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Spinal Cord Gliomas, Glioneuronal, and Neuronal Tumors in the 2021 WHO CNS Central Neural System Classification:

A Pictorial and Pathological Review

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Disclosures

None

Background:

WHO 2021 adopted integrated histo-molecular diagnosis

Brain-based models may not apply to spine

Rare, Uncommon

New entities are molecularly defined

Spine tumors lack specific imaging features

Diagnostically challenging

Purpose:

Highlight the imaging spectrum and radiologic–pathologic correlations of spinal cord gliomas, glioneuronal, and neuronal tumors within the WHO 2021 framework.

Category	Tumor Entity	Key Molecular Profiling
GLIOMAS		
Adult-type diffuse gliomas	Astrocytoma, IDH-mutant	IDH1, IDH2, ATRX, TP53, CDKN2A/B
	Oligodendroglioma, IDH-mutant, 1p/19q-codeleted	IDH1, IDH2, 1p/19q codeletion, TERT promoter, CIC, FUBP1, NOTCH1
	Glioblastoma, IDH-wildtype	IDH-wildtype, TERT promoter, chromosome 7 gain / 10 loss, EGFR
Pediatric-type diffuse low-grade gliomas	Diffuse astrocytoma, MYB- or MYBL1-altered	MYB, MYBL1
	Angiocentric glioma	MYB
	Diffuse low-grade glioma, MAPK pathway-altered	FGFR1, BRAF
Pediatric-type diffuse high-grade gliomas	Diffuse midline glioma, H3 K27-altered	H3K27-altered, TP53, ACVR1, PDGFRA, EGFR, EZHIP
Circumscribed astrocytic gliomas	Pilocytic astrocytoma	KIAA1549–BRAF fusion, BRAF, NF1
	High-grade astrocytoma with piloid features	BRAF, NF1, ATRX, CDKN2A/B (methylome-defined)
GLIONEURONAL & NEURONAL TUMORS		
	Ganglioglioma	BRAF
	Diffuse glioma with oligodendroglioma-like features & nuclear clusters	Chromosome 14 alteration (methylome)
	Rosette-forming glioneuronal tumor	FGFR1, PIK3CA, NF1
	Diffuse leptomeningeal glioneuronal tumor	KIAA1549–BRAF fusion, 1p alteration (methylation-defined)
	Gangliocytoma	BRAF
EPENDYMAL TUMORS		
	Spinal ependymoma	NF2, MYCN
	Myxopapillary ependymoma	No defining mutation listed (methylation-defined)
	Subependymoma	No defining mutation listed (methylation-defined)

Key updates

Diagnosis

- Shift to histo-molecular classification
- Molecular alterations now define entities

Groups

- Adult vs Pediatric diffuse gliomas separated
- Introduction of molecularly defined entities

Molecular Reclassification

- H3K27 alteration → Diffuse midline glioma
- MYB/MYBL1 → Pediatric diffuse low grade glioma
- MAPK pathway → Diffuse low-grade glioma

Challenges in Implementation in the Spine

Limited Epidemiologic & Molecular Data

- Spinal tumors are rare
- Molecular prevalence often unknown

Brain-Centered Classification Bias

- WHO updates primarily validated in intracranial tumors
- Limited spine-specific radiologic literature

Diagnostic Constraints

- Small intramedullary biopsy samples
- Limited access to advanced molecular profiling

Imaging–Pathology Mismatch

- MRI cannot reliably determine tumor grade
- Molecularly aggressive tumors may appear radiologically indeterminate

Case 1: Diffuse Leptomeningeal Glioneuronal tumor (DLG)

13 yo female; back pain

Imaging:
Diffuse dorsal intradural lesion with extensive leptomeningeal and cauda equina enhancement, causing anterior cord displacement and compression.

Molecular:
FGFR1 and PTPN11 mutations → MAPK pathway activation (WHO 2021 entity).

Treatment/Prognosis:
Indolent tumor; resection when feasible, with potential for targeted MAPK-directed therapy

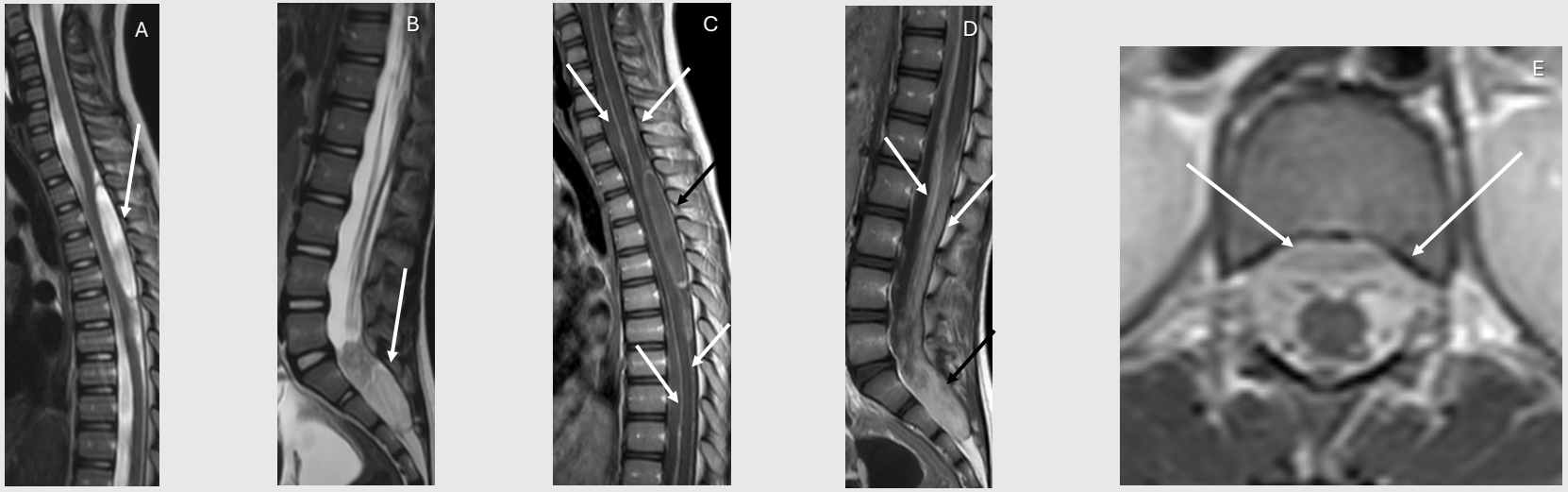

Figure 1. Figure 1A. Sagittal T2WI of the upper spine demonstrates a lesion dorsal to the spinal cord at the level of the upper thoracic spine anteriorly displacing and compressing the spinal cord (arrow). Figure 1B Sagittal T2WI of the lower spine demonstrates a lesion within the dependent portion of the distal thecal sac (arrow). Figure 1C. Sagittal post contrast T1WI of the upper spine demonstrates diffuse enhancement (white arrows) along the surface of the spinal cord with peripheral enhancement (black arrow). Figure 1D. Sagittal post contrast T1WI of the lower spine demonstrates diffuse thick enhancement along the cauda equina nerve roots (white arrows) and an enhancing nodular lesion within the distal thecal sac (black arrow). Figure 1E. Axial post contrast T1WI at the level of the upper thoracic spine demonstrates thick diffuse leptomeningeal disease (arrow).

Case 2: Ganglioglioma

25 yo male; left hand numbness

Imaging:
Long-segment intramedullary mass with mixed cystic–solid components, bone remodeling, and scoliosis; features suggestive of a slow-growing glioneuronal tumor

Molecular:
Typically, MAPK pathway–activated (most commonly BRAF V600E); negative in this case.

Treatment/Prognosis:
Indolent WHO Grade 1 tumor; maximal safe resection is primary therapy with generally favorable prognosis

Spinal gangliogliomas commonly present in young patients and are frequently associated with scoliosis

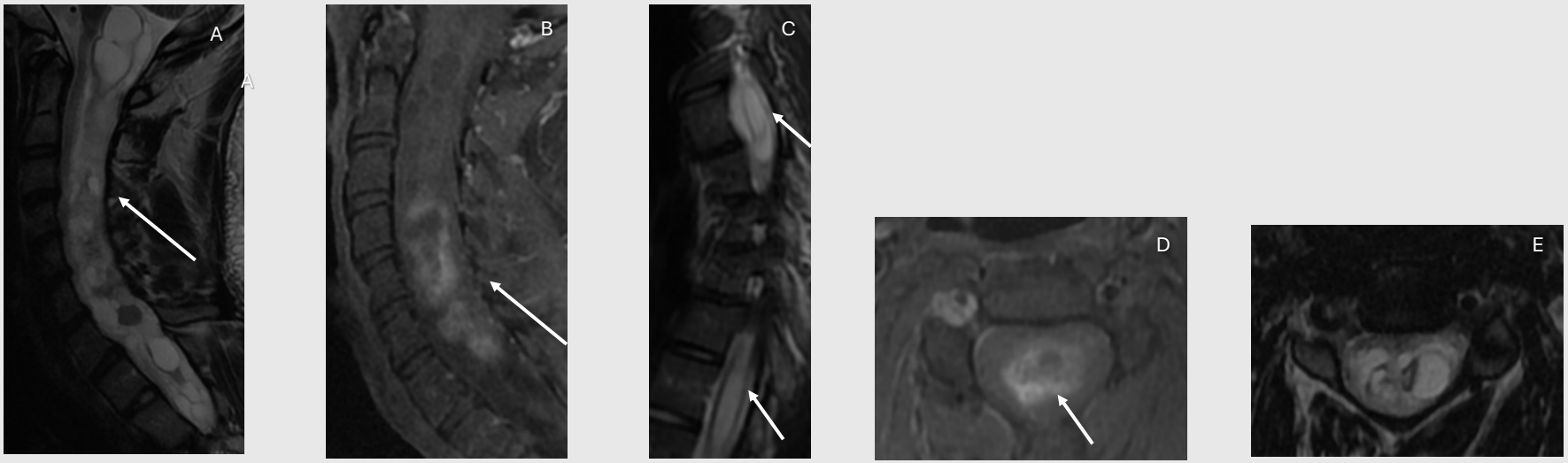

Figure 2. Figure 2A. Sagittal T2 demonstrates a lobulated expansile long segment partially cystic mass spanning the craniocervical junction to the upper thoracic spine (arrow). Figure 2B Sagittal post contrast fat sat T1WI demonstrates enhancement of the solid component solid (arrow). Figure 2C. Sagittal T2WI demonstrates a long segment syrinx within the scoliotic thoracic spine. Figure 2D. Axial T2WI demonstrates the expansile cystic lesion with areas of hemosiderin. Figure 2E. Axial post contrast fat sat T1WI demonstrates the enhancing component of the mass (arrow). Given the long segment cystic mass ganglioma should be considered within the differential.

Case 3: High-grade Astrocytoma with Piloid features (HGAP)

53 yo male; neck pain

Imaging:
T2 hyperintense, heterogeneously enhancing lesion with non-necrotic appearance; imaging may mimic lower-grade glioma despite aggressive biology.

Molecular:
IDH-wildtype with ATRX loss and characteristic DNA methylation profile; frequently associated with MAPK pathway alterations (NF1, BRAF, FGFR1).

Treatment/Prognosis:
Clinically behaves like WHO Grade III/IV glioma; requires aggressive management with resection and adjuvant therapy despite deceptively low-grade imaging features

- *Imaging–pathology discordance: non-necrotic MRI appearance despite high-grade molecular profile*
- *Association with neurofibromatosis type 1*

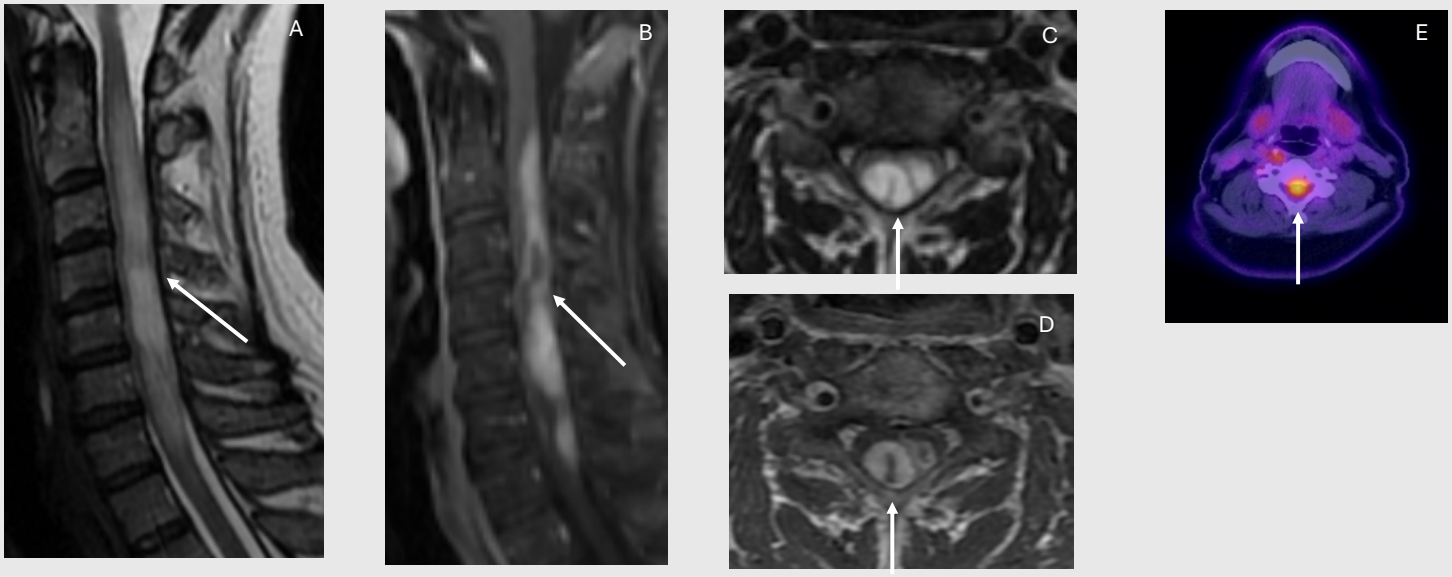

Figure 3. Figure 3A. Sagittal T2WI of the cervical spine demonstrates a long segment mildly expansile T2 hyperintense mass spanning the cervical cord (arrow). Figure 3B. Sagittal post contrast fat sat T1WI demonstrates enhancement within the mass. Figure 3C Axial T2WI demonstrates the expansile hyperintense mass (arrow). Figure 3D. Axial post contrast T1WI demonstrates enhancement of the lesion (arrow). Figure 3D. Axial fused PET/CT demonstrates avidity within the lesion (arrow).

Case 4: Diffuse Midline Glioma

13 yo male; Left sided weakness

Imaging:
Long-segment expansile intramedullary lesion, T2 hyperintense with variable enhancement ± diffusion restriction; hemorrhage may suggest H3 K27 alteration.

Molecular:
Defined by H3 K27 alteration (histone H3 mutation), which establishes WHO Grade 4 diagnosis independent of histology.

Treatment/Prognosis:
Aggressive tumor with poor prognosis (median survival ~11–15 months in pediatric spinal cases); multimodal therapy required with recommended neuroaxis surveillance due to dissemination risk.

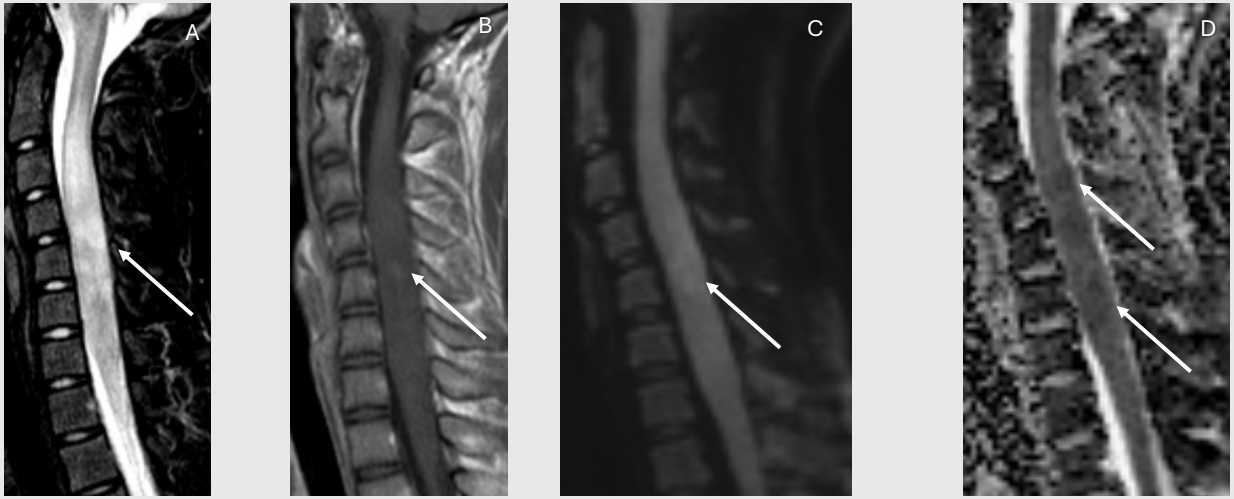

Figure 4. Figure 4A. Sagittal STIR of the cervical spine demonstrates a long segment expansile mass (arrow). Figure 1B. Sagittal post contrast T1WI demonstrates mostly non enhancing mass with a very small enhancing nodule (arrow). Figure 4C and D. Sagittal DWI and ADC demonstrate few areas of diffusion restriction within the mass.

Case 5: Glioblastoma grade IV

31 yo male; numbness and weakness of the lower body

Imaging:
Long-segment infiltrative intramedullary lesion, T2 hyperintense with variable enhancement; imaging overlaps other spinal tumors and rarely allows gross total resection.

Molecular:
IDH-wildtype, WHO Grade 4; spinal GBMs are uncommon and biologically aggressive.

Treatment/Prognosis:
Managed with surgery, radiation, and chemotherapy; median survival is poor (10–14 months) despite aggressive therapy.

Very rare entity (<1.5% of spinal cord tumors)

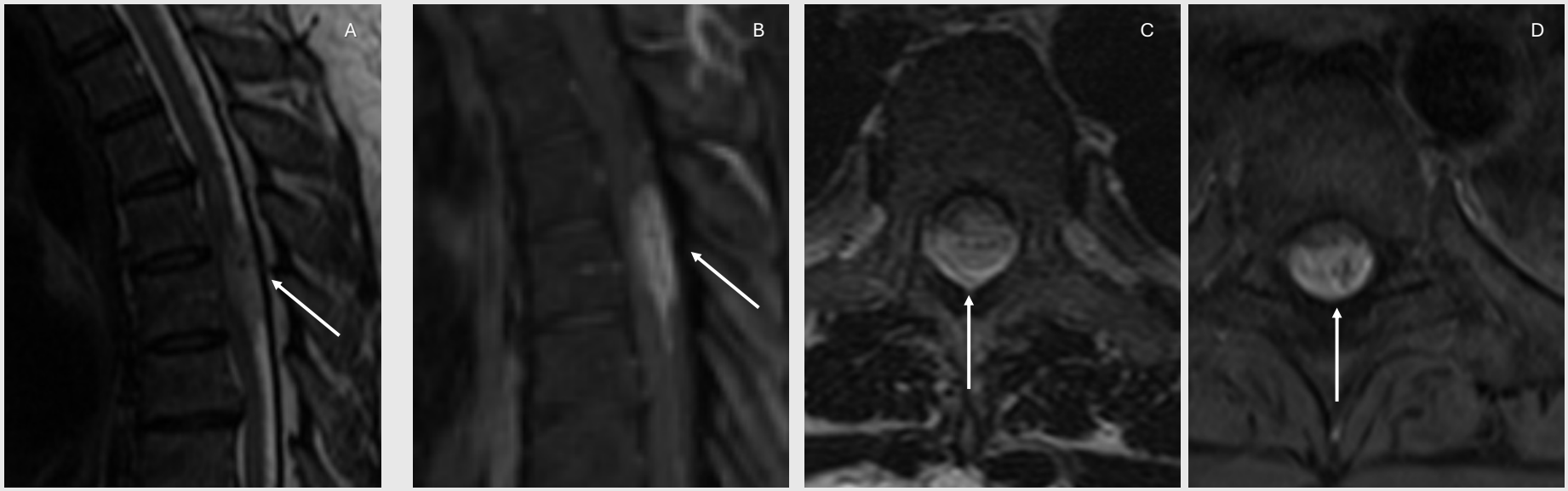

Figure 5A. Sagittal T2WI of the mid thoracic spine demonstrates an exophytic vascular slightly T2 hyperintense mass (arrow). Figure 1B. Sagittal fat sat post contrast T1WI demonstrates enhancement of the mass (arrow). Figure 5C. Axial T2WI demonstrates dorsally exophytic T2 hyperintense mass with spinal cord signal abnormality (arrow). Figure 5D. Axial post contrast fat sat T1WI demonstrates enhancement of the mass (arrow).

Recurrence and Disease Dissemination

- **Recurrence:** 3 years after initial diagnosis.
- **Dissemination:** Follow-up imaging demonstrated diffuse leptomeningeal dissemination involving both spine and brain.
- **Behavior:** Spinal glioblastomas demonstrate aggressive infiltrative biology with high risk of neuraxis spread.

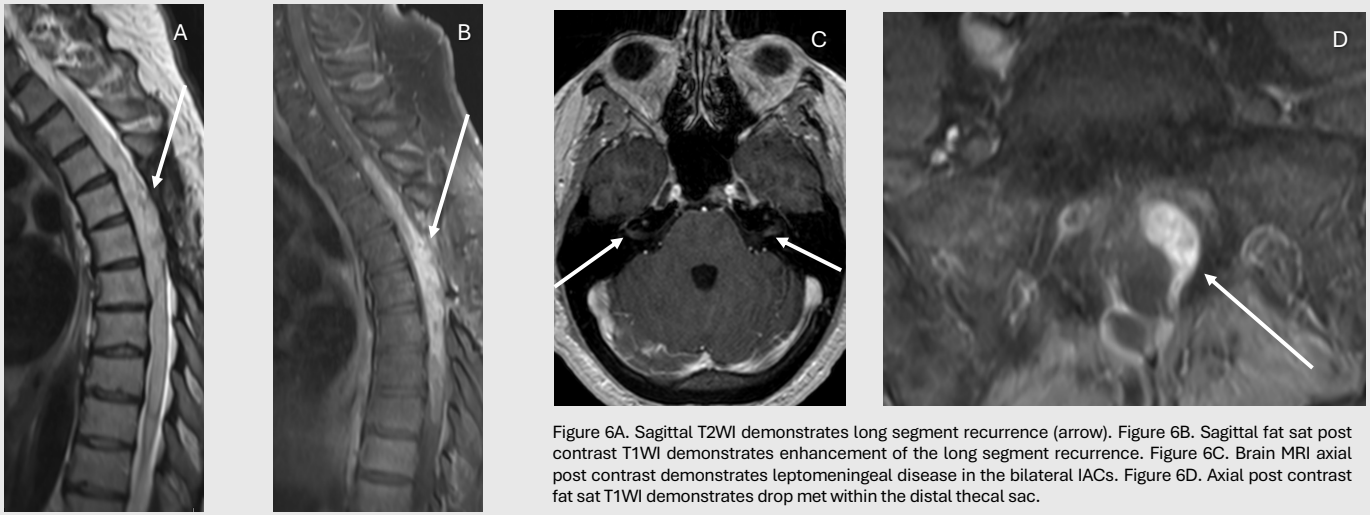

Figure 6A. Sagittal T2WI demonstrates long segment recurrence (arrow). Figure 6B. Sagittal fat sat post contrast T1WI demonstrates enhancement of the long segment recurrence. Figure 6C. Brain MRI axial post contrast demonstrates leptomeningeal disease in the bilateral IACs. Figure 6D. Axial post contrast fat sat T1WI demonstrates drop met within the distal thecal sac.

Case 6: Subependymoma

44 yo male

Imaging:
Well-circumscribed, eccentric intramedullary mass with elongated T2 hyperintense cord expansion: “bamboo leaf sign”.

Molecular:
No characteristic high-grade molecular alterations; distinct from MYCN-amplified or H3-mutant spinal tumors.

Treatment/Prognosis:
Indolent WHO Grade 1 tumor; surgical resection is usually curative with excellent prognosis.

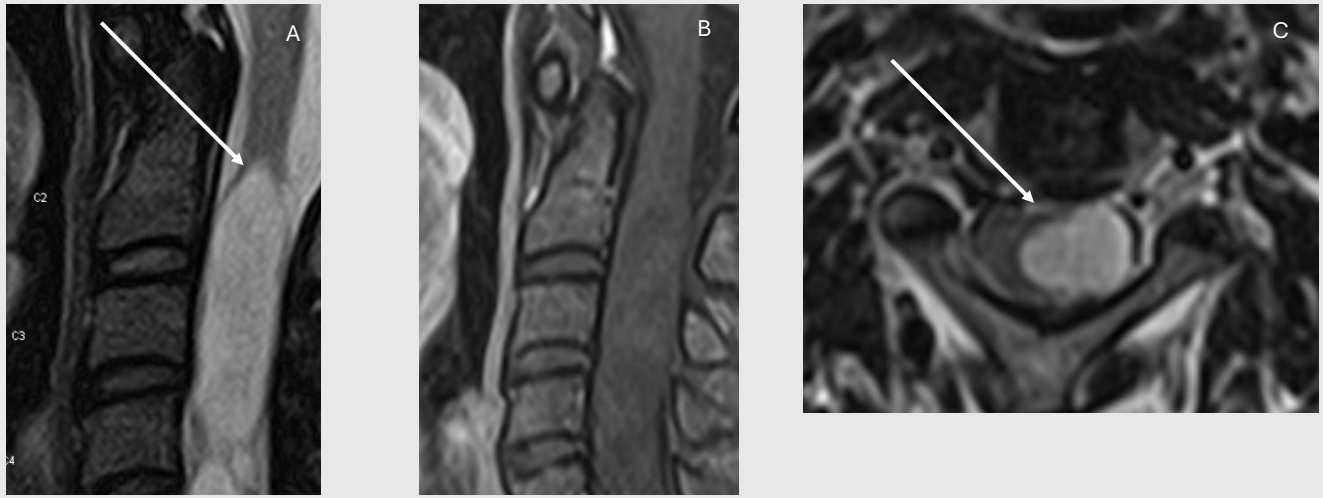

Figure 7. Figure 7A. Sagittal T2WI of the cervical spine demonstrates steep expansile lesion “bamboo leaf sign”(arrow). Figure 7B. Sagittal fat sat post contrast T1WI demonstrates lack of enhancement of the lesion, which is frequently encountered within sub ependymomas. Figure 7C. Axial T2WI demonstrates eccentric growth and rightward displacement and compression of the spinal cord (arrow)

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Thank you