EANS/UEMS European examination in neurosurgery

Variants of questions with answers (compilation - Vyacheslav S. Botev, Department of Neurosurgery, M.Gorky Donetsk National Medical University)

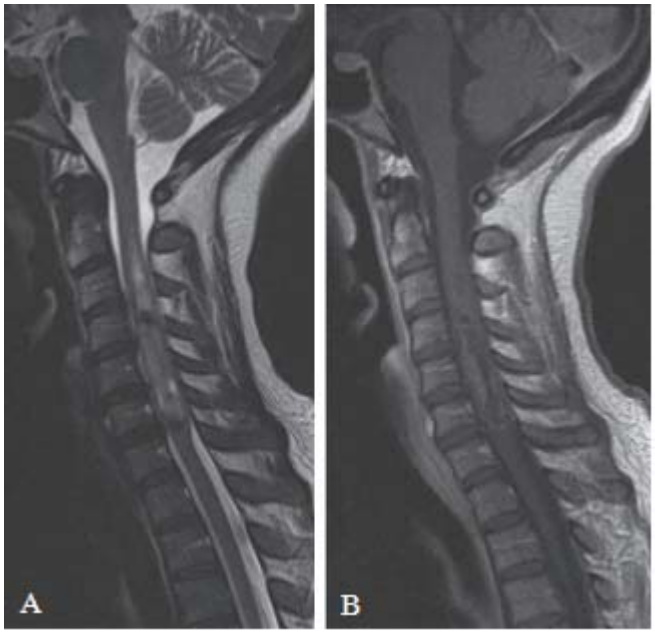
CASE HISTORIES IN SPINE NEUROSURGERY

Case 1

A 37-year-old woman presents with a 2-year history of intermittent tingling of the left hand that has progressively gotten worse and is now also involving the left lower extremity. There was no loss of bowel or bladder function and no headaches. The medical history is remarkable for mitral valve prolapse and two breast biopsies for benign lesions. Neurologic examination was completely normal.

Questions

1. What is the differential diagnosis?
2. What studies do you need to order?
3. The study you ordered is shown below. Describe the findings and the current differential diagnosis.

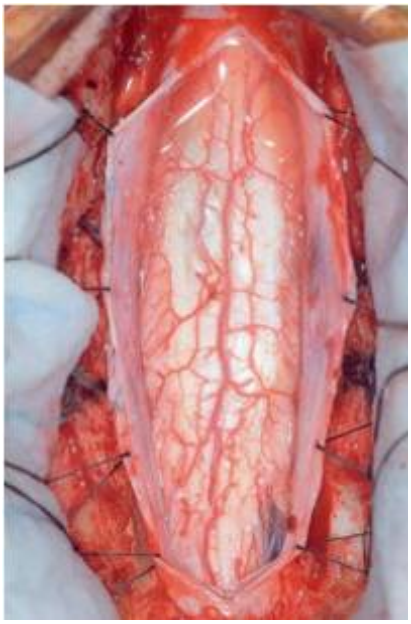


4. MRI of the cord identifies a mass. What finding suggests the tumor is an astrocytoma rather than an ependymoma?

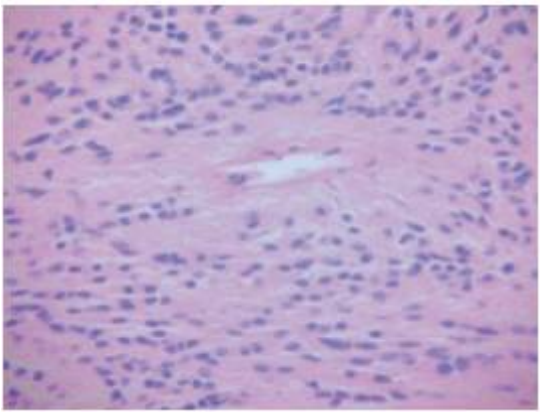
A. Polar cyst
B. Hemorrhage at the cap

C. Seven vertebral segments in length
D. Mixed enhancement
E. Adult patient

5. What treatment would you recommend to the patient? What are the risks?
6. What equipment do you need? Describe the surgical procedure. Figure below is an intraoperative view; how would you proceed?



7. The pathology is shown below. What is your diagnosis?



8. What is the most common spinal location for cord ependymomas?
9. Can an ependymoma present in the extramedullary space?

10. Is it necessary to extend the laminectomy far enough to expose all cysts associated with the lesion?

Case 2

A 39-year-old man presents to your office 2 weeks after he has undergone a hemilaminectomy at L3–L4 for degenerative spinal stenosis. He has a previous history of L4–5 and L5–S1 posterolateral fusion with pedicle screw fixation. He presents now with a 2-day history of severe back pain radiating to the buttocks bilaterally. There is mild erythema around the recent surgical wound. There are no defiits or pain in his lower extremities. Magnetic resonance imaging (MRI) of the lumbar spine is obtained:

Questions

1. Interpret the MRI and give a differential diagnosis.
2. What are the characteristics of epidural abscess on MRI?
3. What is the most common location of a spinal epidural abscess?
4. What are the risk factors for this condition?
5. What are the most common microorganisms involved in epidural abscesses in the immunocompetent patient?
6. What is the incidence of this condition in the postsurgical patient?

7. Which of the following are the *least likely* to be the origin of an epidural abscess?

A. Hematoma after trauma with subsequent degradation of products
B. Instrumentation
C. IV drug abuse
D. UTI in an immune competent patient
E. Psoas abscess

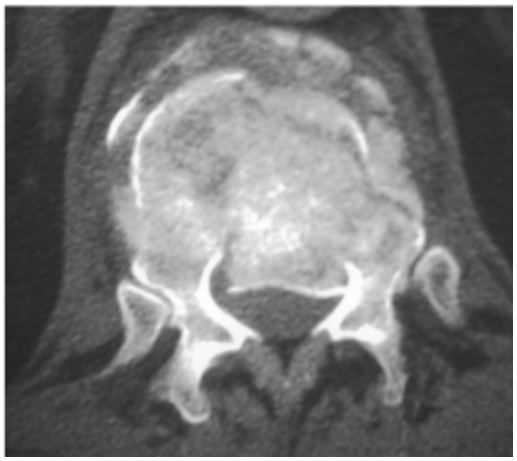
8. How do you manage this condition?

You decide to operate. You decompress the thecal sac and perform a sharp debridement of the wound. During surgery, you send cultures and are told that there are numerous Gram-positive cocci. Once down to the thecal sac, you notice that, for the most part, there is thick granulation tissue over the sac. As you tease this tissue off the dura, you get a dural tear.

9. How do you handle a dural tear in this specific setting?
10. Why do you not remove the instrumentation of the previous fusion?
After you decompress the thecal sac, you notice a piece of floating bone, you remove it, and you notice that it comprises the inferior facet joint of L3 that is detached and free floating (you did not detach it during your current decompression).
11. Why is the piece of facet detached?
12. What condition do you now worry about, given the detached facet, and how do you manage it?

Case 3

1. What is the mechanism of injury in the following spine fracture (axial CT scan)

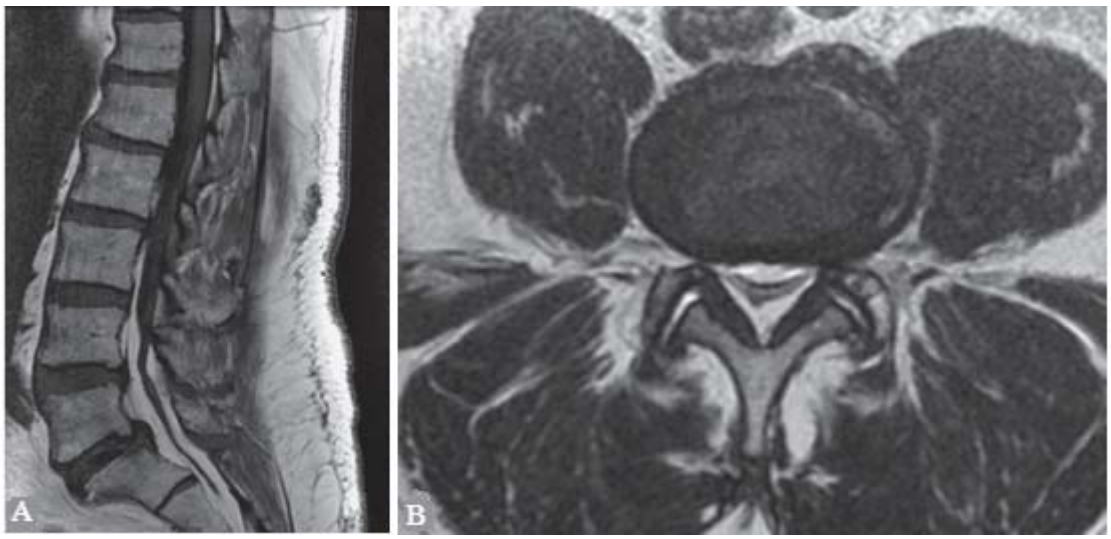


A. Fracture-dislocation
B. Axial compression
C. Flexion-distraction
D. Hyperextension
E. None of the above

2. What is a burst fracture?
3. What are the indications for surgery in patients with burst fracture?
4. What is the difference between a compression fracture and a burst fracture?
5. What is the classification described by Denis?

Case 4

1. What is the most likely diagnosis in a patient with a long history of complicated asthma and the sagittal (A) and axial (B) T1WI MRI at the L4–5 level depicted below?



A. Eosinophilic granuloma
B. Burst fracture
C. Epidural hematoma
D. Epidural lipomatosis
E. Osteomyelitis

2. Is this abnormality more likely to affect males or females?
3. What finding on axial images has been described as pathognomonic of this abnormality when it involves the lumbar spine?
4. Name two other lesions in the differential diagnosis
5. Which contains more fatty tissue, the lumbar or the cervical epidural space?

Case 5

Questions 1 through 3 refer to the following scenario.

A 61-year-old man, who smokes five packs of cigarettes per day and has hypertension, had an abdominal aortic aneurysm repair 8 h ago. The surgery went very well, and there were no reported perioperative complications. Now the patient is unable to move his legs and states that they are “numb.” On examination, he has a flaccid paresis of both lower extremities and has impaired pinprick sensation to a T9 level bilaterally. Joint proprioception is normal.

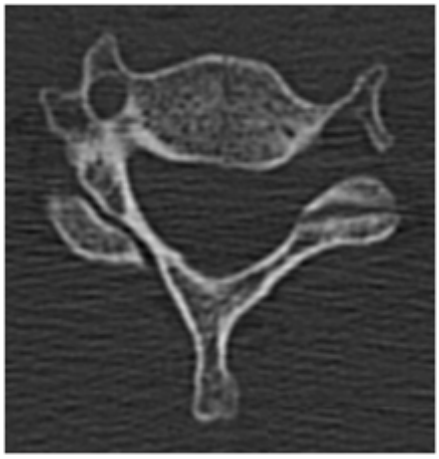
1. The most likely diagnosis in this case is
A. Cerebral stroke
B. Multiple sclerosis
C. Spinal cord infarct
D. Conversion disorder
E. Spinal cord compression

2. The artery of Adamkiewicz is an especially large spinal medullary artery supplementing the arterial blood supply to the spinal cord. Which of the following represents the most consistent location of this vessel?
A. C2–C5
B. C5–C8
C. T2–T8
D. T10–L1
E. L4–S4

3. Which of the following would you expect to find in this patient 6 months from now?
A. Fibrillations
B. Hyporeflexia
C. Fasciculations
D. Spastic paralysis
E. Flaccid paralysis

Case 6

This patient was struck on the back of the head by a heavy barrel. What is the mechanism of injury for the condition depicted in this axial CT scan?



A. Hyperflexion
B. Axial loading
C. Flexion/rotation
D. Hyperextension
E. Distraction

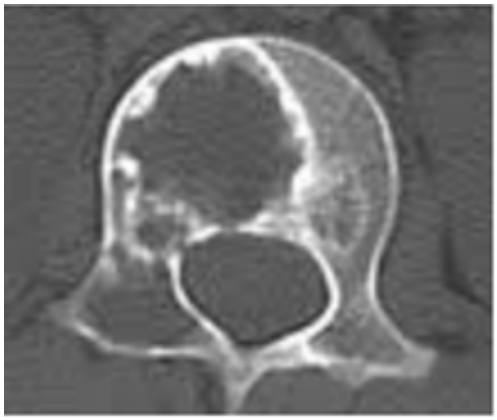
Case 7

What is the most likely diagnosis in an 8-year-old male without a history of trauma?

A. Hemangioma
B. Osteomyelitis
C. Eosinophilic granuloma
D. Giant cell tumor
E. Osteosarcoma

Case 8

A tumor-like lesion arises in bone but is not neoplastic in its cell of origin. Such lesions can cause local vertebral collapse and secondary neural injury.



1. Which of the following is a tumor-like lesion?
A. Lymphoma
B. Epidermoid
C. Neurofibroma
D. Neuroblastoma
E. Aneurysmal bone cyst (ABC)

2. This lesion most frequently involves which part of the vertebra and which region of the spine?

Case 9

Which of the following is associated with Winking Owl Sign?

A. Metastasis
B. Hemangioma
C. Osteomyelitis
D. Aneurysmal bone cyst
E. Fibrous dysplasia

Case 10

A 38-year-old woman presents to the emergency department with 2 weeks of back pain radiating down her left leg. On the previous day, she noticed difficulty passing urine and partial numbness of her buttocks.

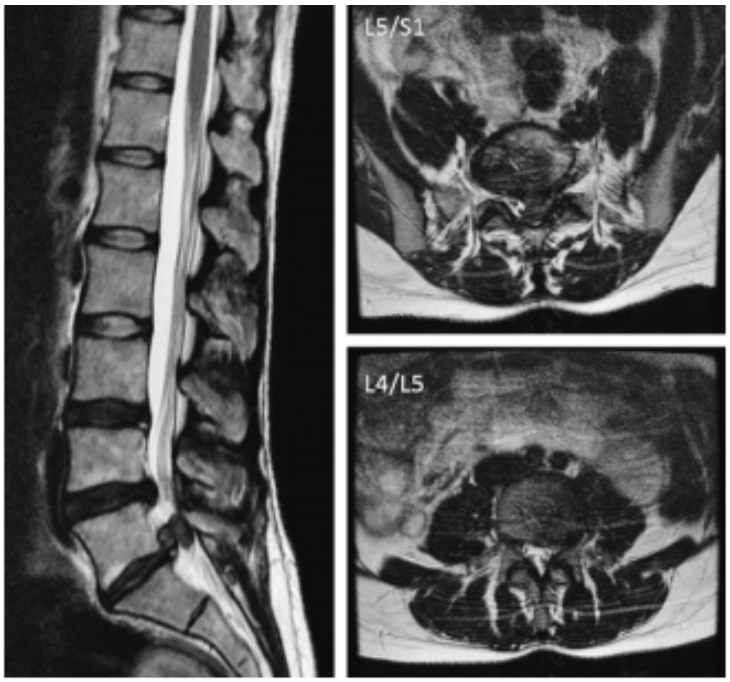
1. What is the diagnosis?
2. What are the important questions to ask?
3. On examination there was some weakness of plantar flexion and hip extension on the left. The left ankle reflex was absent. Pinprick sensation was preserved throughout the lower limbs but sensation was absent in the perineum.

However, when more pressure was applied to the pin, the patient reported that she would feel the sharp pin in the perineum.

(a) Identify the level of the pathology.
(b) Are the clinical findings consistent with the suspected diagnosis?

4. What investigations are required? What would you do if the patient was claustrophobic and unable to tolerate an MRI scan?

5. The patient undergoes an MRI scan of the lumbar spine:



Describe the findings on the scan.
6. What is the management, and on what time scale?

Case 11

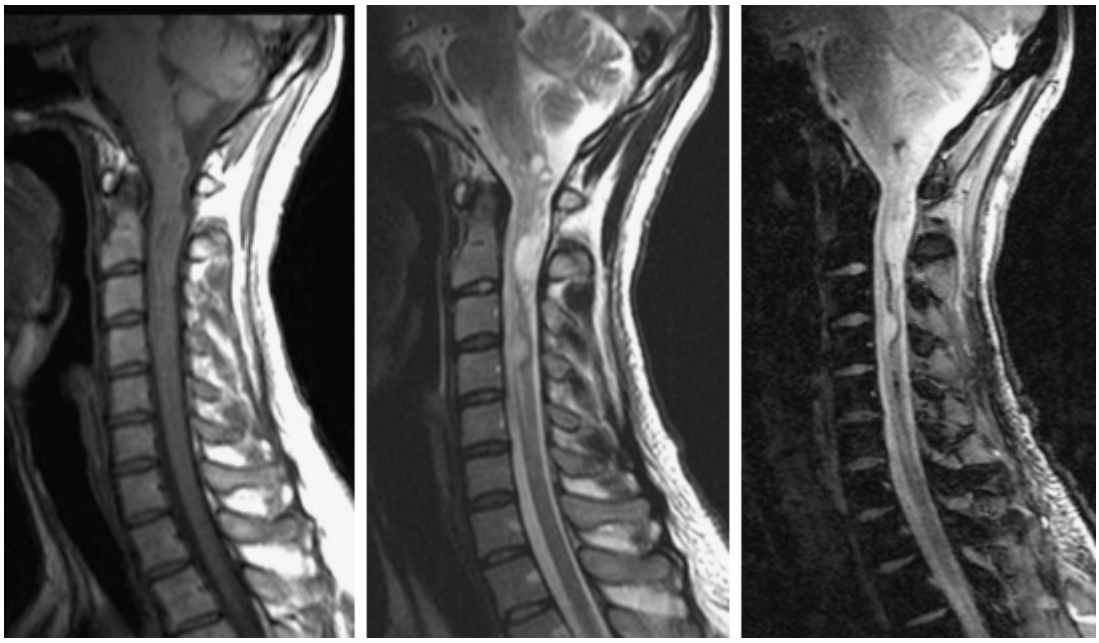
A 12-year-old girl is admitted by the pediatricians. She was previously healthy but recently she was noticed to be running less and tending to walk more slowly or limp at times. She began to complain of neck pain and has started to use her hands more slowly and deliberately. Her GP was concerned about Guillain-Barré syndrome and sent her to the local emergency department.

1. What features of the history are important and what is the differential diagnosis?

2. An MRI scan is done. Describe the labelled abnormality. What is the diagnosis?

3. What types of intramedullary tumors are there?
4. What is the initial management for this patient?
5. What is the role of surgery for intradural intramedullary tumors?
6. What is Weinstein-Boriani-Biagini (WBB) surgical staging system of spine tumors?
7. What are the most common approaches for spine tumors?

Case 12

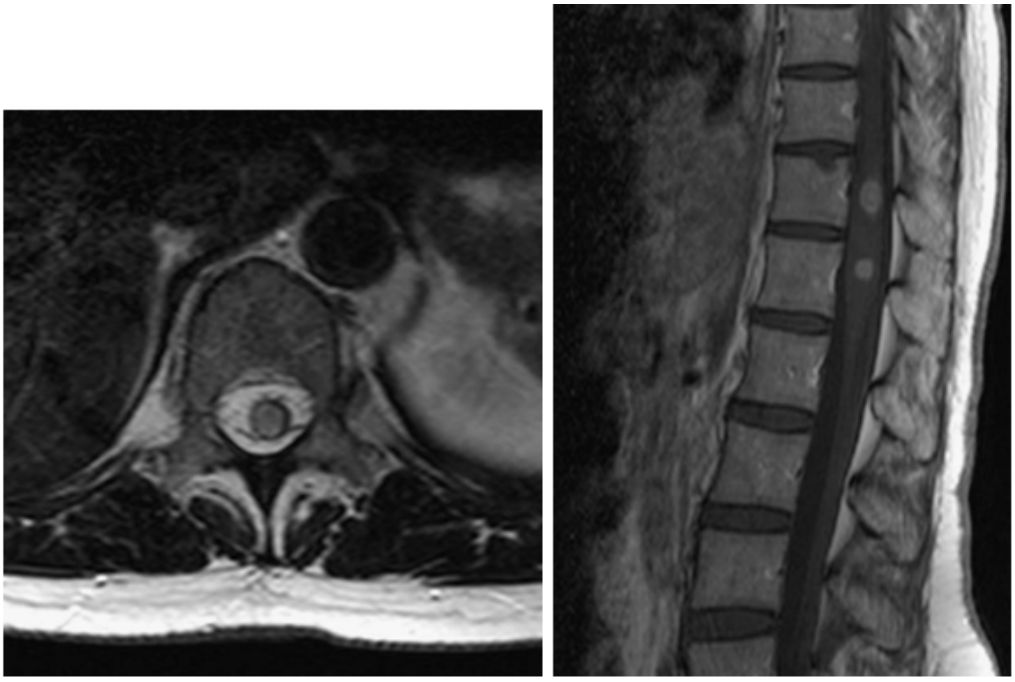


1. Based on the findings on the T1W and T2W spin-echo images and the T2W gradient-echo image, is this lesion intramedullary, intradural extramedullary, or extradural?
2. What is the most common tumor in this location in adults?
3. Name three intramedullary lesions that have been shown to cause superficial siderosis of the CNS.
4. Is this lesion, when small, more likely to cause symmetrical or asymmetrical enlargement of the cord?

Case 13

1. Does the cystic region of the cord at C6–C7 on the T2W and postcontrast T1W images represent an intratumoral cyst?
2. Do spinal ependymomas present earlier or later in life than intracranial ependymomas?
3. Is vigorous enhancement more likely with ependymoma or astrocytoma?
4. Approximately what percentage of ependymomas demonstrate associated cysts?

Case 14



1. Give a differential diagnosis for the findings on these postcontrast T1W images.
2. On the basis of autopsy results of patients with disseminated cancers, are the number of patients with brain metastases approximately 5, 15, or 50 times more frequent than the number of patients with spinal cord metastases?
3. If these were metastatic lesions, would you expect them to represent hematogenous or leptomeningeal spread of disease?
4. True or False: Because of its rich vascularization, the cervical cord is the more common site of intramedullary metastases than the thoracic cord.

Answers

Case 1

1.

• Anatomic
– The sensory symptoms involving the left upper and lower extremities localize the lesion to the cervical spine, brainstem, or right parietal lobe.
– The absence of headache and facial symptoms makes it less likely to be the parietal lobe.
– The absence of cranial nerve involvement makes it less likely to be the brainstem.
– This is probably a cervical spine lesion.
• Etiological
– Degenerative: disc herniation, synovial cyst, cervical stenosis usually associated with pain and/or myelopathy
– Neoplastic: extradural (metastasis, primary bone tumor), intradural extramedullary (meningioma, schwannoma, neurofibroma, ganglioneuroma), or intramedullary (ependymoma, astrocytoma, hemangioblastoma). The chronic progressive symptoms fit with a benign neoplastic etiology.
– Vascular: cavernoma. Arteriovenous malformations (AVMs) usually present acutely or subacutely.
Spinal cord infarcts present acutely, usually width motor symptoms.
– Demyelinating: multiple sclerosis (MS). The remittent course fits with an MS plaque, although the age of onset in MS is usually younger. Transverse myelopathy presents acutely or subacutely with more dramatic deficits.
– Others: trauma, radiation myelopathy, epidural abscess, and acquired immunodeficiency syndrome (AIDS) are less likely from the history. Toxic/metabolic — alcoholism, vitamin B12 deficiency.
Syringomyelia usually presents with bilateral loss of pain and temperature sensation in the upper extremities and spastic paraparesis. Enterogenous cysts, dural ectasia, and arachnoid cysts are rare.

2.

Cervical spine MRI with gadolinium. Laboratory work: vitamin B12 level; methyl malonic acid and homocysteine levels; serum and urine protein electrophoresis; complete blood count; electrolytes.

3.

(A) Sagittal magnetic resonance imaging of the C-spine with T2-weighted image and (B) T1-weighted image with gadolinium revealing an intramedullary tumor. On T2-weighted images, the tumor is isointense to the spinal cord, with hypointensity (hemosiderin deposits) above and below the tumor and a hyperintense overlying syrinx. There is heterogeneous enhancement after gadolinium.

The commonest intramedullary tumor in this are group is ependymoma followed by astrocytoma. Others include hemangioblastoma, schwannoma, and metastasis. The hypointensities on T2-weighted images above and below the tumor are characteristic (though not pathognomonic) of ependymoma.

4. C. Seven vertebral segments in length.
Astrocytomas tend to be longer that six vertebral segments. Their cysts tend to be within the lesion and rarely have hemorrhage at the margins. Astrocytomas occur more often in younger patients and both may have a mixed enhancement pattern.

5. We recommend surgical resection of the tumor (given the extent and appearance of the lesion on MRI and the progressive nature of the symptoms). Risks include neurologic deficits like paraplegia or quadriplegia, loss of bowel or bladder function, respiratory failure, tracheostomy, and ventilatory dependency, cerebrospinal fluid leak, neck pain, cervical instability, tumor recurrence, hematoma, infection, and spinal cord ischemia.

6. Equipment needed intraoperatively includes the following:
– Mayfield (or other kind) head holder
– Jackson radiolucent table or chest rolls
– Neurophysiologic monitoring with motor evoked potentials and somatosensory evoked potentials
– Intraoperative ultrasonography (US) for tumor localization
– Operative microscope and microinstruments
– Ultrasonic surgical aspirator

Details of the surgery are as follows:
– After cervical laminectomy, the dura is opened and tacked. Note the swollen cord and grayish discoloration.
– A midline myelotomy is performed starting in the area of grayish discoloration.
– If the tumor is hard to find, intraoperative US could help.
– A specimen for frozen section should be immediately sent.

– Tumor debulking is then started using suction or ultrasonic aspiration.
– If pathology reveals ependymoma, and a good margin is visualized between the tumor and the spinal cord, every effort should be made for cross total resection.
– In the case of astrocytoma, the margins are usually difficult to define; the goal is to remove as much as can safely be removed.
– After careful hemostasis, the dura is closed with or without patching.
– One may argue to fuse the laminectomized levels.
7. Histological section reveals perivascular pseudorosettes, which are characteristic of ependymoma.

8. Lumbar.
Ependymomas develop from ependymal cells. They are most often seen in adults aging 20 to 40 years. This is a relatively common and usually slow-growing spinal tumor, accounting for more that half of adult spinal cord tumors. Ependymoma can develop anywhere along the spinal cord, but up to 40% are found near the tailbone.

9. Yes, though only rarely.
10. No, because the cyst wall is usually nonneoplastic and the cyst will resolve on their own after neoplasm removal.

Case 2

1.
• There is a posterior epidural collection from L2–L4, hyperintense on T2-weighted images. This would be a cerebrospinal fluid (CSF) collection, abscess, or infected hematoma.
• The disc space looks normal, and so do the vertebral bodies. This finding is important to assess the radiographic presence of concomitant osteomyelitis.
• There is loss of lumbar lordosis.
• Given the clinical presentation, the most likely diagnosis is epidural abscess.

2.
• On T1-weighted images, it has the same intensity as spinal cord or neural elements.
• On T2-weighted images, it shows increased signal.
• On fat-saturated sequences, edema and soft tissue inflammation are clearly

visible and bright.
• Contrast-enhanced images often show a peripheral rim of enhancement due to granulation tissue and hypervascularity.

3. The thoracic spine. The majority are located posterior to the spinal cord. Spinal epidural abscess should be considered in any patient who presents with backache, fever, and spine tenderness.

4.
• Presence of hardware and instrumentation
• Repeated surgery
• Use of steroids
• Diabetes or other underlying medical problems such as renal failure, previous trauma, and urinary tract infections
• Intravenous drug abuse.

5.
• Gram-positive bacteria such as Staphylococcus (S.) aureus (reported in 60% of cases) or S. Epidermidis
• Gram-negative bacteria such as Escherichia coli and Pseudomonas aeruginosa.

6.
• The generally accepted incidence is less than 10%.
• The incidence is reportedly increased with higher complexity procedures. It is in the range of 0.6–3.7% after microdiscectomy and of 3.7–20% after instrumented lumbar cases.

7. A. Hematoma after trauma with subsequent degradation of products. Although a degrading hematoma is the perfect place for bacteria to grow, as long as it is sterile, it is less likely to result in infection. The remainder may seed the epidural space by direct or hematogenous spread.

8.
• Look for any discharge so it can be cultured before starting antibiotics.
• Explore the wound to see if you find a purulent collection or CSF.
• Drain the abscess, sampling tissue, and purulent material for culture.
• Perform a sharp debridement and lavage.
• Place on broad-spectrum intravenous antibiotics until final culture results are available. Vancomycin for the gram-positive covering including methicillin-resistant Gram-positive cocci and cefepime for Gram-negative bacteria are among the most common medications used.

• The closure of the wound is very important and can be done in different ways, but the key concept is the obliteration of the dead space caused by the removal of the infected tissue restoring the blood supply and postoperative drainage by out flow drain or suction/irrigation device.

9.
• A tear in this specific condition can lead to a spreading of the infection in the subdural space because of the disruption of the normal dural barrier.
• Suture with a 7–0 or 8–0 dural suture if accessible in a watertight fashion, using the Valsalva maneuver to confirm the closure.
• Cover with a dry piece of Gelfoam or collagen matrix.
• Cover with fibrin glue or hydrogel sealant.
• You may need to resect more bone to further the exposure and allow access to the whole tear.
• Continue appropriate antibiotics.

10.
• The infection on the MRI does not extend to the previously treated levels.
• The onset of symptoms is very recent as the surgery at the L3–L4 level was performed in the past 2 weeks. Therefore, the formation of the glycocalyx over the rod and other used material is improbable.
• Titanium materials have a porous surface that allows the penetration of antibiotics.
• Good results have been shown when leaving the instrumentation in place, even in cases where the infection was involving those instrumented levels, if thorough debridement is performed and antibiotic therapy is well managed.

11. The bone is likely detached due to erosion from osteomyelitis.

12.
• You worry about two entities:
– Osteomyelitis and spread of the infection to bone
– Instability and the possibility of spinal deformity
• Your management should now involve placing the patient in a brace postoperatively and obtaining flexion-extension lumbar spine radiographs.
• You may repeat radiographs in 2 to 4 weeks as osteomyelitic changes may become visible radiographically only after 2 to 3 weeks.
• He may need a lumbar instrumented fusion once the infection has healed if instability is then demonstrated.
• Treatment with antibiotics does differ in cases of osteomyelitis: he then needs 8–12 weeks of intravenous antibiotics instead of only 4–6 weeks (as in cases of

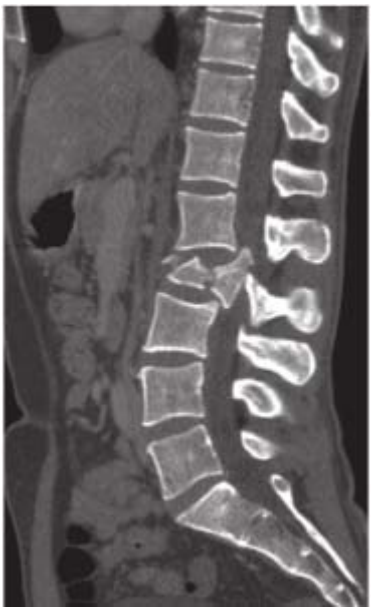
simple epidural abscess with no body involvement).
• Hyperbaric oxygenation can help to promote host immune defense response and stimulate vascularization of the injured tissues.

Case 3

1. B. Axial compression.

Thoracolumbar burst fractures are usually caused by a substantial axial loading force that results in compression failure of the anterior and middle spinal columns. As is the case for compression fractures, falls from a height and motor-vehicle accidents are responsible for the majority of these injuries. The sudden application of a supraphysiological axial load results in vertebral end-plate failure as adjacent disc tissue is driven into the vertebral body. An earlier theory that axial loading creates a sudden increase in internal vertebral body pressure that results in a burst fracture appears to be incorrect. Like compression fractures, burst fractures have a predilection for the thoracolumbar (T11-L2) spinal segments.

2. It is a fracture of the vertebral body caused by axial loading.
It occurs with failure of the anterior and middle column typically in the thoracolumbar spine.



Sagittal reconstructed CT scan of the lumbar spine demonstrating L2 burst fracture with severe canal compromise and retropulsion.

3. Indications for surgery are as follows:
1. Greater that 50% involvement of the canal.
2. Greater than 50% compression of the vertebral body.

3. Greater than 20 degrees of angulation with relation to the vertebral body above and below.
4. Significant neurological deficits or nerve root impingement.
5. Progressive deformity or kyphosis on serial imaging studies.

4.
• In a compression fracture the anterior column is involved, but the middle column is intact.
• Typically these fractures are stable and do not require surgical fixation.
• The mechanism of injury is often more benign and involves simple falls in elderly patients or osteoporotic patients.

5. The system that is most commonly utilized for classification of thoracolumbar burst fractures is that described by Denis. According to this system, a type-A fracture involves failure of both the superior and the inferior end plate, type B involves failure of the superior end plate only, type C involves failure of the inferior end plate only, type D results in an axial loading and rotational injury, and type E results in an axial loading and lateral flexion injury. Type B is the most frequent fracture pattern, followed by type A. The other types are relatively rare.

Case 4

1. D. Epidural lipomatosis.
Prominent epidural fat is seen anterior and posterior to the thecal sac, extending from the L3–4 level to the sacrum in a patient with chronic steroid use for asthma. Note the accentuated tapering of the thecal sac and the Y configuration of the thecal sac on axial T1WI.

2. A strong male predominance exists.
3. The “Y-sign.” This results when epidural fat compresses the thecal sac that is anchored to the osteofibrous walls of the lumbar canal by meningovertebral ligaments. The compression produces a stellate appearance of the sac.
4. Lipoma and angiolipoma.

The most commonly involved levels in the thoracic and lumbar spine are T6–T8 and L4–L5, respectively. Epidural lipomatosis should be differentiated from other extradural mass lesions such as lipoma or angiolipoma. Lipomatosis consists of unencapsulated, diffuse fatty tissue derived from preexisting epidural fat by hypertrophy, whereas lipomas or angiolipomas are typically well-encapsulated, circumscribed masses.

5. Lumbar. The cervical epidural space contains predominantly blood vessels and perivascular connective tissue.

Case 5

1. C. Spinal cord infarct.

This patient probably has a spinal cord infarction from an anterior spinal artery occlusion. The posterior cord may be spared, preserving joint proprioception. Bilateral lower extremity deficits without cranial nerve or mental status findings would be an exceedingly unusual cerebral stroke presentation. There is no information, such as psychological stressors or a non-physiologic exam, to suggest a conversion disorder in this case. Multiple sclerosis causes neurological deficits over space and time. In this case we have a single deficit at a single point in time. History of metastatic cancer or trauma might make the physician suspect spinal cord compression.

2. D. T10–L1.

The artery of Adamkiewicz is a major anterior radicular artery and may supply the lower two-thirds of the spinal cord. It is at risk of occlusion during abdominal aortic aneurysm repair. Other branches off of the aorta or internal iliac arteries may also supply the thoracic and lumbar cord. The upper segments of the spinal cord are usually supplied off the vertebral arteries.
The artery of Adamkiewicz is usually located at the T12-L1 spinal cord levels and is more frequently (about 65% of the time) seen on the left side. The other cord levels listed may have small spinal medullary arteries but not the large diameter vessel characteristic of Adamkiewicz.

3. D. Spastic paralysis.

This patient has an upper motor neuron lesion. The damage has been done proximal to the synapse of the anterior horn of the spinal cord. He will therefore develop a spastic paralysis. Fasciculations, fibrillations, flaccid paralysis, and hyporeflexia are all found following lower motor neuron lesions (at the anterior horn cell or more distally).

Case 6

A. Hyperflexion

Bilateral facet dislocation.
Severe flexion injury.

This axial CT scan depicts bilateral jumped (locked) facets with a concomitant fracture of the lamina. Bilateral jumped (locked) facets results from severe hyperflexion injuries and are usually associated with concomitant spinal cord injury. With jumped facets, the facet capsule, apophyseal joints, ligamentum flavum, and interspinous ligaments are disrupted. Unilateral jumped facets result from flexion/rotation injury mechanism.

Case 7

C. Eosinophilic granuloma.

Eosinophilic granuloma of the spine is most often seen as a lytic lesion without surrounding sclerosis and is a classic cause of a single collapsed vertebral body in patients between the ages 5 and 10 years. Giant cell tumors are highly destructive, lytic masses in patients between the ages of 20 and 40 years, while hemangiomas have the characteristic “polka dot” vertebral body pattern.

Case 8

1. E. Aneurysmal bone cyst (ABC).

Examples include: Aneurysmal bone cyst (shown), Eosinophilic granuloma, Brown tumor of hyperparathyroidism, Giant-cell (reparative) granuloma.

2. The neural arch (posterior elements) and the lumbosacral region.

Comment

Aneurysmal bone cyst is a benign, highly vascular osseous lesion of unknown origin. It accounts for approximately 1% to 2% of primary bone tumors, and 3% to 20% of ABCs involve the spine. Upper cervical lesions are rare. Patients with

ABC are usually in the second decade of life. The pedicles and posterior elements are involved in most cases.

Histologically, the interior of the ABC can be solid and vascular and/or cystic and hemorrhagic (surrounded by a thin bony rim). CT demonstrates the osseous expansion and can detect the multiple small fluid levels, although these are more evident on axial and sagittal T2W MR images. Within the cysts, the dependent fluid is hypointense, whereas the nondependent fluid is hyperintense as a result of the different sedimentation properties and composition of blood products in the two components.

Case 9

A. Metastasis.

On the AP projection the left L1 pedicle is absent. Compare it with the clearly seen right pedicle at the same level and the symmetry of the pedicles at the levels above and below.

The winking eye or Winking Owl Sign refers to the appearances of the spine, when a pedicle is absent - almost always due to a bony metastasis.
The usual appearance of the pedicles representing two eyes on the AP projection is lost, as one pedicle is destroyed - it then appears like one eye is open and the other is winking or shut.
Potential all bony metastases can do this, however breast cancer and lung cancer are the usual culprits.

There is a wide etiology:

- Destroyed pedicle
  - spinal metastases
  - intraspinal malignancies
  - tuberculosis and other infections

    - uncommon: primary bone lesion; lymphoma
- Congenital absence/hypoplasia of a pedicle
- Neurofibromatosis
- Poorly visualised
- Radiation therapy

Case 10

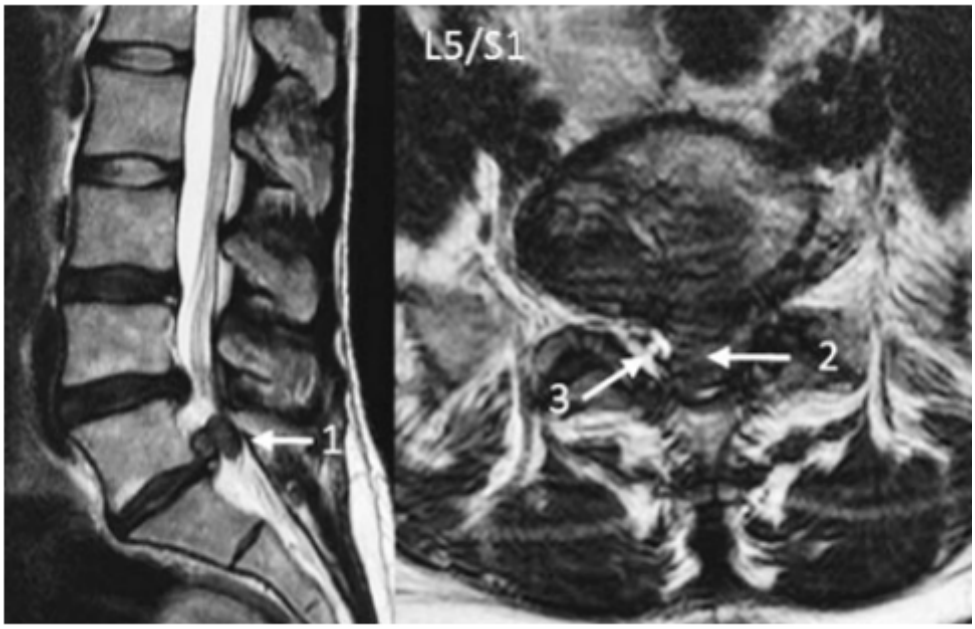
1. Cauda equina syndrome until proven otherwise. This is a clinical syndrome resulting from compression of the cauda equina. It presents with a variable combination of leg pain (classically bilateral), anaesthesia in the sacral distribution, and urinary retention with overflow incontinence. In this case, the most likely cause of cauda equina compression is a prolapsed lumbar intervertebral disc. Other lesions, including tumours, abscesses, and trauma, can also cause cauda equina syndrome.

2**.** The exact nature of the urinary symptoms must be determined because it helps to distinguish cauda equina syndrome from urinary retention due to back pain, urinary tract infection, or pre-existing bladder problems The timing of onset of symptoms is also critical because it influences the timing of surgery. The completeness of the symptoms is strongly correlated with the recovery potential, with patients with incomplete cauda equina syndrome having a better prognosis.

3. (a) The left ankle reflex is affected, suggesting involvement of the S1 nerve root. Ankle plantar flexion and hip extension also have contributions from S1. Therefore the level of compression is likely to be at L5/S1.
(b) Examining perineal sensation can be challenging if the patient's response is inconsistent. As a general rule, if perineal pinprick sensation is less than reported elsewhere when the same pressure is applied, it should be assumed that there is impairment. If a sharp sensation is elicited when the pin is pressed harder, impairment is not excluded. If a sharp sensation is not elicited at all, impairment is confirmed. Therefore these findings are consistent with cauda equina compression.

4. If the clinical diagnosis is suspected, an MRI scan of the lumbar spine is required immediately. Claustrophobic patients may tolerate the MRI scanner with sedation. If not, the options include a CT myelogram or a MRI under general anesthetic.

5. On the sagittal view there is a large disc prolapse at L5/S1 filling the width of the canal



(1). The axial view at this level shows that the disc is laterally sited to the left (2). Note that the thecal sac (surrounded by a small amount of CSF which appears white on this T2 image) is displaced to the right (3). There is a smaller left-sided disc protrusion at the level above, where the canal is more capacious (see L4/L5 image above).

6. This patient presents within 24 hours of onset of urinary symptoms, so surgery should be performed immediately. The operation is a discectomy, involving a midline posterior approach, laminectomy, and removal of the prolapsed segment of disc.

Case 11

1**.** The combination of neck pain with motor deficits in the upper and lower limbs that have evolved over a period of time points to an insidious pathological process affecting the cervical spinal cord. The neck pain is very important as it is classically associated with extradural pathology. In older people this would be degenerative or metastatic disease or trauma. Degnerative disease such as a disc prolapse is almost impossible in a 12-year-old and secondary deposits are very rare in a child with no previous history of malignancy. An abscess should be considered including tuberculosis (TB) depending on the social circumstances. Primary extradural tumors are also a possibility. Intradural pathology can cause

neck pain, particularly if there is a disease process stretching the dura. This includes acute inflammatory processes such as Guillain–Barré syndrome and transverse myelitis as well as intradural tumors.
It would be vital to enquire about any sensory loss or sphincter disturbance, as if present their pattern will provide further clues about the location of the lesion.

2**.** The sagittal T2 image shows a hyperintense circumscribed mass in the spinal cord from C3 to C5 (A). The axial view demonstrates that the lesion (A), appearing hyperintense with normal cord around it which appears as a rim of hypointensity, is located within the spinal cord. This is an intramedullary tumor expanding the cord and there is a little dilation of the central canal of the cord below it.

3**.** The most common types are gliomas. Grade III gliomas (anaplastic astrocytomas) rather than glioblastoma are seen in the spinal cord. The other relatively common tumor (in children) is an ependymoma. These have a range of subtypes from benign (myxopapillary subtype) to more malignant (grade III). Other rare primary CNS tumors can also occur, as well as intramedullary metastases.

4. The initial management is dexamethasone to reduce any associated edema and full neuro-axis MRI for staging.

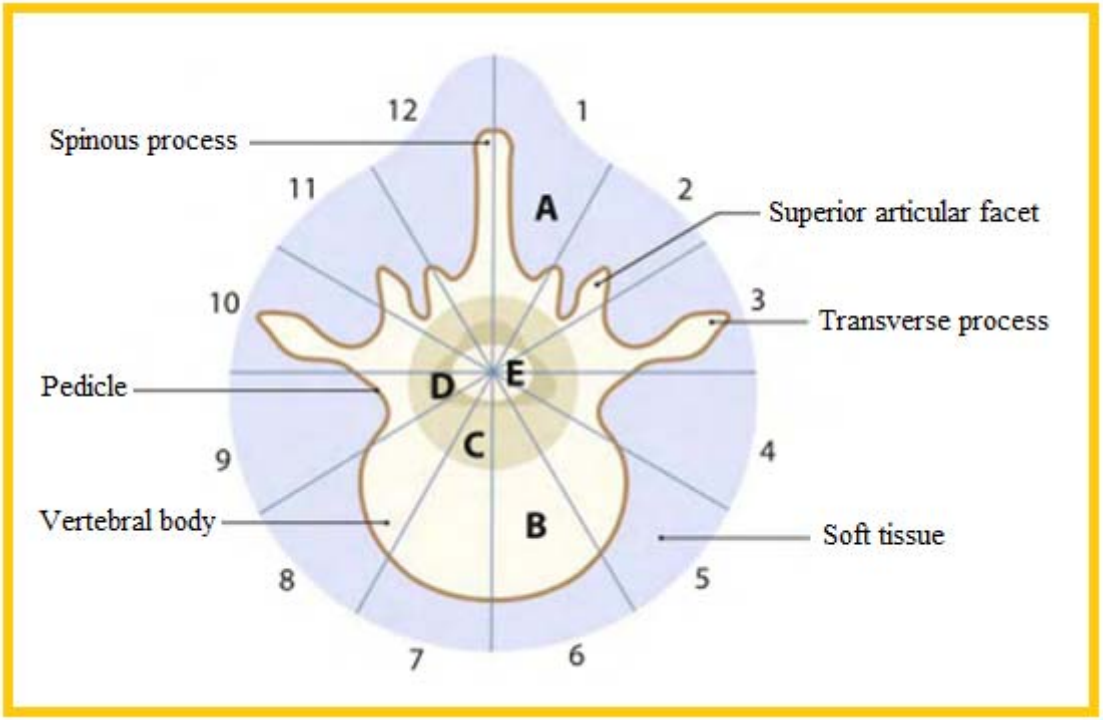
5. The aims of surgery in the treatment of primary intramedullary tumors of the spinal cord should be considered in the following terms.
1. To biopsy tissue where there is diagnostic uncertainty and the result will influence the patient's management, such as the decision to proceed with radiotherapy or chemotherapy.
2. To debulk or excise the tumor where there is a realistic chance of so doing without unacceptable neurological deficit and with good prognosis.
3. To manage associated problems (e.g. syringomyelia) or to drain tumor cysts.
Selecting patients correctly for debulking or excision surgery versus conservative management is important. Where the tumor is well circumscribed (as with many ependymomas), surgery can potentially be curative and more aggressive strategies are justified. Similarly, for malignant tumors (grades III and IV), where the neurological deficit is profound and progressive, aggressive surgery, including cordectomy for patients with complete or near-complete paraplegia, is well described with the occasional long-term survivor reported.

However, those patients with partial slowly progressive deficits presenting earlier in the course of their illness merit a more cautious approach.
In this case, the tumor is well circumscribed radiologically and surgery should

aim for debulking or excision. However, the risks of major deficit are high. Surgery would be performed using continuous electrophysiological monitoring, typically motor and somatosensory evoked potentials (MEPs and SSEPs), to alert the surgeon to potential intraoperative damage.

This child underwent debulking of the tumor after MRI showed no other sites of tumor. MEPs were used and showed severe slowing at one point during dissection of the anterior part of the tumor. She woke up with severe weakness of her hands, bit this improved over 6 days and she was able to feed herself and mobilize independently by day 8. Her bowel and bladder function remained intact. Histology showed a grade III ependymoma and she underwent adjuvant chemoradiotherapy.

6. Weinstein, Boriani, Biagini (WBB) classification describes the vertebral involvement as sections of a clock face ("zones") centred on the spinal cord, from zone 1 (left spinous process and lamina) through zone 6 (left anterior wedge of vertebral body) and back round to zone 12 (right spinous process and lamina). In addition, the prefixes *A*–*E* are used to denote radial levels ("layers") of vertebral involvement, from extraosseous paraspinal tissues (*layerA*) through to extradural (*layer D*) and intradural (*layer E*).



A. Extraosseous soft tissues C. Intraosseous (deep)
B. Intraosseous (superficial) D. Extraosseous (extradural)
E. Extraosseous (intradural)

The staging system can also be useful to plan intralesional surgery, exchanging information among institutions, and evaluating the relationship between treatment and outcome.

7. The most common approaches for spine tumors are as follows:
    1.

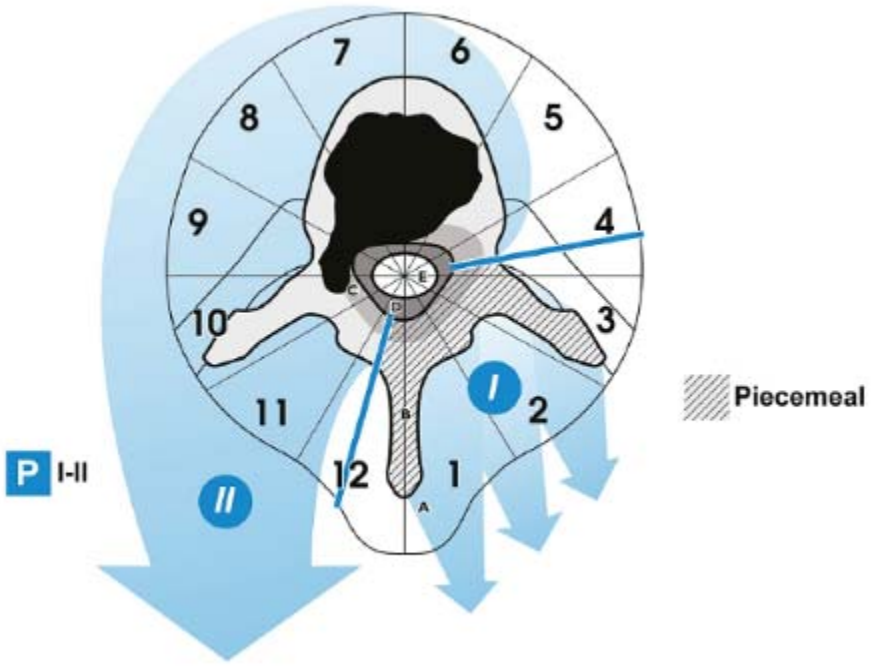


Fig. 1 Single posterior approach.

It enables the removal by en-bloc resection of a tumor arising in the vertebral body of a thoracic vertebra. Criteria to achieve appropriate margins include sector 9 or 4 free from tumor. If the tumor grows in layer D, the margin will be intralesional during the release from the dura. If the tumor grows in layer A, the margin will be intralesional during the release from the anterior structures. This is a two-step procedure, as shown by roman numerals on the figure:

(I) Piecemeal excision of the posterior arch not involved by the tumor. At least four sectors are required, starting from sector 4 or from sector 9. Release from the dura and section of the nerve root(s) involved by the tumor.

(II) Blunt dissection of the anterior part of the vertebral body from the mediastinum, osteotomy, or discectomy above and below the tumor, with full release from the dura and finalizing the resection.

2.

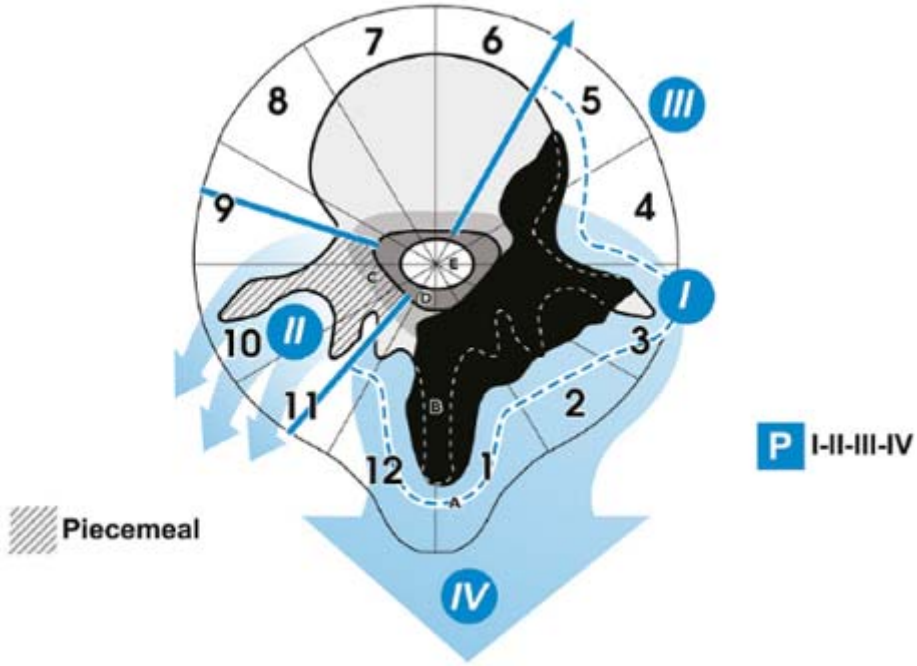


Fig. 2 Single posterior approach with sagittal osteotomy.

A tumor excentrically growing in the lumbar spine can be removed en bloc by a single posterior approach provided that the body is not involved over sector 5 at left and over sector 8 at right. At least three sectors posteriorly must not be involved by the tumor (4 to 1–2 or 12–11 to 9). This is a four-step procedure, as shown by roman numerals on the figure:

(I) Provide the appropriate margin over the tumor posteriorly growing by resecting inside the posterior muscles covering the tumor mass if it is expanding in layer A. The release will proceed laterally until the lateral side of the vertebral body. In the thoracic spine the pleura can be left on the tumor, and in the lumbar spine the posterior part of the psoas must be dissected, but the segmental vessels must be found and ligated.

(II) Piecemeal excision of the posterior arch not involved by the tumor. This step includes the approach to the canal, release of the dura from the tumor (if the tumor grows in layer D, the margin will result intralesional), and section of the nerve root(s) involved by the tumor.

(III) Displace carefully the dura and perform the osteotomy from posterior to anterior in sector 8 or 5.

(IV) The specimen is removed.

3.

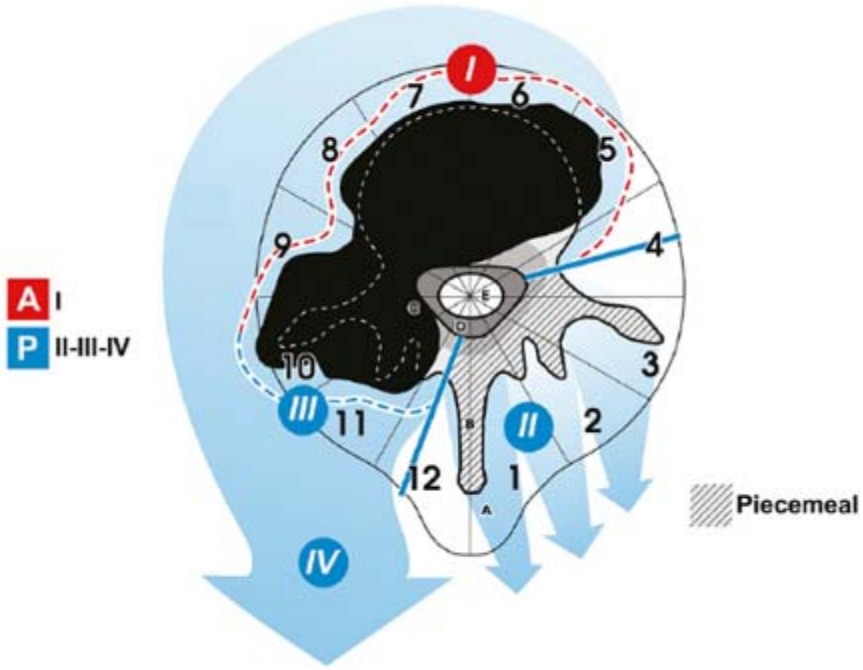


Fig. 3 Two approaches: anterior first, posterior second in the thoracic and in the lumbar spine.

When the tumor is growing anteriorly (layer A), an anterior approach must be performed as the first step to provide a wide/marginal margin under visual control. This is a four-step procedure, as shown by roman numerals on the figure:

(I) In case of tumors mostly occupying the vertebral body, the anterior approach can be the first step to release from mediastinum or retroperitoneal, eventually leaving involved structures as the margin. A sheet of Silastic or similar material can be left as protection.

(II) Posterior approach: piecemeal excision of the posterior arch not involved by the tumor. At least three or four sectors are required, starting from sector 4 or from sector 9.

(III) Release of the dura from the tumor, section of the nerve root(s) involved by the tumor, and then provide the appropriate margin over the tumor posteriorly growing by resecting inside the posterior muscles covering the tumor mass if it is expanding in layer A.

(IV) The specimen is removed by rotating around the dural sac.

4.

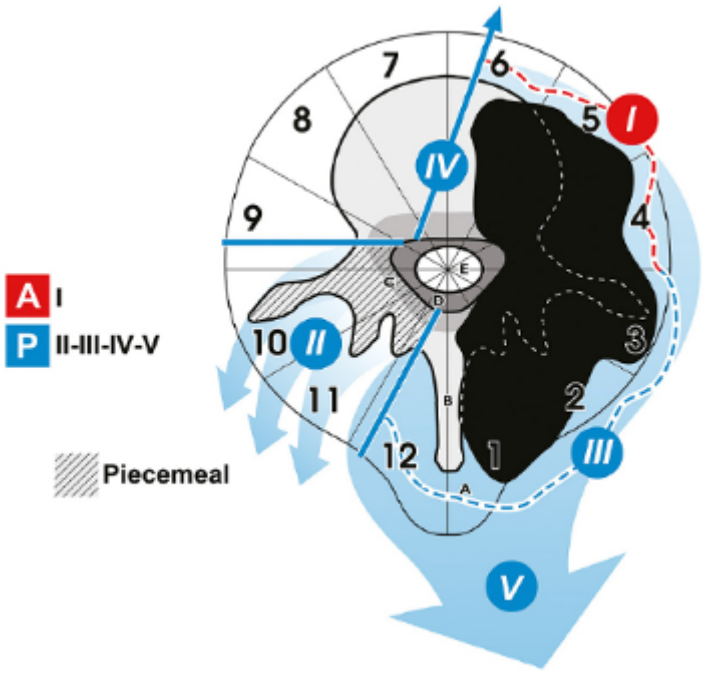


Fig. 4 This procedure can be planned in case the tumor is eccentrically growing.

Anterior first, posterior second in the thoracic and lumbar spine, and sagittal osteotomy posterior to anterior. This is a five-step procedure, as shown by roman numerals on the figure:

(I) In the first stage the anterior approach provides a wide/marginal margin under visual control, releasing from mediastinum or from peritoneal, eventually leaving involved structures as the margin. Discectomies or transversal grooves in vertebral bodies are performed to define the upper and lower margins. A sheet of Silastic or any other tissue can be left as protection. In the second stage, the posterior approach, piecemeal excision of the posterior arch not involved by the tumor is performed.

(II) At least three sectors are required, starting from sector 4 or from sector 9.

(III) Provide the appropriate margin over the tumor posteriorly growing by resecting inside the posterior muscles covering the tumor mass if it is expanding in layer A.

(IV) Release of the dura from the tumor, section of the nerve roots crossing the tumor, and osteotomy posterior to anterior at some distance from the tumor in order to leave uninvolved bone as margin.

(V) The resected specimen can be removed once the upper and lower discectomies or osteotomies are finalized.

5.

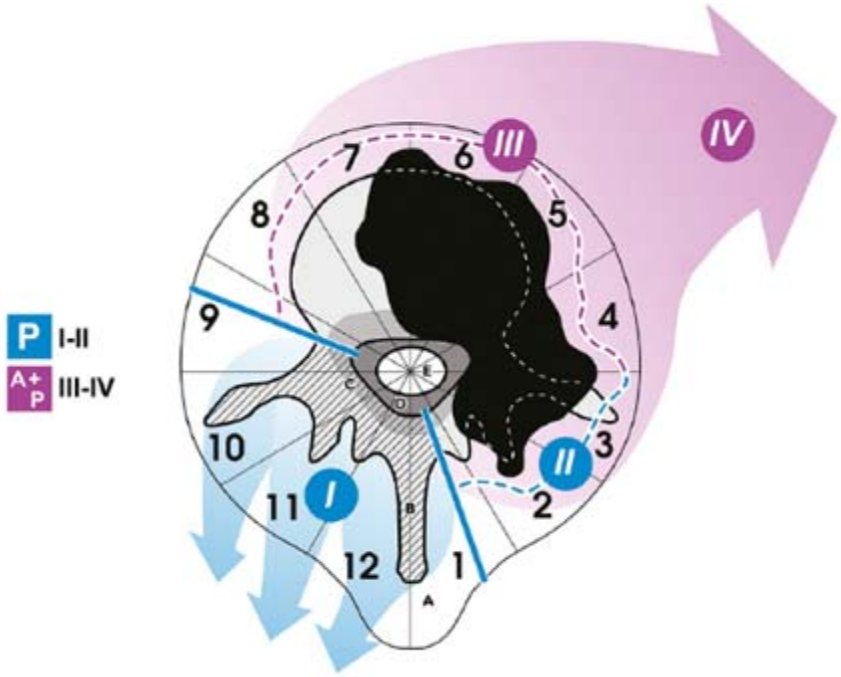


Fig. 5 Two stages: first posterior approach, second contemporary anterior and posterior approaches.

This technique is typical and more appropriate for lumbar tumors and is associated with the highest rate of morbidity and complications. In thoracic spine it is less advisable. The first steps are performed with the patient in the prone position: piecemeal excision of the posterior arch not involved by the tumor. This is a four-step procedure, as shown by roman numerals on the figure:

(I) At least three sectors are required, starting from sector 4 or from sector 9.

(II) In cases of a tumor posteriorly growing and invading layer A, an appropriate margin must be provided by resecting inside the posterior muscles covering the tumor mass. Then release the dura from the tumor (if the tumor grows in layer D, the margin will result intralesional) and section the nerve root(s) crossing the tumor. Discectomies or transversal grooves in vertebral bodies are performed to define the upper and lower margins. The second stage is performed with the patient in the lateral position. Anterolateral approach (thoracotomy, thoracoabdominal, retroperitoneal) and reopening of the posterior approach.

(III) To provide an appropriate margin over the tumor, it must remain covered by pleura or by psoas. Spiral wires are used to embolize the segmental arteries to facilitate the release of the aorta on the contralateral side.

(IV) Once the upper and lower discectomies or osteotomies are finalized, the specimen is removed by combined maneuvers.

Case 12

Ependymoma, Cervical.

1. Intramedullary.
2. Ependymoma.
3. Arteriovenous malformation, ependymoma, hemangioblastoma.
4. Symmetrical, because the tumor arises from ependymal cells in the central canal.

Comment

Spinal ependymomas are the most common intramedullary neoplasms in adults and are most often found in the cervical cord, followed by the thoracic cord and conus.
These masses are well-circumscribed, noninfiltrating, benign tumors that are generally more focal that astrocytomas. They have a propensity for intratumoral hemorrhage and may also produce subarachnoid hemorrhage with leptomeningeal deposition of hemosiderin (superficial siderosis). Cystic degeneration of the tumor (as shown in this case) may be observed, as well as extensive cyst formation rostral and/or caudal to the tumor. Arterial supply to the tumor is most often derived from the anterior spinal artery. Intramedullary ependymomas are manifested clinically as neck or back pain and, less often, numbness or paresthesias. Progressive myelopathy has been attributed to compression of the surrounding spinal cord rather than infiltration into the adjacent cord parenchyma. Given the slow growth and relatively well-defined margins of these tumors, symptoms generally progress slowly, and patients may wave a long history of clinical symptoms prior to diagnosis. Complete surgical resection is the treatment of choice.

On T1W images, ependymomas generally appear isointense relative to the normal cord signal intensity. Less often, they are hypointense. Heterogeneous signal and regions of hyperintensity on T1W images are usually the result of hemorrhagic components of the tumors. On T2W images, ependymomas are generally hyperintense relative to the normal cord, although hemorrhage can result in central and/or peripheral hypointensity owing to susceptibility effects associated width hemosiderin deposition. This hypointensity is usually better

shown on T2W gradient-echo images than on fast-spin-echo T2W images (note the darker lies within the cord tumoral region on the T2W image).

Case 13

Ependymoma with Cyst, Cervical.

1. No, it has the appearance of a caudal cyst.
2. Later.
3. Ependymomas are highly vascular and thus enhance markedly.
4. Estimates range from 50% to 90%, with most favoring the upper end of the range.

Comment

In patients with spinal cord ependymoma, three types of ‘‘cysts’’ have been described:
(1) Intratumoral cysts, which are surrounded by abnormal glial elements, contain blood or xanthochromic fluid, and have enhancing margins on postcontrast T1W images.
(2) Caudal or rostral cysts, which are located at the border of the tumor, have gliotic linings typically, contain CSF-like fluid, and have nonenhancing margins on postcontrast T1W images.
(3) Reactive dilatation of the central canal. This is recognized by its central location within the cord and its location beyond the tumor margins.
Distinction between rostral/caudal cysts and reactive dilatation of the central canal may be difficult, and some authors do not make a clear distinction between (2) and (3). Note the visibility of the central canal at C2–C3 on the T2W image. It is important to differentiate intratumoral cysts from the other two types of cysts on imaging because intratumoral cysts should be excised with the tumor, whereas rostral/caudal cysts do not need to be excised. They contain no tumor cells and may be aspirated and drained. Compared with intratumoral cysts, rostral/caudal cysts and central canal dilatation are more likely to show evidence of intracystic CSF motion. Note that the intramedullary tumor has typical MR findings of hyperintensity on the T2W image, uniform enchancement on the postcontrast T1W image, approximately symmetrical involvement of the whole width of the cord, and cervical location.

Case 14

Intramedullary Metastases (Breast Carcinoma)

1. Hemangioblastomas, metastases, sarcoid.
2. Approximately 15 times more frequent.
3. Hematogenous spread, given their intramedullary location and lack of leptomeningeal enchancement or nodularity.
4. False. The thoracic cord is the more common site.

Comment

The postcontrast T1W sagittal image reveals two enhancing nodular lesions—an oval-shaped one at T11 and a rounded one at T12. The axial image at T11 confirms the intramedullary location of the upper lesion and a lack of cord enlargement. Intramedullary metastases are rare. Of cancer patients, only 1% to 2% have intramedullary metastases at autopsy, compared with 18% to 24% width brain metastases. Carcinoma of the lung is the most common extra–central nervous system (extra-CNS) tumor associated with intramedullary metastases (50%), followed by breast carcinoma (15%), lymphoma (9%), melanoma (7.5%), and colorectal carcinoma (7%). The primary tumor is unknown in less than 2% of cases. On MR imaging, intramedullary metastases are most frequently single, oval shaped, and small, with little or no cord enlargement. They are typically isointense to cord on precontrast T1W images and demonstrate homogeneous, nodular enhancement on postcontrast images.

T2W images show surrounding ‘‘pencil-shaped’’ hyperintensity, representing edema. Larger lesions are more likely to demonstrate central hypointensity on T1W images, peripheral enhancement following contrast administration, extensive edema on T2W images, and cord enlargement. MR of the brain should be recommended because of potential cerebral metastases and also because mimickers of intramedullary metastasis, such as multiple sclerosis or sarcoid, may be favored on the basis of the intracranial findings. Most studies suggest that intramedullary metastases from extra-CNS tumors reach the spinal cord mainly by two routes—(1) arterial circulation to the cord and (2) vertebral venous plexus (Batson’s plexus). Extra-CNS tumors can also extend to the cord by direct invasion from nerve roots or CSF, which may explain some cases of coexistent intramedullary metastasis and leptomeningeal tumor.

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