Department of Diagnostic Radiology
Kingston Health Sciences Centre,
Kingston Ontario, Canada

Rare Case of a Bronchial Endodermal Cyst of the Cauda Equina:

Literature Review & Strategic Imaging Approach

Lauren Mak MSc (MD/PhD Student); Sana Basseri MD, PhD; Christopher Wallace MD, John Rossiter MD, PhD; Omar Islam MD; Ian Silver MD; Benjamin Kwan MD and Donatella Tampieri MD

**Canadian Association of Radiology
Annual Scientific Meeting
April 2019**

Conflict of Interest

The authors report no conflict of interests.

Learning Objectives

1. Review the differential diagnosis of lumbar intradural expansile lesions;

2. Establish a differential diagnostic approach based on location and imaging patterns;

3. Discuss neurenteric cysts of the spine;

4. Present a rare case of a bronchial endodermal cyst of the caudal equina.

Learning Objective #1:

Review the differential diagnosis of lumbar intradural expansile lesions.

Differential Diagnosis

The differential diagnosis of lumbar intradural expansile lesions is vast.

At the lower thoracic and lumbar level the lesions may arise from the cord and filum terminalis, therefore they are considered intramedullary, often with an exophytic component.

If the lesions arise from the nerve roots, the dura or the vascular structures they are considered intradural extramedullary.

Therefore the best strategic approach is to locate the site of origin of the abnormality present in the dural tube.

To follow a summary of the most frequent acquired pathologies we should consider at the time of the assessment of the images.

Differential Diagnosis

Intradural Intra-Medullary Lesions

- **Glial Neoplasms:**
  - Ependymoma, Astrocytoma, Ganglioglioma;
- **Non-Glial Neoplasms and Secondary Neoplasms:**
  - Hemangioblastoma, Paraganglioma, Neurenteric cyst, Metastases and Lymphoma;
- **Inflammatory Processes:**
  - MS, NMO, Sarcoidosis;
- **Infections;**
- **Metabolic Conditions:**
  - Vitamin B12 Deficiency.

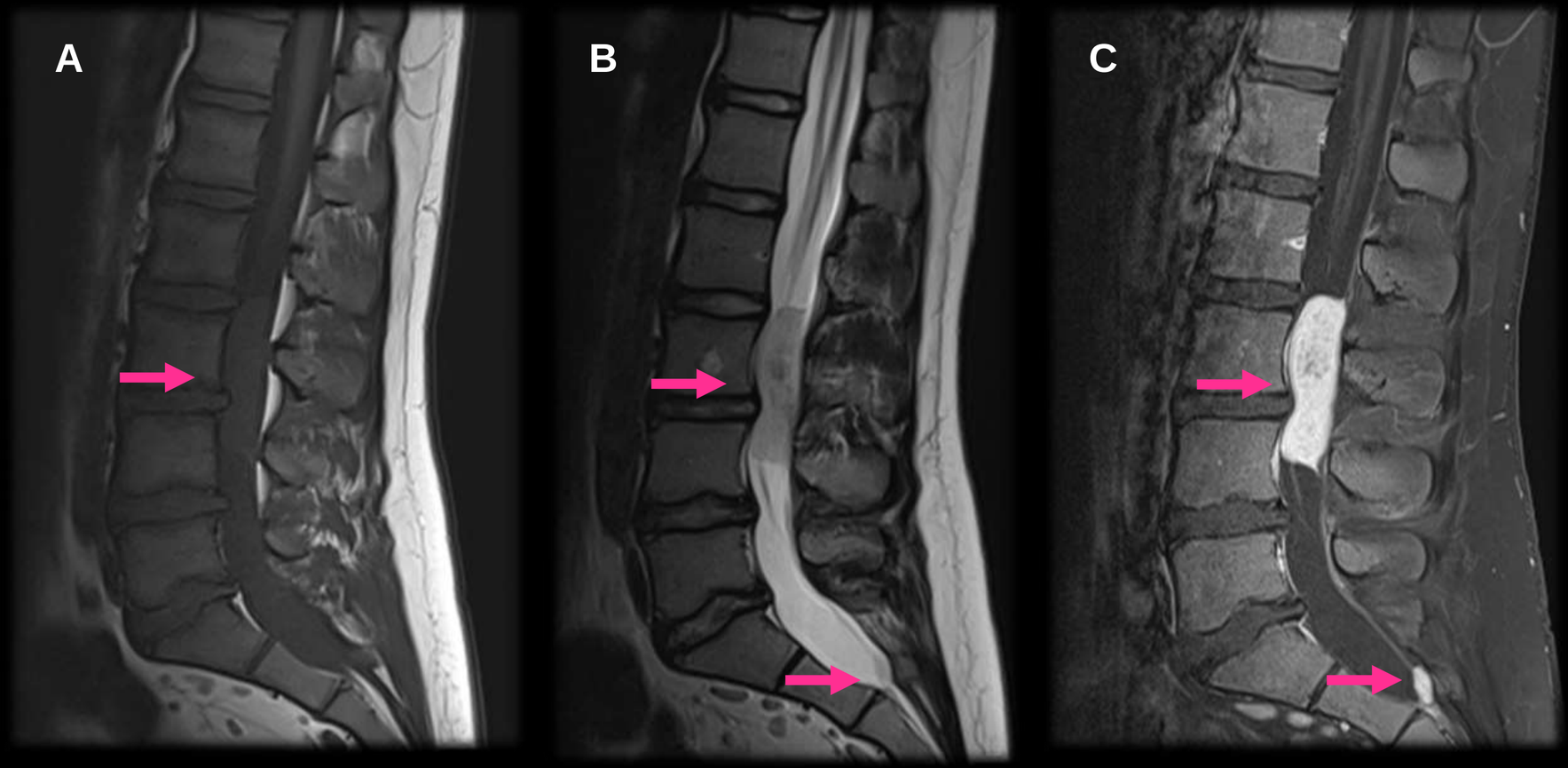
Intradural Extra-Medullary Lesions

- **Primary Neoplasms:**
  - Benign Schwannoma, Meningioma, Myxopapillary Ependymoma in conus, Epidermoid;
- **Secondary Neoplasms:**
  - Metastases, Lymphoma;
- **Inflammatory Processes:**
  - CIDP;
- **Vascular Malformations.**

Learning Objective #2:

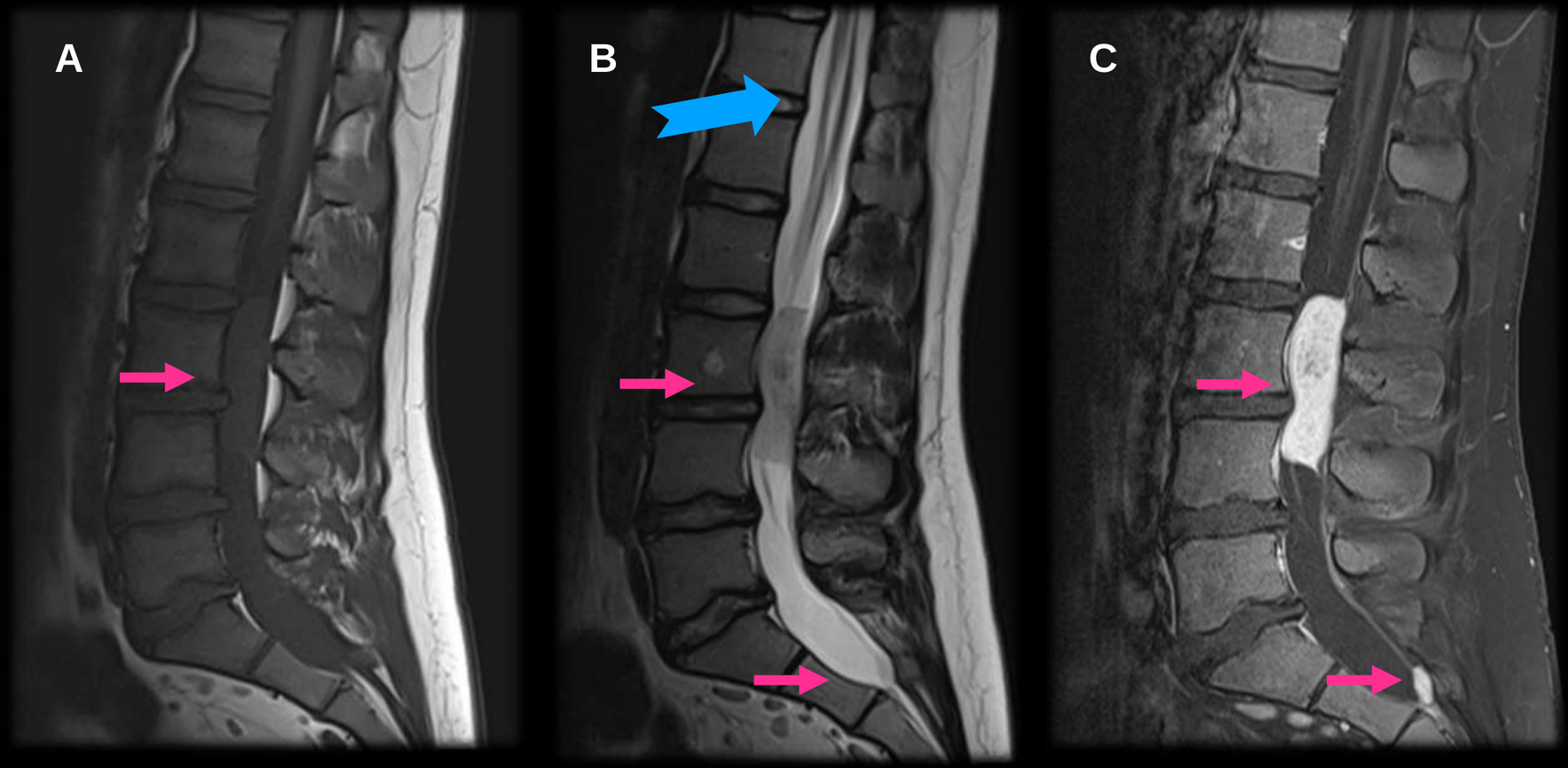
Establish a differential diagnostic approach based on location and imaging patterns.

Intradural lesion located at the L3-L4 level on the midline, isointense in T1 (A) and T2 (B), avidly enhancing (C). Satellite lesion at S2 at the attachment of the filum terminalis. Spinal cord syrinx.



Intradural lesion located at the L3-L4 level on the midline, isointense in T1 (A) and T2 (B), avidly enhancing (C). Satellite lesion at S2 at the attachment of the filum terminalis. Spinal cord syrinx( ).

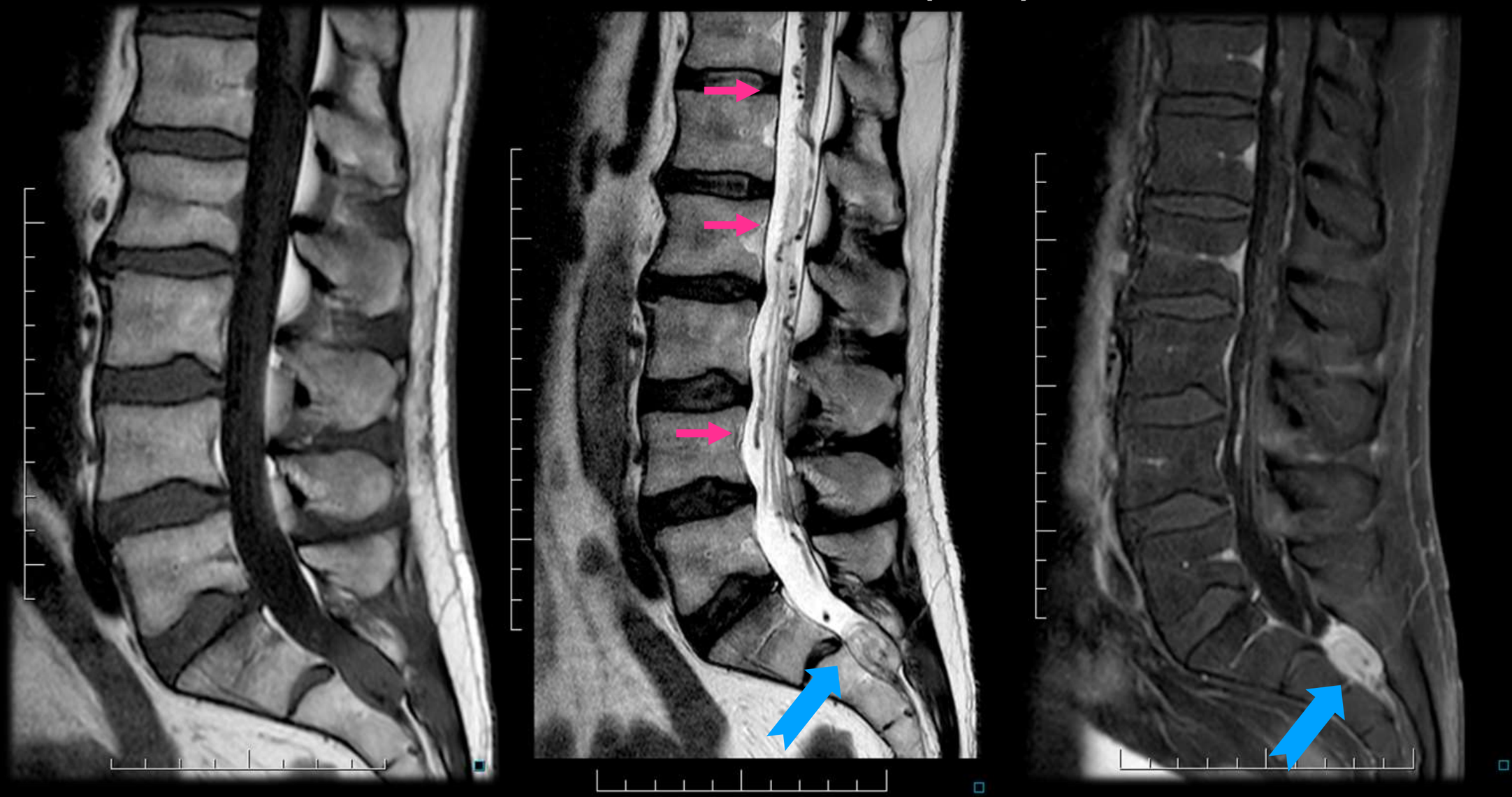
Myxopapillary Ependymoma of the Filum



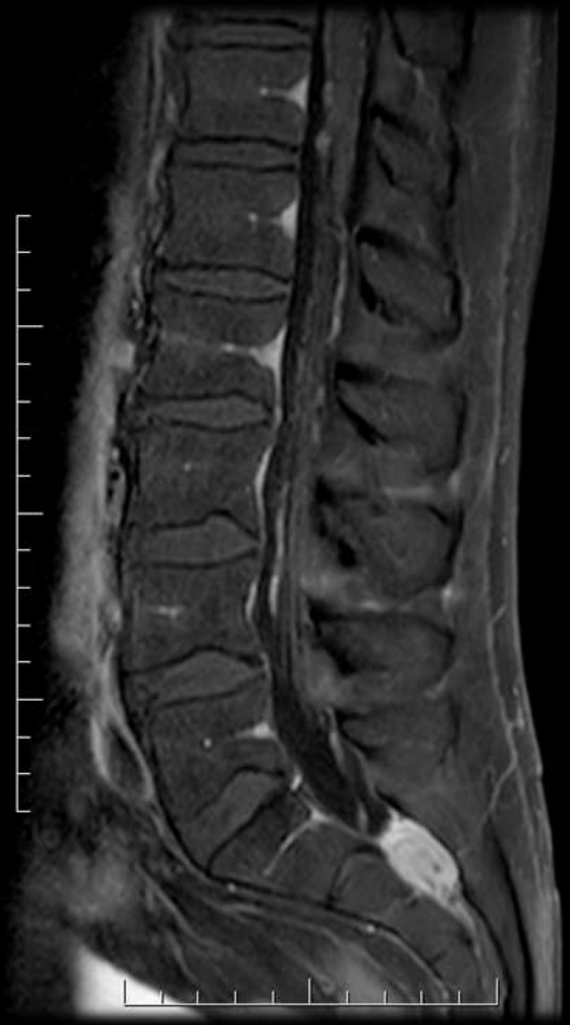
Myxopapillary Ependymoma

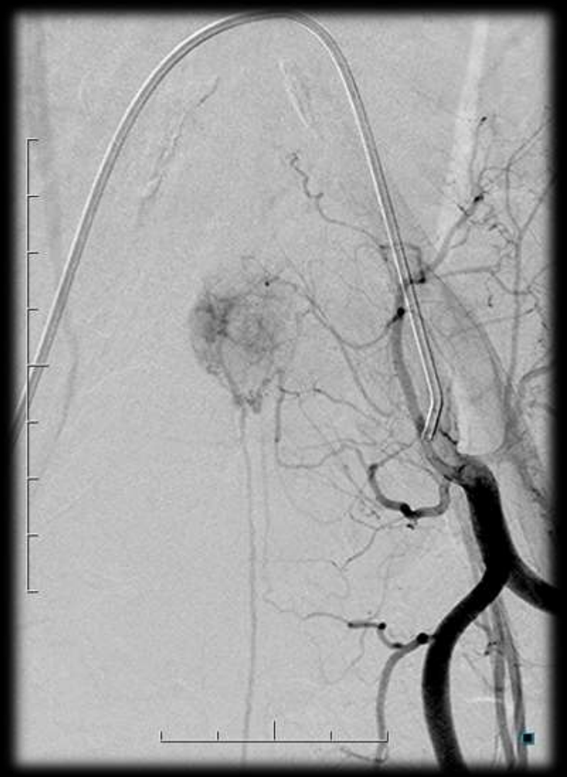
- Ependymoma is the most common intramedullary neoplasm of the adult (60% of glial cord tumors).
- Peak incidence in the 4th decade affecting males more frequently.
- Clinical Presentation: sensory symptoms due to their central location close to the spino-thalamic tracts.
- Pathology: 6 histological types: cellular, papillary, clear cell, tanycytic, melanotic and myxopapillary (filum terminalis).
- Myxopapillary is WHO type I.
- Location: myxopapillary ependymoma is located in the conus or filum terminalis.
- Imaging : well circumscribed lesions usually isointense in T1 and T2 WI , occasionally hyperintensities due to mucin content and hemorrhage can be noted. Often hemosiderin deposition and superficial siderosis is noted, always enhancing after contrast administration.

Intradural lesion isointense in T1 and T2WI located at S2 (➡) at the attachment of the filum terminalis, associated to multiple serpentine vascular structures (→).

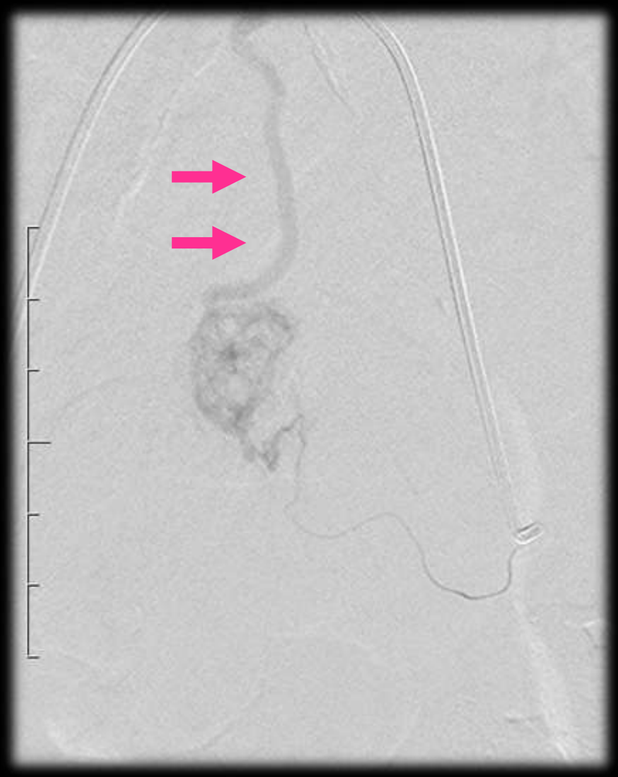


Hemangioblastoma of the Filum Terminalis: these tumours have rich vascularization with large ectatic venous drainage (→)(A-B). The lesion was embolized (C).

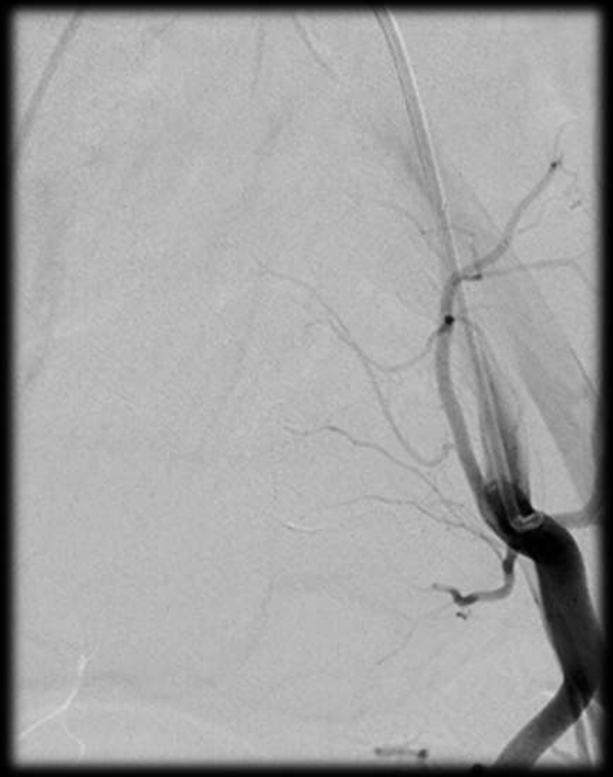




A



B

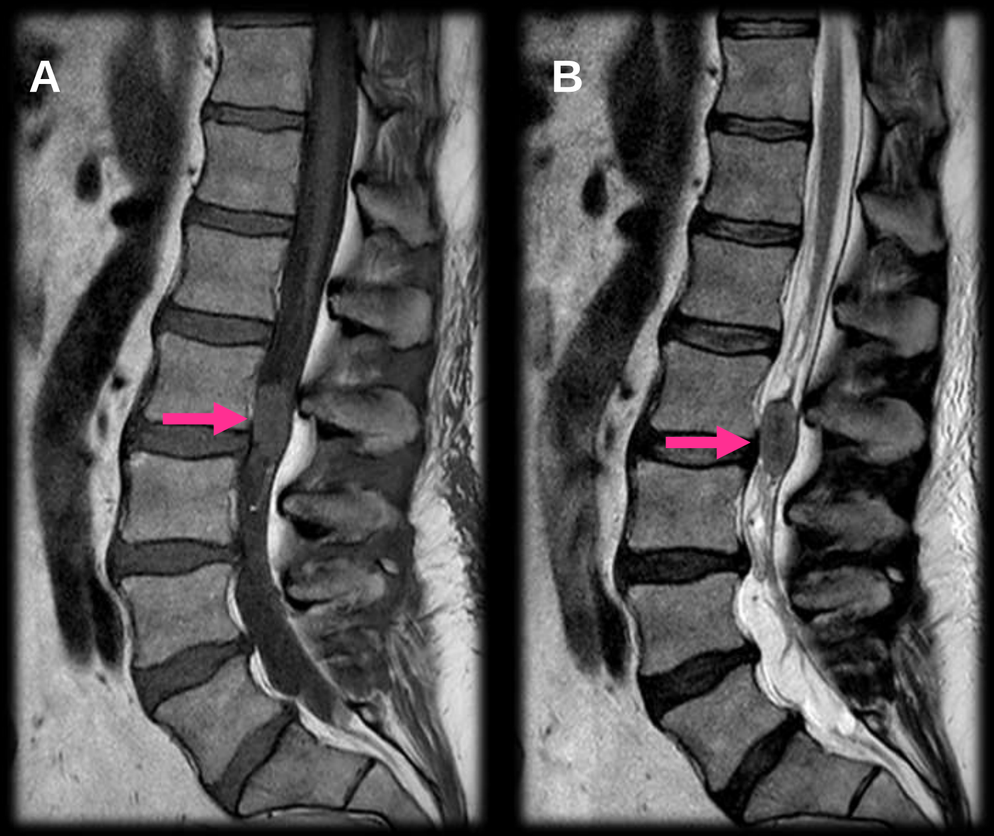


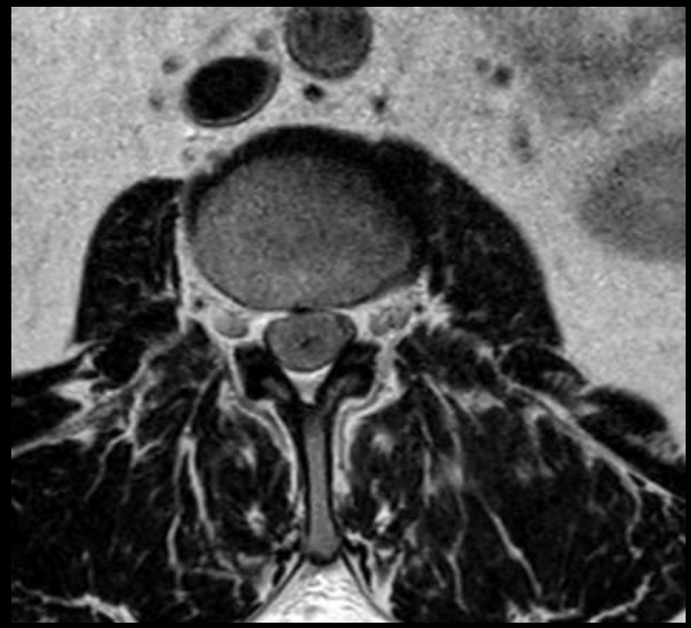
C

Hemangioblastoma

- Represents 2-6% of all intramedullary tumors;
- Sporadic H. have a peak incidence at the 4th decade affecting male and female equally;
- Clinical Presentation: pain, weakness and sensory changes
- Most commonly located in the thoracic cord, they are usually eccentric and exophytic;
- WHO grade I;
- If multiple Von Hippel Lindau syndrome should be suspected;
- Pathology: benign vascular lesions consisting of large pale stromal cells between packed blood vessels.
- Imaging: avidly enhancing tumour, occasionally associated to cyst or syringomyelia, with visible serpentine flow voids (venous drainage).
  - T1: hypo- to isointense.
  - T1 C+ (Gd): enhance vividly.
  - T2: iso- to hyperintense.[4]

Intradural lesion arising from the Filum Terminalis, avidly enhancing after contrast (not available) . No ectatic or abnormal vascular structures.
(A) T1W hypointense; (B) T2W hyperintense.

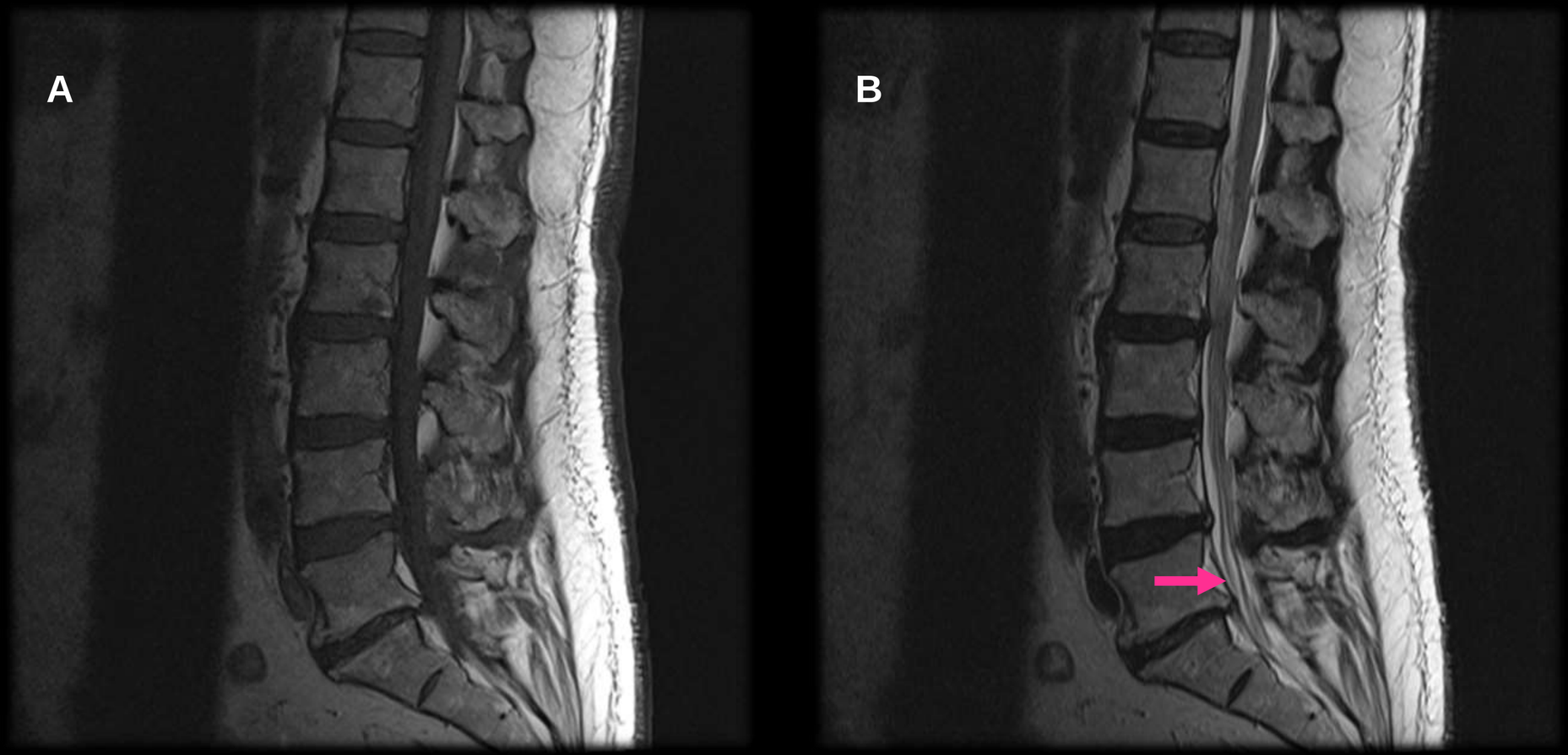




Paraganglioma

- Most cases present in middle age (30-60 years) with males somewhat more affected than females.
- Clinical Presentation: local mass effects (lower back pain and sciatica) or neuroendocrine symptoms.
- Pathology: Indolent and considered WHO grade I. The main components are lobules or nests of chief cells (type I); these structures are known as Zellballen. They are surrounded by a single layer of sustentacular cells (type II).
- Imaging: well-circumscribed masses, inferior to the conus.
  - T1: iso- to hypointense.
  - T1 C+(Gd): intense enhancement.
  - T2: hyperintense with flow voids typically seen along the surface & within.[5,6]

Focal cyst-like lesion isointense to CSF in all sequences in patient with tethered cord. No enhancement following contrast administration

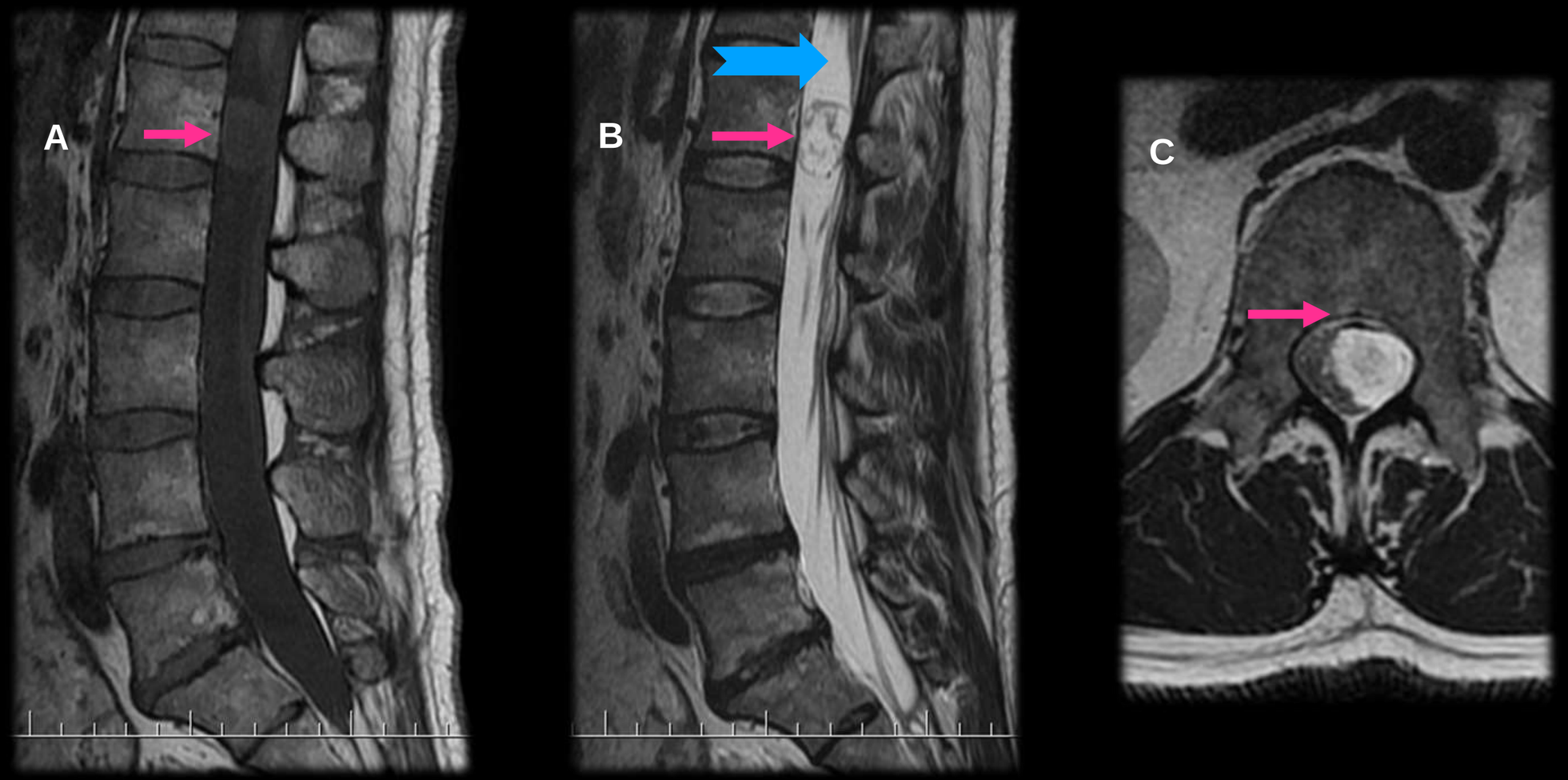


Dilatation of the 5th Ventricle

- An ependymal-lined fusiform dilatation of the terminal central canal of the spinal cord, positioned at the transition from the tip of the conus medullaris to the origin of the filum terminalis (Ventricle of Krause).

- Imaging: appearance becomes increasingly unidentifiable with age.
  - Resembles CSF in signal and density on all sequences.
  - Non-enhancing cystic lesion.[7-9]

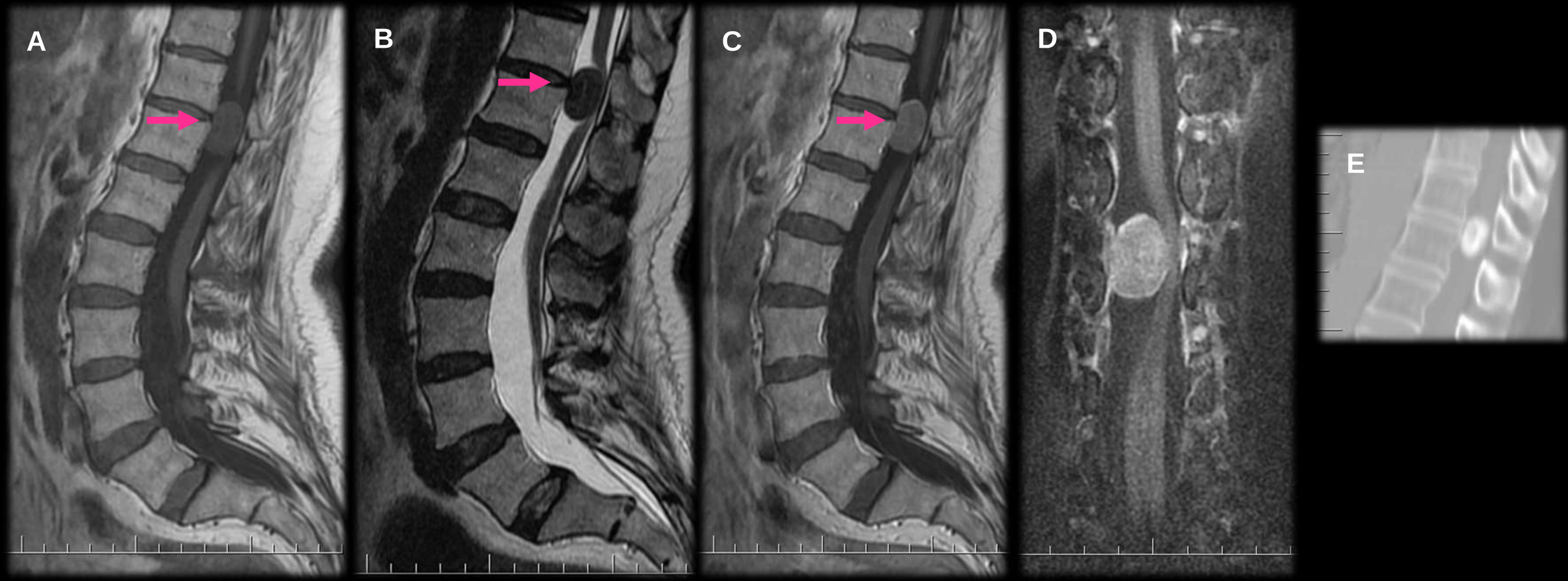
Intradural left-sided lesion, isointense in T1 (A), heterogeneous in T2 (B) and avidly enhancing (C) – displacing the filum terminalis posteriorly ( )



Schwannoma

- Schwannomas and neurofibromas account for 25% of intradural tumors.
- 2.5% of these tumours are malignant, half of them occur in patients with NF.
- Peak incidence in 5th-7th decades with no significant sex predilection.
- Presentation: pain, radicular sensory changes, myelopathy if lesion is large.
- Imaging: often indistinguishable from neurofibromas. Solid, well-defined, rounded lesions and often with associated adjacent bony remodelling.
  - T1: 75% isointense, 25% hypointense.
  - T1 C+(Gd): 100% enhancement.
  - T2: 95% hyperintense, often with heterogenous signal.[10-12]

Intradural extra-medullary lesion at the T11-T12 level in patient with tethered cord. Very hypointense in T2 (B), mild enhancement (C). Calcified, thus the T2 hypointensity.



Spinal Meningioma

- Spinal meningiomas represent around 12% of all meningiomas;
- They have a peak incidence in the 6th-8th decade and account for 25-46% of spinal tumours.
- Risk factors include high dose of ionizing radiation especially in childhood and prior trauma. Most frequently in women.
- Presentation: they are usually small but despite this can result in significant neurologic dysfunction of motor deficits due to spinal cord compression.
- Pathology: 70-90% classified as WHO grade I; they are histomorphic variants of meningiomas.
- Imaging: mostly commonly found in the thoracic spine and often lateral to the spinal cord. They are well-circumscribed with a broad based dural attachment.
  - T1: isointense to hypointense, possibly heterogeneous.
  - T1 C+(Gd): moderate homogeneous enhancement.
  - T2: iso- to hypointense. [13,14]

Learning Objective #3:

Discuss neurenteric cysts of the spine (imaging and pathology)

Case Presentation

- 48-year old female presenting with a 2-week history of lumbar radiculopathy, low back pain and right leg numbness. No reported symptoms of bladder or bowel dysfunction.

- On exam patient had legs weakness.

Neurenteric Cyst

- Rare intra-medullary spinal lesions 0.7-1.3%;
- Typically presents in the 2nd-3rd decade of life with a 2:1 male predominance.
- Usually occur in the lower cervical and upper thoracic spine.
- Presentation: present with progressive focal pain at the level of the spinal axis pathology, fluctuating myelopathic and radicular symptoms.
- Pathology: Hypothesized to be formed by displaced elements of the gastrointestinal or respiratory tract during embryogenesis**.** Bronchogenic are endodermal cysts lined by respiratory-type epithelium.
- Imaging: appearance tends to be variable and is dependent on protein content.
  - T1: iso- to hypointense.
  - T1 C+(Gd): usually non-enhancing.
  - T2: iso- to hyperintense.
  - DW: no restriction of water.[18,19]

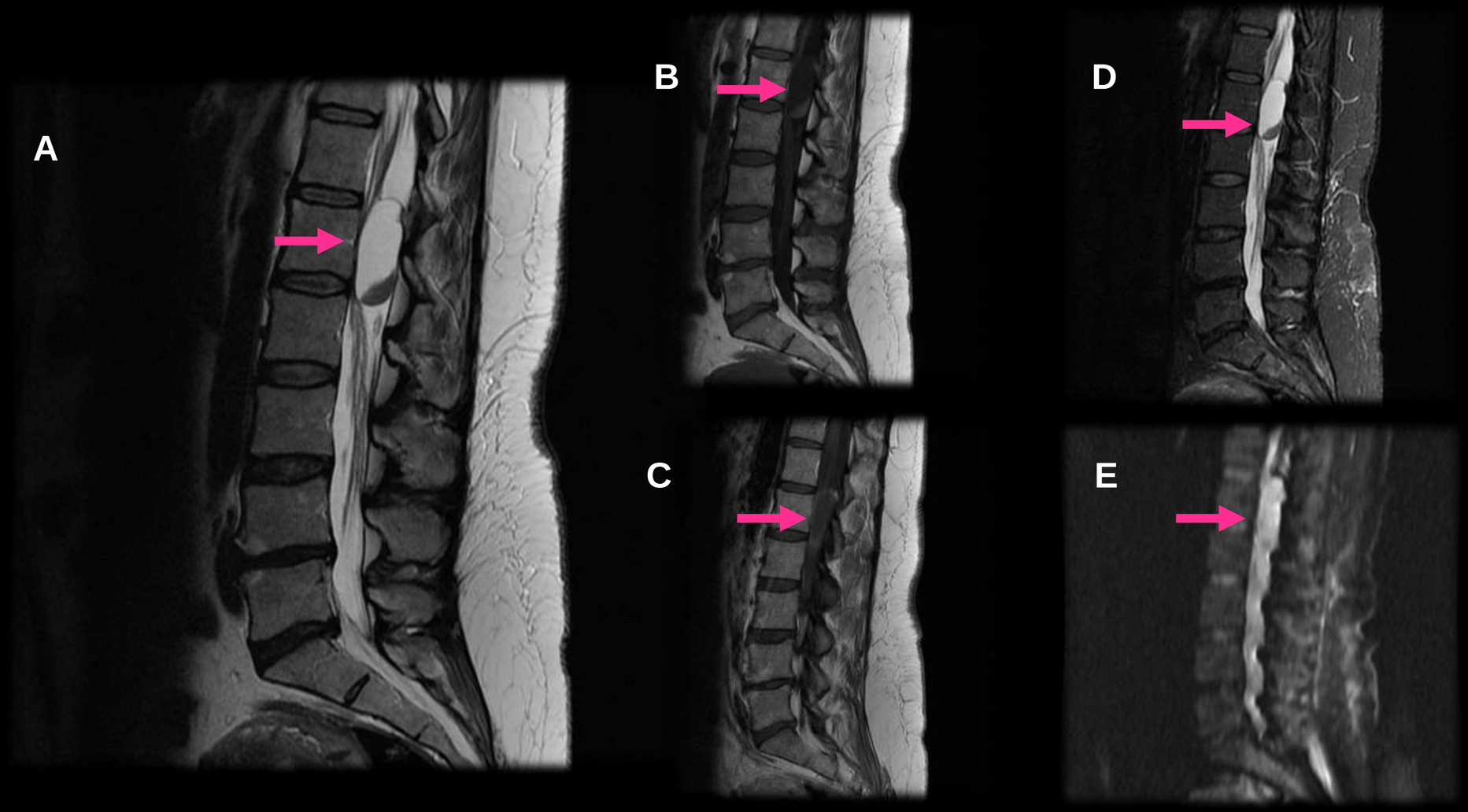
Neurenteric Cyst

Characteristics[20]	Type A	Type B	Type C
Single layer of pseudostratified columnar or cuboidal cells mimicking respiratory of gastrointestinal epithelium	+	+	+
Complex invaginations with glandular organization; mucinous or serous production; nerve ganglion, lymphoid, skeletal muscle, smooth muscle, fat, cartilage, and/or bone elements	-	+	-
Ependymal or glial tissue	-	-	+

Learning Objective #4:

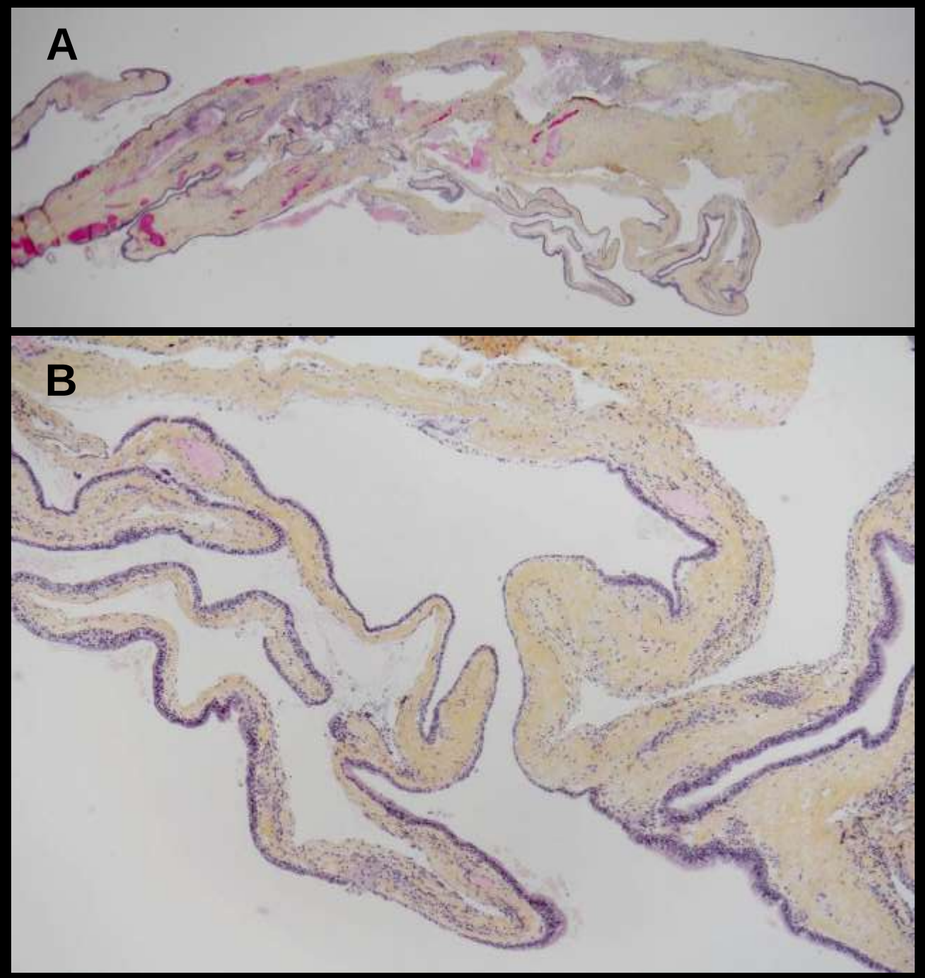
Present a rare case of a bronchial endodermal cyst of the caudal equina

(A) T2W: hyperintense, (B) T1W: hypointense, (C) T1W-post GD: no enhancement, (D) STIR sequence, (E) DW: no water restriction. Intradural cystic lesion measuring 1.6 x 2.1 x 4.1 cm (AP x TR x CC) dorsal to the L1 spinal cord, with a soft tissue appearance in the posteroinferior aspect



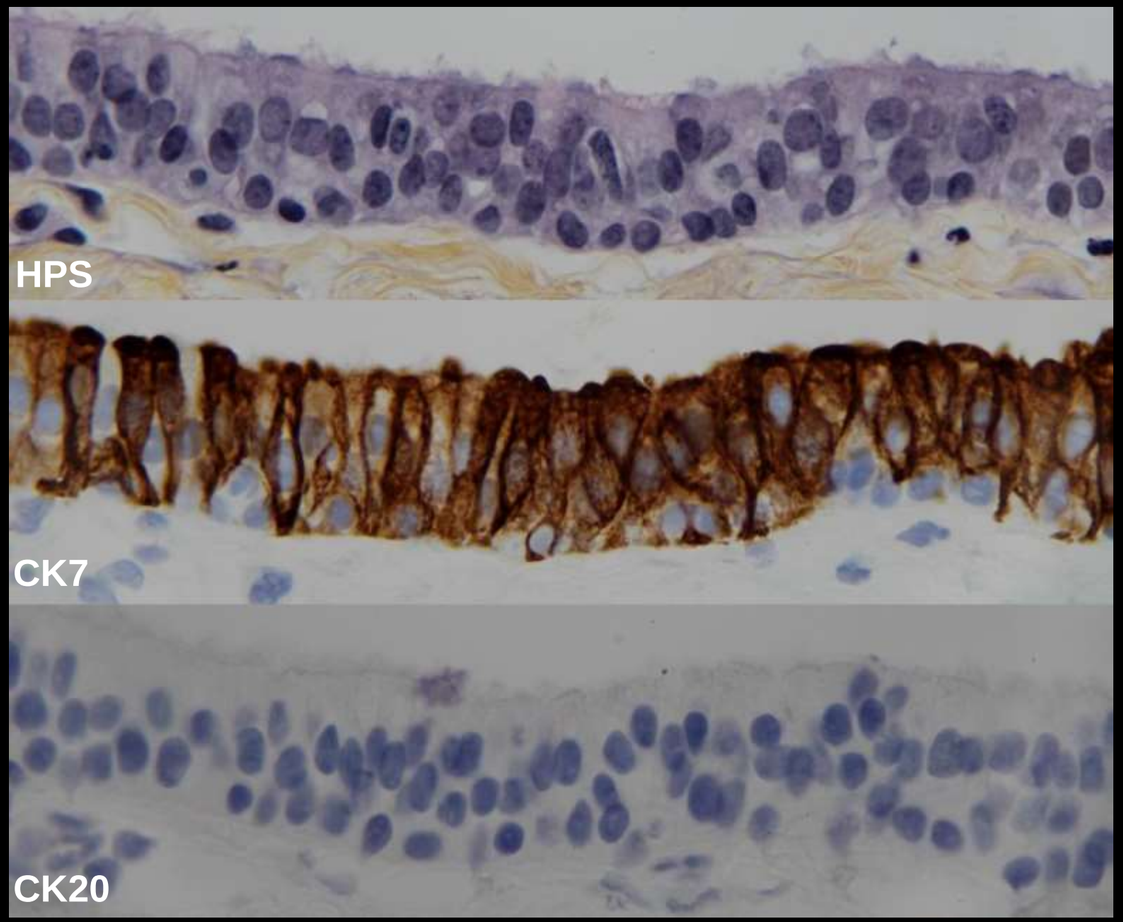
Histopathology

- (A) Low and
- (B) medium power views of the cyst wall, that regionally has a papillary configuration (Hematoxylin Phloxine Saffron stain).



Histopathology

- HPS staining of the cyst wall demonstrating pseudostratified ciliated columnar epithelial cells (respiratory type).
- The epithelial elements are strongly immunoreactive for CK7 and immuno-negative for CK20.



Diagnosis & Management

- The gold standard for diagnosis is MRI. [18]
- CT provides complimentary information about secondary osseous malformations since neurenteric cysts may be associated with congenital vertebral abnormalities.[18,21]
- Often indolent but may become symptomatic with cord compression. [22]
- If possible, total surgical resection is the most effective management option to prevent recurrence. [23.24]

Conclusions

- We have reviewed various lumbar expansile pathologies arising intradurally;
- We have discussed the imaging pattern of various tumors and established an approach to diagnosis which must includes:

  - first step: define lesion location intra vs extramedullary,
  - second step: assess the imaging characteristics including: morphology, signal, post contrast appearance and associated findings (cyst, ectatic vasculature etc.),
  - third step: establish a list of differential diagnosis based on lesion characteristics.

- Finally we have presented a rare case of Bronchial Endodermal Cyst of the Cauda Equina.

References

1. Kahan, H., Sklar, E. M., Post, M. J. & Bruce, J. H. MR characteristics of histopathologic subtypes of spinal ependymoma. *AJNR Am J Neuroradiol* 17, 143–150 (1996).
2. Gaensler, E.H., Purcell, D., and Watanabe, A (2019). Ependymomas. In E. Gaensler and J. Barakis (Ed.), *Brant & Helms' Fundamentals of Diagnostic Radiology, 5th Edition* (pp. 239). Retrieved from https://wolterskluwer.vitalsource.com/#/books/9781496367426/
3. https://radiopaedia.org/articles/spinal-myxopapillary-ependymoma
4. https://radiopaedia.org/articles/spinal-haemangioblastoma?lang=us
5. Leb, J.S. and Kligerman, S (2019). Paraganglioma. In J. Klein (Ed.), *Brant & Helms' Fundamentals of Diagnostic Radiology, 5th Edition* (pp. 689). Retrieved from https://wolterskluwer.vitalsource.com/#/books/9781496367426/
6. https://radiopaedia.org/articles/spinal-haemangioblastoma?lang=us
7. https://radiopaedia.org/articles/ventriculus-terminalis
8. Zeinali, M., Safari, H., Rasras, S., Bahrami, R., Arjipour, M., & Ostadrahimi, N. (2017). Cystic dilation of a ventriculus terminalis. Case report and review of the literature. *British Journal of Neurosurgery*, *17*, 1–5. http://doi.org/10.1080/02688697.2017.1340585
9. Sigal, R., Denys, A., Halimi, P., Shapeero, L., Doyon, D., & Boudghène, F. (1991). Ventriculus terminalis of the conus medullaris: MR imaging in four patients with congenital dilatation. *American Journal of Neuroradiology*, *12*(4), 733–737.
10. https://radiopaedia.org/articles/spinal-schwannoma
11. Kikkawa, Y. *et al.* Spinal endodermal cyst resembling an arachnoid cyst in appearance: Pitfalls in intraoperative diagnosis of cystic lesions. *Surg Neurol Int* 3, 78 (2012).
12. Gaensler, E.H., Purcell, D., and Watanabe, A (2019). Extra-axial Tumours. In E. Gaensler and J. Barakis (Ed.), *Brant & Helms' Fundamentals of Diagnostic Radiology, 5th Edition* (pp. 127-129). Retrieved from https://wolterskluwer.vitalsource.com/#/books/9781496367426/
13. https://radiopaedia.org/articles/spinal-meningioma
14. https://emedicine.medscape.com/article/341870-overview#a4
15. Gaensler, E.H., Purcell, D., and Watanabe, A (2019). Epidermoid and Dermoid Cysts. In E. Gaensler and J. Barakis (Ed.), *Brant & Helms' Fundamentals of Diagnostic Radiology, 5th Edition* (pp. 135). Retrieved from https://wolterskluwer.vitalsource.com/#/books/9781496367426/
16. https://radiopaedia.org/articles/spinal-epidermoid-cyst?lang=us
17. https://radiopaedia.org/articles/spinal-dermoid-cyst?lang=us
18. Savage, J. J., Casey, J. N., McNeill, I. T. & Sherman, J. H. Neurenteric cysts of the spine. *Journal of Craniovertebral Junction and Spine* 1, 58–63 (2010).
19. Lee, S. H., Dante, S. J., Simeone, F. A. & Curtis, M. T. Thoracic Neurenteric Cyst in an Adult: Case Report. *Neurosurgery* 45, 1239–1243 (1999).
20. Wilkens RH, Odom GL. Spinal Intradural Cysts. In: Vinkin PJ, Bruyn GW, editors. Tumors of the spine and spinal cord, Part II. Handbook of Clinical Neurology. Vol. 20. North Holland: Amsterdam; 1976. pp. 55–102.
21. Can A et al. Spinal Neurenteric Cyst in Association with Klippel-Feil Syndrome: Case Report and Literature Review. *World Neurosurgery.* 2015;84(2):592.e9-14.
22. Ma X et al. Intraspinal Bronchogenic Cyst: Series of Case Reports and Literature Review. *The Journal of Spinal Cord Medicine.* 2017;40(2):141-146
23. Weng JC et al. Intradural Extramedullary Bronchogenic Cyst: Clinical and Radiologic Characteristics, Surgical Outcomes, and Literature Review. *World Neurosurgery.* 2018;109:e571-e580.
24. Garg N et al. Is Total Excision of Spinal Neurenteric Cysts Possible? *British Journal of Neurosurgery.* 2008;22(2):241-51