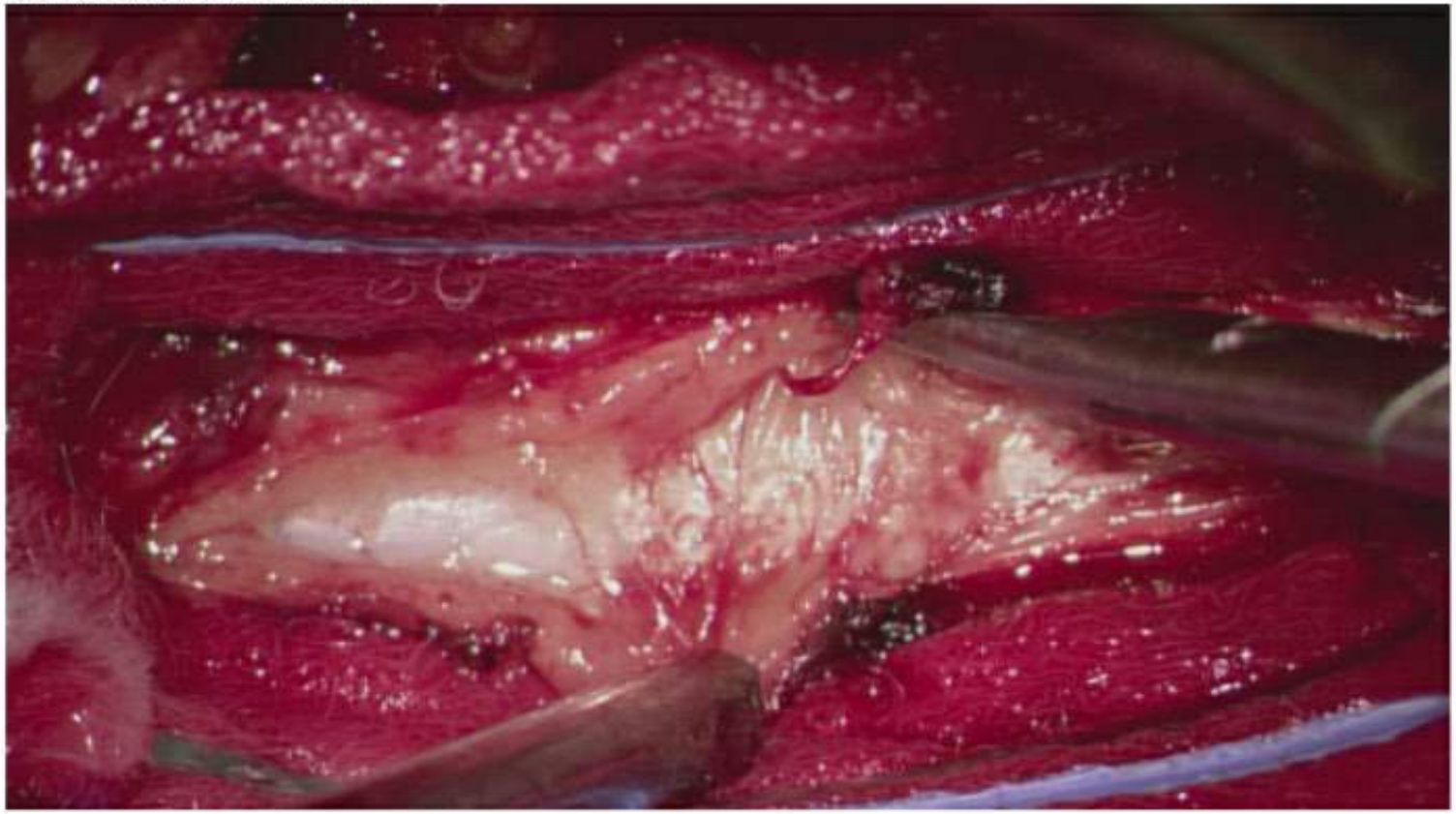


Surgery

- General anesthesia under IONM
- Posterior approach targeted laminectomy at Th8-9 level
- Cleavage and resection of indurated and adherent epidural fat.
- Posterior dural opening and mobilization of rotated spinal cord
- Identification of dural defect and reduction of the cord
- Closure of the anterior dura defect with 8-0 sutures and Tachosil

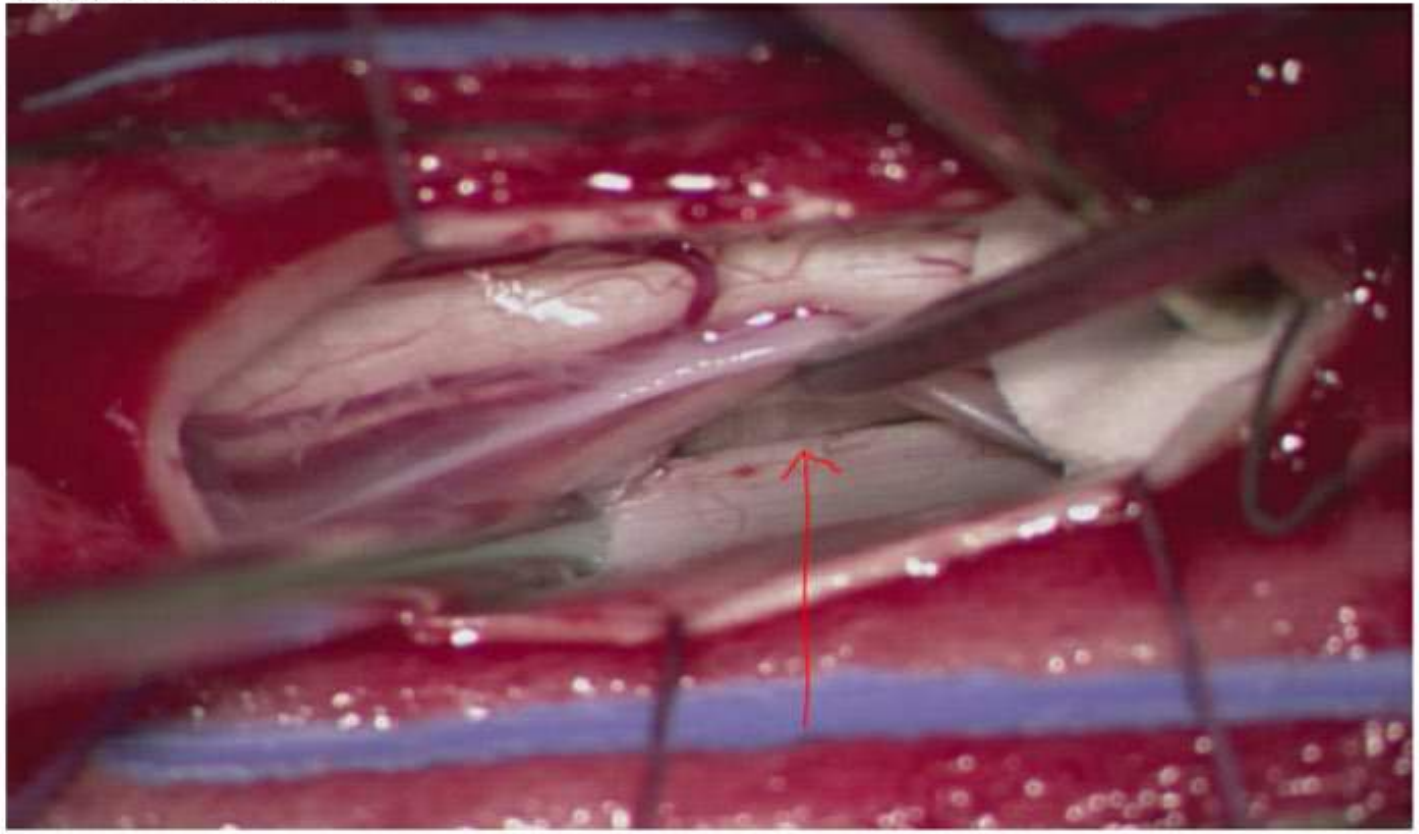
Indurated and sticky epidural fat

Geindureerde 'vetzwoerd':



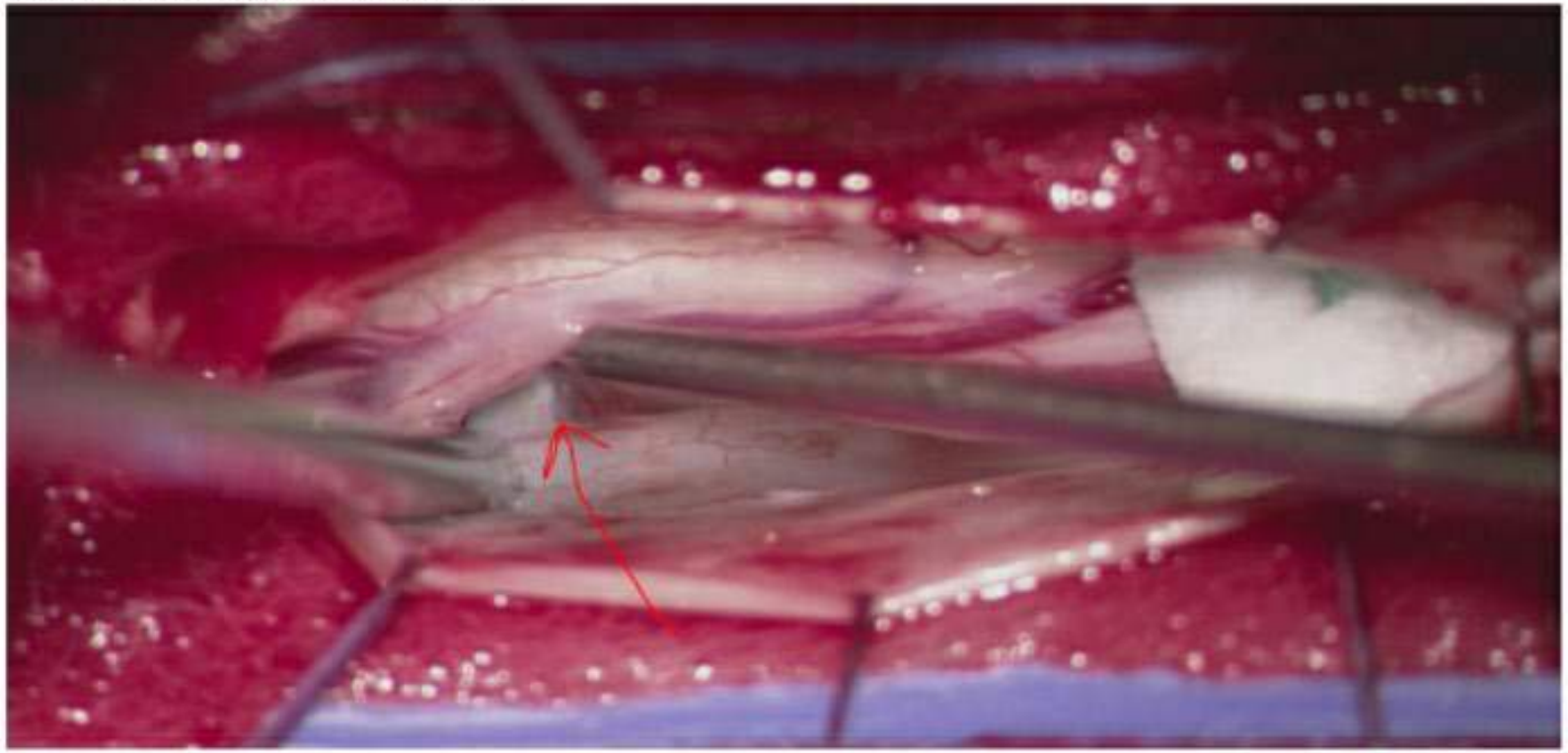
Rim of the anterior tear on the right side

Durascheur rechts:



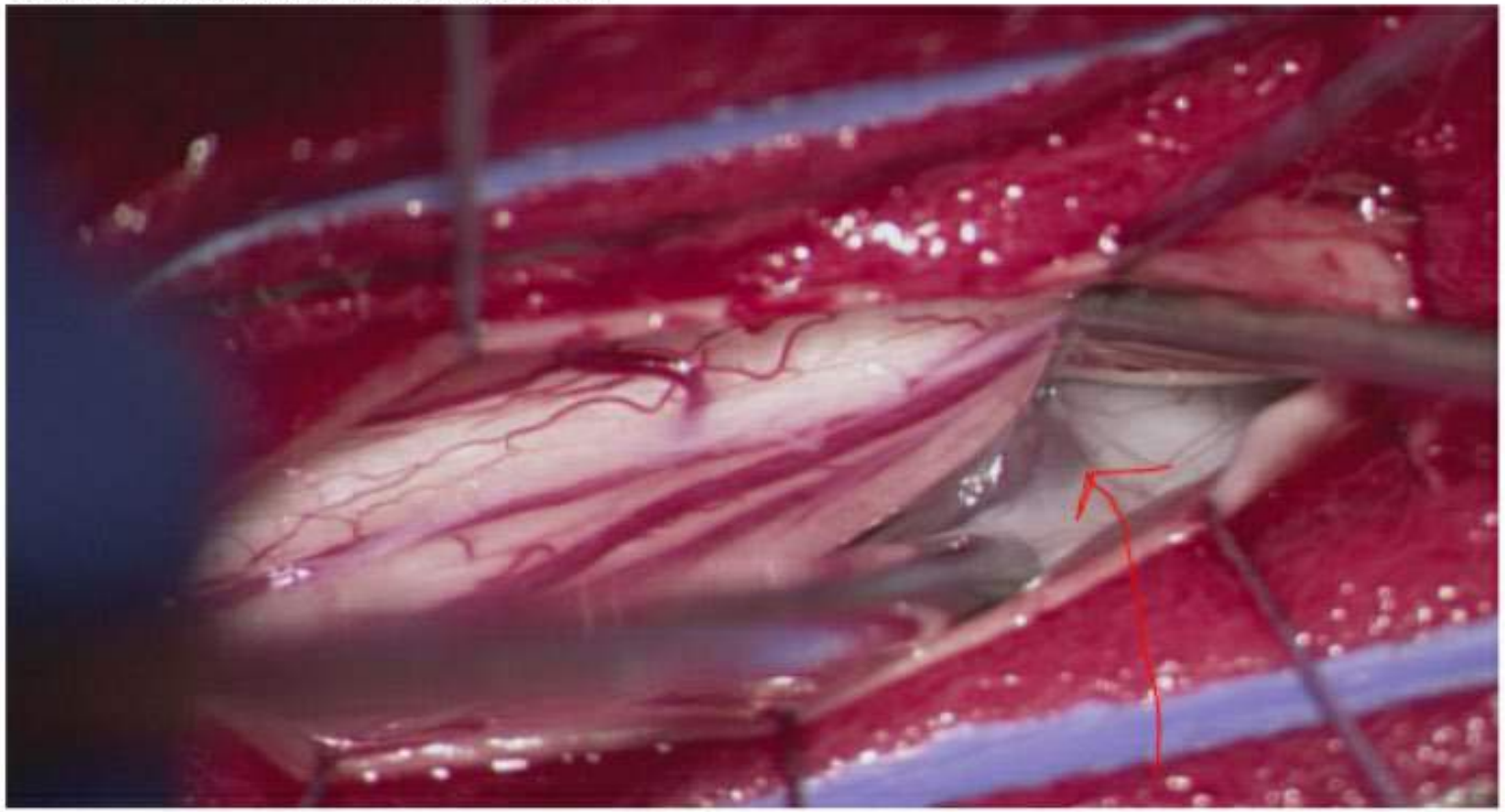
Caudal end of the tear

Caudale einde van de durascheur



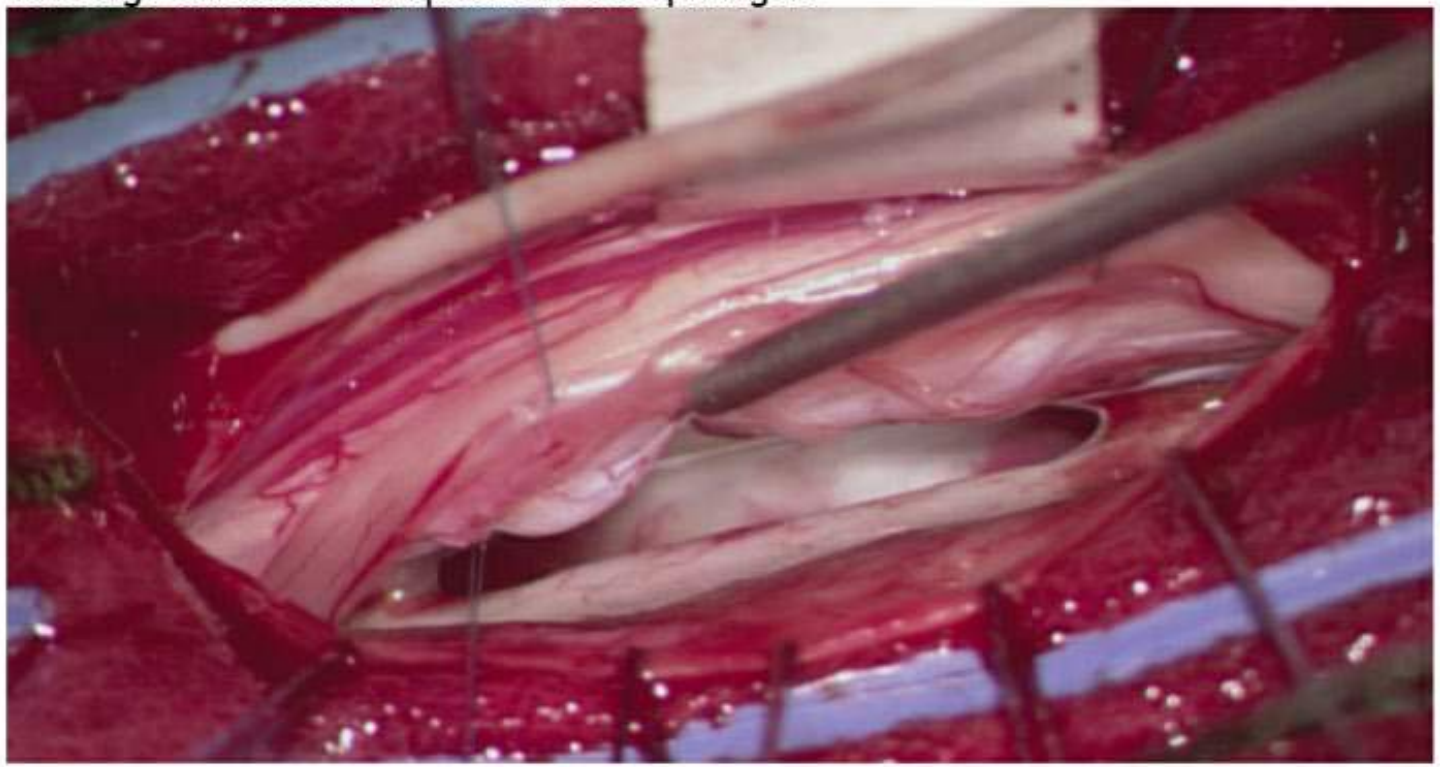
Cranial end of the tear

Craniale einde van de durascheur:



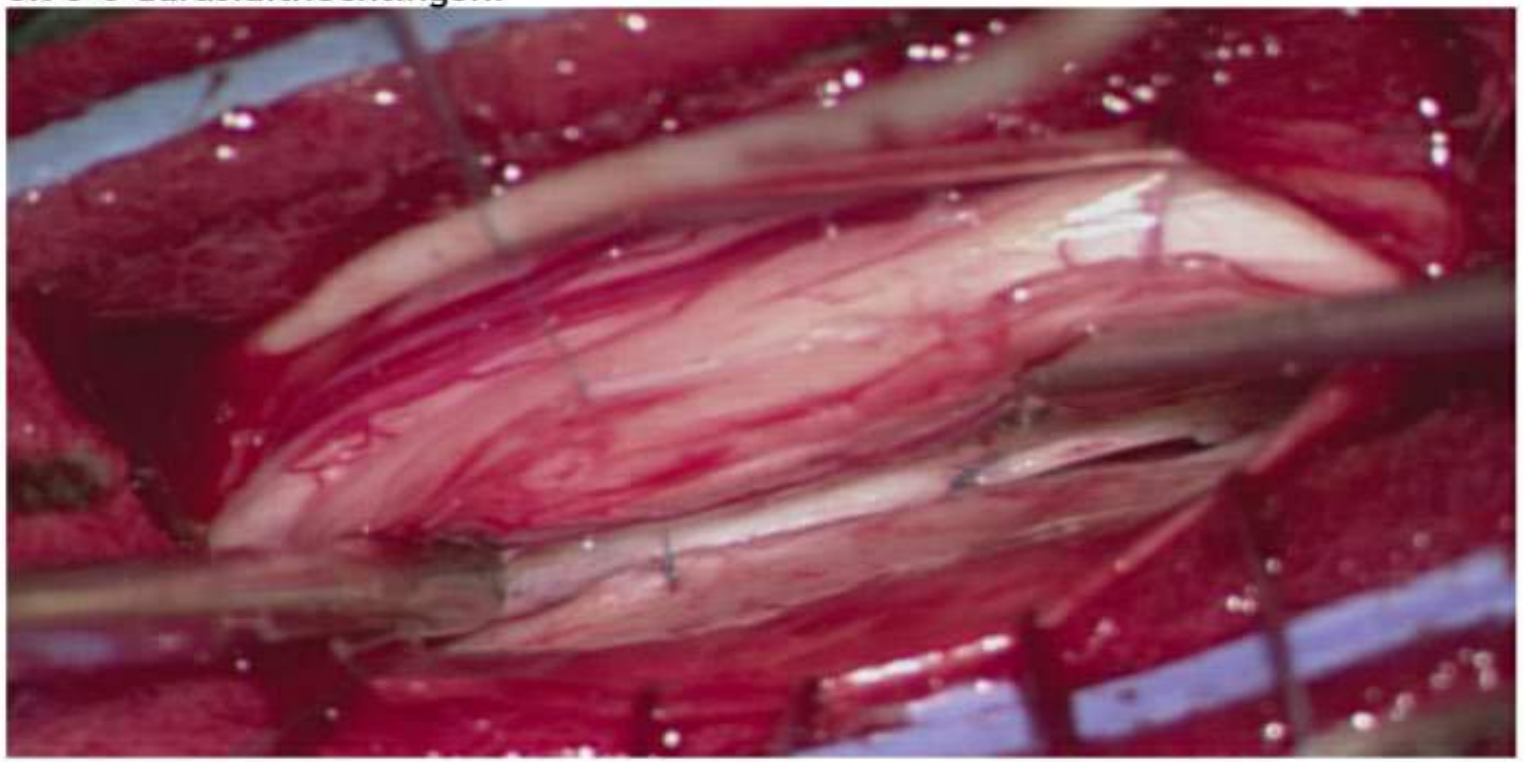
Tenting sutures denticulate ligament

Volledige durascheur na plaatsen 8-0 ophangers



Closure of the tear with sutures

3x 8-0 durasluithechtingen:



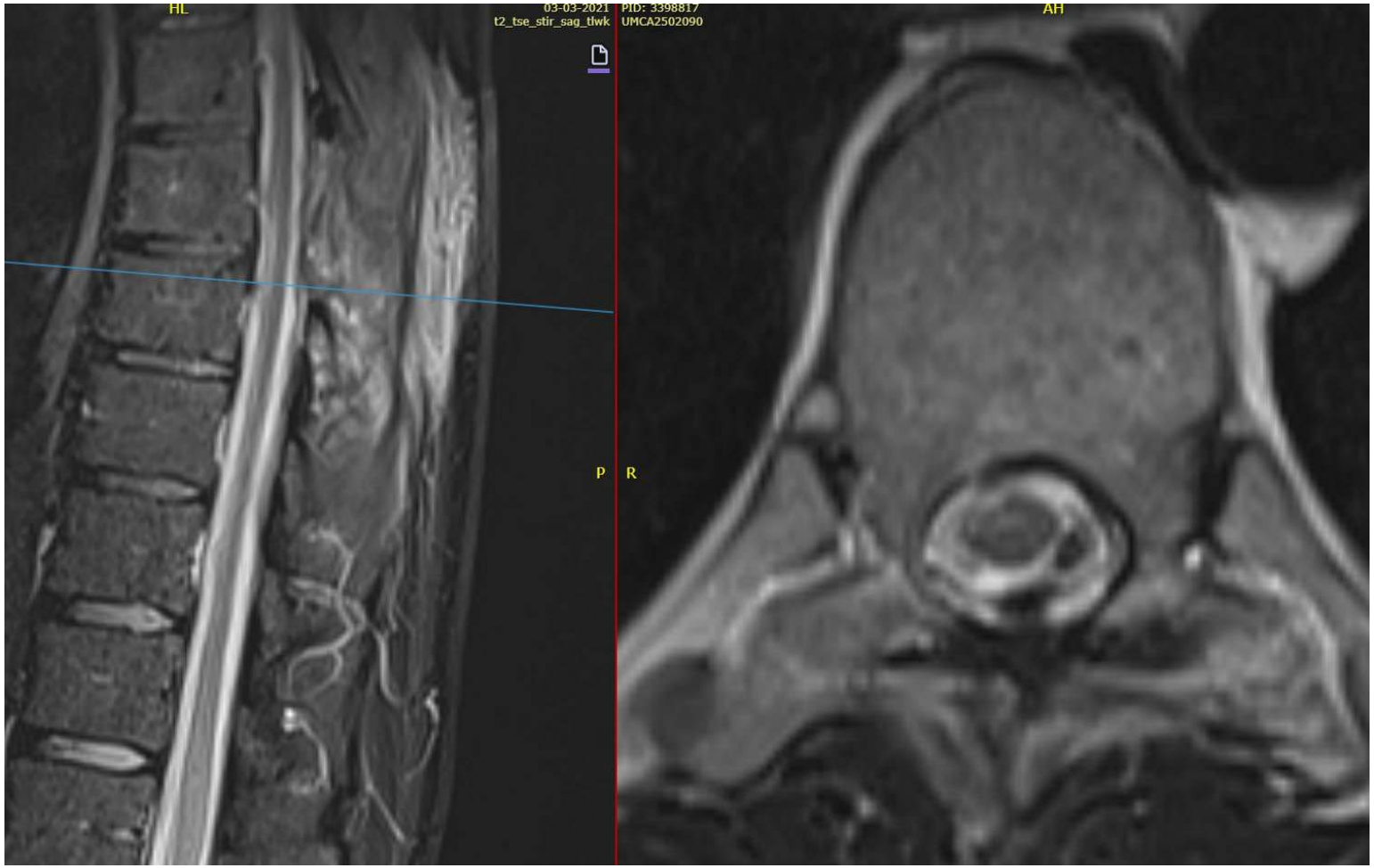
Postoperatively

- Some improvement in perceived power of the leg
- Otherwise no changes

Postop MRI:

- resolution of the extradural fluid collections
- no signs of herniation anymore
- small area of (persisting) myelopathy

Postoperative MRI



Intracranial hypotension and spinal cord herniation

- Common cause, ventral dural defect in thoracic spine
- However rarely seen together at debut
  - Both of course are quite rare
  - Mentioned in literature but no well described case reports
- Spinal cord herniation (probably) tamponades the defect
- IH symptoms may precede and resolve again before the S & S of spinal cord herniation (SCH) are noted

  (Insidious onset of SCH symptoms long after S & S of IH)

Conclusions:

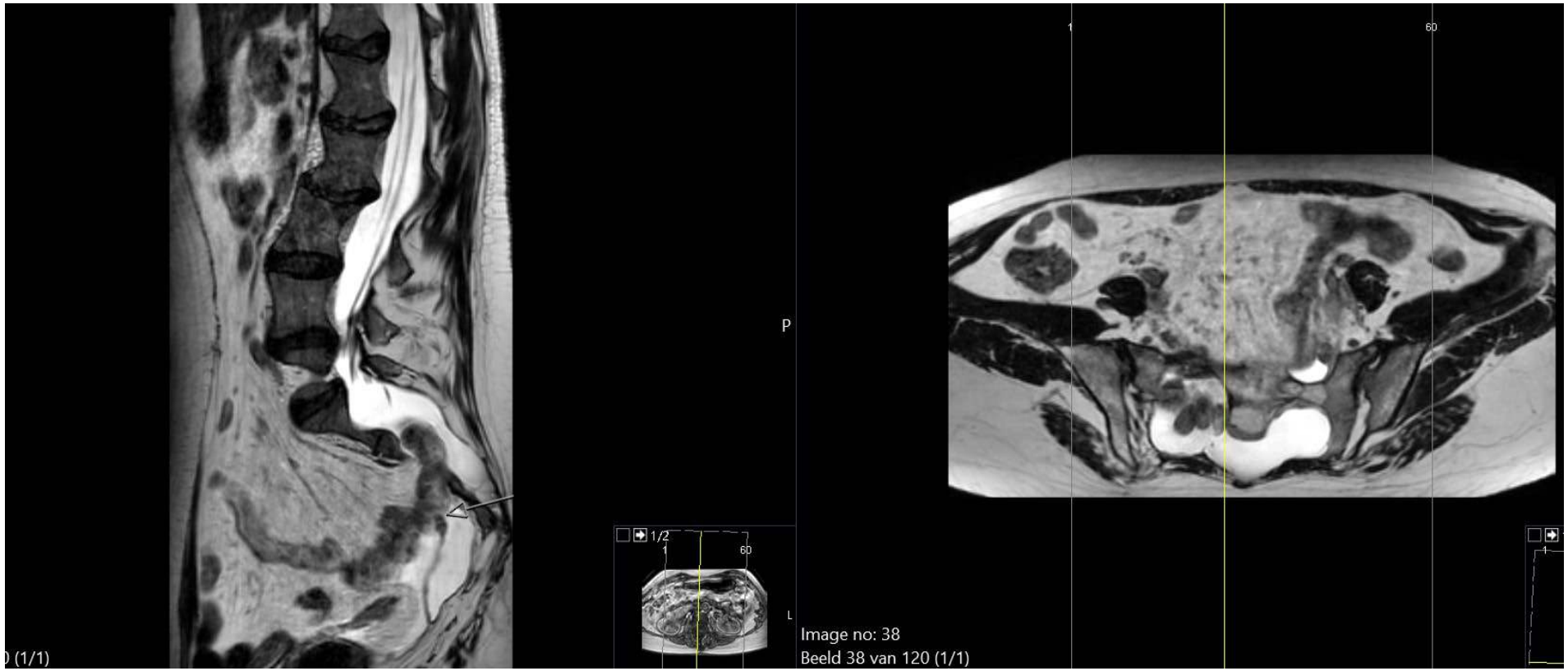
- IH and SCH are both rare entities and related by nature of the underlying pathology
- Combination of both at the same time at presentation time is however even more rare
- Improved awareness and better imaging possibilities (MRI) will lead to more cases

Conclusions:

Surgery can often halt deterioration and/or improve outcome

- Correct localizing diagnosis necessary for reduction the herniated cord/closure of the dural defect:
  - -Primary closure stitches
  - -Dural patches or slings (stay sutures or fibrin glue)
  - -Obliterating the defect with dural substitute, muscle, fat and glue
  - - Combinations
- Complications:
  - Recurrence
  - Arachnopathy with (severe) tethering and syringomyelia

“Spinal” herniation and intracranial hypotension syndrome



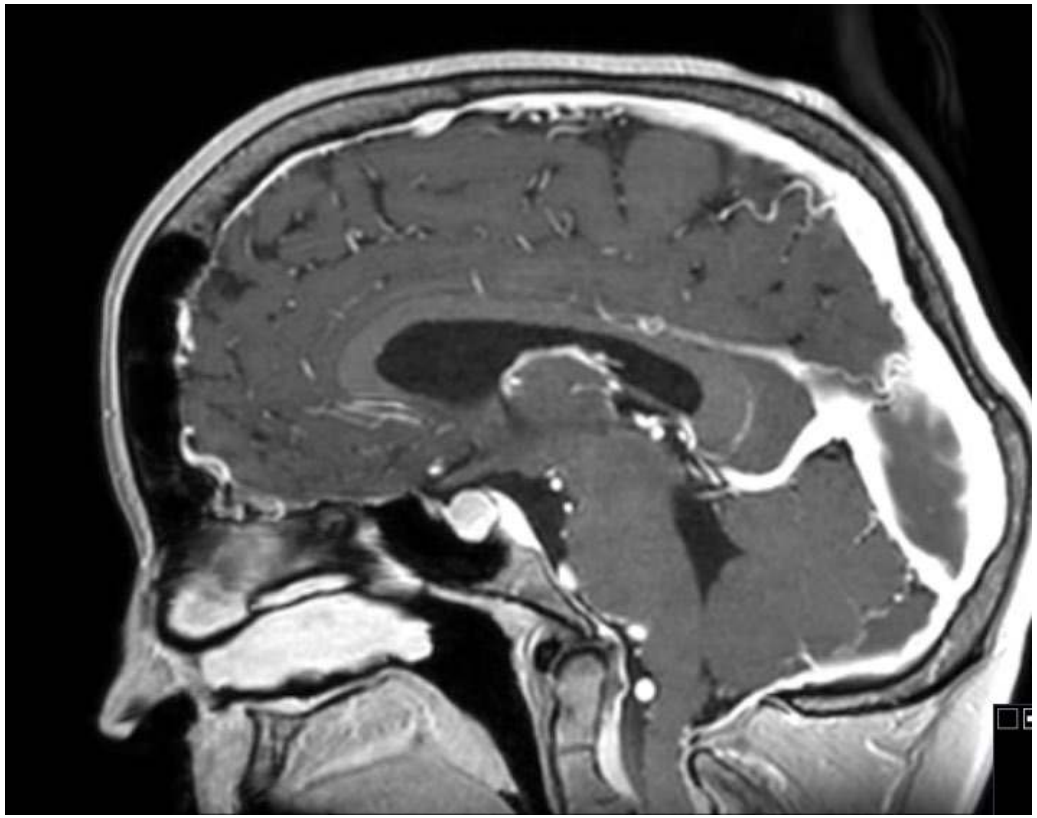
Female 25 y old recovery nurse with Loeys Dietz syndrome

Marfan like genetic disorder of connective tissue impacting multiple organ systems including the heart, blood vessels, bones, joints, skin, and internal organs. It is characterized by a higher risk of aortic and other arterial aneurysms.

June 4th laparoscopic fenestration/biopsy of “ovarian cyst

June 8th re-admitted with severe positional headache, muffled hearing, dizziness , nausea

“Spontaneous” intracranial hypotension



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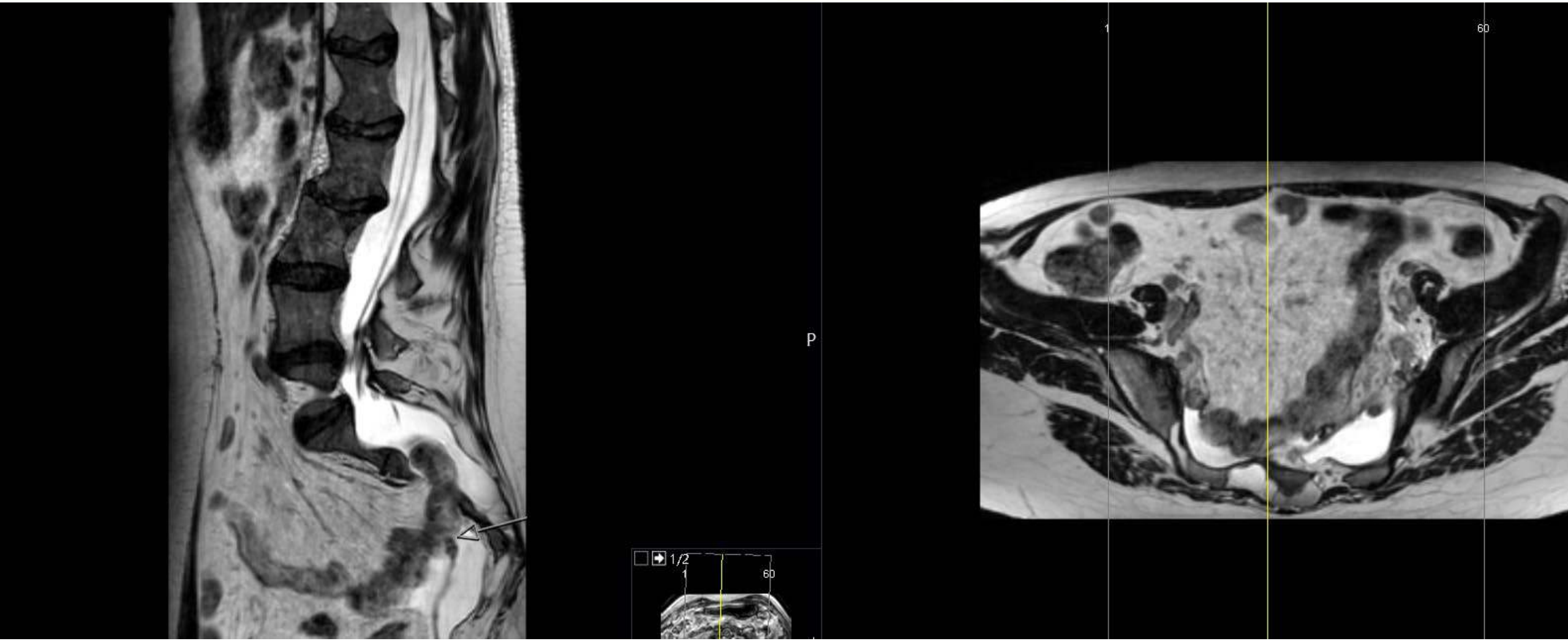

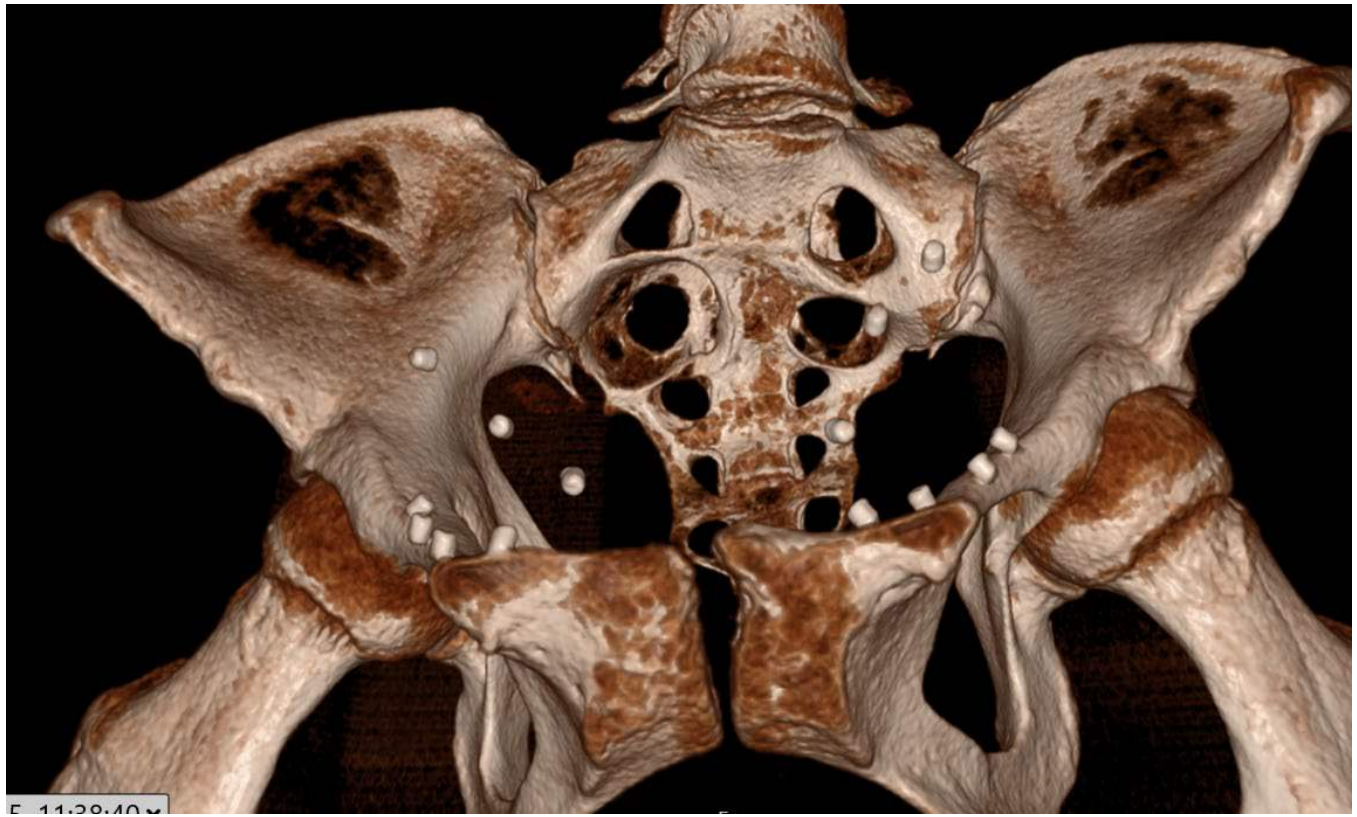

“Ovarian cyst” = large cystic herniation of dural ectasia through right S2 foramen

June 18th combined surgery :

- laparotomy
- removal of the herniated intestine from the sacrum
- coverage of the S2 foramen with artifical dura and tachosil, fibrin glue and pedicled vertical rectus abdominus muscle flap (VRAM)
- prolonged horizontal bedrest