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Spinal tumors

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Abstract

Spinal tumors are uncommon lesions but may cause significant morbidity in terms of limb dysfunction. In establishing the differential diagnosis for a spinal lesion, location is the most important feature, but the clinical presentation and the patient's age and gender are also important. Magnetic resonance (MR) imaging plays a central role in the imaging of spinal tumors, easily allowing tumors to be classified as extradural, intradural–extramedullary or intramedullary, which is very useful in tumor characterization. In the evaluation of lesions of the osseous spine both computed tomography (CT) and MR are important.

We describe the most common spinal tumors in detail. In general, extradural lesions are the most common with metastasis being the most frequent. Intradural tumors are rare, and the majority is extramedullary, with meningiomas and nerve sheath tumors being the most frequent. Intramedullary tumors are uncommon spinal tumors. Astrocytomas and ependymomas comprise the majority of the intramedullary tumors. The most important tumors are documented with appropriate high quality CT or MR images and the characteristics of these tumors are also summarized in a comprehensive table. Finally we illustrate the use of the new World Health Organization (WHO) classification of neoplasms affecting the central nervous system.

Keywords: Spinal tumors; Magnetic resonance imaging; Computed tomography

1. Introduction

Spinal tumors are uncommon lesions and affect only a minority of the population. However, these lesions can cause significant morbidity in terms of limb dysfunction and can be associated with mortality as well. In establishing the differential diagnosis for a spinal lesion, location is the most important feature. Magnetic resonance (MR) imaging, which plays the central role in the imaging of spinal tumors, easily allows tumors to be classified as extradural, intradural–extramedullary or intramedullary. Although this classification is somewhat of an oversimplification, since lesions can reside in several compartments, it is very useful in tumor characterization. MR is also very important in the evaluation of lesions of the osseous spine [1].

In general, extradural lesions are the most common (60% of all spinal tumors), with the majority of lesions originating from the vertebrae. The most frequent extradural tumor is metastasis, while primary bone tumors are much less common. Intradural tumors are rare, and the majority is extramedullary (30% of all spinal tumors), with meningiomas, nerve sheath tumors (schwannomas and neurofibromas) and drop metastases being the most frequent. Intramedullary tumors are uncommon spinal tumors (10% of all spinal tumors), and occur most often in the cervical spinal cord. Astrocytomas and ependymomas comprise the majority of the intramedullary tumors, with ependymomas occurring with about double the frequency of astrocytomas.

In 2000, the World Health Organization (WHO) ratified a new comprehensive classification of neoplasms affecting the central nervous system. This tumor classification is based on the premise that each type of tumor results from the abnormal growth of a specific cell type. To the extent that the behavior of a tumor correlates with the basic cell type, tumor classification dictates the choice of therapy and predicts prognosis. The new WHO system is particularly useful in this regard. The WHO classification also provides a parallel grading system for each type of tumor. In this grading system most named tumors are of a single defined grade. The new WHO classification provides the standard

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for communication between different centers around the world. A modified outline of this classification for spinal tumors is provided in Table 1. An overview of the characteristics of the most important tumors is shown in Table 2.

Table 1
Classification of spinal tumors

- Neuroepithelial tumors
  - Astrocytic tumors (glial tumors)
    - Fibrillary astrocytomas (WHO grade II)
    - Anaplastic (malignant) astrocytoma (WHO grade III)
    - Glioblastoma multiforme (WHO grade IV)
    - Pilocytic astrocytoma (WHO grade I)
    - Pleomorphic xanthoastrocytoma (WHO grade II)
  - Oligodendroglial tumors
    - Oligodendroglioma (WHO grade II)
    - Anaplastic (malignant) oligodendroglioma (WHO grade III)
  - Ependymal cell tumors
    - Ependymoma (WHO grade II)
    - Anaplastic ependymoma (WHO grade III)
    - Myxopapillary ependymoma (WHO grade I)
    - Subependymoma (WHO grade I)
  - Mixed gliomas
    - Mixed oligoastrocytoma (WHO grade II)
    - Anaplastic (malignant) oligoastrocytoma (WHO grade III)
  - Neuronal and mixed neuronal–glial tumors
    - Gangliocytoma (WHO grade I)
    - Ganglioglioma (WHO grade I/II)
    - Anaplastic (malignant) ganglioglioma (WHO grade III)
    - Desmoplastic infantile ganglioglioma (WHO grade I)
    - Dysembryoplastic neuroepithelial tumor (DNET) (WHO grade I)
    - Paraganglioma (WHO grade I)
  - Neuroblastic
    - Neuroblastoma
  - Embryonal tumors
    - Ependymoblastoma (WHO grade IV)
- Peripheral nerve tumors
  - Schwannoma (neurinoma, neurilemoma) (WHO grade I)
  - Neurofibroma (WHO grade I)
  - Malignant peripheral nerve sheath tumor (WHO grade III/IV)
- Hematopoietic tumors
  - Primary malignant lymphomas
  - Plasmacytoma
  - Leukemia
- Germ cell tumors
  - Embryonal carcinoma
  - Teratoma
  - Mixed germ cell tumors
- Tumors of the meninges
  - Meningothelial tumors
    - Meningioma (WHO grade I)
    - Atypical meningioma (WHO grade II)
    - Anaplastic (malignant) meningioma (WHO grade III)
  - Mesenchymal, non-meningothelial tumors
    - Lipoma
    - Angiolipoma
    - Hibernoma
    - Fibrosarcoma
    - Malignant fibrous histiocytoma
    - Chondroma, chondrosarcoma
    - Osteoma, osteosarcoma
    - Osteochondroma
    - Hemangioma
    - Hemangiopericytoma
  - Melanocytic lesions
    - Melanocytoma
    - Malignant melanoma
    - Meningeal melanocytosis
  - Tumors of unclear origin
    - Hemangioblastoma
- Metastatic tumors
- Primary bone tumors
  - Aneurysmal bone cyst
  - Chordoma
  - Chondrosarcoma
  - Ewing sarcoma
  - Fibrosarcoma
  - Giant cell tumor
  - Hemangioma
  - Histiocytosis
  - Lymphoma
  - Myeloma
  - Osteoid osteoma
  - Osteoblastoma
  - Osteosarcoma

2. Magnetic resonance imaging of spinal tumors

In magnetic resonance imaging of the cervical and thoracic spine, patients are positioned supine, head first. When imaging the lower thoracic spine, patients can be positioned feet first if they are not too tall. Otherwise, their feet may reach the end of the magnet bore before they are in the correct position. If the magnet is open on both sides, this may not be a problem. Children are positioned feet first for thoracic examinations. We use a knee support for patient comfort, which in turn is favorable for the image quality since patients are less likely to move during the examination. Moreover, the use of a knee support flattens the lordotic curvature of the lumbar spine, thus diminishing the distance between the spine and the surface coil, which again improves signal-to-noise [2]. MR imaging of the lumbar and sacral spine is best performed with the patient in supine position and feet first. This tends to decrease claustrophobic reactions. The patient should be positioned as comfortably as possible to minimize movement artifacts. On high field magnets with strong gradients, earplugs or a headphone should be provided to the patient.

When imaging the thoracic spine, it can be useful to attach one or more markers to the skin of the patient's back prior to starting the examination. These markers simplify the problem of determining the examined levels afterwards.

In screening spinal MR examinations, for example, in spinal metastases, 'full spine MR' using automatic table movement and image stitching can be very useful.

Table 2
Overview of the most common spinal tumors and their characteristics

Lesion	Location		Incidence	Age	Clinical	Plain film/CT	MR			Cysts	Calcification	Hemorrhage
							T1	T2	Gd			
Astrocytoma	Intramedullary	T	Second most frequent intramedullary	20–40	Local > irradiating pain; cord symptoms sometimes late	−	−/0	+	+	+	−	−
Ependymoma	Intramedullary	Conus; C	Most frequent intramedullary	30–50	Local > irradiating pain; cord symptoms sometimes late	Sometimes scalloping/erosion	−; + (mp)	+	++	+	−	+
Hemangioblastoma	Intramedullary, some extramedullary	T	Rare	30–40	Due to cerebellar lesion; motorsensory	Sometimes widening	−/0	+	++	+	0	+
Meningioma	Extramedullary	T	Second most frequent extramedullary	40–60	Local > irradiating pain	Sometimes calcification	−/0	0/+	++	−	+	−
Nerve sheath	Extramedullary; some extradural	T	Most frequent extramedullary	30–40	Radicular pain; paresthesias, numbness	Scalloping	0	++	0/+	0/+	−	0
Paraganglioma	Extramedullary	Conus	Rare		Local > irradiating pain	−	0	+	++	0/+	−	+
Hemangioma	Extradural	T	Most common benign spinal tumor	All	Sometimes local pain	RX: “jail bar”; CT: “polka-dot”	+	+	+	−	−	−
Plasmacytoma	Extradural	T	Uncommon	40–60	Local > irradiating pain	Lytic, expansile, “mini-brain”	−	+	+	−	−	−
Multiple myeloma	Extradural	T	Most common primary malignant	50–70	Local pain	Punched out lytic, osteopenia, collapse	−	+	+	−	−	−
Metastasis	Extradural	T	Most common malignant	50–70	Significant local pain	Irregular lytic, sometimes sclerotic	−	+	+	−	−	−
Chordoma	Extradural	S	Rare	40–60	Local pain	Bone destruction, amorphous calcific.	0	+	0/+	0/+	−	0/+
Osteoid osteoma	Extradural	L	Common	10–20	Unrelenting pain, worse at night; sensitive to aspirin	Dense sclerosis, lucent nidus, calcifications	−	−/+	++	−	+	−
Aneurysmal bone cyst	Extradural	LS	Rare	10–20	Local pain	Expansile lytic, multiloculated, egg shell	−/+	−/+	0	++	−	+
Giant cell tumor	Extradural	S	Rare	20–50	Local pain	Expansile lytic	−	+	0/+	+	−	+
Eosinophilic granuloma	Extradural	T	Rare	0–10	Variable; sensitive to aspirin	Lytic with sharp borders	−/+	+	++	−	−	−

Patients should be cleared for any MR imaging contraindications before they enter the magnet room. They also should be informed of the benefits and potential risks of MR imaging before the examination is performed. In particular, before administering intravenous contrast products, informed consent (written or oral) should be obtained.

The scenario and length of the examination should be explained in understandable terms. It is helpful to keep the patient informed during the examination about the length of each sequence. That way, swallowing and movement can be minimized since patients will have a better idea about the time they must keep still.

When examining children, we allow a parent(s) to be present in the magnet room. With young children (<6 years), parents are sometimes placed together with their child inside the magnet, lying in prone position, head to head with the child. With anxious patients, it can be helpful to leave a member of the nursing staff inside the magnet room to calm the patient down when necessary. Anxiolytic medication can be beneficial in claustrophobic patients.

Metallic implants used for spinal fusion are not a contraindication for MR imaging. Superparamagnetic materials, e.g. stainless steel, however, will create severe susceptibility artifacts [3]. TSE sequences are less susceptible than SE sequences, which in turn are less susceptible than GRE sequences [4]. Also, shortening the echo time and increasing the bandwidth of the sequence lessens artifacts. If a particular region is not interpretable due to artifacts, it may be worthwhile trying to swap read- and phase-encoding directions. Non-superparamagnetic metals such as titanium only produce RF artifacts, which are less important.

Spinal stimulators and other electronic implant devices (morphine pump, pacemaker, cochlear implant, etc.) in principle constitute an absolute contraindication for MR imaging. Some types of electronic implants, however, are 'MR-compatible'. This should be checked with the manufacturer and the surgeon or clinician who implanted the device, before the patient is brought into the magnet room. In any case, these devices should be switched off before the MR examination. Patients have to be carefully instructed to call during the examination when they have the impression the device is turned on again, or in any case they sense something unusual.

When available, phased-array spine coils should be used since they make coil selection and patient positioning in relation to the coil less critical. The simultaneous use of several phased-array coils allows for a larger FOV. Moreover, new software solutions, known as parallel acquisition techniques (PAT, e.g. SENSE), permit a considerable reduction in acquisition time using phased-array coils in spine imaging with a large FOV.

In general, both sagittal and axial images are obtained in imaging spinal tumors. T1- and T2-weighted images (WI) offer different and complementary information. T2WI are superior in detecting intramedullary spinal tumors. On the other hand, T1WI are more sensitive than conventional non-fat suppression TSE/FSE T2WI in detecting bone-marrow disease, e.g. vertebral metastases, but short T1 inversion recovery (STIR) or other fat suppression T2WI are also able to increase the detection of certain bone marrow diseases. Suppressing the high signal of CSF in T2WI is very useful in detecting subtle intramedullary lesions. The most common technique to obtain this kind of images is FLAIR (fast low angle inversion recovery), an inversion recovery sequence that nulls out CSF signal.

In imaging spinal tumors, one should always perform axial and sagittal TSE T1WI after gadolinium injection. These contrast-enhanced images can be important in tumor detection, delineation and tumor characterization and grading [5]. Some authors recommend contrast-enhanced 3D-GRE T1WI techniques in screening for intradural tumor dissemination [6].

In screening for vertebral metastases, additionally a sagittal GRE so-called out-of-phase sequence can be used. This is a sequence with a specific echo time (TE) corresponding to the time it takes for water and fat protons to move exactly 180° out-of-phase. This time depends on the field strength of the magnet and is 6.9 ms for a 1.5 T imager, and 10.4 ms for a 1.0 T machine. In the normal adult human, the medullary bone of the vertebral bodies contains approximately equal amounts of water and fat protons [7]. In out-of-phase conditions, the signal of both will cancel out, leaving the vertebrae completely black. In case of vertebral pathology, however, the signal will increase and, as such, vertebral metastases (or other lesions) will clearly stand out.

A typical slice thickness for sagittal sequences in the spine is 3–4 mm. We prefer to use an uneven number of slices so that the middle slice is precisely centered on the midpoint of the spinal cord. For axial slices, one can use 3–4 mm slices in the neck and 4–5 mm slices in the lumbar region. Thicker slices, although generating a significantly better SNR, are unsatisfactory in depicting small lesions due to partial volume effects.

Diffusion weighted imaging (DWI) is a special technique using very strong magnetic gradients, effectively canceling signal from protons in free moving water, e.g. CSF. Protons that are more restricted in movement, e.g. in intracellular water, however, still produce a measurable MR signal. The spontaneous movement of protons is known as 'Brownian motion' and results among others in diffusion, hence the term DWI.

This technique is especially useful in detecting cytotoxic edema, where there is cell swelling effectively increasing the amount of intracellular over extracellular water and thereby reducing water diffusion. Since ischemia produces cytotoxic edema very early on (±after 1 h), DWI is capable of early detection of ischemic lesions. However, other applications are emerging, for example, in differentiating tumors from abscesses.

3. Intramedullary tumors

Although a considerable number of different intramedullary tumors exists, only few of them are expected to be encountered in a routine practice. Ependymoma is the most frequent intramedullary tumor, followed by astrocytoma. Together they make up for about 95% of all intramedullary tumors [8]. Astrocytomas are more frequent in the cervical and especially the thoracic region, while ependymomas are mostly seen in the region of the conus medullaris, filum terminale and cauda equina. Hemangioblastoma is the third most frequent intramedullary tumor.

3.1. Astrocytoma

3.1.1. General

The peak incidence for spinal astrocytomas is in the third and fourth decade and the average age at presentation is 30 years. This tumor is also frequently seen in children, where it is the most common intramedullary mass lesion [9]. There is no sexual predilection. More than half of all spinal astrocytomas are seen in the thoracic region, especially the upper thoracic cord. Usually multiple segments are involved.

3.1.2. Clinical presentation

Clinical symptoms are aspecific, including local or less frequently irradiating pain. Clinical evolution is much more rapid than in ependymomas and there often is only a short interval between onset of neurologic symptoms and permanent disability or death. Motor weakness, gait problems and bowel and bladder dysfunction sometimes only present in the later stage of the disease, thereby delaying the diagnosis.

3.1.3. Pathology

The majority (75%) of spinal cord astrocytomas are low grade (WHO I or II). In children, 85–90% are low grade. Glioblastoma multiforme is seen in less than 1.5% of all spinal cord astrocytomas. Astrocytoma diffusely invades the spinal cord and is difficult to delineate (fibrillary type). At the time of diagnosis they have usually progressed over several segments and give rise to a fusiform expansion of the spinal cord. Cystic components are seen in 30% of these tumors. Sometimes these are benign syrinxes, not lined by tumor, that are seen rostral and caudal of the tumor. Calcification is rare.

3.1.4. Imaging

Astrocytomas usually are hypo- to isointense on T1WI and hyperintense on T2WI. They present as focal enlargements of the spinal cord. Despite the fact they are low grade, nearly all spinal astrocytomas enhance following contrast administration (Fig. 1). Although the tumor margins are irregular and not always clear defined, usually after gadolinium administration the tumor is better delineated from edema, cysts and syrinxes. Tumor cysts are generally surrounded

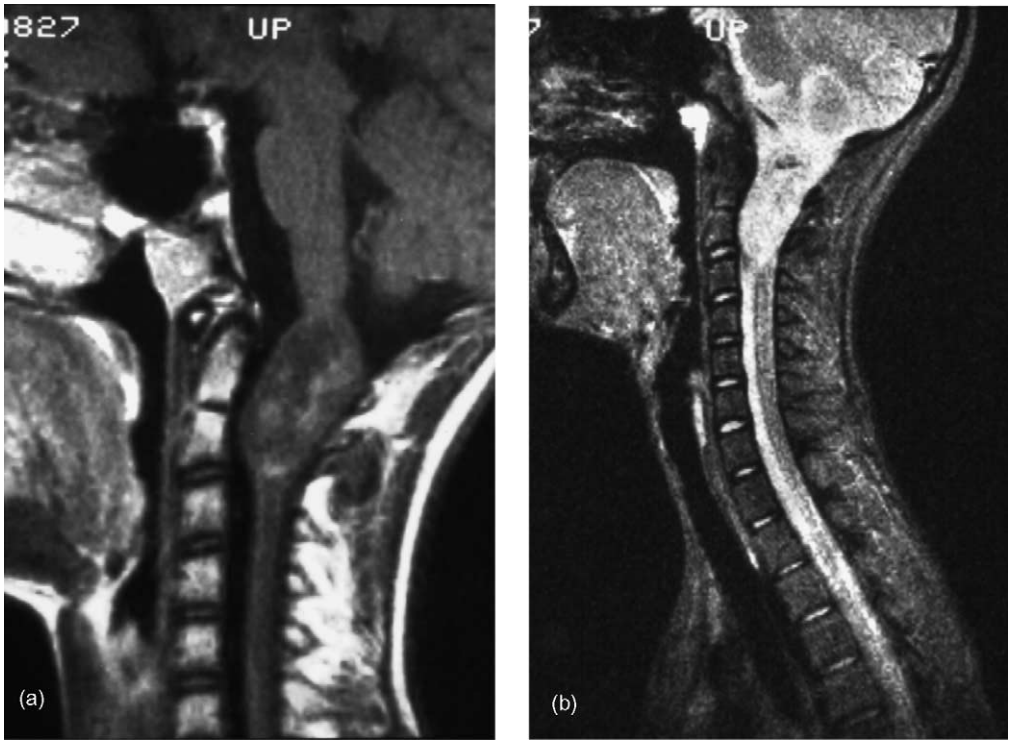


Fig. 1. Astrocytoma. Most spinal gliomas are low grade. Contrary to their cranial counterparts they usually show some enhancement (a). They also tend to be somewhat heterogeneous and often cystic components are recognized (b). Cystic components surrounded by enhancement usually are tumor cysts those not surrounded by enhancement probably represent syrinx formation.

by enhancement, while the lining of a syrinx does not show contrast uptake.

3.2. Ependymoma

3.2.1. General

The peak incidence for spinal ependymomas is in the fourth and fifth decade and the median age at presentation is 35 years [10]. There is no sexual predilection. Ependymomas most often occur in the conus medullaris and the filum terminale. One type of ependymoma, the myxopapillary form, is almost exclusively seen in these locations (95%). More than halve of all tumors of the filum terminale and conus medullaris are myxopapillary ependymomas. Intramedullary ependymoma is most often found in the cervical cord.

3.2.2. Clinical presentation

Clinical symptoms of intramedullary ependymomas are aspecific, including local or less frequently irradiating pain. Neurologic deficits only present in the later stage of the disease, thereby delaying the diagnosis. Myxopapillary ependymomas also present with local back pain and leg or sacral pain. Motor weakness or sphincter dysfunction is only seen in 25% patients at presentation.

3.2.3. Pathology

The most common histology is that of the cellular type. These tumors tend to be intramedullary and are well circumscribed. Cyst formation and hemorrhage is common, especially at the tumor margins. Calcification is extremely uncommon in spinal ependymomas. Myxopapillary ependymomas constitute about 30% of all ependymomas and occur almost exclusively in the conus medullaris and filum terminale. They have an even greater tendency for hemorrhage, which may lead to subarachnoid bleeding.

3.2.4. Imaging

Ependymomas commonly are hyperintense on T2WI. Myxopapillary ependymoma is usually hyperintense on T1, whereas most non-myxopapillary ependymomas are hypointense [11]. They present as focal enlargements of the spinal cord, sometimes visible on plain film as scalloping of the vertebral bodies and/or erosion and widening of the pedicles. These tumors are sometimes heterogeneous due to hemorrhage, although bleeding occurs less often than in brain ependymomas [12]. Sometimes dark caps are seen, especially on T2WI, rostral and caudal of the tumor representing hemosiderin deposits (Fig. 2). After contrast administration intense enhancement is the rule,

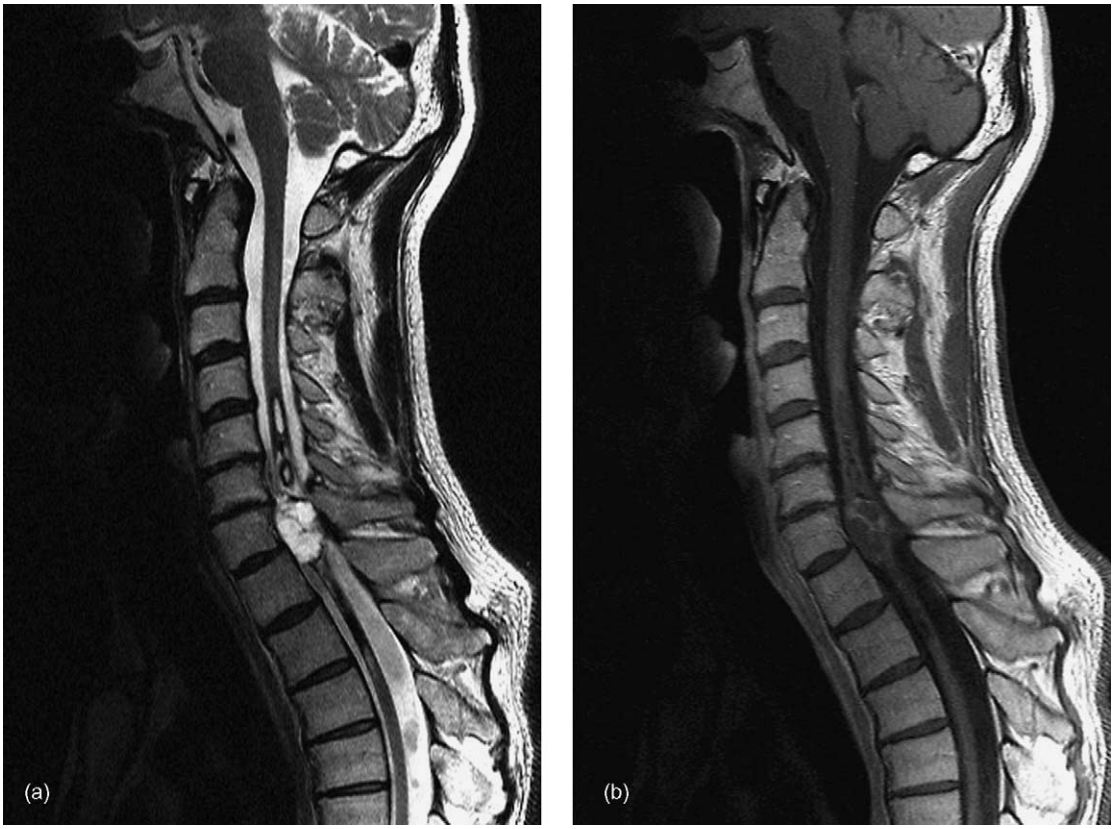


Fig. 2. Ependymoma. Ependymoma is a heterogeneous tumor, especially on T2WI (a). Dark areas above and below the tumor are typical hemosiderin caps after tumor bleeding. Enhancement of spinal ependymoma can be intense, but often also heterogeneous (b) or rim-like.

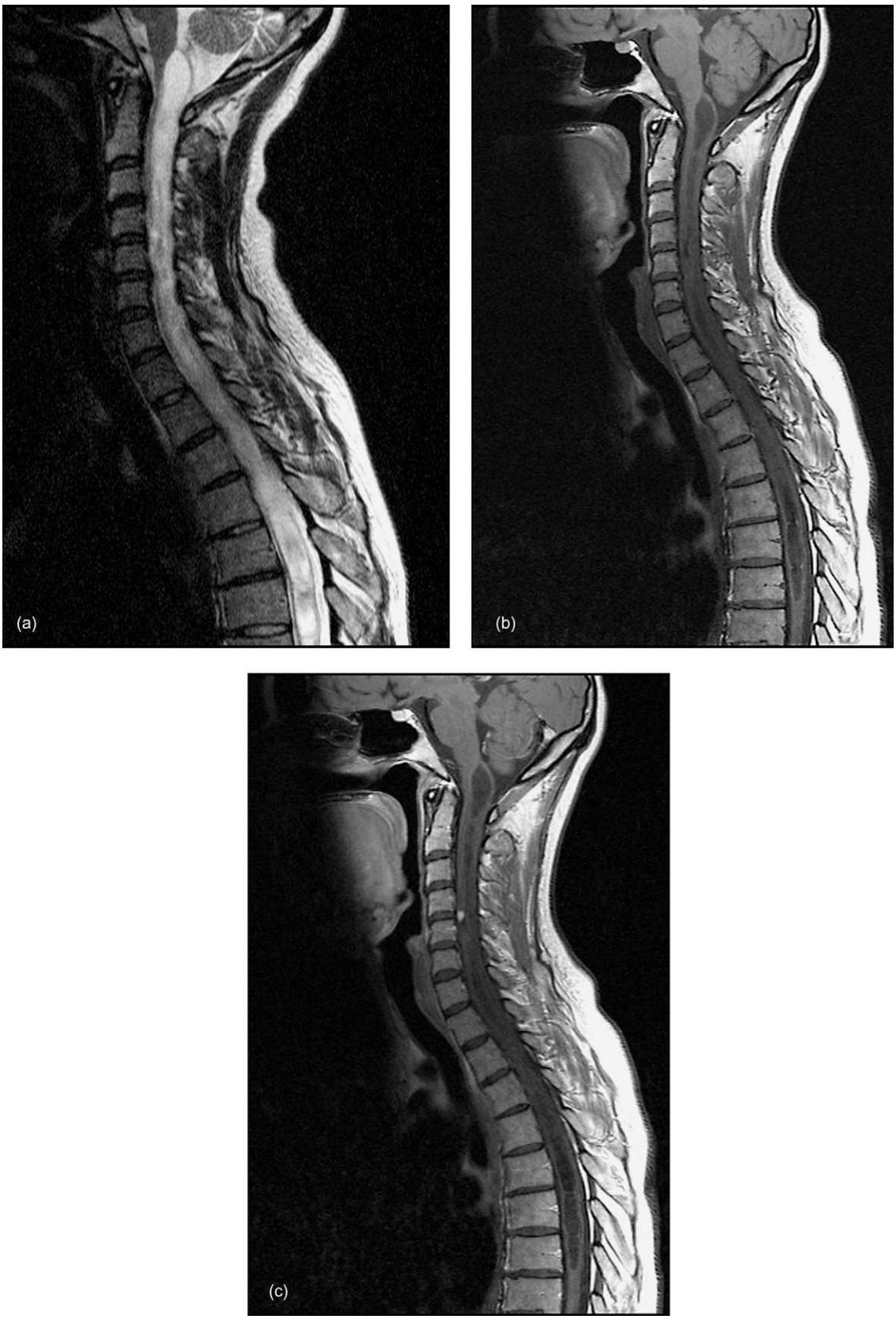


Fig. 3. Hemangioblastoma. Hemangioblastomas almost always have associated cyst formation. Symptomatic small hemangioblastomas have relatively large associated syringes (a), whereas asymptomatic ones do not. After contrast administration intense enhancement of the solid portion of the tumor is the rule (b and c). Delineation of the nidus of the tumor from surrounding edema and cysts is easier after gadolinium enhancement (c).

sometimes homogeneous, but often also heterogeneous of rim-like [11]. Delineation of cysts is easier with gadolinium enhancement.

3.3. Hemangioblastoma

3.3.1. General

The peak incidence for spinal hemangioblastomas is in the fourth decade and the median age at presentation is 35 years [13]. There is no sexual predilection. About one third of all patients with spinal hemangioblastomas have von Hippel-Lindau syndrome. This is an autosomal dominant disease with cerebellar and/or spinal hemangioblastoma, retinal angiomatosis, renal cell carcinoma and/or pheochromocytoma in varying degrees. Hemangioblastomas are found more often in the thoracic than in the cervical region. In about 75% of cases these are intramedullary lesions but sometimes they are extramedullary intradural or even extradural. Most are solitary lesions, but multiplicity is seen in up to 20% of patients.

3.3.2. Clinical presentation

In cases of von Hippel-Lindau syndrome the cerebellar lesions usually produce the first clinical symptoms. Although spinal hemangioblastomas are only seen in 5% of these patients, screening spinal MR imaging should be performed. A spinal hemangioblastoma typically produces motorsensory disturbances.

3.3.3. Pathology

Usually spinal hemangioblastoma is composed of a highly vascular solid portion with small arteries, capillaries and large dilated draining veins. This tumor nidus consists mainly of endothelial cells and interspersed stromal cells. Accompanying this solid portion are large cysts lined by fibrillary cells.

3.3.4. Imaging

Spinal hemangioblastomas may produce focal enlargements of the spinal cord, sometimes visible on plain film as widening of the spinal canal. Small (10 mm or less) hemangioblastomas are mostly isointense on T1WI images, hyperintense on T2WI images, and show homogeneous enhancement. Larger hemangioblastomas tend to be hypointense or mixed hypo- and isointense on T1WI images, heterogeneous on T2WI images, and tend to show heterogeneous enhancement [14]. Feeding arteries or more often large draining veins can be seen around the tumor nidus. Associated cysts may have signal intensities comparable to CSF, but sometimes a rich protein content results in a higher signal intensity on T1WI. Most of these tumors have surrounding areas with high signal on T2WI and low signal on T1WI as a result of peritumoral edema [15]. Symptomatic small hemangioblastomas have relatively large associated syringes, whereas asymptomatic ones do not [14] (Fig. 3). After contrast administration intense enhancement of the solid portion of the tumor is the rule. Delineation of the nidus of the tumor from surrounding edema and cysts is easier after gadolinium enhancement.

4. Extramedullary intradural tumors

Meningiomas and nerve sheath tumors make up for about 90% of all extramedullary intradural tumors. Nerve sheath tumors are the most frequent intraspinal mass lesion, accounting for 30% of all cases. Spinal paraganglioma and metastases are more uncommon extramedullary intradural tumors.

4.1. Meningioma

4.1.1. General

Spinal meningiomas occur most frequently in older patients. The peak incidence for spinal meningiomas is in the fifth and sixth decade. About 70% is found in females. In general 80% of meningiomas are found in the thoracic region, 15% are cervical and 5% are lumbosacral. In men, however, only half of the meningiomas are in the thoracic region and another 40% are cervical. In the thoracic region most meningiomas are posterolateral.

4.1.2. Clinical presentation

Meningiomas usually present with local or less frequently irradiating pain. Motor weakness, paresthesias, gait problems and bowel and bladder dysfunction are less frequent.

4.1.3. Pathology

Spinal meningiomas arise from persistent arachnoid cells and not from the dura. They may attach to nerve roots, spinal cord and especially the dura. Almost all spinal meningiomas are well-circumscribed and slow-growing tumors that are associated with good patient outcomes following surgery. Spinal meningiomas occurring in younger patients may be more aggressive, with a worse prognosis [16]. Histologically, meningothelial, psammomatous, angiomatous and fibroblastic types are recognized. Spinal meningiomas frequently have calcifications (75%).

4.1.4. Imaging

Spinal meningiomas are iso- to hypointense on T1WI and slightly hyperintense on T2WI. There is a strong and homogeneous enhancement with gadolinium, except for the calcified areas. Some meningiomas are heavily calcified and dark on all sequences with only little contrast uptake. These tumors are usually solitary, have a broad attachment to the dura and are usually located posterolateral in the thoracic region and anteriorly in the cervical region (Fig. 4). They are well circumscribed and clearly delineated from the spinal cord.

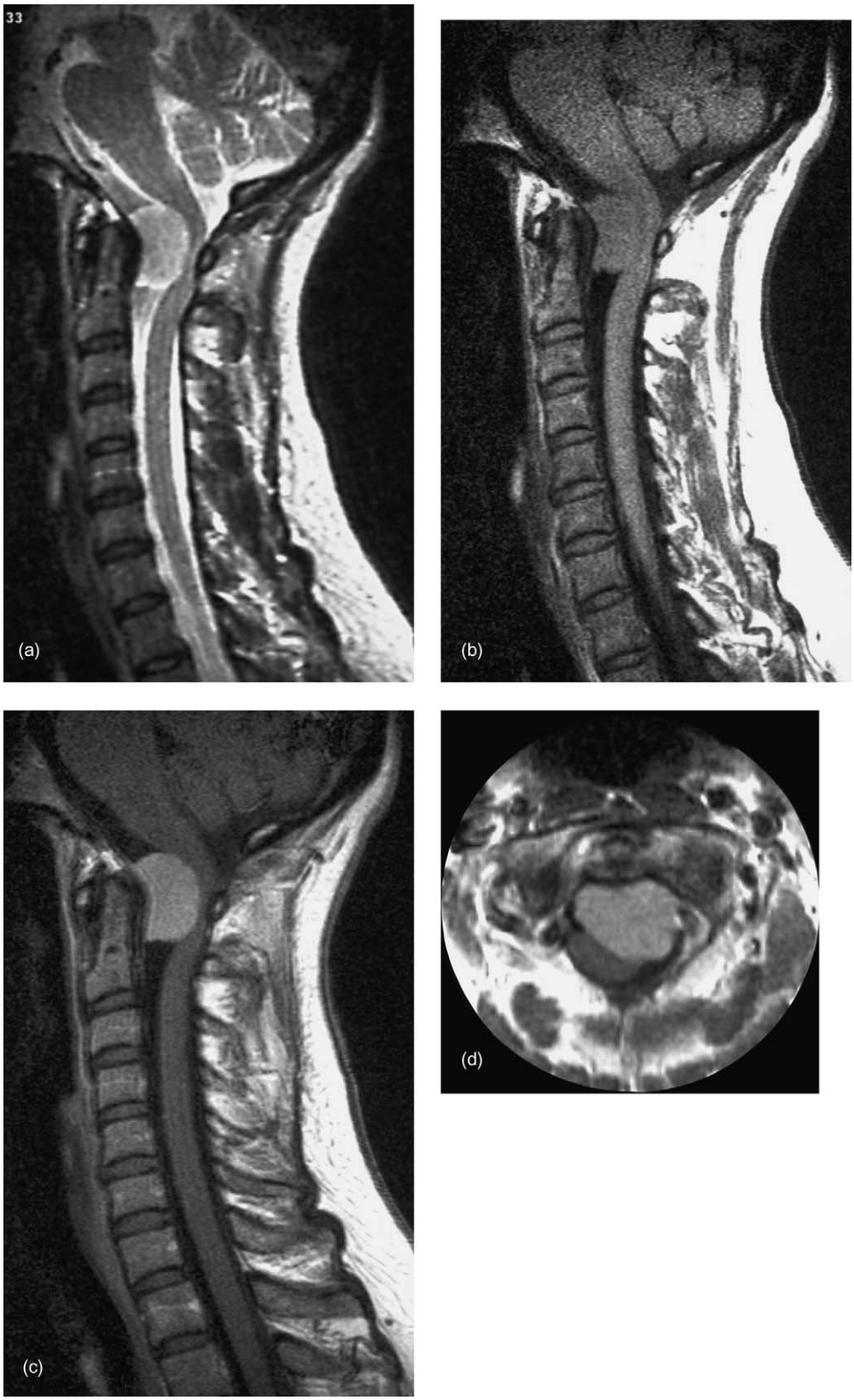


Fig. 4. Meningioma. Meninigiomas are extramedullary tumors, usually with a broad dural base. In the cervical region they have a typical anterior position (a). Non-calcified tumors can have a relatively homogeneous signal intensity, high on T2-WI (a) and isointense on T1WI (b). Also homogeneous contrast enhancement is the rule (c and d).

4.2. Nerve sheath tumor

4.2.1. General

Different types of spinal nerve sheath tumors are recognized. Schwannomas, also known as neuromas or neurilemomas, usually are solitary tumors, and are the most frequent type. By contrast, neurofibromas are sometimes multiple, and even when single are often associated with neurofibromatosis, which is present in 35–45% of all patients with spinal nerve sheath tumors. A third type of neurogenic spinal tumor, ganglioneuroma, is very rare. The peak incidence for spinal nerve sheath tumors is in the fourth decade. These tumors are less frequently seen in children. There is no sexual predilection. These tumors are most often seen in the cervical or lumbar region, followed by the thoracic region. Typically, they are found on the dorsal sensory roots. More than half of all these lesions are extramedullary intradural, about 25% are completely extradural, some (15%) are both intra- and extradural, and very rarely they are seen intramedullary (<1%).

4.2.2. Clinical presentation

Due to their frequent location on the nerve roots patients with nerve sheath tumors most often present with radicular pain or radiculopathy. Paresthesias, numbness or motor weakness can also be present. Typically, this pain is more pronounced at night.

4.2.3. Pathology

The majority (>90%) of spinal nerve sheath tumors are benign. Sarcomatous degeneration is especially seen in patients with neurofibromatosis. Neurofibromas encase the nerve roots, while neurinomas more frequently develop asymmetrically and displace the nerve root. *Neurinomas* are composed of Schwann cells associated with fibrous material. Nerve fibers do not run through the tumor. They often show cystic degeneration and hemorrhage, but no calcification. *Neurofibromas* are composed of Schwann cells, fibroblasts and nerve fibers in a matrix with mucopolysaccharides, fluid and fibrous material. Cyst formation is rare. Plexiform neurofibromas occur in patients with neurofibromatosis type 1 (NF1). In NF1, spinal tumors cause neurological symptoms in about 2% of patients. Only (spinal) neurological deficits in patients with NF1 should prompt further (spinal) diagnostic clarification [17].

4.2.4. Imaging

Nerve sheath tumors are isointense on T1WI and have a typical marked high signal on T2WI (Fig. 5). Sometimes a target appearance with centrally less high signal is noticed. Enhancement is variable and can be intense and homogeneous in some lesions, while others may only show faint peripheral enhancement. Differentiation of neurinoma and neurofibroma on imaging is not reliable. Inhomogeneity can be seen in both benign and malignant nerve sheath tumors. Malignant lesions do tend to have more irregular and invading margins compared to benign lesions, and usually are larger at the time of diagnosis. Multiple tumors are typical of neurofibromatosis type 1 (Fig. 6).

4.3. Paraganglioma

Paraganglia are the accessory organs of the peripheral nervous systems. Paraganglioma are most often seen in the adrenals (known as pheochromocytoma), carotid body and glomus jugulare. Spinal paraganglioma are usually found in the conus medullaris, cauda equina or filum terminale (Fig. 7). They present as inhomogeneous lesions, isointense on T1WI and hyperintense on T2WI. Hemorrhage, intratumoral and adjacent vessels with flow voids and strong enhancement are the rule. Sometimes there are associated cysts [18]. Even though paragangliomas in general are benign and slowly growing their growth pattern can vary and be more aggressive, to the point of metastatic spread [19].

5. Extradural tumors

In general, extradural lesions constitute the most frequent group of spine tumors. Most originate from the vertebrae. The most frequent extradural tumor is metastasis, while primary bone tumors are much less common. Solitary vertebral lesions are less common than tumors with multiple locations. Benign lesions are usually asymptomatic, incidentals findings, malignant vertebral tumors give back pain and sometimes manifest neurologic symptoms, especially in children. MR imaging is the best imaging modality, but bone scintigraphy is useful in detecting multiple lesions and distant metastases. Computed tomography (CT) is also important in imaging these lesions and sometimes shows typical patterns of bone destruction, sclerosis and/or remodeling (Fig. 8).

The most frequent malignant extradural tumor is metastasis. Other malignant tumors include lymphoma–multiple myeloma, chordoma, Ewing sarcoma and chondrosarcoma. The most frequent benign lesions are hemangioma, enostosis (simple bone island), osteoid osteoma–osteoblastoma, aneurismal bone cyst, giant cell tumor, eosinophilic granuloma (histiocytosis) and osteochondroma. Besides imaging findings, the age of the patient and the multiplicity and location of the lesion(s) are most important in the differential diagnosis.

5.1. Vertebral hemangioma

5.1.1. General

The incidence of vertebral hemangiomas increases with age, peaking around the fourth to sixth decade. Two types of vertebral hemangioma should be discerned: asymptomatic lesions, and aggressive, symptomatic types, with compression of the spinal cord [20]. Especially the symptomatic lesions occurs somewhat more frequently in women. Vertebral hemangioma is the most common benign spinal tumor.

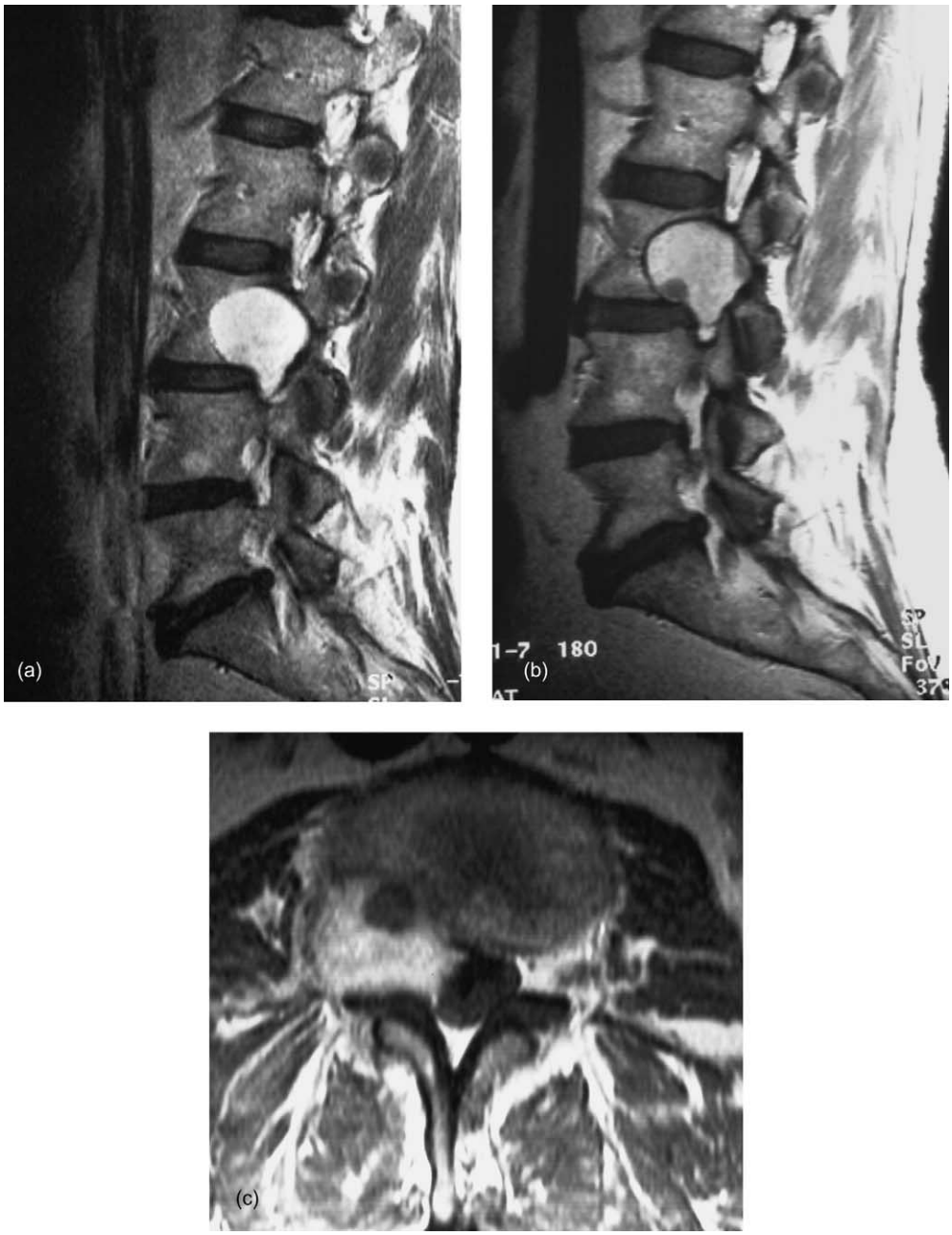


Fig. 5. Neurinoma. Spinal neurinomas can be very small and located on the intraspinal nerve roots or may be larger and have a typical dumbbell shape with an intra-/extraspinal localization (c). Besides this typical location, they also have a characteristically high signal on T2WI (a). Because it are slow-growing lesions, bone erosion and remodeling is not uncommon (b).

More than half of all vertebral hemangiomas are seen in the thoracic region, one third in the lumbar region and the remainder in the cervical and sacral region. About one third is multiple. Most occur in the vertebral body, but about 10% extend in the posterior elements. Extraosseous lesions are rare (1%).

5.1.2. Clinical presentation

The vast majority of vertebral hemangiomas is discovered incidentally. Sometimes local pain of tenderness is present. Symptoms of cord or nerve compression are rare and due to tumor extension rather than vertebral collapse. Compression fractures are unusual because of the thickened trabeculae.

5.1.3. Pathology

Vertebral hemangioma consists of endothelium lined vascular structures. They can be of either capillary, cavernous or venous origin. Because of destruction of some of the bone trabeculae the remainder tends to increase in thickness.

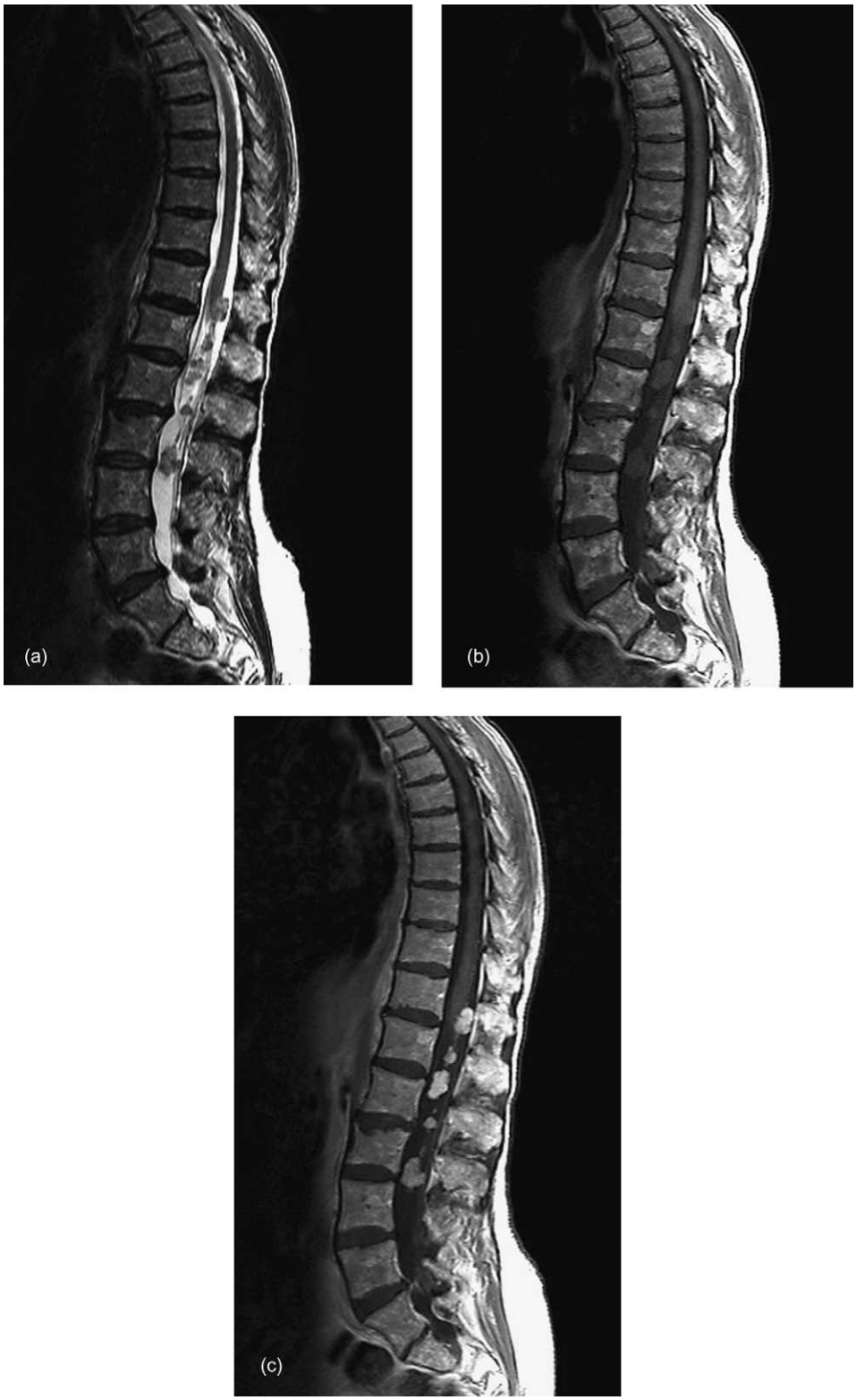


Fig. 6. Neurofibromatosis 1. Multiple spinal root tumors are a typical representation of neurofibromatosis 1 (NF1). They are iso- to hyperintense on T2WI (a), isointense on T1WI (b) and show strong homogeneous enhancement (c). Complete neuro-axis MR is mandatory in the work-up of these patients.

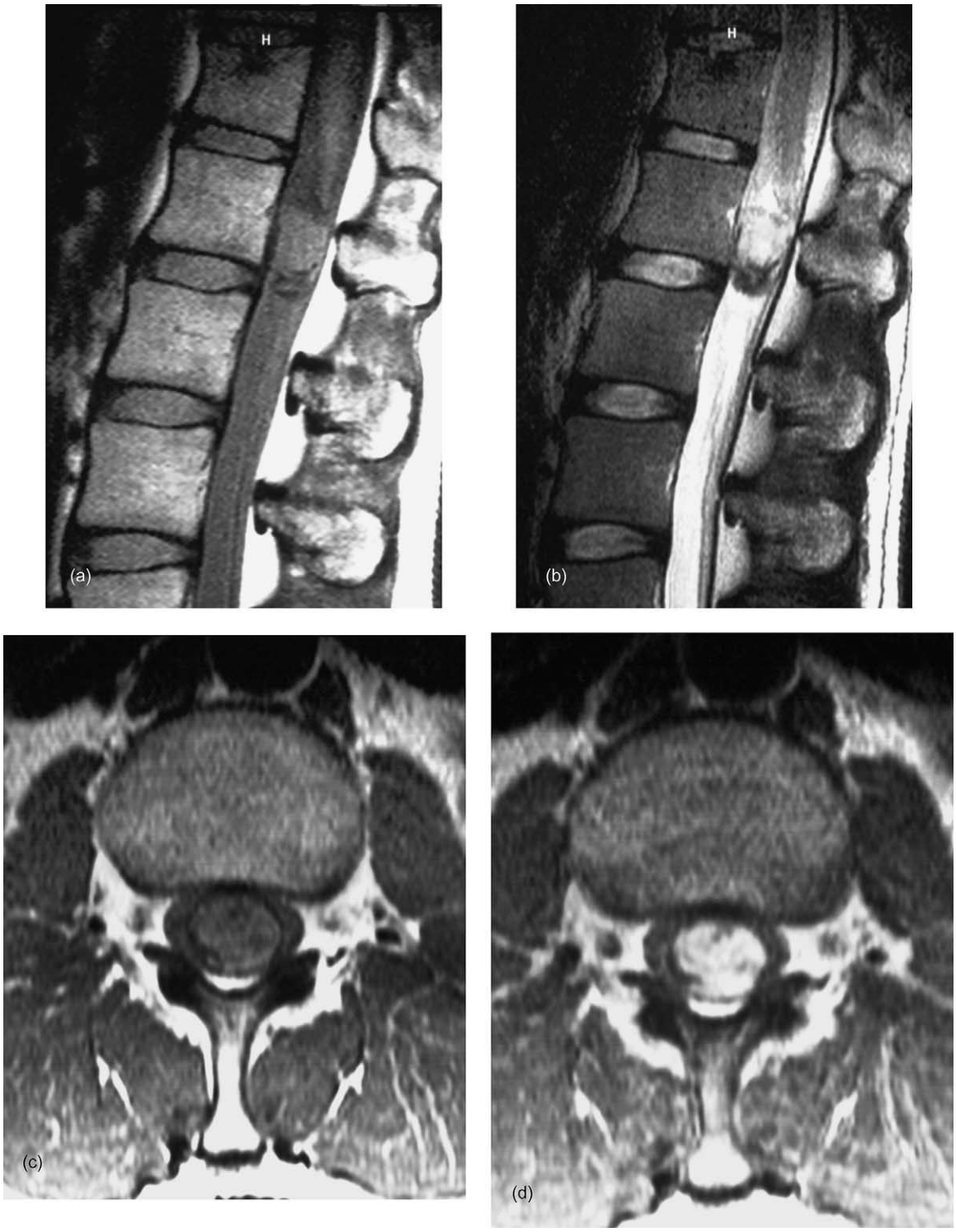
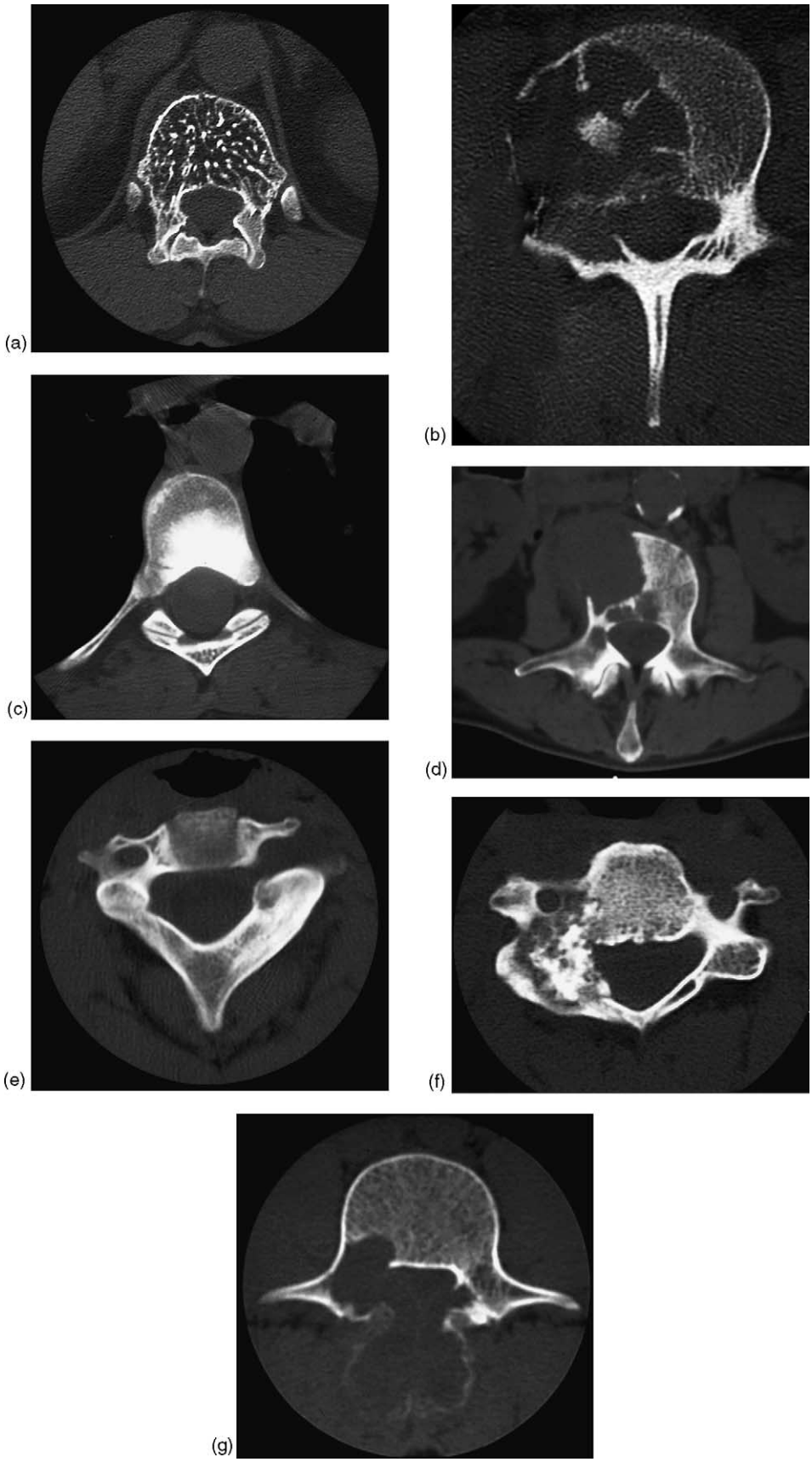


Fig. 7. Paraganglioma. Although uncommon, spinal paraganglioma can have a typical MR representation. Isointense on T1WI (a), heterogeneous high signal on T2WI (b), but especially surrounding flow-voids are suggestive for a highly vascularized tumor (c). Strong enhancement is the rule (d).

5.1.4. Imaging

The thickened vertical bone trabeculae in vertebral hemangioma give rise to its typical "jail bar" or "honeycombing" appearance on plain films. On axial CT, this is seen as a spotted appearance, known as "polka-dot" (Fig. 8a). On MR imaging these lesions are well circumscribed and have a typical high signal intensity both on T1WI and T2WI. This high signal is caused by a high fatty content [21]. Some lesions though have a low signal on T1WI, and these tend to be more aggressive. Hemangiomas


(a)
(b)
(c)
(d)
(e)
(f)
(g)

have a contrast uptake, which is especially noticeable in the more aggressive lesions that have a lower signal on T1WI to start with.

5.2. Plasmacytoma

5.2.1. General

The peak incidence for spinal plasmacytoma is in the fifth to sixth decade. Men are more affected than women. Solitary plasmacytoma is an uncommon tumor occurring in 3–7% of patients with plasma cell neoplasms. The lesion is commonly found in the axial skeleton (25–60%). Plasmacytoma is a lesion of the vertebral body but involvement of the posterior elements is the rule. Often, the lesion is present for many years as an isolated lesion, but on occasion, multiple plasmacytomas can develop.

5.2.2. Clinical presentation

Local or irradiating pain is the most common presenting complaint. Sudden intense pain may be caused by acute compression fracture. Radicular or cord symptoms occur with compression of neurologic structures often after vertebral collapse. The thoracic spine is the commonest site involved, followed by lumbar spine, cervical spine and sacrum in descending order. Most patients have an indolent course with a median survival rate of 10 years.

5.2.3. Pathology

Plasmacytomas are considered to represent the early stages of multiple myeloma, but plasmacytoma can precede laboratory evidence of multiple myeloma for many years. There are no signs of myeloma cells on bone marrow examination.

5.2.4. Imaging

On plain film plasmacytoma presents as a lytic and usually expansile bone lesion with thickened trabeculae and multicystic appearance [22]. Often fractures or even frank collapse exists. MR imaging shows a low signal intensity on T1WI and high signal intensity on T2WI involving the entire vertebral body. Diffuse enhancement with intravenous contrast medium is the rule, but in some cases only peripheral enhancement is noted. Curvilinear low-signal-intensity structures on all imaging sequences extending partially through the vertebral body and resembling sulci seen in the brain, causing a “mini brain” appearance on axial images is very typical [23] (Fig. 8b). These low-signal-intensity structures are likely caused by thickened cortical bone caused by the slow-growing nature of plasmacytoma. This appearance can also be seen on CT of plasmacytoma.

5.3. Multiple myeloma

5.3.1. General

The peak incidence of multiple myeloma is in the sixth to seventh decade. The spine and especially the vertebral bodies are the most common location, frequently with epidural extension. It is the most frequent primary malignant tumor of the spine.

5.3.2. Clinical presentation

Bone pain, usually involving the back or sternum, from tumor expansion is the most common presenting symptom. Less common presentations of multiple myeloma include vague symptoms of fatigue, weakness, or weight loss. Often, however, the disease is discovered while the patient is being examined for other medical problems. Routine radiographs, demonstrating bony lesions can be the first sign of multiple myeloma.

5.3.3. Pathology

Multiple myeloma is a monoclonal proliferation of malignant plasma cells usually affecting bone. Monoclonal gammopathies are a premalignant condition of multiple myeloma.

5.3.4. Imaging

The typical imaging findings on conventional radiography and CT include punched out lytic bone lesions, diffuse osteopenia, fractures, and, rarely, osteosclerosis. On MR imaging different signal patterns, ranging from normal-appearing bone marrow to focal lesions or diffuse bone marrow infiltration are seen. On T1WI, a low signal intensity is typically noted, with marked enhancement after the administration of contrast material. Even in advanced stages of this disease, up to 20% of radiographs and MR examinations can have normal findings. Spiral CT allows imaging of the entire spine and provides detailed information on osseous involvement in multiple myeloma. Especially in anatomically complex regions like the pelvis and the thoracic spine, it is superior

Fig. 8. Typical CT patterns of spinal bone tumors. (a) Hemangioma. The polka-dot pattern represents thickened bone trabeculae as a reaction to stress in areas of bone weakening due to tumor involvement. It is a typical pattern in benign slow-growing vertebral hemangioma. (b) Plasmacytoma. The appearance of an osteolytic lesion resembling a “mini-brain” is typical, but not specific, for a vertebral plasmacytoma. CT shows an expansile lytic lesion with marginal sclerosis. (c) Osteoblastic metastatic disease. Osteoblastic metastatic disease most commonly originates from breast carcinoma and prostate cancer, although other malignancies can also be evolved (carcinoid, testis, stomach, colon, adrenal neuroblastoma). (d) Osteolytic metastatic disease. Osteolytic bone metastasis is much more common than osteoblastic lesions. One of the smaller lesions in this patient shows no or minimal sclerotic reaction, while especially the larger metastatic location shows clear marginal bone sclerosis. (e) Osteoid osteoma. The typical CT appearance of osteoid osteoma is the sclerotic bone lesion with a small central lucent nidus and dense calcified center. (f) Osteoblastoma. CT shows a large expansile lesion with preserved cortex and internal calcification. These lesions are predominantly located in the neural arch. (g) Aneurysmal bone cyst. Marked expansile lytic lesion. The typical fluid levels are not visible on this image with bone windows. Formation of new bone between the original cortex an the expanded part is often seen in aneurysmal bone cysts.

to conventional radiography. Compared with conventional radiography and MR imaging, spiral CT provides more detailed information on the risk of vertebral fracture [24]. For evaluating diffuse bone marrow changes, MR imaging is still the imaging modality of choice. Because MR imaging of the spine shows normal findings in up to 20% of patients with proven bone marrow infiltration, the initial staging of multiple myeloma should include MR imaging in combination with spiral CT.

5.4. Metastatic disease

5.4.1. General

Metastatic disease to the spine is the most frequent spinal tumor in general. Any malignant tumor can metastasize to bone, but the most common metastatic spinal tumors found in women are from the breast and lung and in men from the prostate and lung. The tumors that most often affect the spine, regardless of their proper incidence, are myeloma, breast, prostate and stomach. Generally, metastatic seeding appears in the thoracic spine (accounting for about 70% of cases), with the lumbar spine being the next most involved site (20% of cases). The cervical spine is affected in approximately 10% of cases. Multiple spinal levels are affected in about 30% of patients. Most frequently metastasis affects the vertebral body, but all parts of the vertebra may be affected.

5.4.2. Clinical presentation

Tumors that have spread to the spine usually cause a significant amount of pain. If they are large enough, they may put pressure on the spinal cord, which causes increasing pain and loss of normal functioning in the bowel, bladder, and legs. Thecal sac impingement is seen in 5% of all patients with systemic cancer.

5.4.3. Imaging

Spinal metastasis usually presents as lytic lesions on plain film, but they can also be sclerotic (osteoblastic), especially in prostate and breast carcinoma (Fig. 8c). On CT metastatic disease presents as multiple lytic lesions of different size, with irregular non-sclerotic margins, often with cortical breakthrough and paravertebral or epidural extension (Fig. 8d). On MR imaging most metastases are hypointense on T1WI. On T2WI they can be either dark (sclerotic) or more frequently show high signal. Marked enhancement after contrast administration is the rule. Diffusion WI has been proposed by some to differentiate metastatic compression fractures from benign vertebral collapse [25,26].

5.5. Chordoma

5.5.1. General

The peak incidence for chordoma is in the fifth to sixth decade. The average age at presentation is 55 years [27]. Men are more affected than women (2:1). Half of these tumors is found in the sacrum, 35% in the clivus and the rest in the vertebrae.

5.5.2. Clinical presentation

Local pain is the most frequent presenting symptom. Symptoms of spinal cord or nerve root compression can occur if they grow large enough.

5.5.3. Pathology

Chordomas arise from remnants of the notochord. They are composed of large vacuolated physaliphorous cells with intervening fibrous septae [28]. Fluid and gelatinous mucoid substance (associated with recent and old hemorrhage) and necrotic areas are found within the tumor; in some patients, calcification and sequestered bone fragments are found as well. In addition to conventional chordomas, chondroid chordomas are identified, which are composed of cartilaginous hyaline tissue.

Chordomas are malignant tumors that show local invasion, and especially vertebral body cases show distant metastasis.

5.5.4. Imaging

On plain film bone destruction often with amorphous calcification is seen. CT can show paravertebral and especially epidural extension [29]. Due to the variety of components most lesions are heterogeneous on MR imaging. They are isointense on T1WI and hyperintense on T2WI. Chondroid forms have higher signal on T1WI and lower signal on T2WI because of low water content. Enhancement after contrast injection varies from little to prominent. Often internal septations and a low signal surrounding capsule can be seen. Sometimes hemorrhage and cyst formation is present.

5.6. Osteoid osteoma

5.6.1. General

The peak incidence for osteoid osteoma is in the second decade. The average age at presentation is 17 years. Men are more affected than women (2–4:1). It accounts for about 10% of all bone tumors involving the spine. An osteoid osteoma is a benign lesion that has a unique tendency to affect the posterior part of the vertebra and occurs primarily in the pedicle and the posterior elements, not in the vertebral body. The lumbar spine is the most affected, followed by the cervical, thoracic and sacral regions [30].

5.6.2. Clinical presentation

It typically presents as unrelenting pain, clearly worse at night, which is exquisitely sensitive to aspirin or non-steroidal anti-inflammatory drugs (NSAIDs). Not infrequently patients with spinal osteoid osteoma have a painful scoliosis [31].

5.6.3. Pathology

Osteoid osteoma has a central nidus of vascular fibrous connective tissue with a surrounding osteoid matrix. Multinucleated giant cells and osteoclasts are frequently observed. They are surrounded by bone sclerosis. Lesions larger than 1.5 cm are usually categorized as osteoblastoma.

5.6.4. Imaging

Plain film shows a lucent nidus, frequently with small calcifications. Surrounding the nidus is variable bone sclerosis. Bone scintigraphy is highly sensitive for osteoid osteoma. Especially when there is extensive bone sclerosis the nidus is much easier distinguished on CT [32] (Fig. 8e). On MR imaging the calcifications and bone sclerosis have a low signal both on T1WI and T2WI. The non-calcified portion of the nidus has high signal on T2WI. After contrast administration intense enhancement of the nidus is seen. Sometimes adjacent bone marrow and soft tissue changes are seen.

5.7. Osteoblastoma

Osteblastomas are related to osteoid osteomas, and by definition, are larger than 1.5 cm in diameter. They also have the tendency to affect the posterior part of the spine and present with pain (Fig. 8f). Osteoblastomas can be more aggressive than osteoid osteomas and more often require surgical resection. The recurrency rate is about 10%, which is also higher than that seen with osteoid osteomas.

5.8. Aneurysmal bone cyst

5.8.1. General

The peak incidence for aneurysmal bone cyst (ABC) is in adolescence, and 80% occurs before age of 20 years. Women are somewhat more affected than men. The lumbosacral spine is the most affected and the tumor may affect the posterior elements of the spine or expand to the pedicles and the vertebral body itself.

5.8.2. Clinical presentation

These tumors may present with pain and in some cases, fractures and cord or nerve root compression with neurological symptoms.

5.8.3. Pathology

Aneurysmal bone cysts are benign tumors of unknown etiology. In almost half of all cases they are associated with a pre-existing bone lesion. It are large, expansile and multiloculated lesions with high vascularization.

5.8.4. Imaging

Plain film shows an expansile, osteolytic lesion with thin surrounding cortical bone (Fig. 8g). CT and MR imaging typically show a multiloculated lesion with fluid–fluid levels. Sometimes internal septations and lobulations are present. MR imaging demonstrates blood degradation products.

5.9. Giant cell tumor

5.9.1. General

The peak incidence for giant cell tumor is in the third through fifth decade. there is no sex predilection. In spinal giant cell tumors there is a female preponderance. Spinal giant cell tumors are especially seen in the sacrum and are rare in other spinal locations. It is the most frequent benign tumor of the sacrum. They tend to affect the vertebral body (front of the spine).

5.9.2. Clinical presentation

Giant cell tumors present with local pain. Neurologic complications are not uncommon.

5.9.3. Pathology

Despite being technically “benign”, they can be very aggressive and sometimes spread elsewhere. They extend to the cortex, but usually do not transgress the periosteum. Giant cell tumors contain monocytes, macrophages and multinucleated giant cells. On pathologic examination they are indistinguishable from brown tumors and may resemble chondroblastoma, chondromyxoid fibroma, ABC and osteosarcoma. Incomplete removal of a vertebral giant cell tumor can lead to possible malignant transformation. Malignant transformation is seen in about 10% of cases.

5.9.4. Imaging

Plain films show a lytic and expansile spinal lesion, typically in the sacrum. MR imaging demonstrates an inhomogeneous multiloculated cystic mass. They frequently contain blood degradation products.

5.10. Eosinophilic granuloma

5.10.1. General

The peak incidence for eosinophilic granuloma of the spine is in the first decade. There is a clear male predilection. Eosinophilic granuloma is one of the presenting forms of Langerhans cell histiocytosis. Lesions may be single or multiple. Most lesions are seen in the cervical spine, especially mid-cervical in children and at C2 in adults [33].

5.10.2. Clinical presentation

Eosinophilic granuloma has a highly variable clinical presentation ranging from non-existent to very painful, sometimes worsening at night and sensitive to aspirin or non-steroidal anti-inflammatory drugs. The presenting symptoms of cervical eosinophilic granuloma are usually pain and restricted range of motion. In contrast to eosinophilic granuloma of the thoracic spine and lumbar spine, the neurologic symptoms are less frequent.

5.10.3. Pathology

Eosinophilic granuloma is a benign non-neoplastic disorder with unknown etiology. They tend to evolve from small cystic and hemorrhagic to larger fatty and fibrous lesions. Initially they contain many eosinophils and lymphocytes.

5.10.4. Imaging

Plain film shows a lytic lesion with sharp borders. It is a classic cause of a single collapsed vertebral body (vertebra

plana). Nevertheless, vertebra plana is a rare sign in cervical eosinophilic granuloma [33]. On MR imaging it has a high signal on T2WI and a variable signal on T1WI. It enhances strongly with gadolinium. When the vertebral body is involved it is usually affected in its entirety.

6. Conclusion

Spinal tumors are relatively rare and affect only a minority of the population. Because they may cause significant morbidity in terms of pain and limb dysfunction, and are associated with mortality as well, their correct diagnosis and prompt treatment can be very important. In establishing the differential diagnosis, location is the most important feature. MR imaging, which plays the central role in the imaging of spinal tumors, easily allows tumors to be classified as extradural, intradural–extramedullary or intramedullary. CT also has a major role especially in the imaging of tumors of the vertebral column.

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