

CONTINUING EDUCATION PROGRAM: FOCUS...

Tips and traps in spinal cord pathology

G. Cosnard

Saint-Luc University Clinics, Louvain Catholic University, Department of Radiology and Medical Imaging, avenue Hippocrate 10, 1200 Brussels, Belgium

KEYWORDS

Nervous system;
Spinal cord;
MRI

Abstract While reviewing major pathological conditions, the radiologist must learn to adapt his technique to the indication and look for multifocal lesions. In conditions involving malformation, transdural cord herniation and diastematomyelia may be discovered late. In vascular diseases, a dural arteriovenous fistula with perimedullary venous drainage is the most common vascular malformation and a source of diagnostic error. On discovering a medullary cavity, the radiologist needs to know when to discount focal distensions of the ependymal canal and how to detect tumoral syringomyelia. In the case of a tumour, he should know the characteristics of common tumours such as astrocytomas, ependymomas, haemangioblastomas and cavernomas. In inflammatory diseases, he should know when a brain examination is required. When faced with images appearing to show a tumour, he should consider the possibility of pseudotumours and in particular of granulomatoses.

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Traps and tips are discussed in the context of conditions involving malformation and vascular and chronic ischaemic, medullary cavity, tumoral, inflammatory and infectious diseases.

Malformation

A transdural spinal cord herniation [1] (Fig. 1) must not be confused with a posterior arachnoid cyst. In both cases, the cord is ventral in the canal, and the conditions are usually in the thorax. In transdural spinal cord herniation, the cord is drawn to and trapped in a gap in the dura mater between T2 and T8 (the condition is often congenital, more

E-mail address: guy.cosnard@yahoo.fr

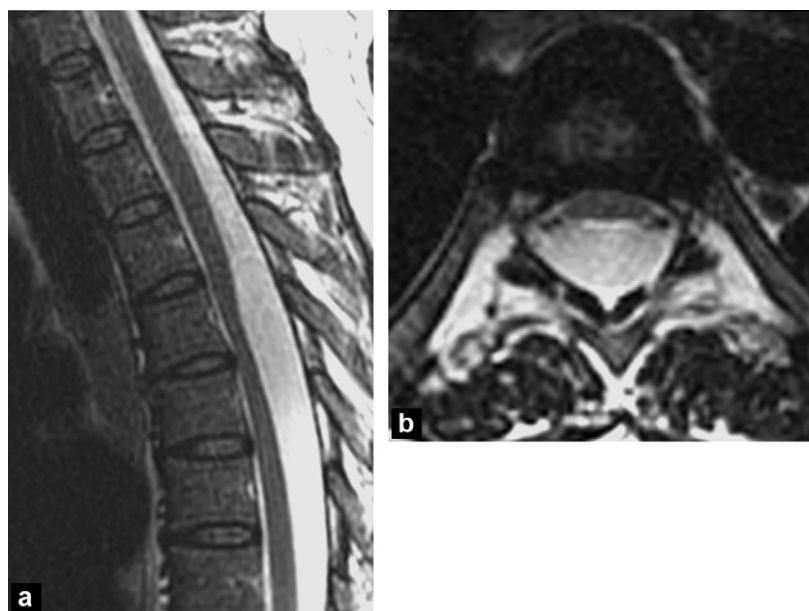


Figure 1. Transdural spinal cord herniation: a: MRI, sagittal T2-weighted image: the spinal cord shows 'bayonet' deformity and is drawn towards the gap in the duramater; b: MRI, axial T2-weighted image: the spinal cord appears to be pressed against the posterior surface of the vertebral body.

rarely traumatic) and shows a clear bayonet deformity with expansion of the posterior subarachnoid spaces. This is a rare condition, since only 100 cases have been reported in the literature in English, but its frequency is underestimated. Transdural spinal cord herniation appears clinically at about 50 years of age due to gradual myelopathy or more often Brown-Sequard syndrome secondary to microtrauma or ischaemia related to adhesions to the gap in the dura mater. Sagittal MRI shows narrowing of the spinal cord for 1 to 3 cm (its bayonet deformity), the thinning and focal disappearance of the anterior subarachnoid spaces and their

enlargement posteriorly. Very thin DRIVE and CISS sequence images show invagination (usually parasagittal and lateral) of the spinal cord and the focal amputation of CSF hyperintensity at the point of cord entrapment. If the cord is damaged, it may show an area of myelomalacia as hyperintensity in a T2-weighted image.

In arachnoid cysts, which are often posterior and in the thorax, the spinal cord is displaced and pressed against the vertebral bodies but the posterior subarachnoid spaces are partitioned, and this shows up well in flow imaging.

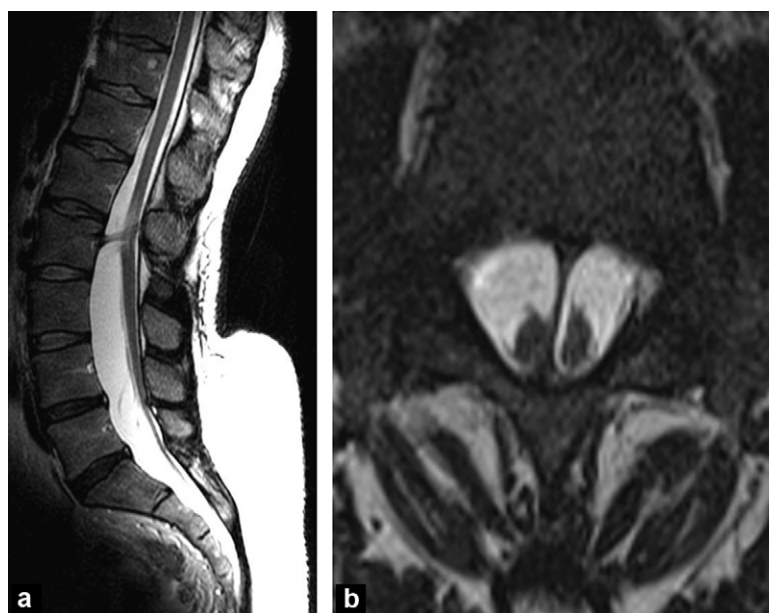


Figure 2. Diastematomyelia: a: MRI, sagittal T2-weighted image: there is low attachment of the spinal cord; b: MRI, axial T2-weighted image: the spinal cord is focally divided into two hemicords separated by an osteofibrous spur.

Diastematomyelia [2] (Fig. 2) should not be ignored. This malformation is usually found in children but is sometimes discovered later. It occurs above all in the thoracolumbar region. Focally the cord divides into two hemicords each with an ependymal canal, an anterior and a posterior root and sometimes one of the contralateral roots. In type I diastematomyelia, each hemicord is surrounded by its own leptomeningeal and dura mater sheath. It is separated from the other hemicord by an extradural space containing an oblique sagittal spur which is osteocartilaginous or fibrous in nature. In type II diastematomyelia, there is a single arachnoid and dura mater meningeal sheath containing the two hemicords. They are separated in this case by a fibrous or fibro-adipose strip. In 85% of cases, other anomalies are associated with the condition:

- intramedullary cavities in half of the cases;
- caudal fixation of the cord by a thick filum or distal spur;
- meningocele, hemimyelocoele;
- cutaneous abnormalities.

There is scoliosis in 60% of cases, this being the main reason for consulting. A unilateral club foot with motor weakness in the homolateral leg is a characteristic in 50% of cases of lumbar diastematomyelia. Urinary complaints are rarely in the forefront but urodynamic tests are often abnormal. X-ray images of the spine show scoliosis, spina bifida, increase in the transverse diameter of the vertebral canal, abnormal vertebral segmentation, intersegmental fusion of opposite lamina of two adjacent levels, and sometimes an ossified spur in the centre of the canal. MRI shows the site and extent of the cleft in the cord, its unifocal or multifocal character, its type, I or II, the position of the conus medullaris and the roots of the cauda equina relative to the diastematomyelia, any associated malformations and the main complications, such as an intramedullary cavity or myelopathy.

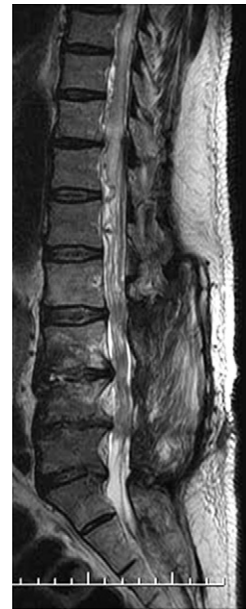


Figure 4. Dural arteriovenous fistula. MRI, sagittal T2-weighted image: traces of laminectomy; conus medullaris hyperintense. Nodular and serpiginous images of signal voids in the subarachnoid spaces and around the distal cord.

Vascular and chronic ischaemic disease

The radiologist must not fail to diagnose a dural arteriovenous fistula with perimedullary drainage [3] (Figs. 3–5). It is the most frequent vascular malformation, three times more common in men, particularly at around 60 years of age. It appears insidiously as walking difficulties, sensory disorders and sphincter problems reflecting the ascending myelopathy secondary to medullary venous hypertension. The symptoms

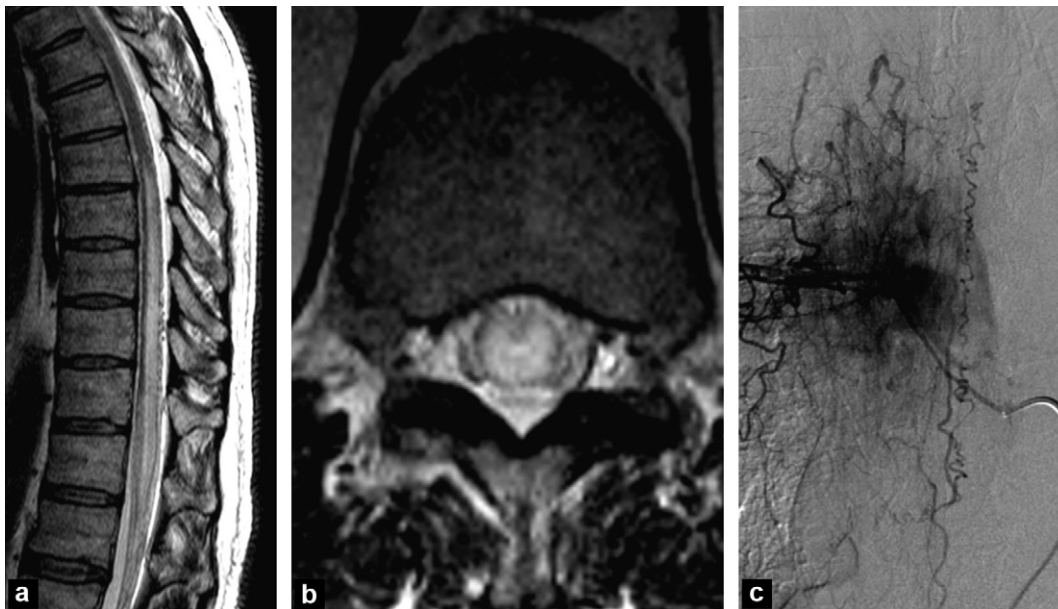


Figure 3. Dural arteriovenous fistula: a: MRI, sagittal T2-weighted image: homogeneous hyperintensity of the conus medullaris and small nodules of flow voids around the distal cord; b: MRI, axial T2-weighted image: conus medullaris hyperintense with a chronic ischaemic aetiology. Peripheral border of healthy cord; c: angiography: arteriovenous fistula and distension of the perimedullary veins.

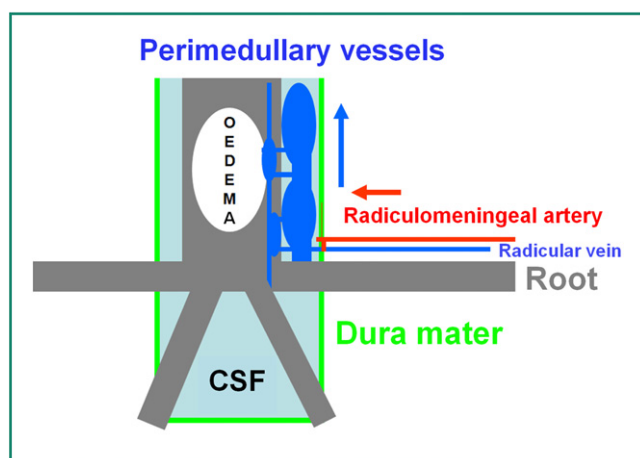


Figure 5. Diagram representing a dural arteriovenous fistula with perimedullary venous drainage with the oedemato-ischaemic consequences this has for the conus medullaris.

are often attributed to degenerative narrowing of the canal which, moreover, is often associated with it. This is the cause of frequent diagnostic errors and delay in appropriate management, which is very detrimental for the patient. The dural fistula, the topography of which is often foraminal, is a slow-flow fistula. It is a micro-shunt between a meningeal branch of a radicular artery and a vein with abnormally retrograde drainage to the perimedullary or intramedullary veins. The fistula is usually thoracolumbosacral between T3 and S1, L1 and T10 being the most common sites (but there is sometimes a multiple shunt). More rarely the fistula may be cervical or even intracranial.

In MRI the conus medullaris appears swollen with a high intensity signal in T2-weighting, the height of which is sometimes extended. This hyperintensity is homogeneous,

centromedullary, with a narrow border of spinal cord with normal intensity. Abnormal perimedullary vascular images with nodular or serpiginous 'no signal' voids in the sub-arachnoid spaces are not always visualised because of the very slow flow, but their absence does not mean that the diagnosis can be eliminated.

Diagnostic medullary arteriography is always essential. Slowing of venous circulation is one of the important elements in the diagnosis, with the absence of visible venous return from the Adamkiewicz artery within a normal time. The fistula must be closed, either by surgery or by endovascular embolisation. Clinical improvement occurs at the same time as regression or disappearance of the T2-weighted hyperintensity. Therapeutic results of surgical and endovascular series are identical. Best results are obtained when there is only a short time between the first symptoms and treatment, but often without improvement in sphincter and sexual disorders. On the other hand, it is important to emphasize that partial recovery may be seen even in paraplegic patients. Given the low risk associated with treatment of these fistulas, medullary arteriography and treatment should be widely indicated, even in elderly patients with many problems.

Intramedullary cavities

A focal distension of the ependymal canal may be found by chance in 1–2% of spinal cord explorations, in two cases out of three in the thoracic region [4] (Fig. 6). For it to be dismissed, it must be filiform or fusiform and less than 3 to 4 mm in diameter and must be situated at the junction between the anterior third and posterior two thirds of the anterior-posterior diameter of the spinal cord, except in the conus where it must be central.

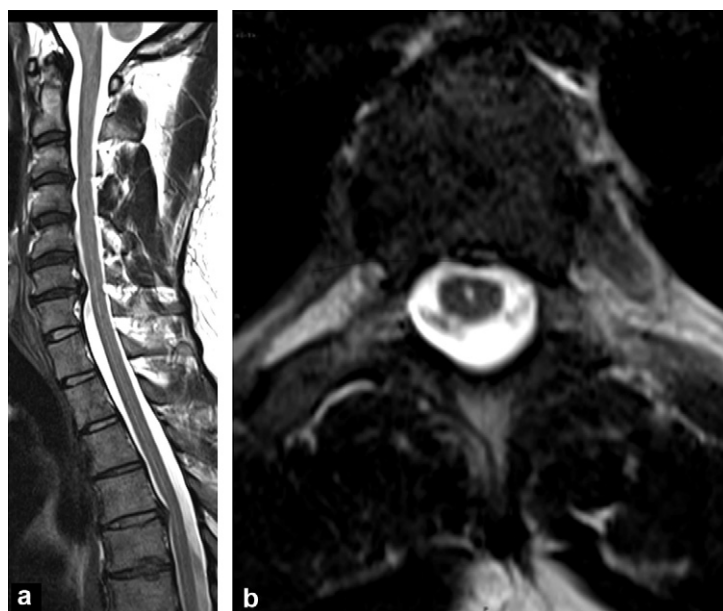


Figure 6. Focal distension of the central canal: a: MRI, sagittal T2-weighted image: high intensity linear image at the C6 level; b: MRI, axial T2-weighted image at C6: the cavity is narrow (less than 3 mm) and situated at the junction between the anterior third and the posterior two thirds of the anterior-posterior diameter of the spinal cord.



Figure 7. Intramedullary cavity secondary to lumbosacral arachnoiditis: a: MRI, cervicothoracic sagittal T2-weighted image: intramedullary cavity extending in height to T3; b: MRI, lumbosacral sagittal T2-weighted image: wide lumbosacral canal, discrete vertebral scalloping, inconspicuous intrathecal roots; c: MRI, axial T1-weighted image after IV gadolinium injection: 'empty bag' appearance, with roots attached to the hyperaemic duramater that enhances after injection.

The cause of any intramedullary cavity, other than this possibility of a benign focal distension of the central canal, must be determined: a Chiari malformation, spinal dysraphism, a tumour, a trauma sequela, arachnoiditis [5] (Fig. 7), etc.

Technical recommendations are to explore the entire spinal canal including the craniovertebral and lumbosacral joints. T1- and T2-weighted sequences should be obtained. To assess CSF movement in the subarachnoid spaces and especially in the cavities, with T2-weighting it is best not to use flow compensation or cardiac or peripheral gating. Axial slices show the topography of the cavity and any communications with the subarachnoid spaces and also allow the residual thickness of the medullary parenchyma to be evaluated. It is not necessary to inject an IV contrast agent if a spinal malformation is found, particularly of the craniovertebral joint and especially a Chiari malformation with cerebellar tonsil ectopia. On the other hand, any medullary cavity of unclear origin requires IV gadolinium injection to detect any enhancement of a cystic wall or tumour nodule (Fig. 8).

Tumour diagnosis

An intramedullary tumour appears as a fusiform swelling of the spinal cord or an intracanal mass, with irregular borders with the spinal cord, which appears as an area of hyperintensity in T2-weighting with or without partial or complete enhancement, and in T1-weighted sequences following gadolinium injection. The location of the tumour is defined according to its cervical, thoracic or conus medullaris topography or sometimes with a pan-medullary extension. The height of the tumour is measured in centimetres and the number of vertebrae it crosses. The shape and the contours of the cord should be described. The diameters



Figure 8. Peritumoral intramedullary cavity on a haemangioblastoma in Von Hippel Lindau's disease: a: MRI, cervicothoracic sagittal T2-weighted image: intramedullary cavity extending in height to the cervicothoracic junction; b: MRI, lumbosacral sagittal T1-weighted image after IV gadolinium injection: peritumoral cysts and tumour nodules enhancing in the conus medullaris.

of the spinal cord and the canal should be measured and erosion of the bone of the vertebral bodies (scalloping) or pedicles described. One of the key points is to determine whether the axial topography of the lesion is central, lateral or exophytic.

MRI provides information on the macroscopic constitution of the lesion: methaemoglobin and melanin (T1-weighted

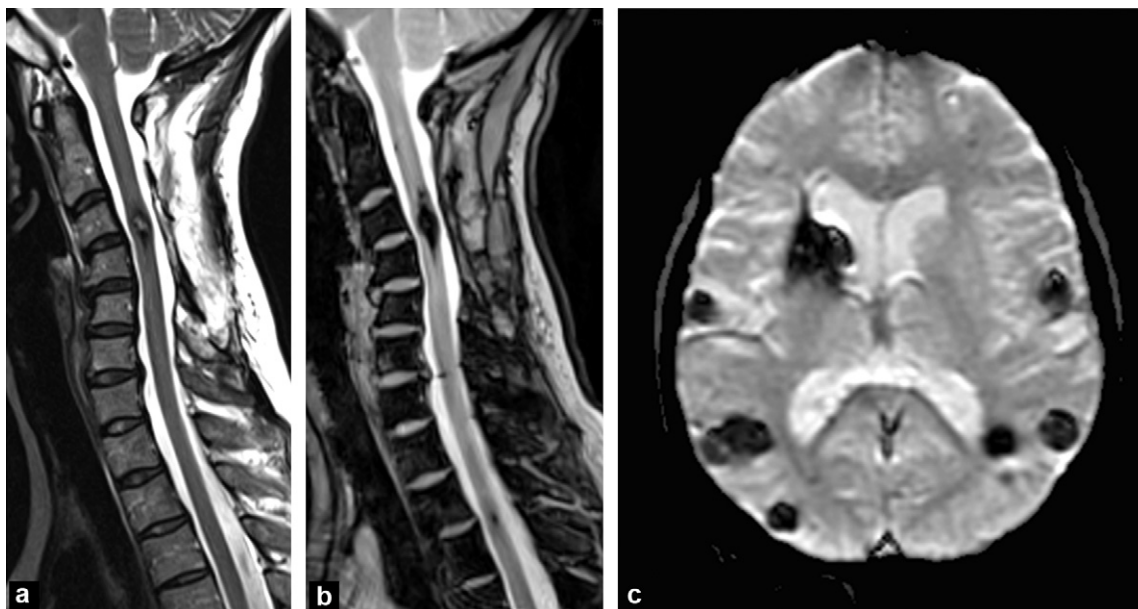


Figure 9. Intramedullary cavernoma: a: MRI, sagittal T2-weighted spin-echo image: nodule level with C3, with centromedullary hyperintensity and a hypointense shell due to haemosiderin deposition; b: MRI: T2*-weighted gradient-echo with the slice in the same plane: hypointensity accentuated on this sequence sensitive to the magnetic susceptibility effect and another lesion level with T2-T3 revealed; c: cerebral MRI, T2*-weighted GE-EPI: cavernomatosis.

spontaneous hyperintensity), haemosiderin (T2-weighted hypointensity, a larger area with T2*-weighting), vascular pedicle and vascular distensions (hypointense intratumoral serpiginous or perimedullary images due to rapid flow), hydro or syringomyelic cavities (iso-intense with CSF in all sequences), associated reaction cysts (no enhancement following IV gadolinium injection), tumoral cysts (mural enhancement following injection), or oedema (discrete hypointensity with T1-weighting, hyperintensity with T2). Barrier rupture will be seen by partial or complete enhancement following injection, which may be nodular or in clumps, and may or may not be intense, while fibres can be imaged with diffusion tensor imaging: an infiltrating tumour entangled in the fibres makes them appear amputated (astrocytoma) while a space-occupying mass pushes aside the fibres (ependymoma). However, a pilocytic astrocytoma also pushes the fibres aside like an ependymoma.

Recognising a cavernoma [6]

A cavernoma is an angiographically occult vascular pseudotumour discovered by chance, if it is not complicated, or by symptoms of progressive deficit or during an acute episode in one in three cases (Fig. 9). The haemorrhagic risk is 1 to 2% per year or more in the case of familial cavernomatosis (10% of cases). It is more commonly located in the thoracic than the cervical region. In one out of four cases, it is associated with cerebral lesions. A cavernoma appears as a nodule or a well-circumscribed polynodular mass, with calcifications and nodules spontaneously hyperintense in T1-weighted sequences. It is surrounded by a hypointense shell in T2-weighted sequences, which is greater in T2*-weighted gradient-echo. Following injection there is no or late enhancement.



Figure 10. Low grade intramedullary ependymoma: a: MRI, sagittal T2-weighted image: the cord is swollen at the cervicothoracic junction and occupies the whole canal at the level of T1-T2, the site of the tumour, which has a superior and inferior polar cap of haemosiderin. Superjacent and subjacent oedema; b: MRI, same sagittal plane in T1-weighting after IV gadolinium injection: the tumour is partially enhanced following the injection.

Suspecting an ependymoma [7]

Ependymomas represent 65% of the spinal tumours in adults between 30 and 40 years of age (Fig. 10). They are very often low grade and their prognosis is better when in the spine than when cerebral. They are especially found in

the cervical region. In two thirds of cases, the tumour is centromedullary, well defined with a cleavage plane (it can be completely ablated more often than an astrocytoma) and evolves slowly. The tumour causes the cord to swell and enlarges the canal. It is heterogeneous, with cysts in 60% of cases, medullary cavities, calcifications, and haemorrhagic foci. The image of the polar tumour cap of haemosiderin, hypointense with T2-weighting, is very characteristic. Enlargement of the canal creates images of 'scalloped' vertebral bodies, and sometimes erosion of the pedicle. Oedema contributes to swelling of the cord. In 80%

of cases, enhancement is complete, and in 70%, homogeneous. In diffusion tensor imaging, the fibres appear to be displaced by the tumour [8].

Suspecting an astrocytoma [7]

Astrocytomas account for 30% of (spinal) tumours in young (20–30 years old) adults, where they are low grade in 75% of cases, and are the most frequent tumour in children, representing 85% of cases (Figs. 11 and 12). They are low grade in 90% of these cases. They occur above all in the thorax

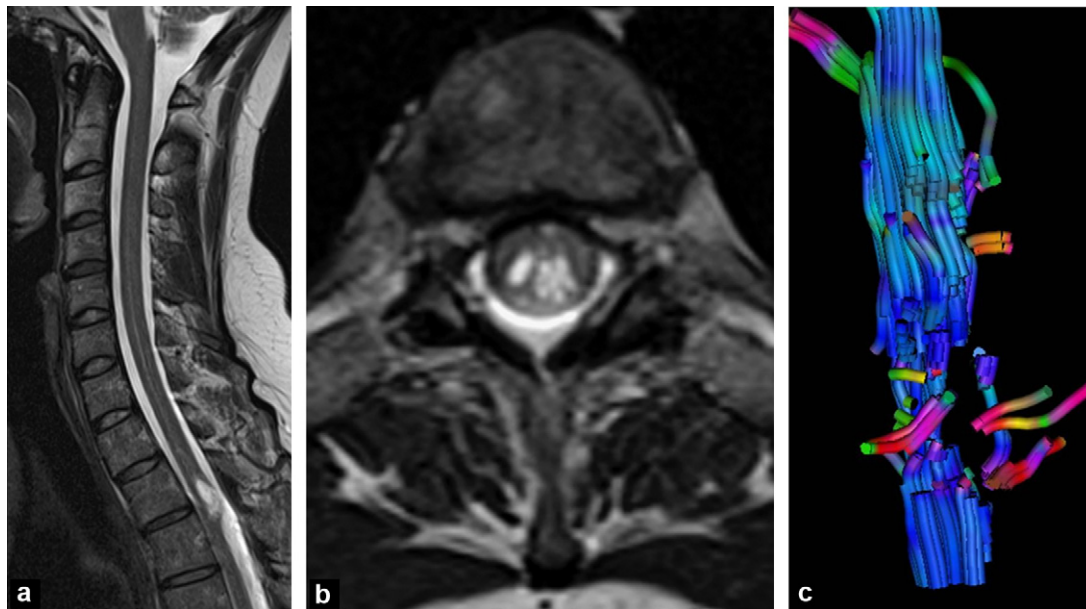


Figure 11. Astrocytoma: a: MRI, sagittal T2-weighted image: the spinal cord is focally swollen and has a hyperintense area; b: MRI, axial T2-weighted image showing the posterior asymmetric location not sparing the funiculi; c: MRI, diffusion tractography: apparent amputation of the fibres at the tumour site.

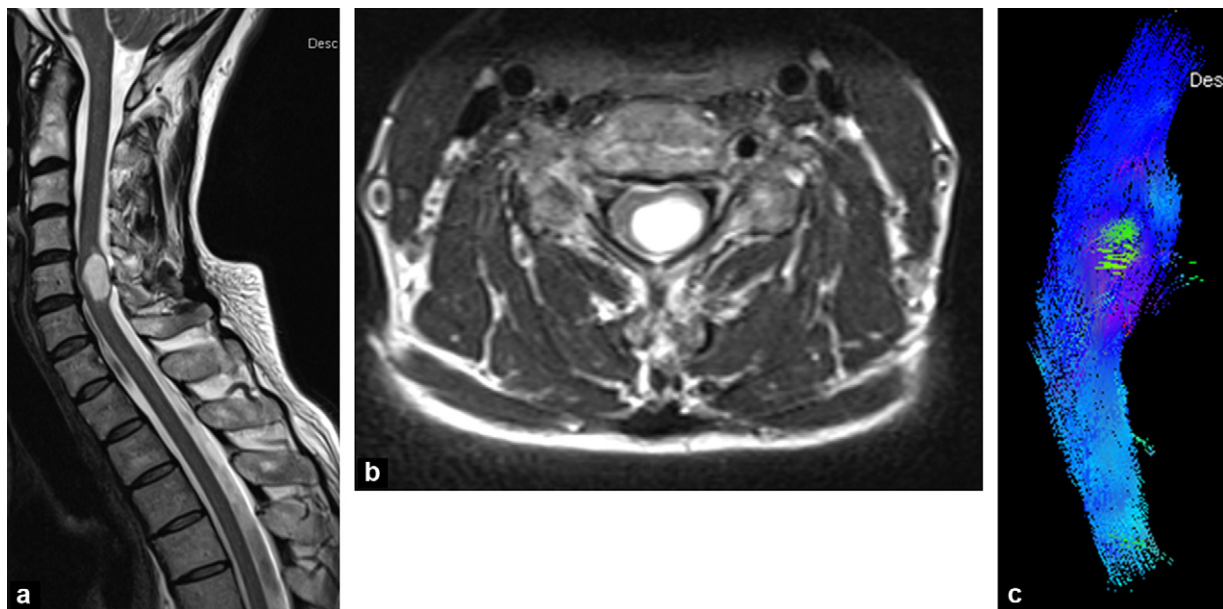


Figure 12. Pilocytic astrocytoma: a: MRI, sagittal T2-weighted image: the spinal cord is focally swollen and has a hyperintense area; b: MRI, axial T2-weighted image showing the left posterior-lateral location of the tumour; c: MRI, diffusion tractography: fibres pushed aside to an equal extent at the site of the tumour, identical to that found with ependymomas.

or at the cervicothoracic junction, but much more rarely can be pan-medullary. In 75% of adult cases, and still more in children, the tumour is grade I or II. It is an infiltrating tumour, with cysts in less than one in three cases. Due to the infiltration, the spinal cord shows fusiform expansion. The lesion is hyperintense in T2-weighted sequences and frequently shows enhancement (70%). It is eccentric in two out of three cases, cystic in only one case in three and exophytic in one in five cases. With diffusion tensor imaging, the fibres appear entangled in the tumour and because of this, complete resection is only possible in one case in three.

Recognising a haemangioblastoma [9] (Fig. 8)

Haemangioblastomas represent 2% of all intramedullary tumours. Seventy-five per cent of them are intramedullary, 20% are radicular and 5% extramedullary. This is a grade I benign tumour in young subjects, and is discovered sporadically or in the context of Von Hippel Lindau's disease. It occurs above all in the cervical region, the cervicothoracic junction and the conus medullaris. It is rarely centromedullary but rather posterolateral opposite the dorsal root. It is an encapsulated tumour presenting as an intensely enhancing hypervascularised nodule after injection and, when the tumour is larger than 2 cm, it has a hypointense tumoral vascular pedicle with a high flow and sinuous, dilated dorsal veins. It is often associated with cystic lesions, intramedullary cavities (9 times out of 10) and considerable oedema. Excision of the enhancing nodule is sufficient for complete ablation.

Pseudotumours

The radiologist's role is to assist in differentiating between tumours and pseudotumours in the epidemiological and clinical (oncological history, HIV) context and using the results of laboratory tests (blood and CSF), and above all to reveal any associated cerebral lesions. There may be:

- inflammatory lesions: MS (often several medullary lesions including some that are atrophying), neuromyelitis optica (a history of optic neuritis, a very extensive lesion in terms of height), ADEM (acute disseminated encephalomyelitis: cerebral lesions with synchronous enhancement), acute transverse myelitis (clinical signs more sudden and rapid than for a tumour);
- neurosarcoidosis granulomatous lesions with predominantly leptomeningeal involvement, dissociation of radioclinical findings with extensive lesions and few clinical symptoms, since isolated medullary locations are exceptional [10];
- tuberculous granulomatous lesions usually as part of a systemic condition with associated pulmonary or bone lesions;
- infectious and parasitic lesions: a bacterial abscess (suggestive diffusion-weighted imaging, a reduced apparent diffusion coefficient (ADC), extension in height along the ependymal axis), schistosomiasis (a characteristic image of conus medullaris hyperintensity in T2-weighted sequences with central linear enhancement and punctate arborised enhancements) [11], cysticercosis (a cystic

image with scolex in the vesicular stage), toxoplasmosis (the nodule enhancing in an immunosuppressed subject, and associated cerebral lesions), cryptococcosis (a nodule in the thoracic cord often hypointense with hyperintense microfoci in T2-weighting, with nodular or annular intense enhancement after injection surrounded by oedema and associated cerebral lesions of identical appearance).

In any exploration of the spinal cord, it should be kept in mind that the latter is only a small part of the central nervous system and that there are sometimes associated cerebral lesions which often more readily suggest a diagnosis than the lesions of the spinal cord.

Moreover, we need to know how to look around the spinal cord. Some associated lesions sometimes help diagnosis: the discovery of spinal metastases in the case of cord lesions or metastatic enhancing and nodular leptomeningeal lesions, the association of bone infarction with ischaemic cord lesions, images of fatty replacement of irradiated vertebrae and post-therapeutic necrotic cord lesions, mediastinal or retroperitoneal lymphadenopathy and pseudotumoral lymphoma lesions or sarcoidosis, cord lesions and pulmonary tuberculosis lesions, pheochromocytoma and haemangioblastoma in Von Hippel Lindau's disease, etc.

TAKE-HOME MESSAGES

- Always think vascular and always think of eliminating this diagnostic hypothesis.
- During exploration of the lumbar spine, always look at the conus medullaris and ensure that it is normal. Take care not to delay diagnosis of a dural arteriovenous fistula with perimedullary drainage, a delay which too frequently occurs.
- The spinal cord is part of the central nervous system and the pathology is often multifocal, hence it is usually useful to perform an additional cerebral examination.
- A lesion of the spinal cord is not always isolated. The key to the diagnosis is sometimes on the spine, in the mediastinum, the retroperitoneum, the lungs, etc.
- Some inflammatory and granulomatous lesions may have the appearance of a tumour. The radiologist needs to know when to put forward these diagnoses and when to repeat the examinations after a therapeutic trial.

Clinical case

Observation

Forty-five-year-old patient presenting paraesthesia and weakness of the legs. MRI has provided sagittal T2-weighted slices (Fig. 13a) and T1-weighted images after gadolinium injection (Fig. 13b and c).

Questions

1. What are the main differential diagnoses?

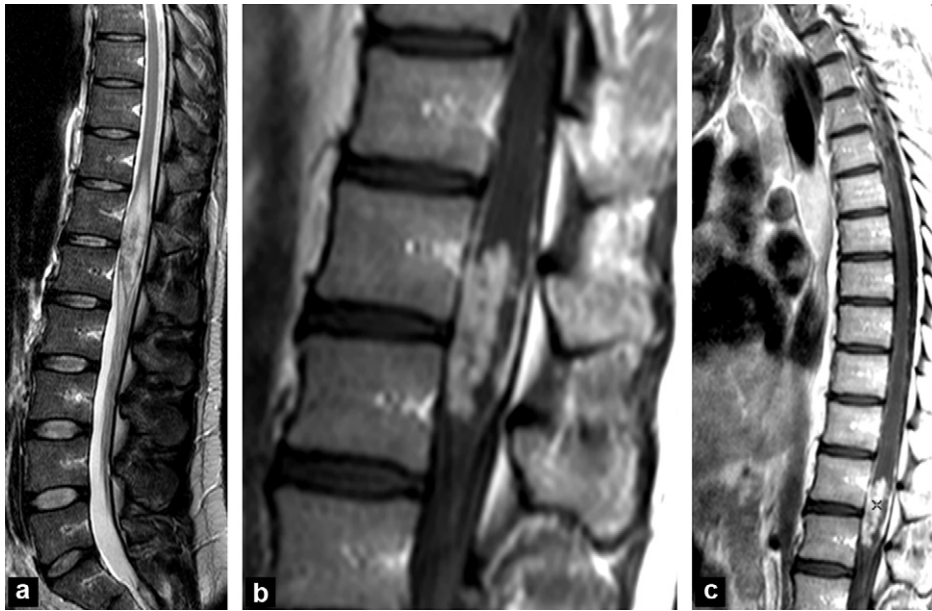


Figure 13. MRI, sagittal T2-weighted image (a), T1-weighted after gadolinium injection (b, c).

2. What diagnoses can be excluded or dismissed, and on what grounds?
3. What are the most likely diagnostic hypotheses and the arguments for or against them?
4. Would another lesion point towards a diagnosis?

Answers

1. There is swelling of the conus medullaris extending over three vertebrae. The lesion area is heterogeneous. There are no intrathecal dilated vessels, no enlargement of the canal, no vertebral scalloping. Enhancement is intramedullary and leptomeningeal perimedullary. The principal differential diagnoses are:
 - vascular, in particular, a dural fistula;
 - tumoral: benign or malignant primary, secondary;
 - inflammatory and infectious: MS, ADEM, vasculitis, viral myelitis, a parasitic condition, etc.;
 - granulomatous: tuberculosis, sarcoidosis.
2. The diagnosis of vascular malformation: no dilated vessels, no signal voids, too great enhancement of the conus medullaris. The inflammatory and secondary tumoral diagnosis: because of the radioclinical differences between a large lesion and the limited clinical symptoms, with no acute phenomena.
3. Tumoral: an astrocytoma or ependymoma is possible. But enhancing leptomeningeal lesions are in this case very unusual.

Parasitic: schistosomiasis is possible but a previous stay in an endemic zone would be necessary.

Granulomatosis: tuberculosis and sarcoidosis.
4. Yes, the patient presents mediastinal lymphadenopathy. Diagnosis: neurosarcoidosis.

Disclosure of interest

The author declares that he has no conflicts of interest concerning this article.

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