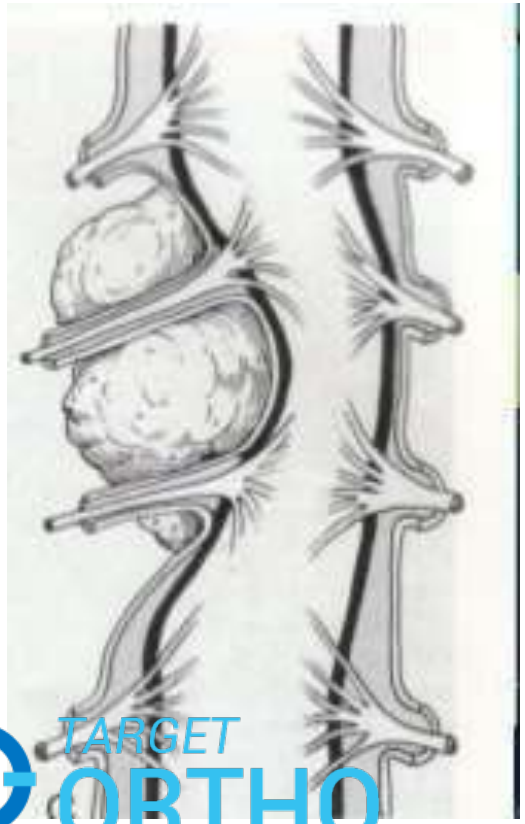
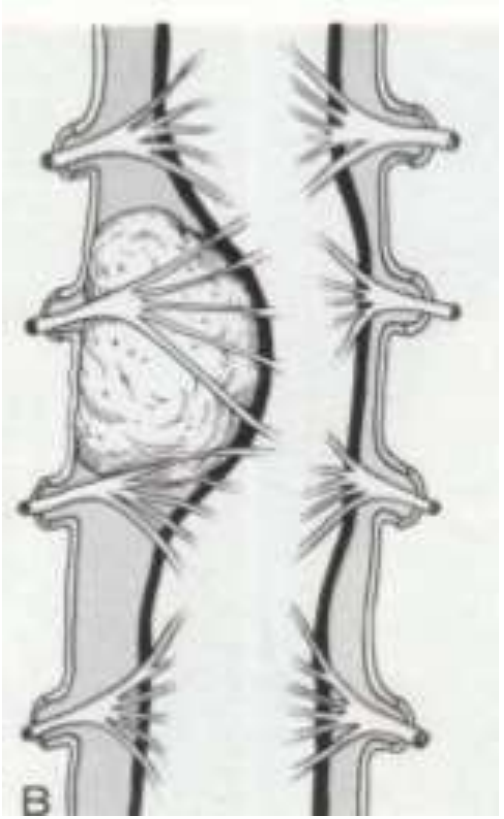
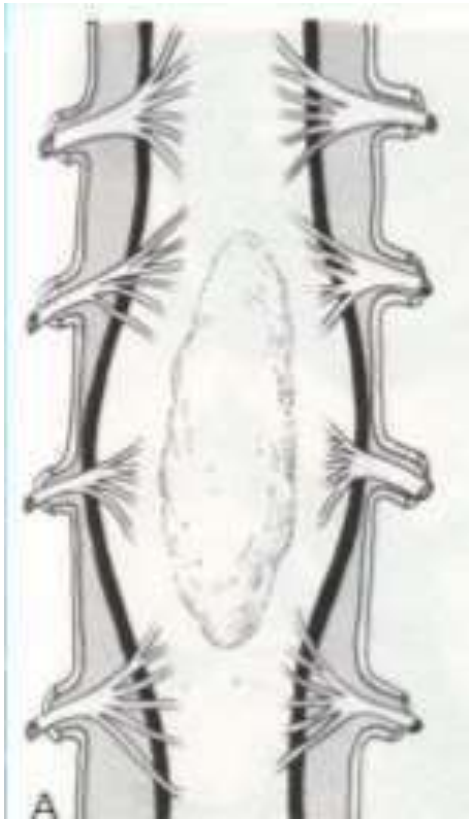
SPINAL CORD TUMOURS







General Information

- Most primary CNS spinal tumors are benign (unlike the case with intracranial tumors).
- Most present by compression rather than invasion.

Adams RD, Victor M. In: Intraspinal Tumors. Principles of Neurology. 2nd ed. New York: McGraw-Hill; 1981:638–641

Compartmental Locations Of Spinal Tumors

- Classified into 3 groups based on the compartment involved.
- Metastases can be found in any compartment

1. Extradural (ED) (55%): arise outside cord in vertebral bodies or epidural tissues

2. Intradural extramedullary (ID-EM) (40%): arise in leptomeninges or roots. Primarily meningiomas and neurofibromas (together =55% of ID-EM tumors)

3. Intramedullary spinal cord tumors (IMSCT) 5%: arise in SC substance. Invade and destroy tracts and grey matter

Differential Diagnosis

Extradural spinal cord tumors (55%)

- Arise in vertebral bodies or epidural tissues

1. Metastatic:
    - Mostly are osteolytic (causes bony destruction) Lymphoma, lung, breast , prostate
    - Mets may be osteoblastic , prostate in men, breast in females

2. Primary spinal tumors (very rare)

a) chordomas

b) osteoid osteoma

c) osteoblastoma

d) aneurysmal bone cyst (ABC)

e) Chondrosarcoma

f) osteochondroma (chondroma)

g) vertebral hemangioma

h) giant cell tumors (GCT) of bone

i) giant cell (reparative) granuloma

3. Miscellaneous:

a) Plasmacytoma

b) Multiple myeloma

c) Ewing's sarcoma

BOX 90.1 Incidence of Tumors in Adults

Extramedullary (Two-Thirds of Cases)

Nerve sheath tumor: 40%
Meningioma: 40%
Filum ependymoma: 15%
Miscellaneous[a]: 5%

Intramedullary (One-Third of Cases)

Ependymoma: 45%
Astrocytoma[b]: 40%
Hemangioblastoma: 5%
Miscellaneous[c]: 10%

Intradural Extramedullary Spinal Cord Tumors (40%)

1. **Meningiomas**: usually intradural, but may be partly or, in 15% wholly extradural
2. **Neurofibromas**: usually intradural, but may be partly or wholly extradural
3. **lipomas**
4. Miscellaneous: only ≈ 4% of spinal metastases involve this compartment

Intramedullary spinal cord tumors (5%)

1. astrocytoma : 40%
2. ependymoma: 45%
3. miscellaneous: 10%, includes:
    A) malignant glioblastoma
    B) dermoid
    C) epidermoid
    D) teratoma
    E) lipoma
    F) hemangioblastoma-5%
    G) neuroma (very rare intramedullary)
    H) syringomyelia (not neoplastic)

Intradural Extramedullary Spinal Cord Tumors

Spinal meningiomas

Epidemiology:

- Peak age: 50–70 years.
- Female:male ratio =4:1 overall, 1:1 in the lumbar region.
- 82% Thoracic, 15% cervical, 2% lumbar.
- 90% are completely intradural, 5% are extradural, and 5% both
- Intra- and extra-dural :68% are lateral to the spinal cord, 18% posterior, 15% anterior.

Spinal Meningiomas

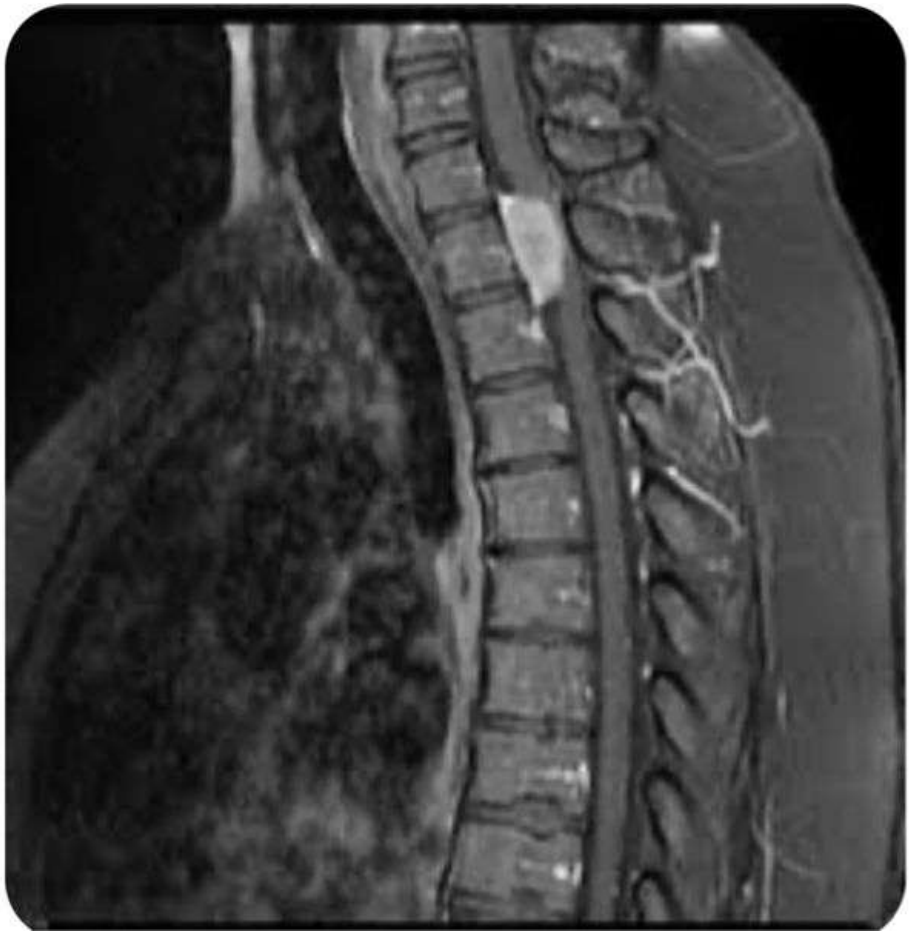
Clinical Findings:

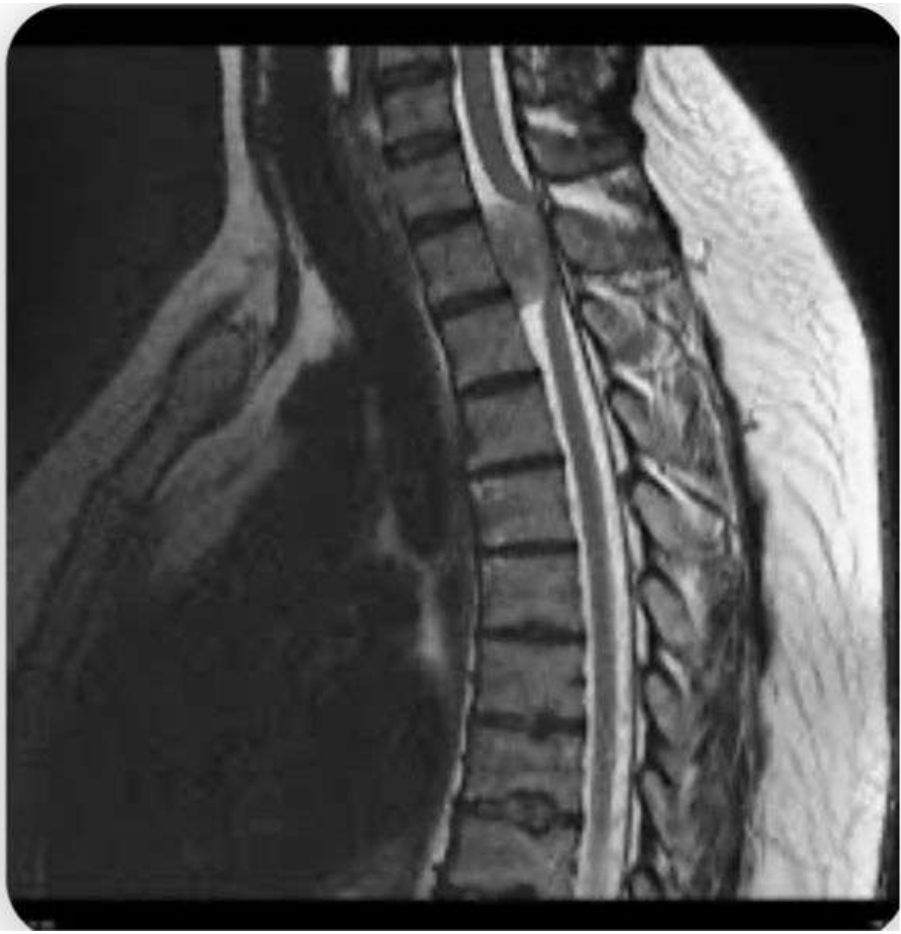
1. **motor**
    a) pyramidal signs only: 26%
    b) walks with aid: 41%
    c) antigravity strength: 17%
    d) flexion-extension with gravity removed: 6%
    e) paralysis: 9%

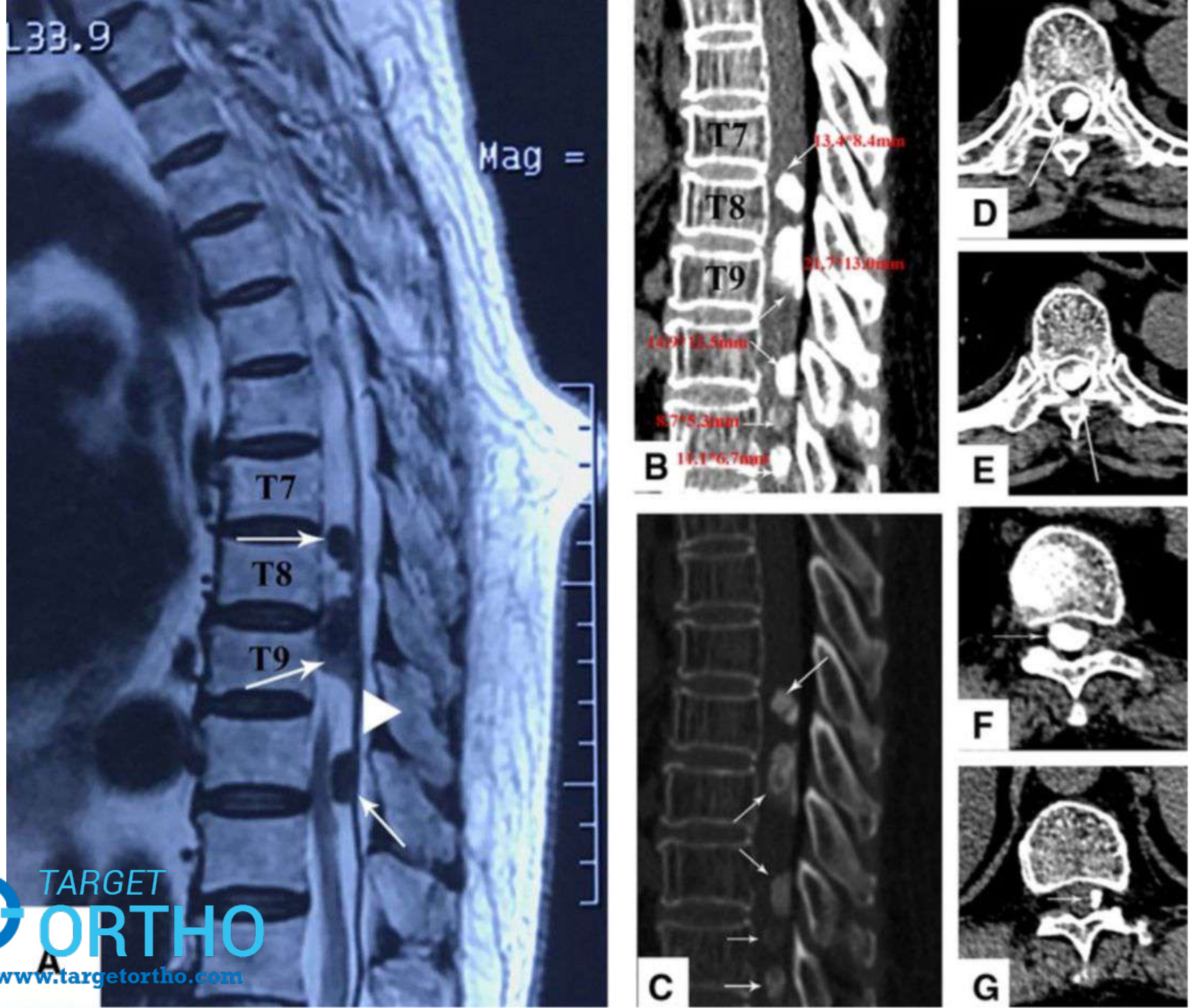
2. **sensory**
    a) radicular: 7%
    b) long tract: 90%

3. **sphincter deficit**: 51%

4. Meningioma during pregnancy- Progestrone

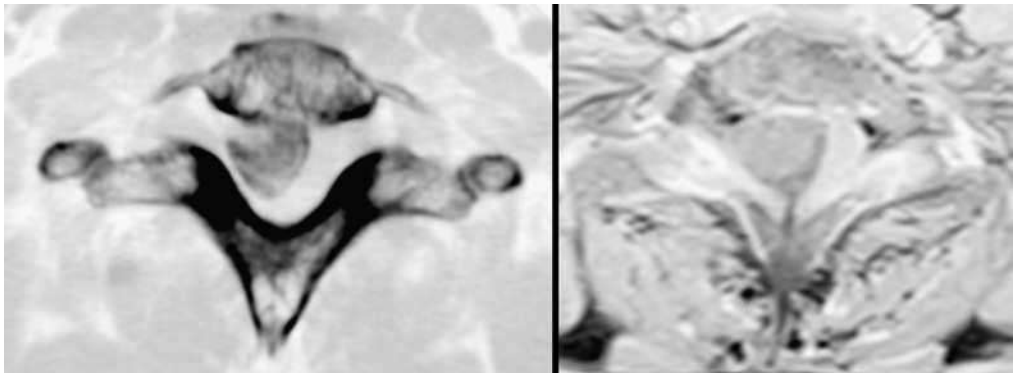






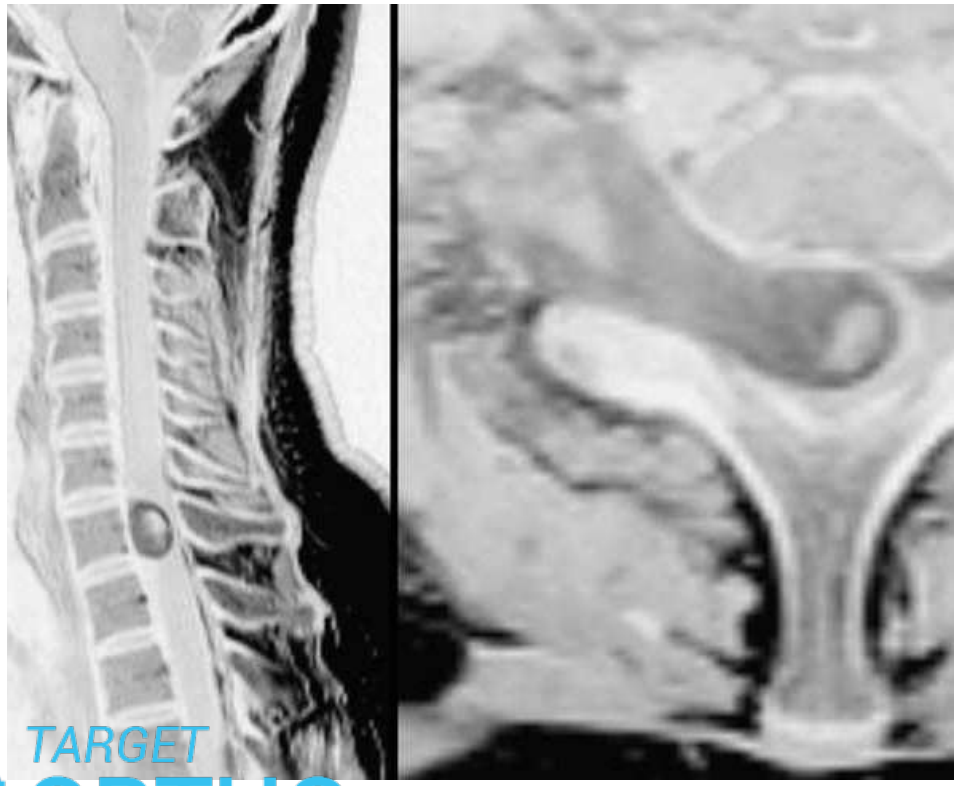
Outcome

Recurrence rate with complete excision is 7% with a minimum of 6 years follow-up (relapses occurred from 4 to 17 years post- op).



Solero CL, Fornari M, Giombini S, Lasio G, Oliveri G, Cimino C, Pluchino F. Spinal meningiomas: review of 174 operated cases. Neurosurgery. 1989; 25:153–160

Spinal Schwannomas



•**General Information:**

- Incidence:
  0.3–0.4/100,000/yr.
  Most occur sporadically and are solitary,
- may also be associated with neurofibromatosis primarily type 2 (NF2), but can occur with type 1.

- Schwannomas
- 85%
- NF2
- MRI- Cystic region
- Easily seperated from nerve root

- Neurofibromas
- 15%
- NF 1
- solid
- Nerve root sacrifice

Key Concepts

- Slow growing benign tumors
- Most (75%) arise from the dorsal (sensory) rootlets
- Early symptoms are often radicular
- Recurrence is rare after total excision (except in neurofibromatosis)

Configurations

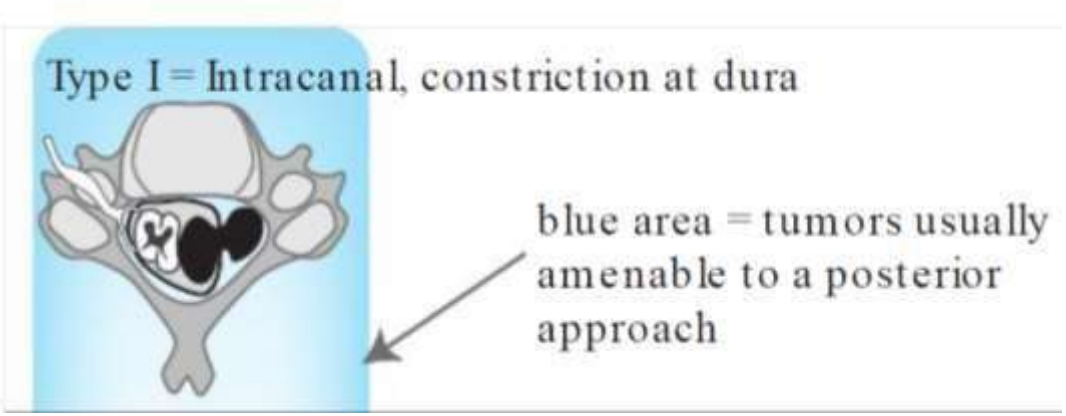
- Most are entirely intradural.
- 8–32% may be completely extradural
- 1–19% are a combination,
- 6–23% are dumbbell
- 1% are intramedullary.

Conti P, Pansini G, Mouchaty H, Capuano C, Conti R. Spinal neurinomas: retrospect ive analysis and longterm outcome of 179 consecutively operated cases and review of the literature. Surg Neurol. 2004; 61:34–43; discussion 44

Dumbbell Tumors

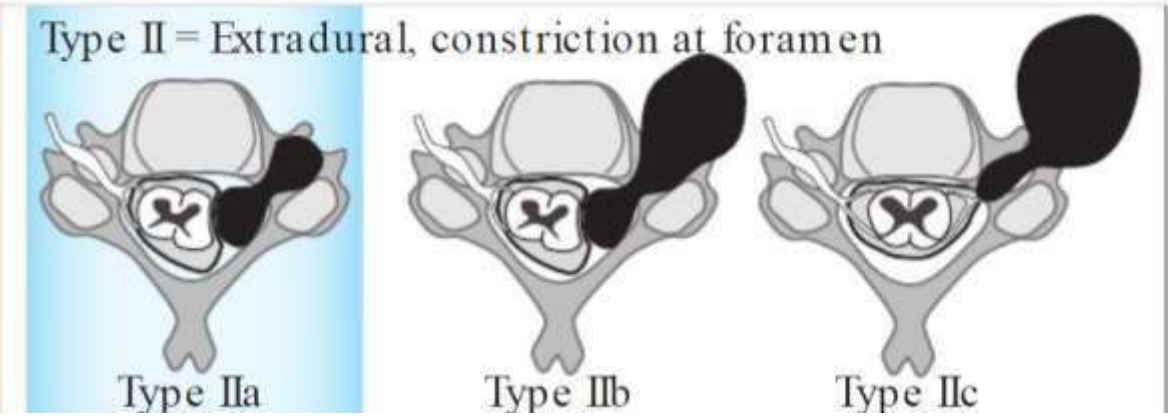
- Tumors that develop an “hourglass” shape as a result of an anatomic barrier encountered during growth.
- Not all dumbbell tumors are schwannomas, e.G. Neuroblastoma
- Most have a contiguous intraspinal, foraminal (usually narrower) and extraforaminal components
- Widening of the neural foramen is a characteristic finding, can be recognized even on plain films.

Asazuma Classification System For Dumbbell Spinal Schwannomas



Type I tumors are intradural and extradural and are restricted to the spinal canal. The constriction occurs at the dura.

Asazuma T, Toyama Y, Maruiwa H, Fujimura Y, Hirabayashi K. Surgical strategy for cervical dumbbell tumors based on a three-dimensional classification. Spine. 2004; 29:E10–E14



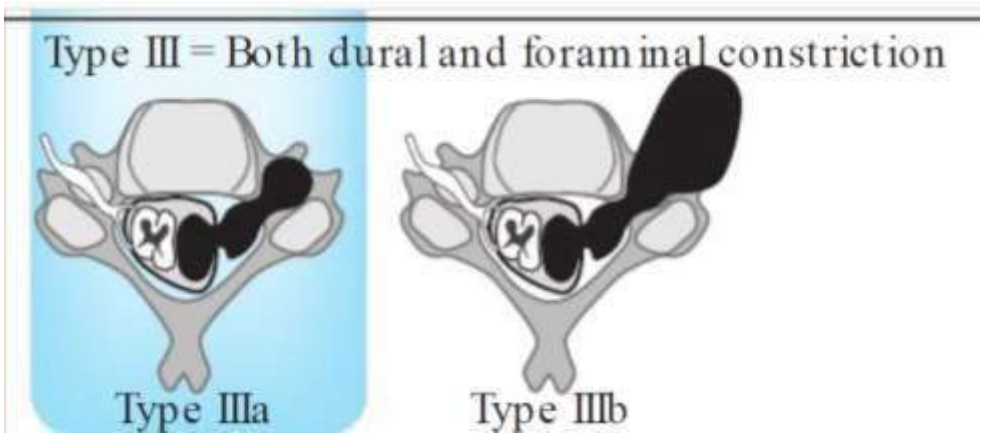
Type II are all extradural, and are subclassified as:

IIa do not extend beyond the neural foramen,

IIb = inside spinal canal + paravertebral

IIc = foraminal + paravertebral.

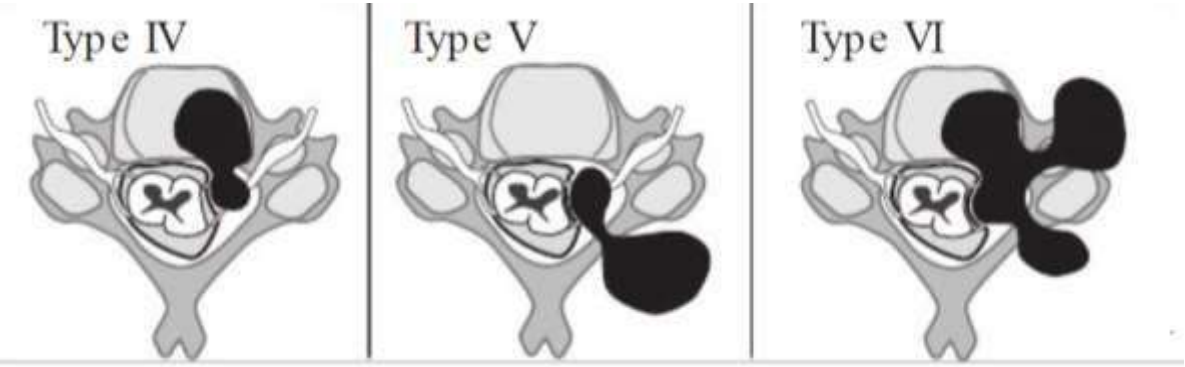
Asazuma T, Toyama Y, Maruiwa H, Fujimura Y, Hirabayashi K. Surgical strategy for cervical dumbbell tumors based on a three-dimensional classification. Spine. 2004; 29:E10–E14



Type IIIa are intradural and extradural foraminal

IIIb are intradural and extradural paravertebral.

Asazuma T, Toyama Y, Maruiwa H, Fujimura Y, Hirabayashi K. Surgical strategy for cervical dumbbell tumors based on a three-dimensional classification. Spine. 2004; 29:E10–E14



Type IV are extradural and intravertebral.

Type V are extradural and extralaminar with laminar invasion.

Type VI show multidirectional bone erosions

Asazuma T, Toyama Y, Maruiwa H, Fujimura Y, Hirabayashi K. Surgical strategy for cervical dumbbell tumors based on a three-dimensional classification. Spine. 2004; 29:E10–E14

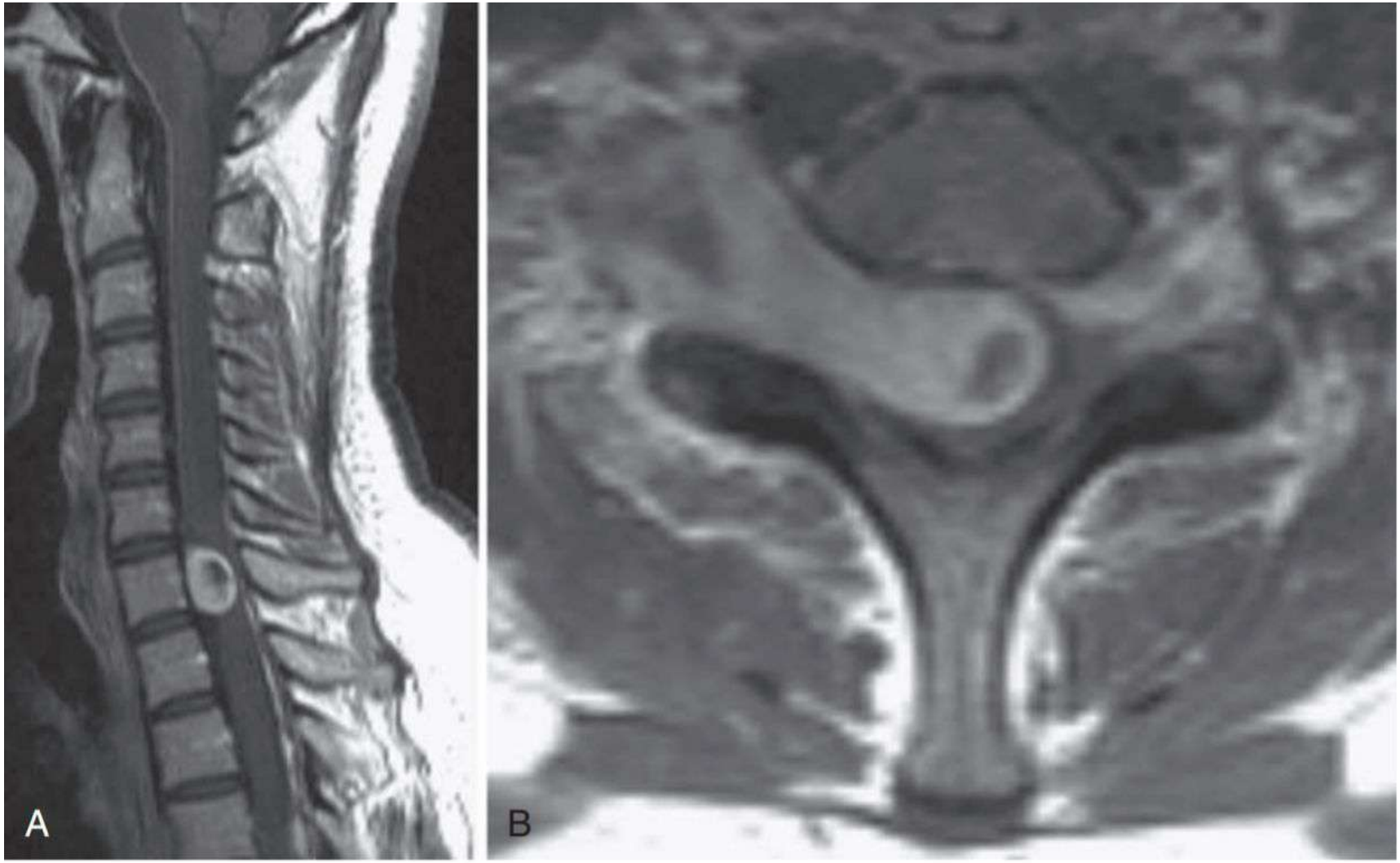


FIG. 90.2 (A) Sagittal T1-weighted gadolinium-enhanced magnetic resonance imaging (MRI) scan showing heterogeneously enhancing, well-demarcated, extradural schwannoma. (B) Axial T1-weighted gadolinium-enhanced MRI of the same cervical schwannoma showing dumbbell-shaped tumor extending through the right neural foramen.

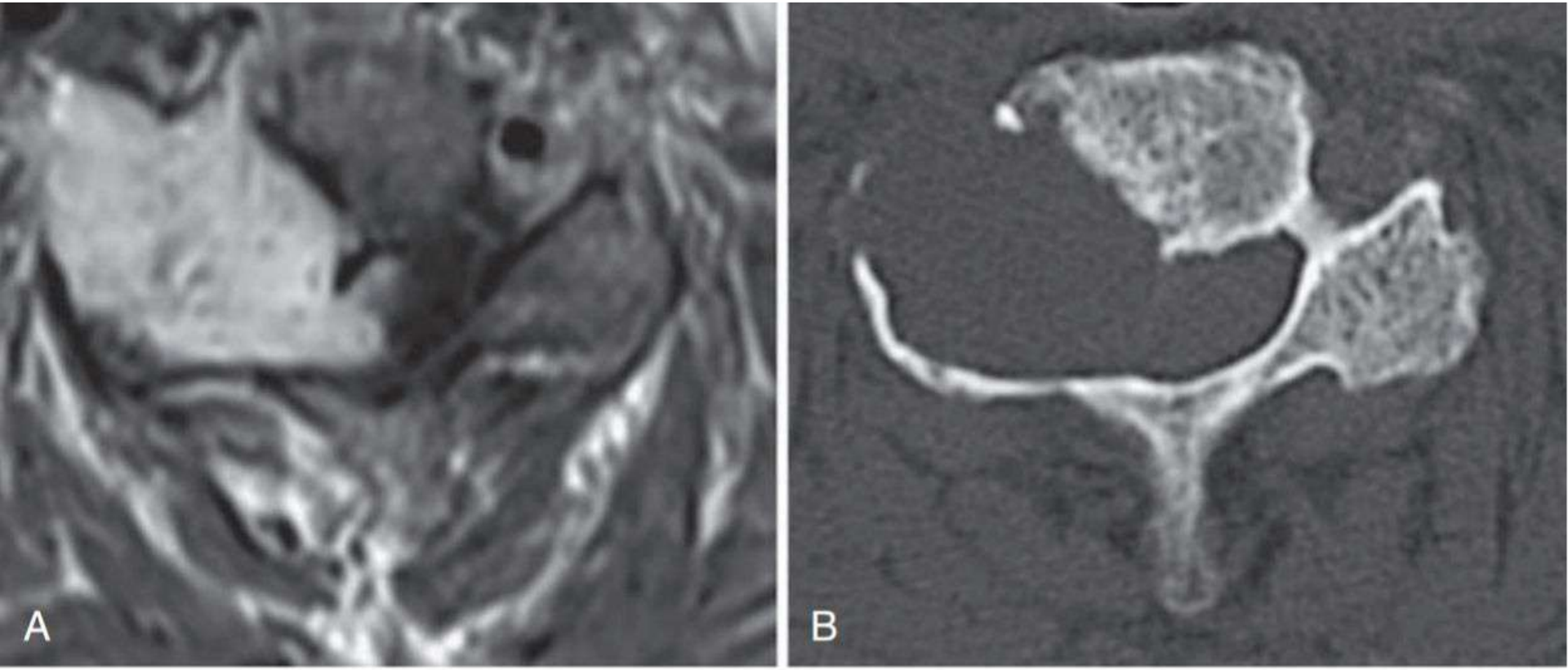


FIG. 90.3 (A) Axial T1-weighted gadolinium-enhanced magnetic resonance image of a malignant neurofibroma expanding the right neural foramen and extending into the extraspinal space. (B) Axial computed tomography of the same mass showing the extensive bony erosion caused by the tumor.

Clinical Features:

- Patients typically present with local pain.
- Neurologic deficits develop late. Tumors may cause radiculopathy (from nerve root compression),
- Myelopathy (from spinal cord compression), radiculomyelopathy (from compression of both), or cauda
- Equina syndrome (for tumors below conus medullaris).

Surgical Approaches

Posterior approaches:

- Types I, IIa IIIa, some upper cervical IIIb and some VI are generally amenable to a posterior approach.

- IIa & IIIa usually require total facetectomy for complete removal.
- Reconstruction with instrumentation may be needed if substantial posterior disruption occurs

Asazuma T, Toyama Y, Maruiwa H, Fujimura Y, Hirabayashi K. Surgical strategy for cervical dumbbell tumors based on a three-dimensional classification. Spine. 2004; 29:E10–E14

- **Anterior and combined anterior/posterior approaches**: combined approach for Type IIb, IIc and IIIb lesions where the extraforaminal extension is large (viz. beyond the vertebral arteries).

- Reconstruction with instrumentation was required for some tumors (≈ 10% of all patients treated) which were type IV (2 pat ients), IIIb (1 pt) and VI(1 pt).

Asazuma T, Toyama Y, Maruiwa H, Fujimura Y, Hirabayashi K. Surgical strategy for cervical dumbbell tumors based on a three-dimensional classification. Spine. 2004; 29:E10–E14

Nerve sacrifice

- It is usually possible to preserve some fascicles of the nerve root, although sometimes section of the
- Entire nerve root is required. New deficits may not occur since involved fascicles are often nonfunctional, and adjacent roots may compensate.
- The risk for motor deficit is higher for neurofibromas than for schwannomas, for cervical vs. Lumbar tumors, and for cervical tumors with extradural extension.

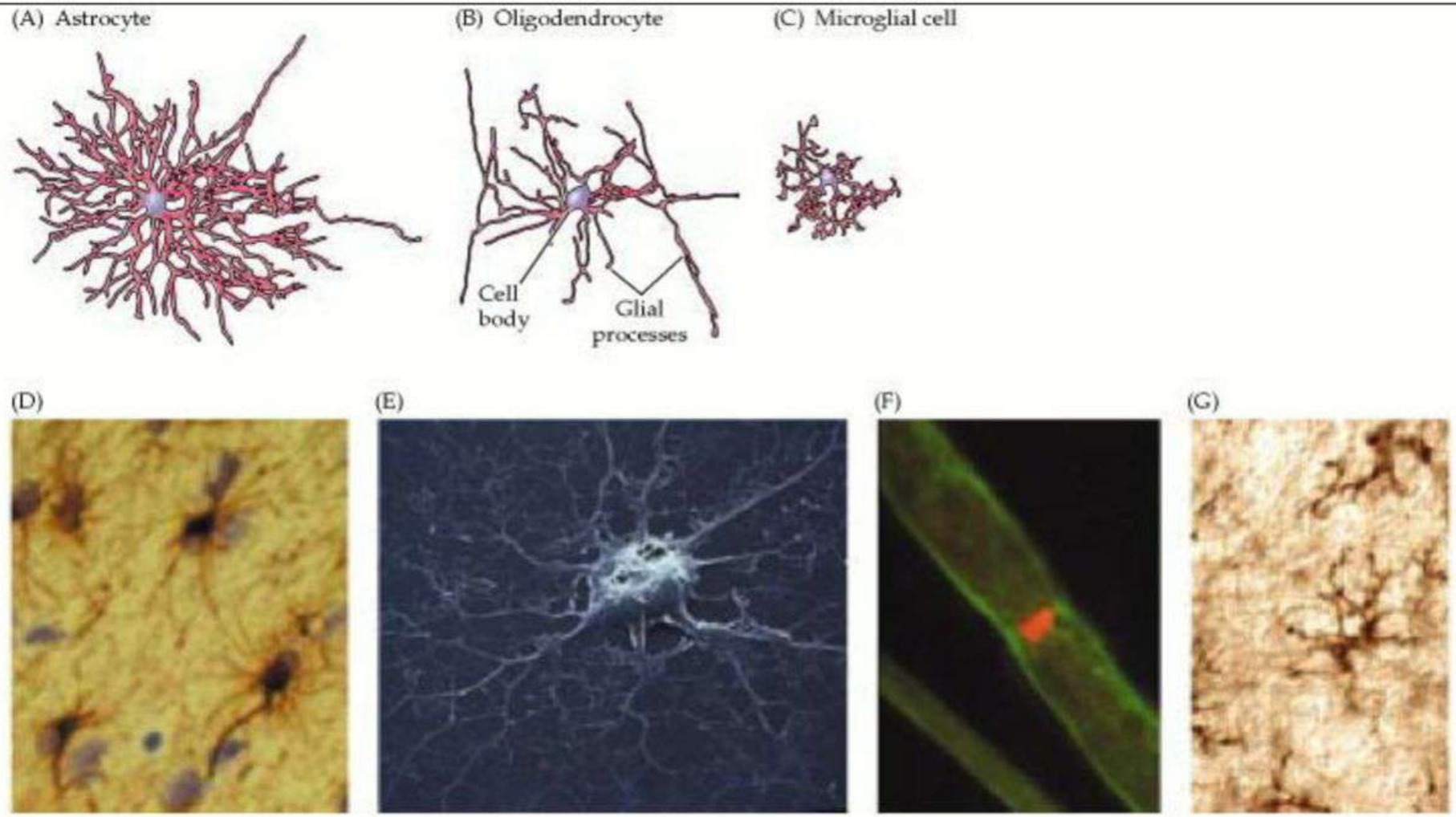
Outcome

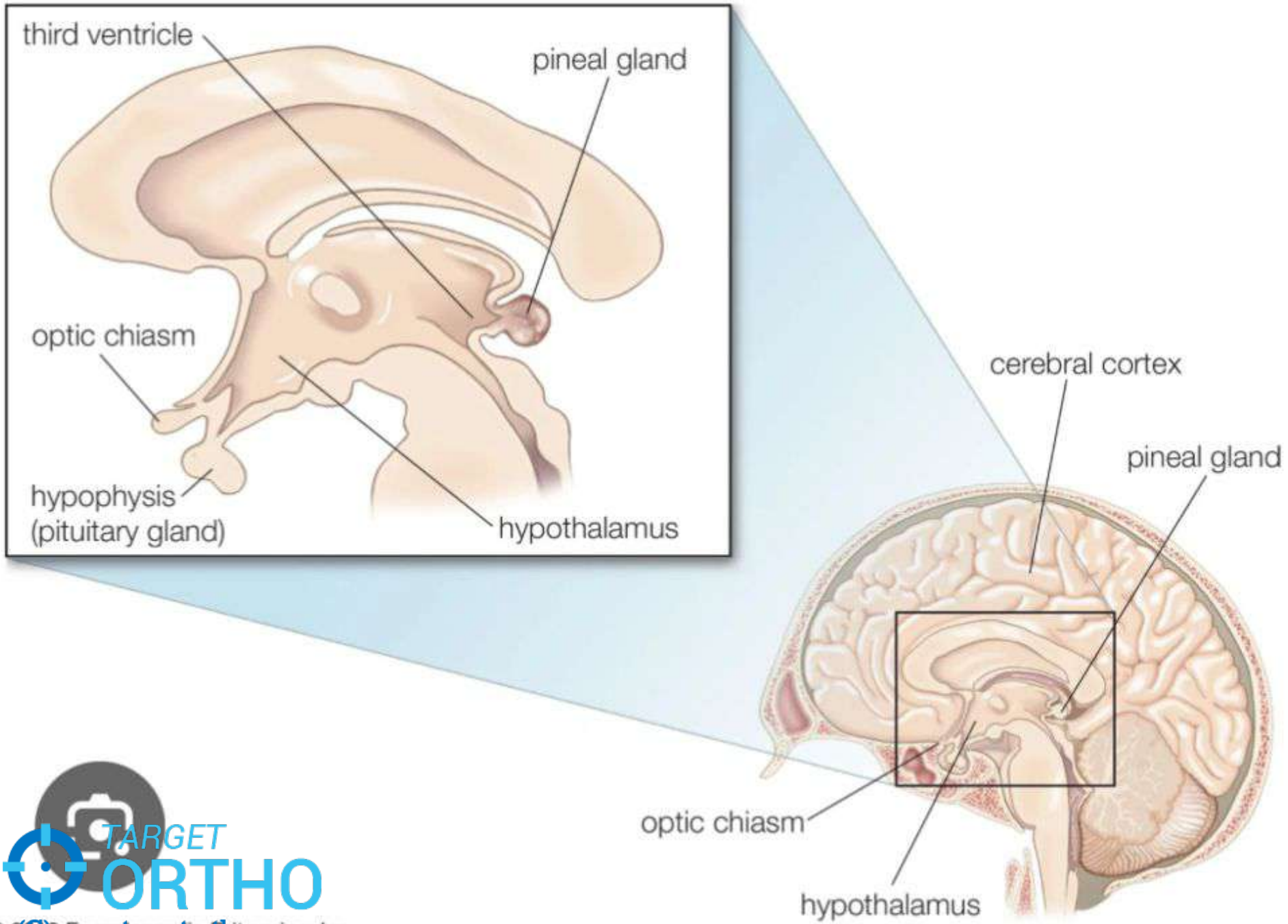
- Recurrence is rare following gross total excision, except in the setting of NF2

Asazuma T, Toyama Y, Maruiwa H, Fujimura Y, Hirabayashi K. Surgical strategy for cervical dumbbell tumors based on a three-dimensional classification. Spine. 2004; 29:E10–E14

Intramedullary spinal cord tumors

- In pediatrics, astrocytoma and ependymoma constitute 90% of intramedullary spinal cord tumors (IMSCT).



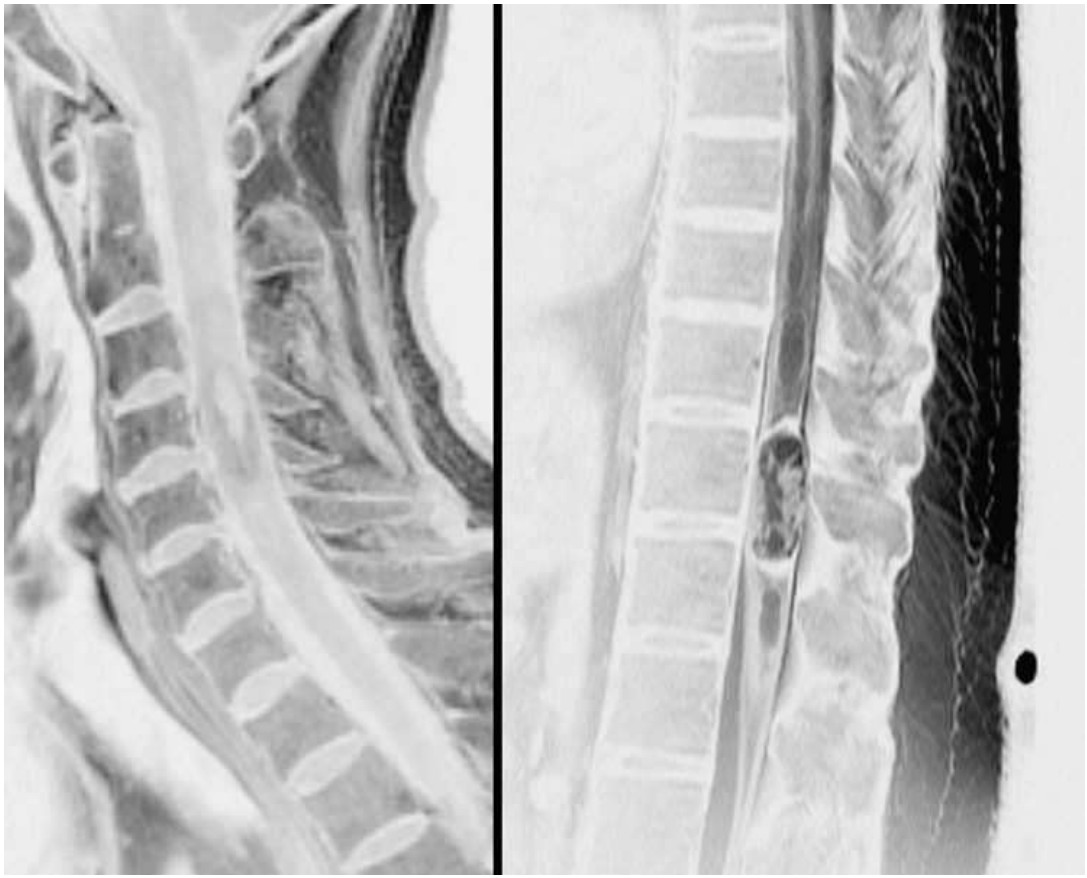


Ependymoma

•General Information:

- The most common glioma of the lower spinal cord, conus and filum (below). Slow-growing. Benign

- Slight male predominance; slight peak in 3rd to 6th decade. Over 50% in filum, next most common location is cervical.



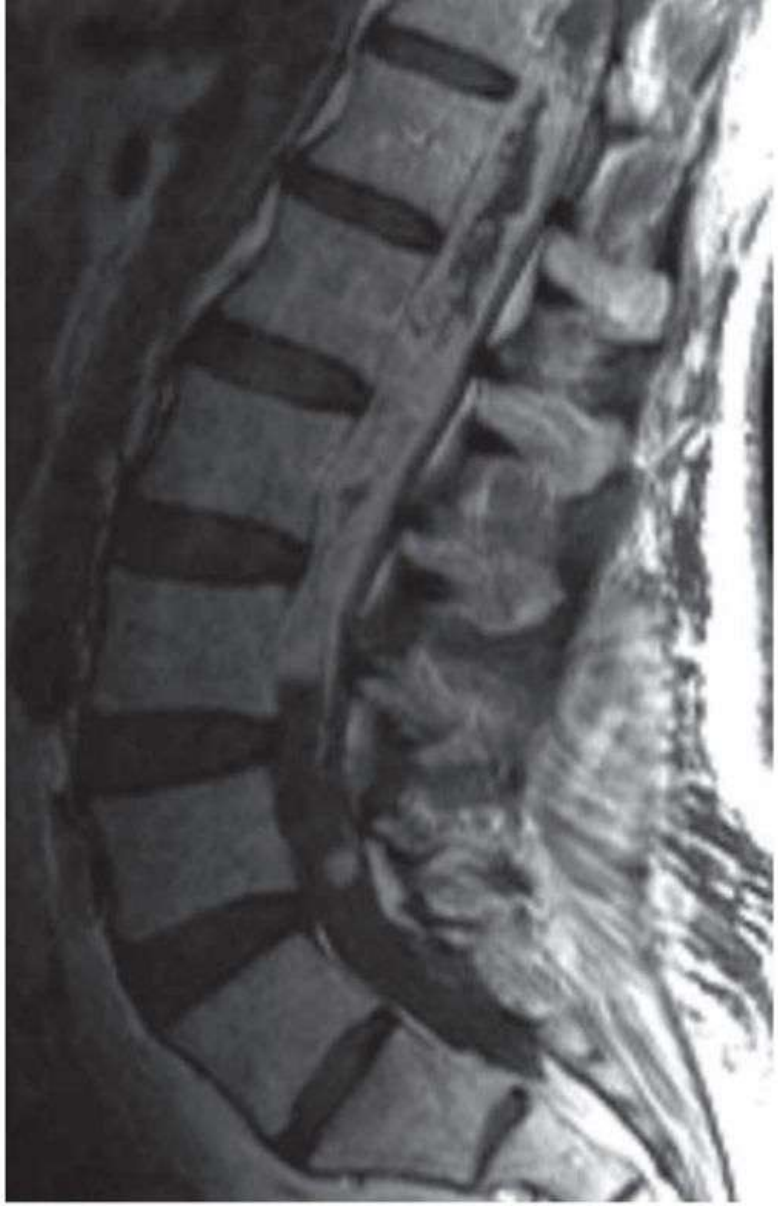


FIG. 90.4 Sagittal T1-enhanced magnetic resonance image demonstrating a large filum terminale ependymoma filling the lower thoracic–upper lumbar spinal canal and encasing the cauda equina. Note the more inferior tumor nodule at the L4 level junction representing a drop metastasis.

- **Histologically:** papillary, cellular, epithelial, or mixed (in filum, myxopapillary ependymoma is most common, see below).

- Cystic degeneration in 46%. May expand spinal canal in filum. Usually encapsulated and minimally vascular (papillary: may be highly vascular; may cause SAH).

- Symptoms present > 1 yr prior to diagnosis in 82% of cases.28

Key concepts

- The most common glioma of lower cord, conus and filum (most ependymomas in conus and filum are myxopapillary ependymomas). More common in adults
- Evaluation: includes imaging the entire neuraxis (usually with enhanced MRI: cervical, thoracic, lumbar & brain) because of potential for seeding through CSF
- Associated cysts are common
- **Treatment**: surgical excision (most are encapsulated)

- Surgical removal of filum tumors consists of coagulating and dividing the filum terminale just above and below the lesion - and excising it in total. The filum is first cut above the lesion to prevent retraction upwards.

Distinguishing Features Of The Filum Terminale Intraoperatively

- The filum is differentiated from nerve roots by presence of characteristic squiggly vessel on surface of filum.

- the filum has a distinctively whiter appearance than the nerve roots, and ligamentous-like strands can be seen running through it.

Astrocytoma

- Uncommon in first year.
- Peak: 3rd – 5th decade.
- Male:female =1.5:1.
- The ratio of low-grade:high grade =3:1 in all ages.
- Occurs at all levels, thoracic most common, then cervical.

  38% are cystic; cyst fluid usually has high protein

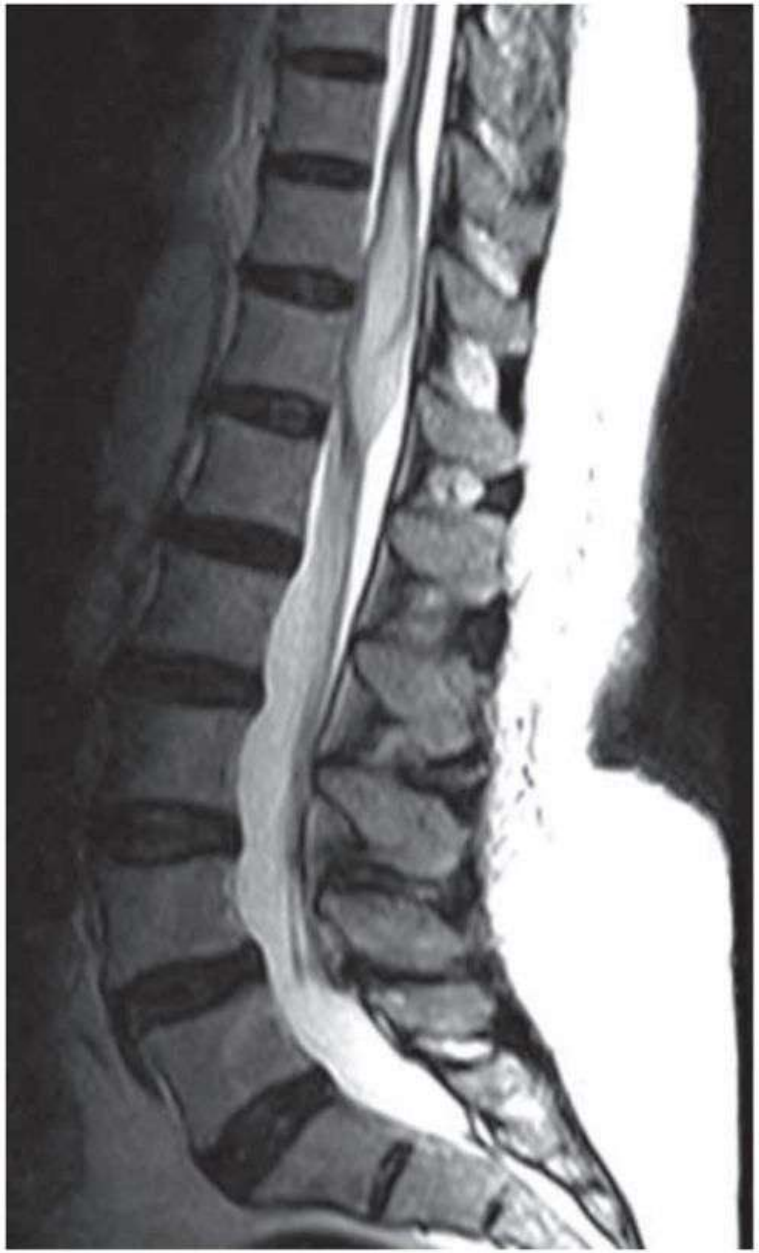


FIG. 90.7 Sagittal T2-weighted magnetic resonance image showing an astrocytoma with high signal expanding the spinal cord and conus medullaris.

Dermoid and epidermoid

- Epidermoids are rare before late childhood. Slight female predominance.
- Cervical and upper thoracic rare; conus common.
- Usually ID-EM, but conus/cauda equina may have IM component (completely IM lesions rare).

Lipoma

- May occur in conjunction with spinal dysraphism
- Peak occurrence: 2nd, 3rd and 5th decade.
- Technically hamartomas.
- No sex predominance.
- Cervicothoracic region is the most common location. NB: unlike other IMSCT's, most common symptom is ascending mono- or para-paresis (c.f. pain).
- Sphincter disturbance is common with low lesions. Local subcutaneous masses or dimples are frequent.

Hemangioblastoma

- Usually non-infiltrating, well demarcated, may have cystic caps.
- 33% have von Hippel-Lindau disease
- Requires microsurgical approach similar to AVM, possibly with intraoperative hypotension.

Metastases

- Most spinal mets are extradural, Intramedullary metastases are rare, accounting for 3.4% of symptomatic metastatic spinal cord lesions.

- Primaries include: small-cell lung Ca, breast Ca, malignant melanoma, lymphoma and colon Ca.

- Ca rarely presents first as an intramedullary spinal met.

Presentation

1. **Pain:**
   - The most common complaint.
   - Almost always present in filum tumors (exception: lipomas).

Possible Pain Patterns:

a)radicular: increases with Valsalva maneuver and spine movement. Suspect SCT if dermatome is unusual for disk herniation

b) local: stiff neck or back, Valsalva maneuver increases pain. Pain during recumbency (“nocturnal pain”) is classic for SCT

c)medullary (as in syrinx): oppressive, burning, dysesthetic, non-radicular, often bilateral, unaffected by Valsalva maneuver

2. Motor Disturbances

a)weakness is 2nd or 3rd most common complaint. Usually follows sensory symptoms temporally

b) children present most frequently with gait disturbances

c) syringomyelic syndrome: suggests IMSCT. Findings: UE segmental weakness, decreased DTR, dissociative anesthesia

d)long-tract involvement →clumsiness and ataxia (distinct from weakness)

e) atrophy, muscle twitches, fasciculations

3. Non-painful Sensory Disturbances

a) Dissociated sensory loss: decreased pain and temperature, preserved light touch, as in brown-séquard syndrome.

B) paresthesias: either radicular or “medullary” distribution

4. Sphincter Disturbances

a) usually urogenital (anal less common) →difficulty evacuating, retention, incontinence, and impotence. Early in conus/cauda equina lesions, especially lipomas (pain not prominent)

b) sphincter dysfunction common in age <1 yr due to frequency of lumbosacral lesions (dermoids, epidermoids, etc.)

5. Miscellaneous Symptoms:

a) scoliosis or torticollis

b) SAH

c) visible mass over spine

Time course of symptoms

- Onset usually insidious, but abruptness occurs (benign lesions in children occasionally progress in hours)

- The onset is often erroneously attributed to coincidental injury.

- Temporal progression has been divided into 4 stages:
  1. Pain only (neuralgic)
  2. Brown-séquard syndrome
  3. Incomplete transectional dysfunction
  4. Complete transectional dysfunction

Diagnosis

- It is usually diffcult to distinguish IMSCT, ID-EM and ED on clinical
  grounds.

- Schwannomas often start with radicular symptoms that later progress to cord involvement.

- Most IMSCTs are located posteriorly in cord which may cause sensory findings to predominate early.

Diagnostic Studies

MRI: mainstay of diagnosis.

Ependymomas enhance intensely and are often associated with hemorrhage and cysts. Cord edema may mimic a cyst.

Plain radiographs:

- Vertebral body destruction
- Enlarged intervertebral foramina
- Increases in interpedicular distances suggests ED SCT.

Lumbar Puncture:

- Elevated protein is the most common abnormality seen in ≈ 95%.
- Glucose is normal except with meningeal tumor.
- SCT can cause complete block, indicated by:
  - Froin's syndrome: clotting (due to fibrinogen) and xanthochromia of CSF
  - Queckenstedt's test (failure of jugular vein compression to increase CSF pressure, which it normally does in the absence of block)
  - Barrier to flow of myelographic contrast media

CT SCAN

- Some IMSCTs enhance with IV contrast. Myelo-CT distinguishes IMSCT from ID-EM (poor in differentiating IMSCT subtypes).

Management

General Information

- Asymptomatic lesions may be followed since there is significant risk of neurologic deficit with surgery.

- For symptomatic lesions, surgery should be performed as soon as possible

Astrocytomas

- **For low grade lesions**, if a plane can be developed between the tumor and spinal cord an attempt at total excision is an option.

- **For high grade** astrocytomas or for low-grade astrocytomas without a plane of separation, biopsy alone or biopsy plus limited excision is recommended.

- For high-grade lesions, post-op RTX (± chemotherapy) is recommended.

- RTX is not supported following radical resection of low grade gliomas.

Ependymomas

- An attempt at gross total removal should be attempted. XRT is not recommended following gross total removal.

Adjunctive options include:

a) intraoperative spinal cord monitoring (SSEP, and motor evoked potentials (MEPs)

b) intraoperative ultrasound: also controversial, favored by some experts. Astrocytomas are usually iso-echoic with spinal cord, whereas ependymomas are usually hyperechoic

A myelotomy is performed either in the midline or just to one side of the dorsal midline to avoid the posteromedian vein.

Tumors may cause distortion and displacement of the midline – look for dorsal root entry zones on both sides to identify the midline as the midpoint between root entry zones

Either laser or ultrasonic aspiration (USA) are used to debulk tumor from within until the glial tumor interface is reached. Charring from laser may make it more difficult to recognize the glial/ tumor interface than USA, and the laser tends to be slower when debulking larger tumors

Watertight dural closure is critical

Key Concepts In Surgical Removal Of IMSCT

, imscts should be debulked from within using ultrasonic aspirator or laser (to avoid manipulation of neural tissue), and no attempt should be made initially to develop a plane between tumor and spinal cord (even for ependymomas, which of the 3 most common imscts is the only one that actually has such a plane)

If meps are monitored: it is suggested that tumor removal should be discontinued if the amplitudes drop to ≤ 50% of baseline

Prognosis

No well-designed studies give long term functional results with microsurgery, laser and radiotherapy.

Better results occur with lesser initial deficits. Recurrence depends on totality of removal, and on growth pattern of the specific tumor.