Tumors and pseudotumors of the spine: a review of the main aspects in computed tomography and magnetic resonance imaging.

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Learning objectives

The purposes of the exhibit is:

- Describe the primary tumors and pseudotumors of the spine and the most common radiological findings.
- Review the main differential diagnoses according to the location of the lesions.
Background

Spinal cord tumors represent approximately 10-20% of primary central nervous system tumors.

Tumors and pseudotumors of the spine cause various signs and symptoms. Frequently initially present with non-specific symptoms such as back pain and clumsiness and are therefore often associated with a prolonged period between symptom onset and diagnosis. Other symptoms are: progressive weakness that can lead to palsy, sphincter disorders, erectile dysfunction, even to the present syndrome of cord compression, according to the type and location of the tumor.

Spinal cord neoplasms are rare lesions in children. Many children present frequently to healthcare services before a diagnosis is made, and appropriate imaging is often only instigated once a child has developed neurological deficits.

The diagnosis of spinal tumors is based on patient age, topographic features of the tumor, and lesion pattern as seen at CT and MR imaging. A systematic approach is useful for recognizing tumors of the spine with characteristic features.

The patients with cervical tumors tend to have delayed diagnoses due to the time taken before visiting a physician. Thoracic spine tumors were undiagnosed due to the symptoms mimic degenerative disorders. The possibility of thoracic spinal tumor should be remembered when unexplained low back pain persists, even in response to various treatments.
Imaging findings OR Procedure details

The differential diagnosis of tumors of the spine, the location is the most important data, as well as the clinical presentation and age of the patient.

As for location, are classified as extra-dural, intradural-extramedullary and intramedullary.

Extradural metastases account for about 60% of tumors of the spine vertebral.

Meningiomas and nerve sheath tumors (intradural-extramedullary tumores) account for 30% and astrocytomas and ependymomas (intramedullary tumors) account for 10%.

1. **EXTRADURAL SPINAL TUMORS:**

   - Extradural tumors that are outside the dural sac. The main tissues involved are those located in the bone marrow, epidural space, including the paraspinal soft tissues. Examples of injuries that affect the extradural space: herniated disc (FIG 1), syndesmophytes, fractures, primary bone tumors and metastases.
   
   - Vertebral hemangioma represents the most common benign tumor of the spine. Metastases, lymphoma, and multiple myeloma are the most frequent malignant spinal tumors. Primary osseous tumors of the spine, in contrast, are rare conditions but may demonstrate typical imaging findings.

   - Metastases account for about 60% of extradural tumors (FIGs 2-5). The spine is the commonest site for bone metastases, and the incidence of spinal metastases is increasing, with increasingly older populations, longer life expectancy, and improvements in medical treatment.
   
   - Primary tumors whose metastases frequently involve the spine are breast, lung, renal, prostate, thyroid, melanoma, myeloma, lymphoma and colorectal cancer.
   
   - The use of paramagnetic contrast agent is very useful for the diagnosis of metastases in the spine. Osteolytic metastases compose about 70% of lesions. There is hyperintense signal on T2-weighted sequences and STIR, and post-contrast the enhancement is intense. In osteoblastic metastases, enhancement is heterogeneous and peripheral.

   - In diffusion-weighted magnetic resonance imaging (DWI), the observed MRI signal intensity is attenuated by the self-diffusion of water molecules. DWI has been used in particular for the differentiation of benign and malignant vertebral compression fractures and has high accordance with skeletal
scintigraphy in detecting bone metastases. DWI with low $b$ value provided excellent distinction between metastatic tumor infiltration and benign vertebral fracture edema. Hyperintense signal intensity on DWI was highly specific for the diagnosis of metastatic tumor infiltration of the spine (FIG 6).

- Diagnosis and management of metastatic spine disease requires disease categorization into the compartment involved, pathology of the lesion, and anatomic region involved. The diagnostic approach should commence with careful physical examination, and the workup should include plain radiographs, magnetic resonance imaging, computed tomography, and bone scintigraphy (FIG 7, 8). Handling vary from palliative non-operative treatment to aggressive surgical ones. There is an international group of spinal surgeons, called the Global Spine Tumour Study Group, who are dedicated to studying the techniques and outcomes of surgery for spinal tumours, to build on the existing evidence base for the surgical treatment of spinal tumours. Optimal management requires proper patient selection to individualize the most appropriate treatment modality.

- Primary vertebral tumors, although less common than metastases to the spine, make up a heterogeneous group of neoplasms that can pose diagnostic and treatment challenges. They affect both the adult and the pediatric population and may be benign, locally aggressive, or malignant. These lesions encompass a wide variety of tumor types classified by their cell of origin: bony tumors, cartilaginous tumors, vascular tumors, plasma cell dyscrasias, and tumors that arise from embryonic rests. Further classification of these tumors into malignant or benign subtypes is based on their clinical progression, histopathological evidence of invasiveness, and response to therapy.

- The most common benign primary vertebral tumors are: hemangioma, aneurysmal bone cyst, chondroma and enchondroma, osteoid osteoma, and osteoblastoma and lesions related to eosinophilic granuloma and fibrous dysplasia (FIGS 9-13).

**EXTRADURAL PSEUDOTUMOR:**

The epidural lipomatosis is an example of this pseudotumoral lesion topography. It’s an excessive accumulation of intraspinal fat causing cord compression and neurologic deficits. The best diagnostic clue is abundant epidural fat compressing thecal sac. The natural history and prognosis is excellent. > 80% with post-surgical symptomatic relief (FIG 14).

**2. INTRADURAL EXTRAMEDULLARY TUMORS:**
Included in this classification tumors that are inside the dural sac but outside the spinal cord. Examples of tissues intradural-extramedullary tumors: nerve roots, meninges, CSF space.

Spinal intradural extramedullary tumors account for 2/3 of all intraspinal neoplasms and are mainly represented by meningeomas and schwannomas, with the former accounting for the 25-46% of all primary intraspinal tumors.

THE NERVE SHEATH TUMORS:

- Higher incidence in the 4th. decade of life, being less frequent in children. No sex predilection.
- More common: schwannomas (synonym: neurinoma or neurilemmomas)
  2o type: neurofibromas
  3o. type (RARE): ganglioneuroma
- **SCHWANNOMA**: well-circumscribed, "dumbbell" shaped, enhancing spinal mass. Most are small. Giant schwannoma extend over 2 vertebral segments (FIG 15-19).
- **NEUROFIBROMA**: bulky multilevel spinal nerve root tumors in patient with stigmata of neurofibromatosis type I (NF1). Variable size and has three morphologic features: localized, diffuse and plexiform.

MENINGEOMAS:

- The meningiomas occur predominantly in the 5th. and 6th. decades which 70% are women. Regarding the topography predominates in the thoracic spine (70%), followed by cervical (15%) and lumbar (5%) Among men, the meningiomas occurring about 50% in the thoracic spine and 40% in the cervical spine (FIG 21,22)
- The most common clinical symptom is local pain. Less frequently there may be pain radiated. Changes motor and sphincter and tingling are more rare.
- Pathology: grow from cels. arachnoid rather than dura Young patients: it can be more aggressive with a worse prognosis
- The treatment of choice for most of these tumors is a complete surgical resection. In particular if neurological deficits appear, a fast surgical intervention is indicated, since the prognosis depends on duration and severity of the preoperative existing deficits (FIG 23, 24).

INTRADURAL EXTRAMEDULLARY PSEUDOTUMOR:

ARACHNOID CYST:
The arachnoid cyst is one of intradural-extradural pseudotumors, and it’s a intraspinal extradural-extradural loculated cerebral spinal fluid collection.

Most patients are asymptomatic. Others signs/symptoms are: pain, paraoaeresis, paresthesia, hyperreflexia, bladder and bowel incontinence. Worsening neurologi deficits with enlarging cyst.

May present at any age.

There is a Nabors classification of this lesions:

Type I: extradural cyst without spinal nerve root fibers

- A: Extradural
- B: Sacral meningocele

Type II: extradural cyst with spinal nerve root fibers

- Tarlov perineural cyst (FIG 25)
- Spinal nerve root diverticulum

Type III: Intradural cyst

- Intradural
- Acquired intradural cyst also known as subarachnoid cyst

Type I thoracic arachnoid cyst more common in adolescents (FIG 26).

This lesion has an excellent prognosis. The treatment is laminectomy with complete cyst wall resection. If complete resection not possible: wide marsupialization of cyst or shunting.

3. **INTRAMEDULLARY TUMORS:**

- Location: inside the spinal cord
- Tissues: spinal cord parenchyma, pia mater
- Examples: ependymoma, astrocytoma, hemangioblastoma, spinal cord metastases.
- Rare: oligodendroglioma, ganglioglioma and intramedullary schwannoma
- In relation to intramedullary tumors, MRI shows lesions that determine swelling of the spinal cord and has characteristic features of post-contrast enhancement on MRI, which help in the differential diagnosis of astrocytomomas and ependymomas.
In tumors with infiltrative growth into the intramedullary area, a marginal tumor tissue has to be left in situ in order to avoid additional neurological deficits. In particular if neurological deficits appear, a fast surgical intervention is indicated, since the prognosis depends on duration and severity of the preoperative existing deficits.

Among the intradural-intramedullary tumors the most frequent ones are ependymomas and astrocytomas.

EPENDYMOMA

- Well-circumscribed intramedulary mass, with homogeneous enhancement in the post-contrast sequences.
- Peak incidence: 4th. and 5th. Decades (35 years)
- No sex predilection
- More common locations: cervical > thoracic > conus (FIG 27, 28)
- Pathology: Form Myxopapillary (95%). Most are WHO grade II.
- Nonspecific clinical presentation (local or radiating pain, motor dysfunction or sphincter only 25% of cases)
- Cyst formation and hemorrhage are common.

Multiple ependymomas were present in 58% of patients with neurofibromatosis Type 2. The most common site of involvement was the cervical cord or cervicomedullary junction, followed by the thoracic and lumbar cords. The majority of patients had no symptoms related to their tumors. Neurofibromatosis Type 2-related ependymomas exhibit an indolent growth pattern with tumor progression limited to a minority of patients (FIG 29).

ASTROCYTOMA:

- Most common intramedullary tumor in children/young adults. No sex predilection
- Location: Cervical> thoracic.
- Clinical data: local or radiating pain. Motor abnormalities, urinary incontinence or faecal occur in later stages of the disease.
- Most (75%) are low grade (WHO I or II)
  - Children: 85-90% low-grade
  - GBM: <1.5%
- In the spinal cord astrocytomas, the mass tends to be poorly defined, with imprecise limits, and heterogeneous enhancement after contrast, and expanding cord. Cystic component occur in 30% tumors.
• Survival varies with tumor histology/grade. Post-operative neurologic function determined largely by degree of pre-operative deficit (FIG. 30, 31)

HEMANGIOBLASTOMA (FIG 32):

• Hemangioblastomas are highly vascularised tumors of the central nervous system and account for 1.5-2.5% of all spinal cord tumors, predominantly in the 4th. decade (mean age 35 years).
• 1/3 patients Syndrome Von Hippel-Lindau (autosomal dominant disease, with cerebellar hemangioblastoma / medullar angiomatosis of the retina, renal cell carcinoma and / or pheochromocytoma in varying degrees)
• Signs and symptoms: sensory and motor disturbances
• Pathology: solid portion, highly vascularized with small arteries, capillaries and venules drainage. May have associated cystic component.
• Operation was recommended to every symptomatic patient as early as possible. In VHL patients, surgery was recommended if tumor growth was observed on MRI in the next practicable time.
• Patients without VHL most frequently require hemangioblastoma resection for diagnostic purposes and/or because symptoms prompted an imaging work-up that lead to the discovery of the tumor.

PARAGANGLIOMA (FIG 33,34):

• Extra adrenal, spinal paraganglioma composed of chromaffin cells (groups of cells associated to the autonomous system). WHO grade I.
• Symptoms: Cauda equina syndrome and compression of nerve roots. Chronic back pain and sciatica is common. Spinal paragangliomas have little/no secretory activity. Symptom duration varies from days to years. Late diagnosis is common because it mimics degenerative conditions.
• Best diagnostic clue: vascular cauda equina mass.
• Differential diagnosis: ependymomas, schwannomas, metastases, and dermoid.

INTRAMEDULLARY METASTASES:

• Very rare (1% -3% of cancer patients).
• Primary tumors: breast, lung, central nervous system (glioblastoma, medulloblastoma, ependymoma)
• Hematogenous spread.
• Intramedullary tumors of leptomeningeal metastasis: penetration through the spinal cord.

INTRAMEDULLARY PSEUDOTUMORS:

MULTIPLE SCLEROSIS (FIG 35):

• Primary demyelination disease of central nervous system with multiple lesions disseminated over time and space. Spine cord: an early site. 10-20% isolated spinal cord disease. Cervical is the most commonly affected spinal cord segment.
• Prevalence in women. 15-50 years. Peak: 3rd. and 4th. decades
• Demyelinating plaque: dorso-lateral spinal cord at any segment. Less than half cross sectional area of spinal cord and less than two vertebral segments in length.
• Demyelinating as well as neoplastic spinal cord diseases can cause paresthesia, progressive sensomotoric deficits and bowel and bladder dysfunction.
• Imaging of the spine, especially with magnetic resonance imaging (MRI), is an essential component in the diagnostic assessment of myelopathy and makes a substantial contribution to achieving the correct diagnosis.

INFECTIONS INTRAMEDULLARY LESIONS:

• Among the intramedullary infectious and inflammatory lesions are described: intramedullary abscesses, viral myelitis (acute transverse myelitis), myelopathy, HIV, schistosomiasis, cysticercosis and others.
• In these lesions is important to know the clinical-laboratory, because the imaging findings are varied.
• Intramedullary spinal cord abscess: ring-enhancing mass within cord, variable size, usually less than 2 cm.
• Schistosomiasis: myelopathy, cord enhancement, edema in patient from endemic area. Location: thoracic cord, conus.
• Cysticercosis: cystic lesion, size variable, in thoracic segment in 60-75%. Imaging varies with developmental stage of infection, host response.
• Myelopathy resulting from primary HIV infection: spinal cord T2 hyperintensity which may show patchy enhancement. In these patients, can be revealed opportunistic infections such as abscesses including toxoplasmosis (FIG 36).
Fig. 1: Disc herniation at C5-C6 with compression of the spinal cord
Fig. 2: MyeloCT: demonstrating sclerotic lesions in the dorsal vertebral body, with injury to the pedicles and the cortical disruption, being observed also marked extrinsic compression of the dural sac and spinal cord in correspondence. Metastatic prostate cancer.

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Fig. 3: MRI: extradural metastasis of lung tumor in dorsal vertebra.

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**Fig. 4:** Osteolytic metastases of breast cancer: extradural mass with cortical disruption and damage to the pedicles, causing spinal cord compression.

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Fig. 5: Metastasis of breast cancer, with partial collapse of the body of D2 and mass with marked extradural compression of the spinal cord and the dural sac.

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Fig. 6: Male patient 70 years old: Partial collapse of thoracic vertebra for metastatic lung cancer, showing hypersignal in diffusion sequence, featuring high cellularity and eliminating the possibility of osteoporotic collapse.

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Fig. 7: Large extradural metastasis of kidney cancer in the lumbar spine.

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Fig. 8: Same patient: MRI 3 months after surgical resection of the extradural mass, showing disease progression, with the appearance of lytic lesions in the vertebral bodies of L3 and L5 with cortical rupture.

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Fig. 9: Hemangioma in the vertebral body of L2.

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Fig. 10: Aneurysmal bone cyst: insuflantes lytic lesions in the body and right pedicle and posterior arch of lumbar vertebra.

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Fig. 11: Aneurysmal bone cyst in cervical vertebra, causing severe compression of the spinal canal.

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Fig. 12: Osteoblastoma: expansile mass occurring in posterior elements in cervical spine.

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**Fig. 13:** Osteoid osteoma: small radiolucent tumor nidus with surrounding sclerosis in cervical spine.

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Fig. 14: Epidural lipomatosis extending from C2 to D2, causing severe spinal cord compression.

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Fig. 15: CT: Schwannoma of L3, with intense enhancement after contrast
**Fig. 16:** Coronal reconstruction of the same case, showing the thickening of the left L3 root.

**Fig. 17:** Schwannoma: CT and MRI. Comparison between the methods, demonstrating improved visualization of the lesion on T1 post-gadolinium sequence in the transverse plane, with the right neural foramen enlargement. Note that the sagittal sequence is not useful to characterize the topography of the intradural extramedullary lesion, serving only to report their level in the spine (L4).
Fig. 18: Schwannoma: MR sagittal, coronal and axial views. The sagittal plane indicates that the lesion is at the level of C3 and C4. The coronal and axial views shown to be an intradural-extramedullary tumor with spinal cord compression and increased right neural foramen.
Fig. 19: Giant Schwannoma determining great compression of the cervical cord: MRI in the coronal and axial planes.

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Fig. 20: Multiple neurofibromas in the cauda equina in a patient with neurofibromatosis type 1.

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**Fig. 21:** Meningioma in the thoracic spine (D1-D2): MRI sagittal, axial and coronal. Note the dural base of implantation, the homogeneous contrast-enhancement and compression of the spinal cord.

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Fig. 22: Cervical meningioma. intradural-extramedullary mass with intense homogeneous enhancement by contrast and in contact with the dural surface.

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**Fig. 23:** Male patient, aged 20, with meningioma in the lumbar spine at the level of the cauda equina.

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Fig. 24: Same patient fig 23, postoperative control after wide laminectomy, showing excellent surgical outcome.

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**Fig. 25:** Multiple periradicular sacral cysts, causing enlargement of the sacral foramina. Tarlov perineural cysts.

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Fig. 26: Boy 12 years old, with spinal cord compression syndrome. MyeloCT and MRI of the thoracic spine showed arachnoid cyst extending from D6 to D9, compressing the spinal cord.

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Fig. 27: Cervical ependymoma: a well-defined intramedullary mass with hypointense area on T2-weighted sagittal sequence, suggesting hemorrhage.

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Fig. 28: Conus ependymoma: a well-defined mass, hyperintense on sagittal T2-weighted sequence, hypointense on T1-weighted sequence and post-contrast enhancement
Fig. 29: Patient with neurofibromatosis type 2, with cervical meningioma in the level of C2-C3, multiple neurofibromas in the cauda equina and ependymomas in lombar spine, and bilateral acoustic schwannomas.
**Fig. 30:** CERVICAL ASTROCYTOMA: intramedullary mass with hypointense signal on T1-weighted sequence and heterogeneous post-contrast enhancement, which extends throughout the cervical spine to the level of D2.

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**Fig. 31:** Evolution of the same case FIG 30, showing increased intramedullary tumor volume, which extends to the level of the brainstem, observing cystic areas.

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**Fig. 32:** Cervical hemangioblastoma: a patient with von Hippel-Lindau disease. Note the heterogeneous intramedullary mass with areas of prominent vessels inside, best evidenced in T1-weighted sequences after contrast.

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Fig. 33: Paraganglioma: Vascular cauda equina mass filling thecal sac and obliterating normal CSF signal. Flow voids present along superior margin.

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Fig. 34: Postoperative control of the same case of Figure 33, showing complete resection of the paraganglioma and pseudomeningocele.

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Fig. 35: MULTIPLE SCLEROSIS: Demyelinating plaques in cervical spinal cord.

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Fig. 36: HIV-positive patient presenting intramedullary lesion with peripheral enhancement after contrast and aspect eccentric target: laboratory-confirmed toxoplasmosis.

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Conclusion

- The location of spine tumors is the most important information for the differential diagnosis and the MRI is the best method of image in this evaluation, especially in the coronal and transverse sequences after intravenous administration of paramagnetic contrast.
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