The Acute Pediatric Spine and Spinal Cord

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Introduction

Acute lesions involving the spine and spinal cord in the pediatric age group are rare, but comprise several different pathomechanisms and entities. Patients presenting in the emergency room with acute back pain and/or signs of acute neurological involvement including sensorimotor deficit of the upper and/or lower limbs and sphincter dysfunction warrant emergent clinical evaluation and neuroimaging studies. The risk of a spinal cord compression must always be borne in mind by referring clinicians, since neurological recovery often depends primarily on surgical decompression and stabilization, and the impact of permanent paralysis in an otherwise normal child may result in severe psychological trauma to the child as well as the parents. Often, however, neurological deterioration occurs subacutely in the context of preexisting conditions. It is not uncommon for patients with acute spinal injury to complain with back pain or, in younger children, pain equivalents such as refusal to crawl, sit, or walk. The presence of back pain in prepubertal children almost always heralds a serious underlying disorder; however, families and even physicians overlooking the problem may cause a significant delay in the diagnosis. Red flags for children with back pain include age less than 4 years, presence of a functional disability, duration of greater than 4 weeks, presence of a fever, postural shift of the trunk caused by the pain, limitation of motion due to the pain, and presence of a neurologic abnormality. Head tilt, or torticollis, may be indicative of injury to the craniocervical injury which, if unstable, can further lead to severe neurological deterioration and even death.

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Neuroimaging: General Principles

Diagnostic imaging in patients presenting with an acute spinal injury is aimed at identifying the location, degree, and quality of primary injury in order to institute an immediate treatment and to limit or prevent secondary injury. Imaging should be fast, be readily available, and not interfere with the emergent treatment, and be highly sensitive and specific [1]. Among available imaging modalities, consisting of conventional X-rays, computerized tomography (CT), and magnetic resonance imaging (MRI), the choice should be made based on the patient's clinical status and the available equipment.

In the setting of spinal trauma, X-rays are still often used as the first imaging modality, although helical CT with multiplanar reformat and including bone and soft tissue algorithms should be preferred, especially in patients with serious trauma. MRI is typically performed rapidly as a second line of imaging if focal neurological signs are present, suspecting a spinal cord lesion. Conversely, non-trauma patients presenting in the emergency room with acute neurological signs compatible with a spinal cord lesion should directly be referred for emergent MRI with the goal of identifying potentially curable causes of spinal cord compression. The MR field of view should always cover the entire spine on the sagittal plane and include both T1- and T2-weighted images; axial imaging is typically focused on any focal pathological finding. Coronal imaging with STIR sequences is also useful to obtain a panoramic coverage of the spine while exploiting the exquisite sensitivity of STIR to vertebral bone edema. Post-contrast imaging is also strongly advised in the context of acute neurological presentations, in order to characterize lesions and to evidence areas of blood-cord or blood-root barrier breakdown. In the pediatric age group, sedation is often required to carry out

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MRI studies if the patient is too young or too ill to cooperate.

Spine Trauma

The type, severity, and consequences of trauma significantly depend on the peculiar properties of the immature pediatric spine [2]. These include the predominantly cartilaginous composition of the spine and the relatively lax ligaments, which render the pediatric spine mobile and deformable compared to adults. As a consequence, vertebral fractures are less frequent than dislocations, ligamentous injuries, epiphyseal detachments, and lesions of the ossification centers. The craniocervical junction of infants and small children is also especially vulnerable because of their comparatively larger head with respect to the torso and the weaker neck musculature, especially in the context of sudden acceleration and deceleration. Only from about 10 years of age does the more typical adult distribution start to prevail, with predominantly cervicothoracic or lumbar injuries. Knowledge of the peculiar biomechanics and structure of the pediatric spine, as well as of normal variants or pitfalls (such as the physiological C2–C3 subluxation), is essential for a correct interpretation of imaging studies in the pediatric age group.

The leading cause of spinal trauma in children is motor vehicle accidents (52 %), followed by sports-related injuries (27 %) [3]. Patients with craniocervical junction or cervical trauma have a typically acute presentation with neurological deficit and have a poor prognosis. Intracranial lesions such as hematomas or diffuse axonal injury may coexist and also have a profound impact on the clinical picture and prognosis.

SCIWORA The term indicates a form of spinal cord injury without radiographic abnormality, i.e., lacking evidence on either X-ray or CT scan [4], but evident on MRI studies. This form of spinal cord injury is typical of infants and children and is caused by the high mobility and flexibility of the pediatric spine, which allows for an

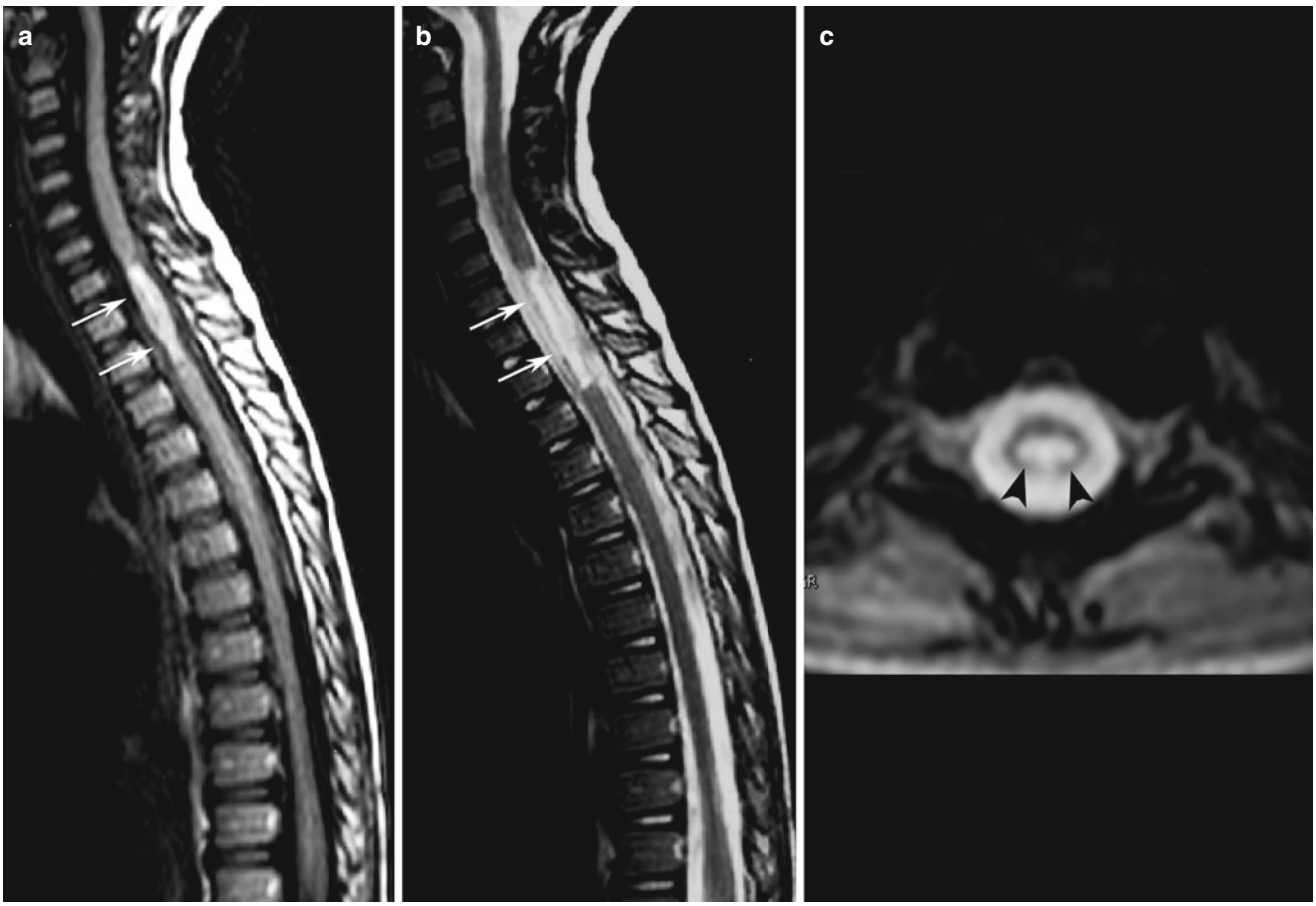

Fig. 1 SCIWORA in a 2-year-old girl involved in a motor vehicle accident. Sagittal FLAIR (**a**) and T2-weighted images (**b**) show intramedullary hyperintense lesion at the C7–T2 level (*arrows*). The osteocartilaginous spine is unremarkable. Axial T2-weighted image (**c**) shows cross-sectional spinal cord involvement (*arrowheads*)

intermittent luxation with consequent impact and contusion of the spinal cord. MRI usually shows a swollen, T2 hyperintense spinal cord in the affected segment (Fig. 1); petechial hemorrhages or frank hematomyelia may complicate the picture.

Atlanto-occipital and Atlanto-axial Dislocation In these severe cases of dislocation, there is typically an anteriorization of the atlas with acute narrowing of the spinal canal, which in turn causes injury to the bulbomedullary junction and spinal cord (Fig. 2). This form of injury is almost invariably severe and leads to a very guarded, if not constantly dismal, prognosis, especially when a complete ligamentous rupture causes a true axial/vertical atlanto-occipital dissociation. Less severe forms of progressive atlanto-axial dislocation may occur also outside the context of trauma, and especially in patients with inflammatory disorders of the craniocervical junction (especially juvenile idiopathic arthritis) or with common otolaryngeal conditions including pharyngitis, adenotonsillitis, tonsillar abscess, cervical abscess, and otitis media that results in hyperemia and pathological relaxation of the transverse ligament of the atlanto-axial joint (Grisel syndrome) [5].

Vertebral Fractures Most vertebral fractures seen in the pediatric clinical practice are compression fractures are characterized by wedge-shaped deformity of the involved vertebral body with interruption/fracture of the anterior vertebral contour [1]. These fractures typically result from falls and are stable if they only involve the anterior column. More severe fractures include burst fractures, in which both the anterior and posterior contour of the vertebral body are involved, resulting in an intrinsically unstable condition which can further be complicated by intraspinal dislocation of bony fragments causing direct spinal cord damage. Peculiar regional fractures can also be encountered, such as the Jefferson fracture of the atlas and the Anderson and Hangman fractures of the axis. In the lumbar region, the Chance fracture occurs due to flexion distraction, typically as a result of a seat or lap belt

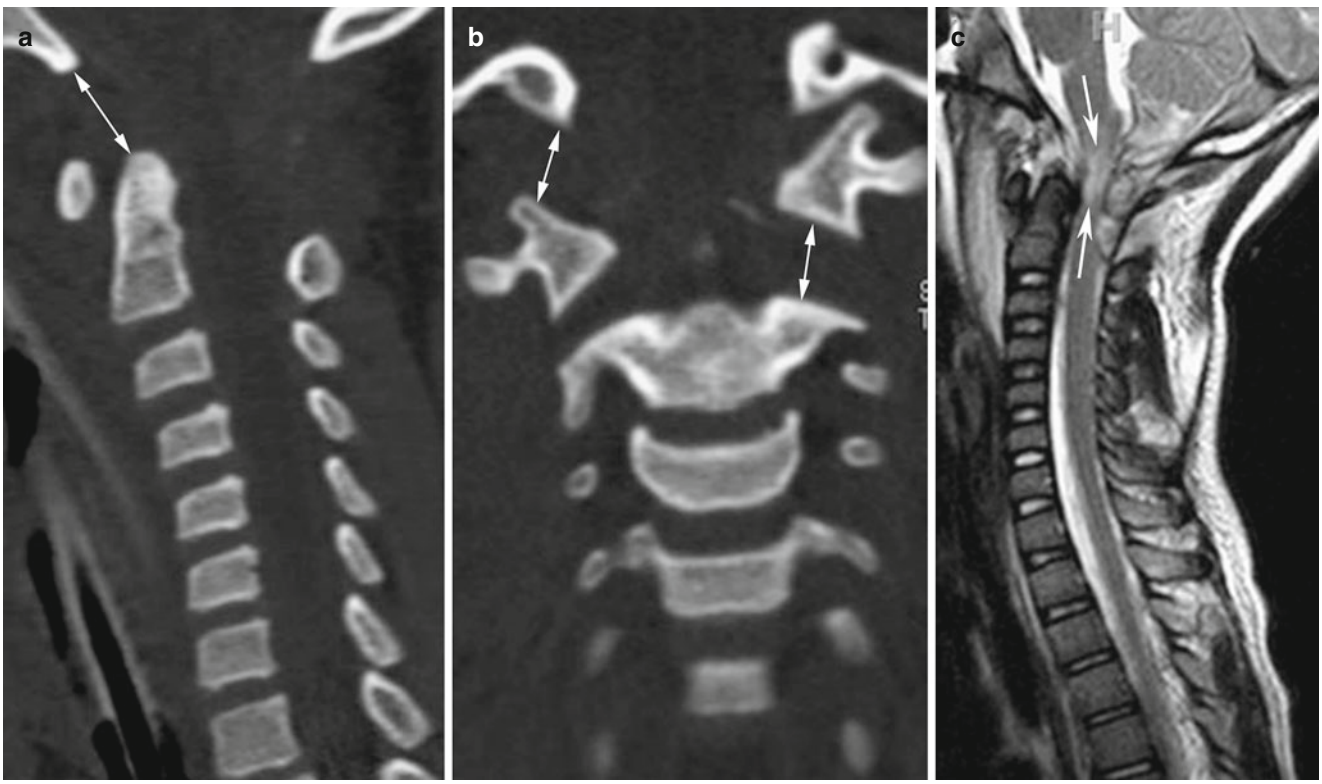

Fig. 2 Atlanto-occipital dislocation. Sagittal (**a**) and coronal (**b**) CT images show asymmetrical dislocation of C1 in relation to C0/skull base with an increased distance between the basion and the tip of the dens (*double arrow*, **a**) and widened right atlanto-occipital and left atlanto-axial joints (*double arrows*, **b**). Sagittal T2-weighted MR image (**c**) shows focal contusion of the lower brain stem and upper cervical spinal cord (*arrows*) as well as a significant ligamentous injury of the craniocervical junction (Case courtesy of T.A. Huisman, Johns Hopkins, Baltimore, USA)

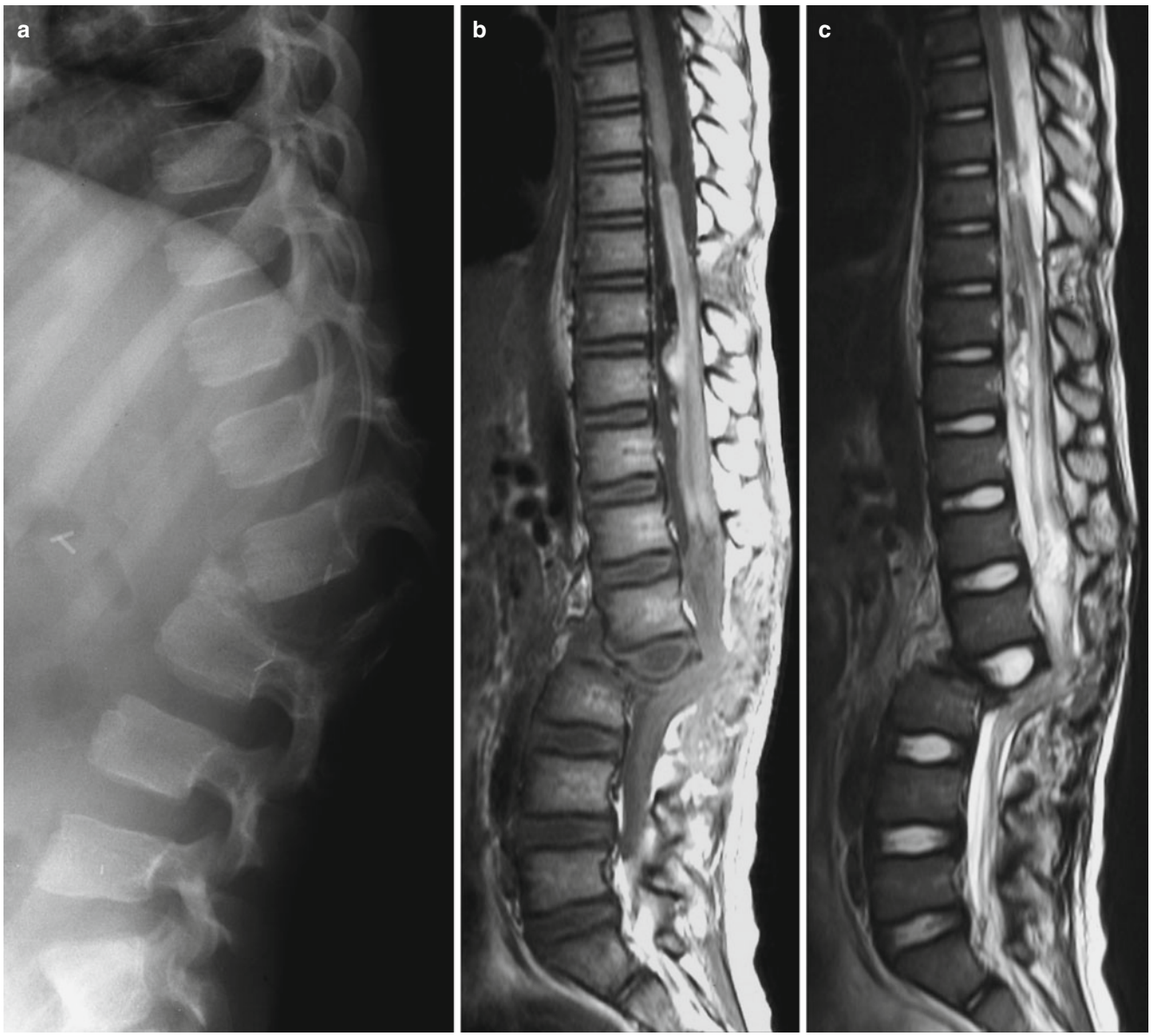

Fig. 3 Lumbar Chance fracture with spinal cord damage in a 7-year-old boy involved in high-speed car accident. Lateral radiogram (**a**) shows disruption of the superior L3 end plate with marked dislocation and kyphosis. Sagittal T1-weighted (**b**) and T2-weighted (**c**) MR images obtained 2 days after emergency decompression show extensive spinal cord and caudal nerve root damage

injury, and is intrinsically unstable as it involves all three longitudinal vertebral columns (Fig. 3).

CT scan is obviously dictated to detect the characteristics of vertebral fracture and to plan surgical management wherever needed; however, MRI including STIR sequences is especially sensitive to even minimal degrees of bone marrow edema (Fig. 4) and is helpful to confirm minimal compression fractures, which may sometimes escape detection on conventional X-ray. MRI is mandatory to assess spinal cord damage or intraspinal hematomas in patients who experience neurological deficit as a result of their trauma.

Infectious/Inflammatory Diseases

Inflammatory and infectious disorders of the spine and spinal cord are less common in children than in adults. The most common infectious spinal disorder in children is bacterial meningitis, which generally does not require imaging studies and is diagnosed and treated on a clinical and physical

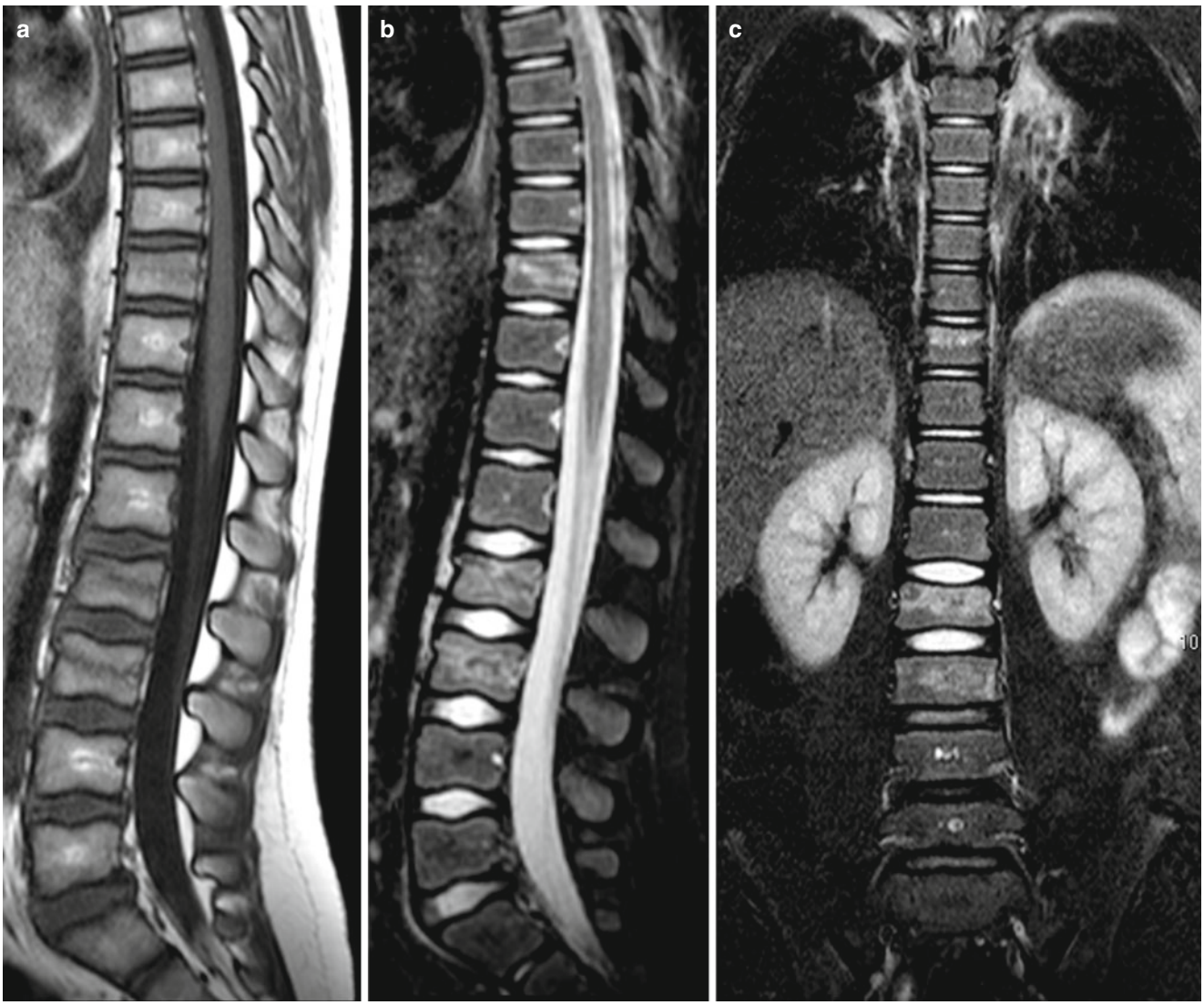

Fig. 4 Compression fractures in a 9-year-old boy. Sagittal T1-weighted image (**a**) shows ill-defined hypointensity of the T10, L2, and L3 vertebral bodies, with a mild reduction in height. Sagittal (**b**) and coronal (**c**) STIR images exquisitely depict hyperintensity of the involved vertebral bodies consistent with bone marrow edema

examination basis. MRI is essential for diagnosing spinal infectious/inflammatory conditions of the spine and spinal cord, while CT does not play a role outside certain specific conditions. Although many of these disorders also show brain abnormalities, I will focus on the abnormalities found in the spine and spinal cord.

Acute Transverse Myelitis Acute transverse myelitis (ATM) is a focal inflammatory disorder of the spinal cord characterized by an acute onset of motor, sensory, and autonomic dysfunction. Clinical presentation is with pain, paresthesias, leg weakness, and sphincter dysfunction, all of which more or less rapidly progress in the hours to days following the onset. Cerebrospinal fluid (CSF) analysis shows pleocytosis or elevated IgG index. The diagnosis of idiopathic ATM involves exclusion of other causes of acute neurological involvement such as extrinsic spinal compression, ischemia, tumor, arteriovenous malformation, and toxicities [6]. Identified causes of ATM such as connective tissue disease, ADEM, multiple sclerosis, and neuromyelitis optica (NMO) must also be ruled out.

MRI plays a fundamental role in patients suspected of harboring ATM and should be done in emergency, because the presentation of compressive lesions such as extramedullary tumor or hemorrhage can be identical [7]. It is wise to also include the brain in order to assess possible additional lesions (such as those typical of ADEM or multiple sclerosis) without delay while minimizing the need for additional seda-

tion [7]. Spinal MRI (Fig. 5) should include high-resolution sagittal T1- and T2-weighted images; sagittal short-tau inversion recovery (STIR) is also extremely useful to detect subtle signal intensity abnormalities of the spinal cord. Optimal slice thickness for sagittal studies should be 3 mm or less. Axial T2-weighted images across lesional areas help to determine the cross-sectional extent of the spinal cord involvement, an important element in the differential diagnosis [8]. Post-contrast images should be acquired in the three planes of space. In ATM, MRI shows normal or slightly expanded, T2-hyperintense spinal cord segment involving more than 3–4 vertebral levels in length and more than 2/3 of the cross-sectional area. Gadolinium enhancement depends on the degree of inflammation and timing of imaging with respect to clinical onset, and can be thoroughly absent [3]. Brain imaging is normal in patients with idiopathic ATM.

Acute Disseminated Encephalomyelitis Acute disseminated encephalomyelitis (ADEM) involves a first episode of inflammatory demyelination with polyfocal neurological signs implicating involvement of multiple sites of the CNS. Patients present with a rapid onset of encephalopathy and motor and/or sensory deficits with brainstem signs and symptoms and ataxia, and mandatory presence of encephalopathy [9, 10]. Most patients with ADEM present in the aftermath of infection or vaccination. The disorder commonly begins 1–2 weeks after a viral, and seemingly minor, illness. CSF analysis may show increased proteins and leukocytosis.

On MRI, multiple, more or less well-defined or demarcated areas of increased T2 signal intensity within the cord are found [11]. Usually, the lesions do not enhance with gadolinium administration. The single most common differential feature from ATM is the presence of brain involvement, as evidenced by clinical signs of encephalopathy and MRI evidence of signal abnormalities, in ADEM but not in ATM (Fig. 6).

Neuromyelitis Optica Neuromyelitis optica (NMO) is a rare, severe, mono- or multiphasic demyelinating disease of the CNS that preferentially affects the optic nerves and spinal cord. The diagnosis of NMO is made in the presence of optic neuritis and acute myelitis, associated with either a spinal MRI lesion extending over three or more segments or positive NMO serology [10]. The presence of at least two of three findings, (1) contiguous spinal cord MRI lesion extending over 3 vertebral segments, (2) brain MRI not meeting criteria for MS, and (3) NMO-IgG seropositive status, is 99 % sensitive and 90 % specific for NMO [12]. The neuromyelitis optica immunoglobulin G (NMO-IgG) is an autoantibody found in the serum of patients affected by NMO that binds to aquaporin 4 (AQP4), the main channel that regulates water

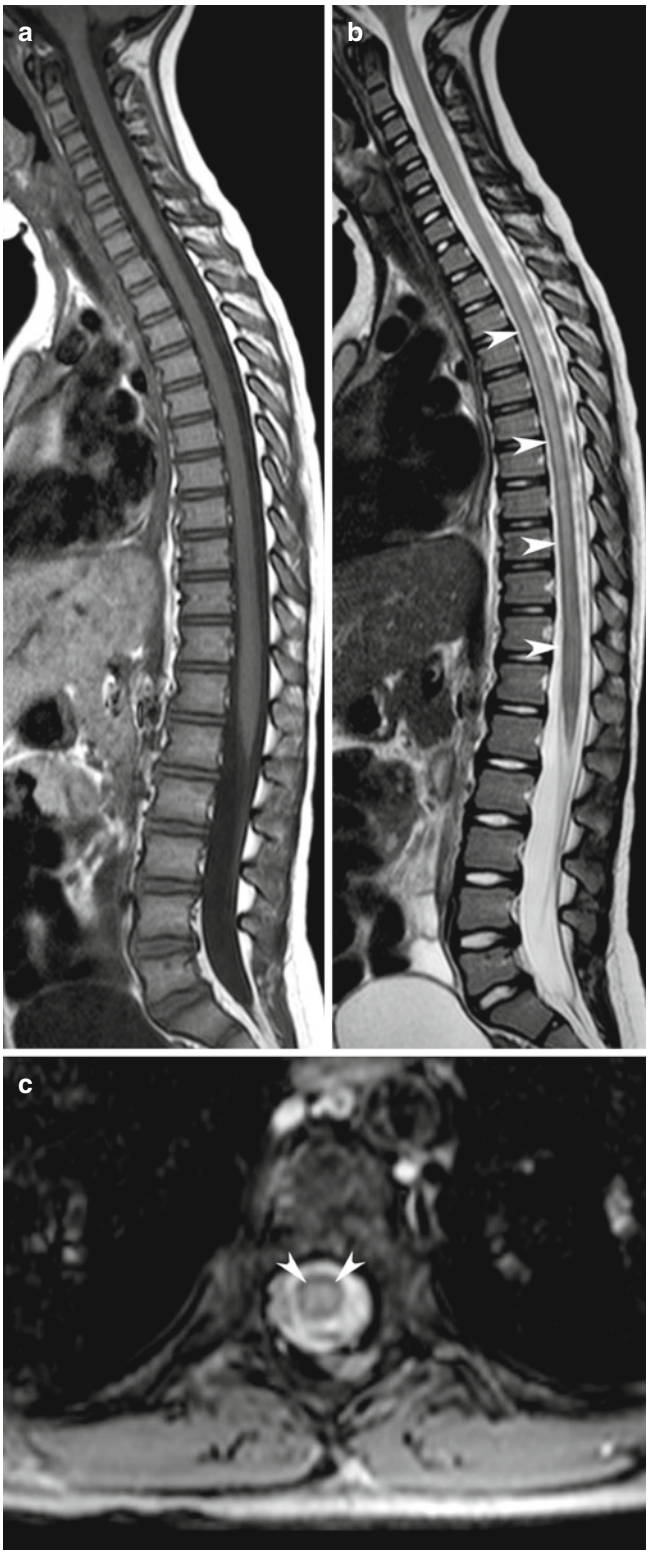

Fig. 5 Acute transverse myelitis in a 5-year-old girl. Sagittal T1-weighted image (**a**) is unrevealing. Sagittal T2-weighted image (**b**) shows ill-defined central hyperintensity of the spinal cord involving most of the thoracic cord and conus medullaris (*arrowheads*). There is not significant swelling of the cord in this case. Axial T2*-weighted image (**c**) confirms hyperintensity of the central portion spinal cord (*arrowheads*), exceeding 2/3 of the cross-sectional area of the cord

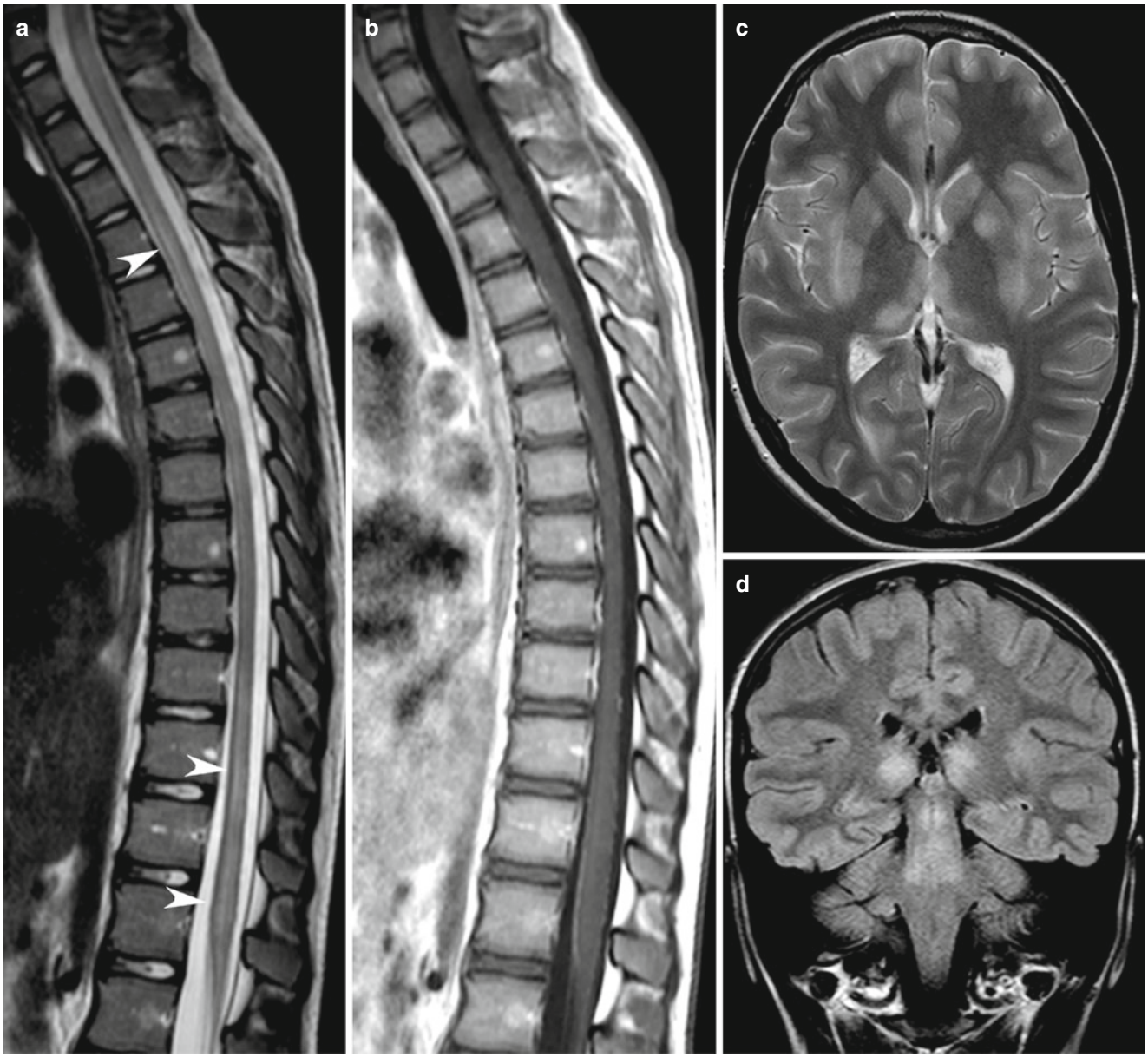

Fig. 6 Acute disseminated encephalomyelitis (ADEM) in a 12-year-old girl. Sagittal T2-weighted image (**a**) shows ill-defined areas of hyperintensity involving the spinal cord at multiple levels (*arrowheads*), while Gd-enhanced sagittal T1-weighted image (**b**) is unrevealing. The imaging findings at level of the spinal cord are not unlike those of isolated ATM. However, axial T2-weighted (**c**) and coronal FLAIR (**b**) images of the brain show multiple hyperintense areas at level of the nucleobasal regions and brainstem, consistent with a diffuse neuraxial process

homeostasis in the CNS, causing disruption of water homeostasis, demyelination, and necrosis with little or no inflammation. In NMO, attacks of acute myelitis precede optic neuritis in only 20 % of cases; in these cases, patients present with severe symmetric paraplegia, sensory loss below the lesion, and bladder dysfunction. The majority of children with an eventual diagnosis of NMO present with isolated LETM, optic neuritis, or brainstem encephalitis, thus failing to meet the diagnostic criteria at the time of their first presentation.

Spinal MRI shows large areas of high T2 signal intensity involving the spinal cord extensively and enhancing inhomogeneously, with a necrotic-cystic pattern that may simulate an intramedullary tumor (Fig. 7) [13, 14]. Imaging of the entire craniospinal axis should always be performed in patients suspected of harboring NMO, and the optic nerves and chiasm should be specifically evaluated. Hypothalamic, periaqueductal gray, and area postrema lesions can also be found in the brain of NMO patients.

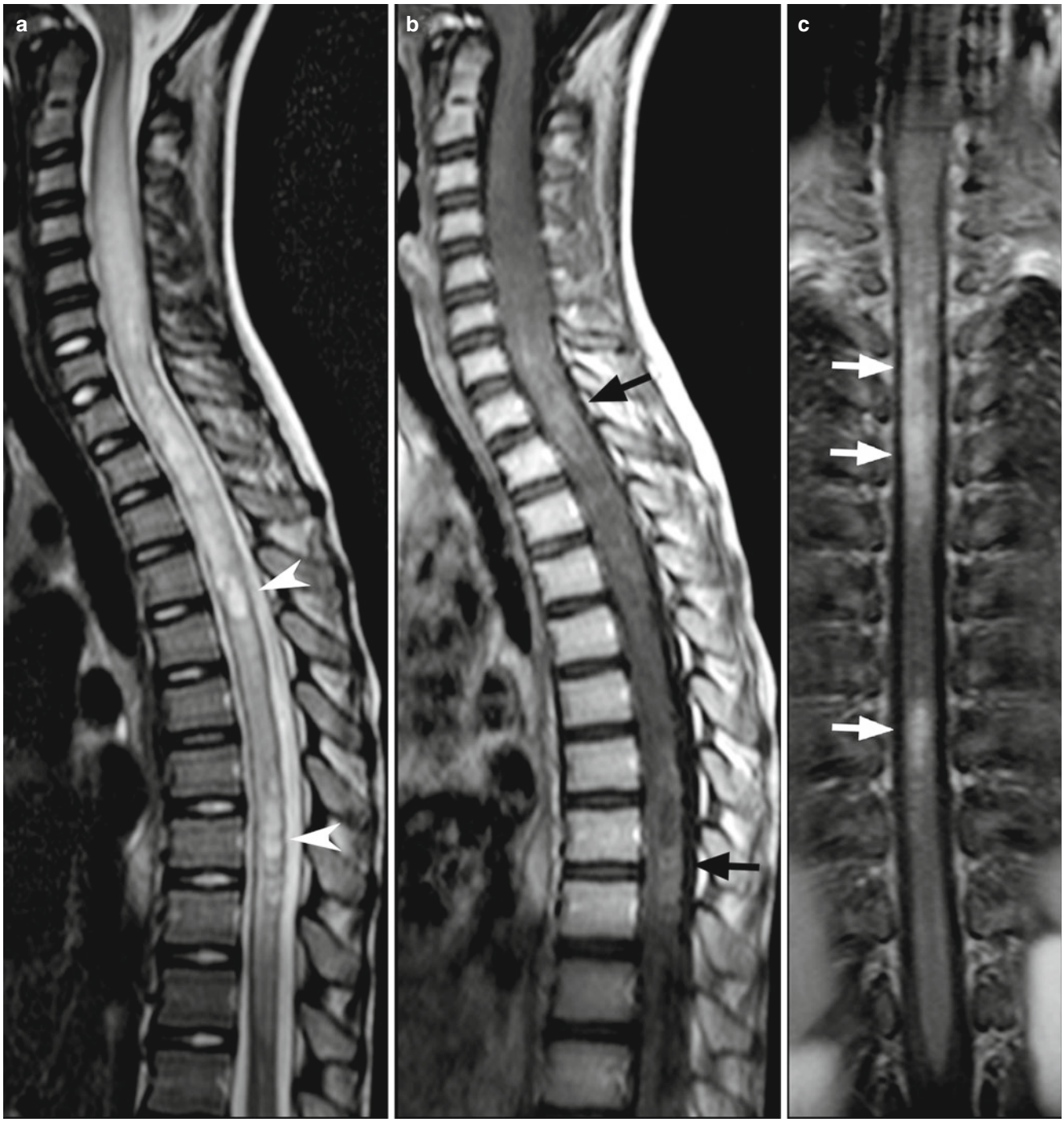

Fig. 7 NMO-IgG-positive longitudinally extensive transverse myelitis (neuromyelitis optica spectrum) in a 9-year-old girl. Sagittal T2-weighted image (**a**) shows marked swelling and hyperintensity of the spinal cord with a holocord extension. Areas of necrotic-cystic changes are shown (*arrowheads*). Gd-enhanced sagittal (**b**) and coronal (**c**) T1-weighted images show irregular areas of enhancement. This patient did not have optic neuritis, and only positive serology revealed the diagnosis, effectively ruling out neoplasm

Spinal Cord Abscess Spinal cord abscesses are extremely rare; predisposing conditions are usually required for germs to colonize the spinal cord, including congenital heart disease, disorders of the immune system, underlying spinal cord tumors, and especially dermal sinuses [15], which allow for a direct communication between the skin and the CNS.

MRI shows increased T2 signal intensity and expansion of the cord; more or less well-defined marginal enhancement occurs after gadolinium administration. A large part or even the whole length of the spinal cord may be involved in the most serious cases (Fig. 8).

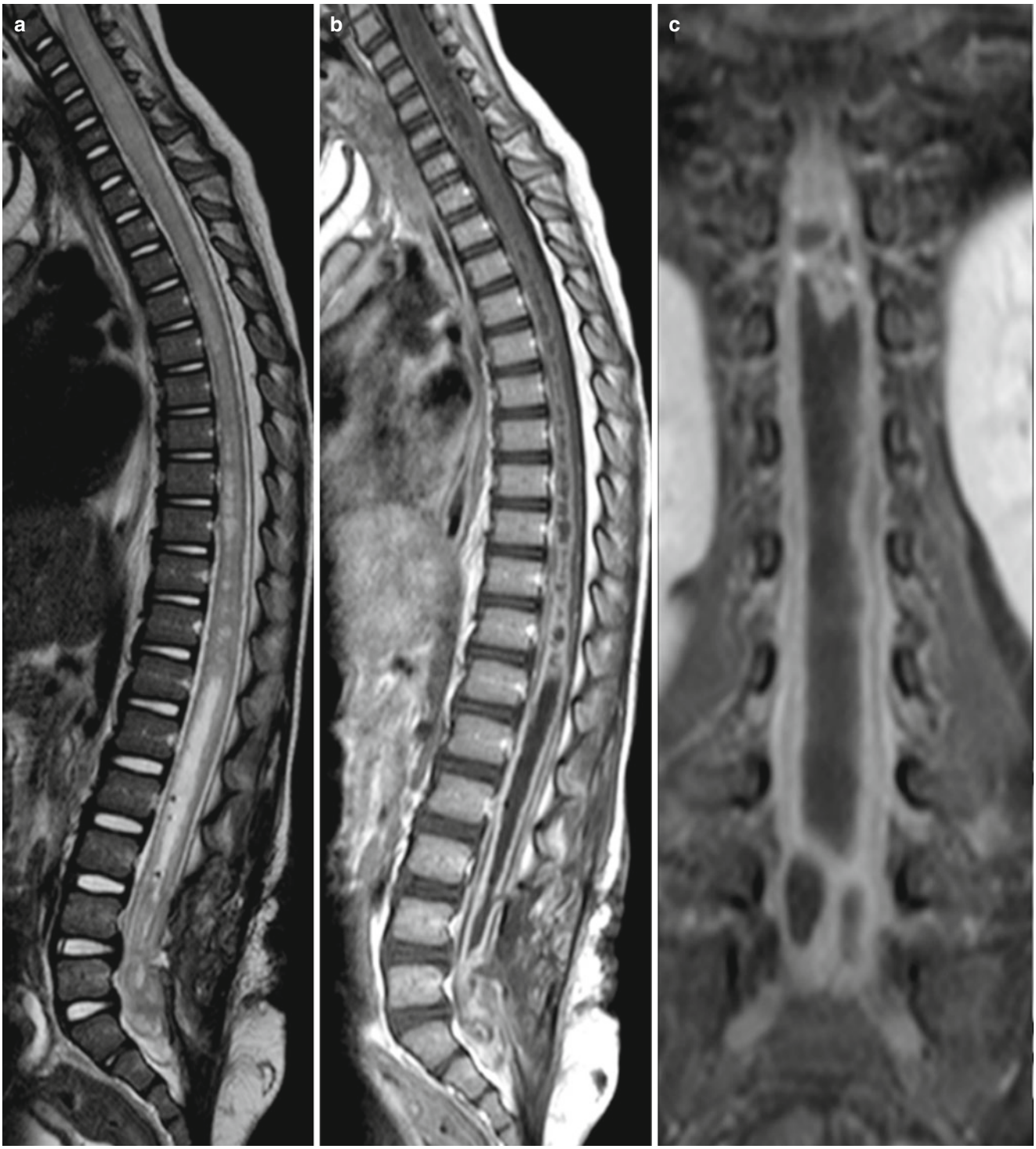

Fig. 8 Diffuse spinal cord abscess in a 2-year-old with prior surgery for lipomyelomeningocele. Sagittal T2-weighted image (**a**) shows diffuse swelling of the whole spinal cord, with signs of cavitation at the lumbar level. The termination of the spinal cord is low due to the pre-existing malformation. Gd-enhanced sagittal (**b**) and coronal (**c**) T1-weighted images show diffuse enhancement of the spinal cord, forming a syrinx-like abscess at the T12–L4 level

Guillain-Barré Syndrome Guillain-Barré syndrome (GBS) is an acute inflammatory disorder involving the spinal and peripheral nerves [16, 17], typically presenting after a recent viral disease (usually a respiratory illness or gastroenteritis). Patients present acutely with lower extremity weakness progressing to flaccid paralysis, often accompanied by sensory disturbances such as pain and paresthesia but rarely associated with sphincter dysfunction.

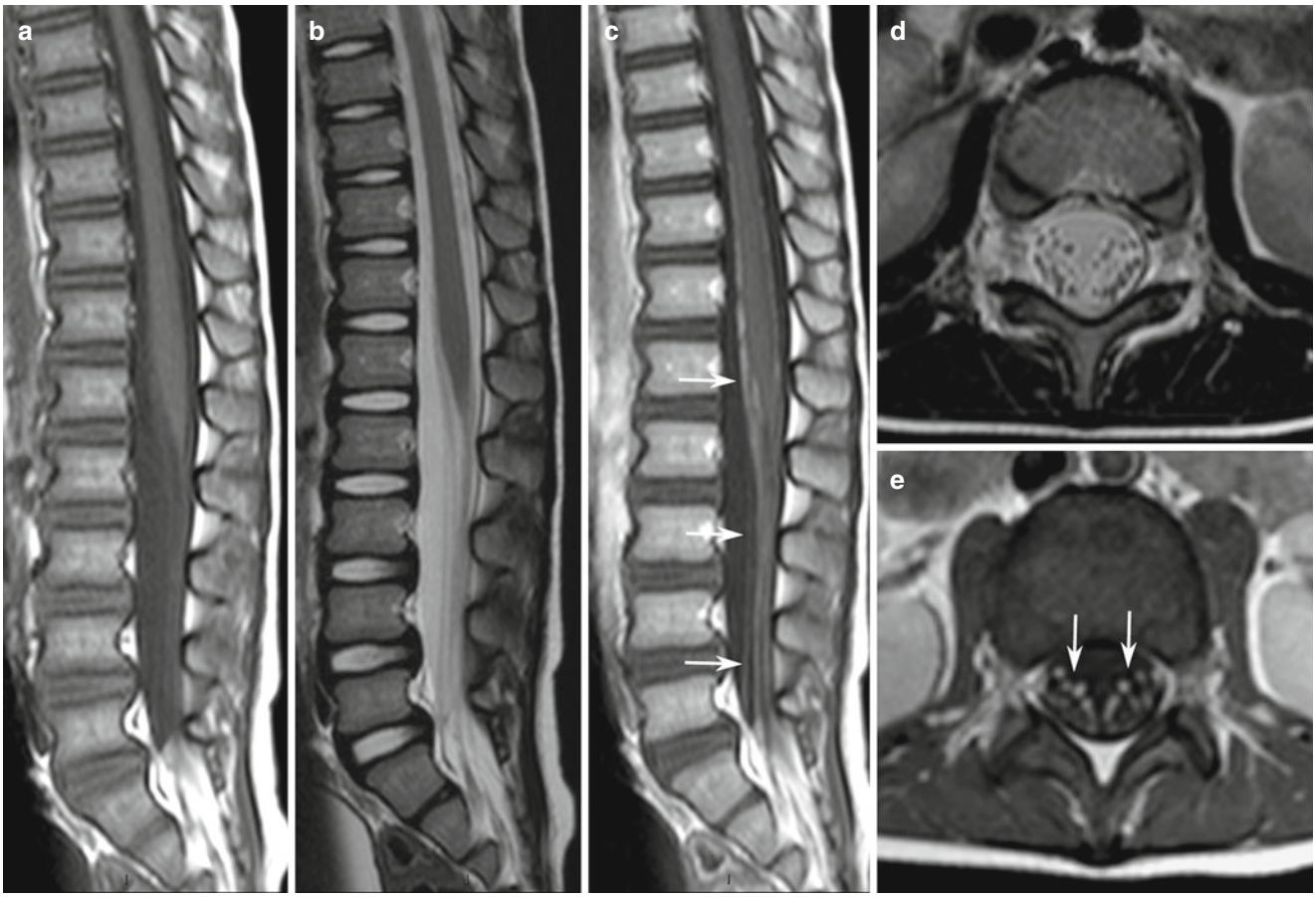

Fig. 9 Guillain-Barré syndrome in a 2-year-old girl presenting with rapidly progressive paraparesis. Sagittal T1-weighted (**a**) and T2-weighted (**b**) images are unremarkable, while Gd-enhanced sagittal T1-weighted image (**c**) shows enhancement of nerve roots of the cauda equina (*arrows*). Axial T2-weighted image (**d**) shows the caudal nerve roots are not thickened, while Gd-enhanced axial T1-weighted image (**e**) shows enhancement of anterior nerve roots in the cauda equine (*arrows*). The diagnosis would be missed without contrast material administration

CSF analysis shows elevation of proteins reflecting nerve root demyelination and a lack of inflammatory cells. GBS progresses rapidly, reaching a maximal deficit within 4 weeks of the onset, during which time approximately 40 % of children become non-ambulant and up to 20 % require ventilatory support [17].

The peculiarity of MRI in patients with GBS is that unenhanced scans are typically unrevealing, as spinal root thickening may be difficult to appraise or even totally absent. On the other hand, gadolinium administration reveals enhancement predominantly of the anterior nerve roots of the cauda equina (Fig. 9) and sometimes global thickening and enhancement of the whole cauda equina. Involvement of cranial nerves in the same inflammatory process is called Miller Fisher syndrome [18]. Affected patients complain with ophthalmoplegia, ptosis, facial weakness, and ataxia, and MRI shows enhancement of multiple cranial nerves.

Spondylodiscitis In children, spondylodiscitis can be pyogenic or tubercular (TB); infection typically originates in the vertebral body adjacent to the end plate and spreads to the disc secondarily. Patients present with nonspecific findings such as failure or refusal to walk, abdominal pain, fever, and chronic back pain; back pain is often the predominant complaint. When the cervical spine is affected, manifestations may include dysphagia and stiff neck. Progression to weakness and paralysis suggests the formation of an epidural abscess with compression of the spinal cord and nerve roots. Abscesses can be particularly large in TB spondylodiscitis and involve the paravertebral region extensively.

Conventional X-rays have very low sensitivity and specificity [19]; loss of definition and irregularity of the vertebral end plate, narrowing, and erosion of the disc space are seen. On MRI, reduction in the height of the intervertebral disc, swelling of the annulus, and T2 hyperintensity of the disc

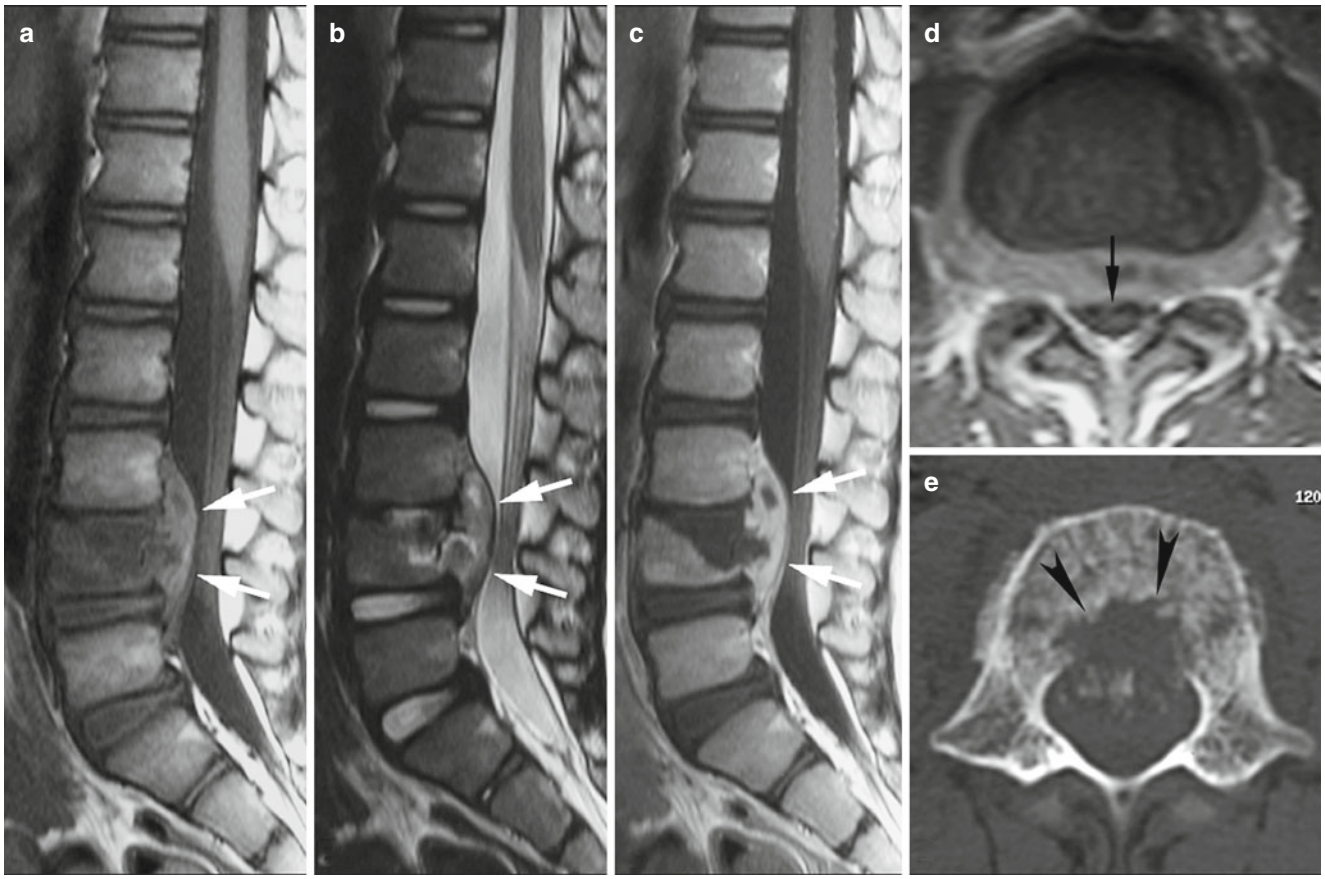

Fig. 10 Tubercular spondylodiscitis in an 8-year-old boy. Sagittal T1-weighted (**a**), T2-weighted (**b**), and Gd-enhanced T1-weighted (**c**) images show involvement of the central portion of the L4 vertebral body with disruption of the posterior vertebral wall and propagation into the spinal canal below the posterior longitudinal ligament, forming an epidural abscess (*arrows*). The L3–L4 disc space is irregular and probably already involved, albeit without frank, diffuse enhancement (*arrow*). Gd-enhanced axial T1-weighted image (**d**) shows the thecal sac is markedly compressed by the ventrally located collection, while axial CT scan (**e**) shows centroposterior vertebral body necrosis (*arrowheads*) with disruption of the posterior body wall

with enhancement after gadolinium administration are seen in the early stage of the disease. Signal changes of the vertebral end plates and subchondral regions are initially very subtle and are more easily picked up by STIR sequences. As the disease progresses, loss of integrity and definition of the vertebral end plates are detected.

Vertebral collapse can occur, especially in TB cases. Epidural abscesses are frequently seen and appear as a rim-enhancing abnormality in the epidural space whose nonenhancing center generally corresponds to pus (Fig. 10) [20].

Vascular Lesions

Spinal Cord Ischemia Spinal cord ischemia is rare in children and requires a predisposing condition such as cardiovascular surgery, arterial dissection, or fibrocartilaginous embolism. Patients experience a stroke-like presentation, with signs of acute spinal cord dysfunction which progress to nadir more rapidly than in inflammatory ATM, typically within 4 h of the onset. The neurologic presentation of spinal cord infarction is largely defined by the vascular territory involved [21]. The severity of the impairments can vary widely, from paraplegia to minor weakness On MRI (Fig. 11), typical anterior spinal artery distributions will result into involvement of the anterior horns of the gray matter, giving the so-called "snake's eye" appearance on axial T2-weighted images. Diffusion-weighted imaging (DWI) will show areas of restricted diffusion as in cerebral ischemia; however, implementation of DWI sequences is usually more cumbersome and less reliable in the spinal compartment than in the brain [22].

Spinal Cord Hemorrhage Hemorrhagic lesions of the spinal cord are exceedingly rare in the pediatric age group. Apart from exceptional idiopathic cases (Fig. 12), hematomyelia can be caused by bleeding diathesis due to

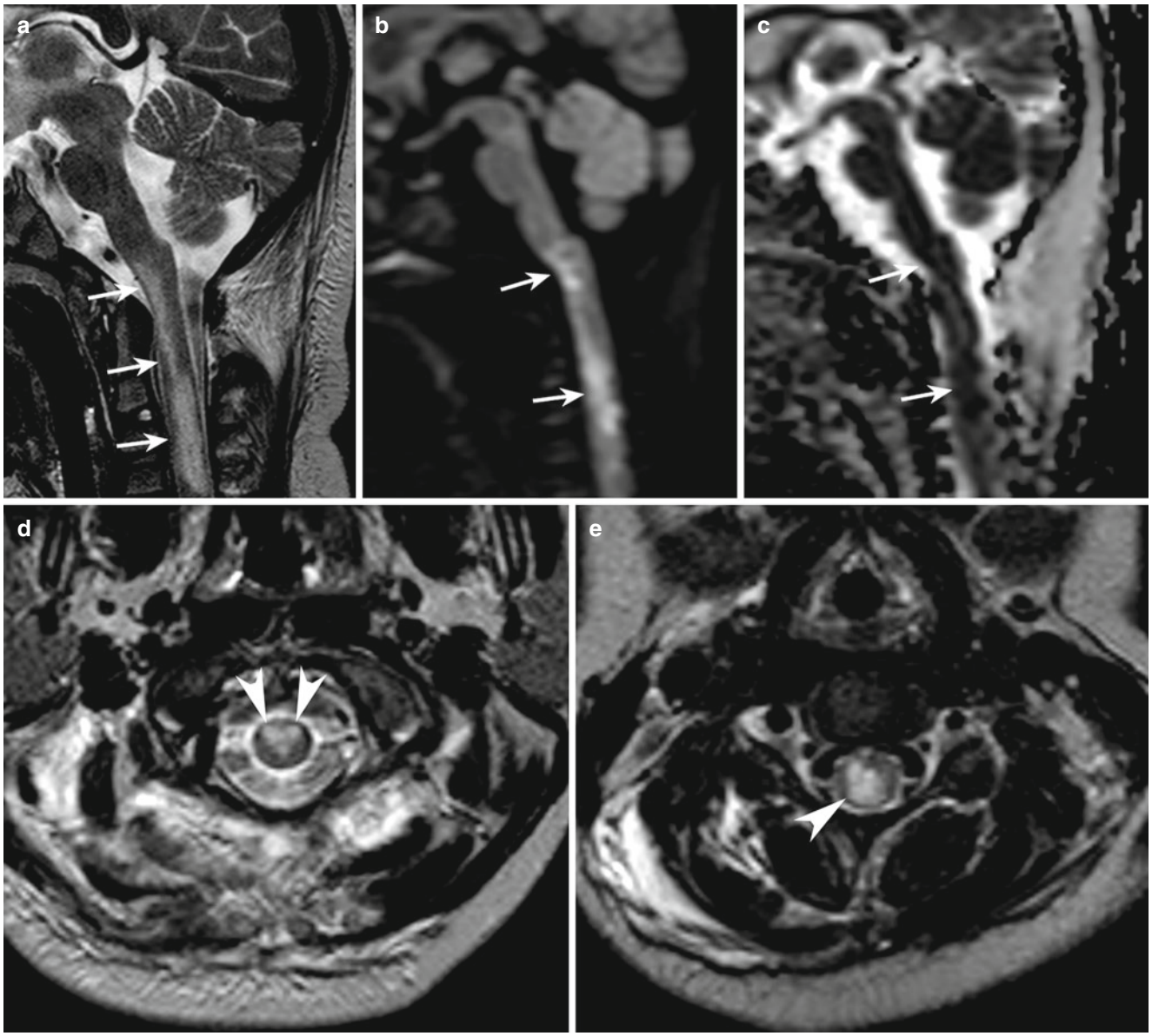

Fig. 11 Spinal cord ischemia in an 8-year-old girl with Down syndrome, craniocervical instability, and a stroke-like presentation with tetraparesis. Sagittal T2-weighted image (**a**) shows hyperintense signal (*arrows*) that involves the anterior aspect of the cervical cord cranially and has a more diffuse distribution caudally. Sagittal diffusion-weighted image (**b** 1000 m/s^2) (**b**) and corresponding ADC map (**c**) show areas of restricted diffusion. Axial T2-weighted images (**d**, **e**) show a typical "snake's eye" sign (*arrowheads*, **d**) due to the involvement of the anterior gray matter horns corresponding to the anterior spinal artery territory; at a more caudal level, the cross-sectional area of the cord is more diffusely involved (*arrowheads*, **e**). MR angiography (not shown) revealed left vertebral artery dissection

coagulation imbalance, trauma, or vascular malformations, such as arteriovenous malformations (AVM) or cavernous malformations. AVMs are usually identifiable because of the presence of tangles of dilated vessels forming an intramedullary or pial nidus [23]. Cavernomas are exceptionally found in the spinal compartment; spinal cord cavernomas produce blooming T2* hypointensities due to hemosiderin staining, similar to their cerebral counterparts.

Tumors

Tumors of the spine and spinal cord may present with chronic back pain; however, frequently these patients present in the emergency room despite the long-standing duration of their complaints, mainly when additional neurological deficits related to either compression or infiltration of the spinal cord and nerve roots occur. Spinal

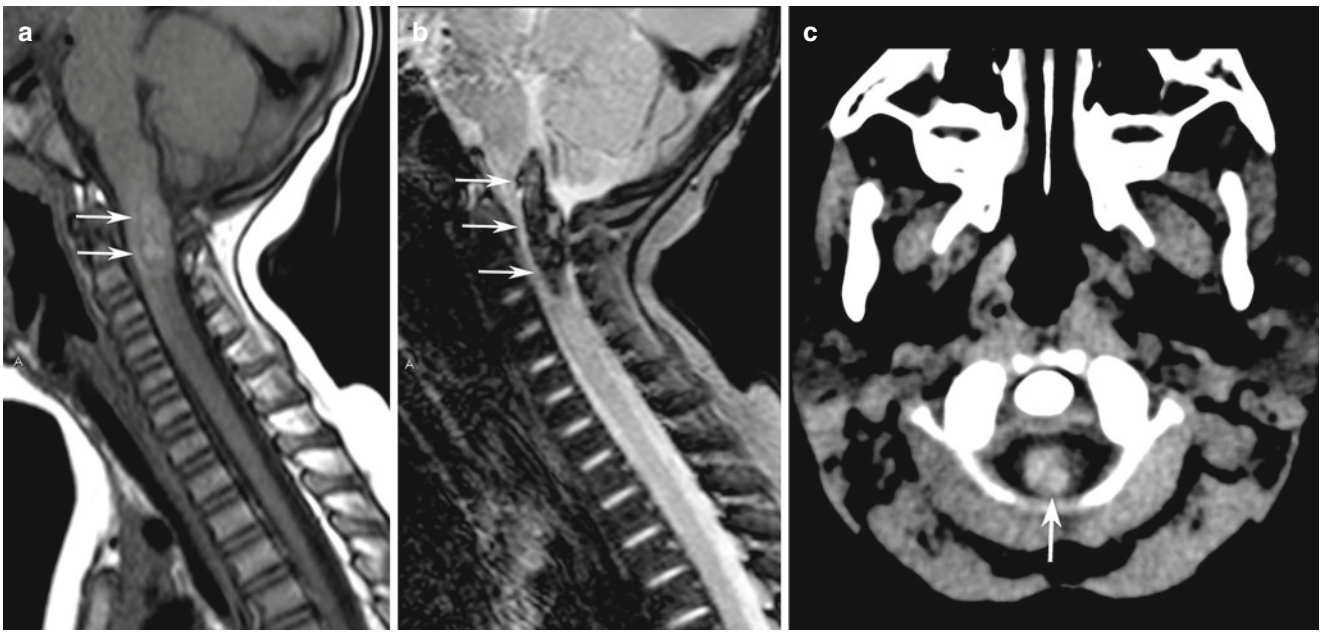

Fig. 12 Bulbo-medullary hemorrhage in a 1-year-old girl. Sagittal T1-weighted image (**a**) shows ill-defined hyperintensity at the bulbo-medullary junction (*arrows*). Sagittal gradient-echo T2*-weighted image (**b**) shows extensive hypointensity (*arrows*) consistent with fresh blood. Hemorrhage is confirmed by unenhanced CT scan (*arrow*, **c**). No cause could be identified in this case

tumors are classified according to the involved compartment into intramedullary, intradural extramedullary, and extradural [24].

Intramedullary Tumors They account for 25 % of pediatric spinal tumors and prevail in children between 1 and 5 years of age [24]. Astrocytomas (especially pilocytic) are the most common intramedullary tumor in the pediatric age group (82 % of cases) followed by gangliogliomas, whereas ependymomas are distinctly uncommon in children outside the setting of neurofibromatosis type 2. The presentation, duration, and course of the disease may be variable. Affected patients may have a prolonged duration of symptoms before a diagnosis is established. Back pain is often the earliest and most persistent complaint, and should prompt to MRI in order to rule out intraspinal pathology. Rigidity and contracture of the paravertebral muscles may result from thecal sac enlargement, involvement of adjacent bone, and impairment of cerebrospinal fluid (CSF) dynamics. Progressive scoliosis may cause delays in the diagnosis if underestimated. Head tilt and torticollis, as well as lower cranial nerve palsies with dysphagia, dyspnea, and dysphonia, may represent early signs of cervicomedullary neoplasms due to involvement of the spinal roots of the accessory nerve. Hydrocephalus with raised intracranial pressure may rarely represent the clinical presentation of intramedullary tumors and is caused by obstruction of the spinal subarachnoid spaces, CSF seeding, or increased CSF protein content.

On MRI, intramedullary neoplasms (Fig. 13) produce enlargement of the spinal cord giving heterogeneous signal intensity. They may be solid or associated with cysts, either neoplastic or nonneoplastic, which are better defined after gadolinium administration. They frequently involve a large portion of the cord, spanning multiple vertebral levels in length. Among intramedullary tumors, ependymomas are especially prone to spontaneous hemorrhage, which may cause abrupt clinical presentations.

Intradural Extramedullary Tumors In the pediatric age group, primitive tumors in this location are the least common among spinal tumors and are mostly represented by schwannomas and neurofibromas in neurofibromatosis patients. A host of other neoplasms, including filar ependymomas, meningiomas, and atypical teratoid rhabdoid tumors, can also be found. Clinical features basically are represented by pain and signs of cord or nerve root compression, depending on the location of the mass.

Extradural Tumors Extradural tumors account for about 2/3 of all spinal tumors in the pediatric age group and may be grouped into bone tumors, tumors of the epidural space, and extraspinal tumors invading the spine. Affected children usually complain with back pain and myeloradiculopathy. Neuroimaging of extradural tumors requires both MRI and CT. MRI depicts extradural soft tissue components,

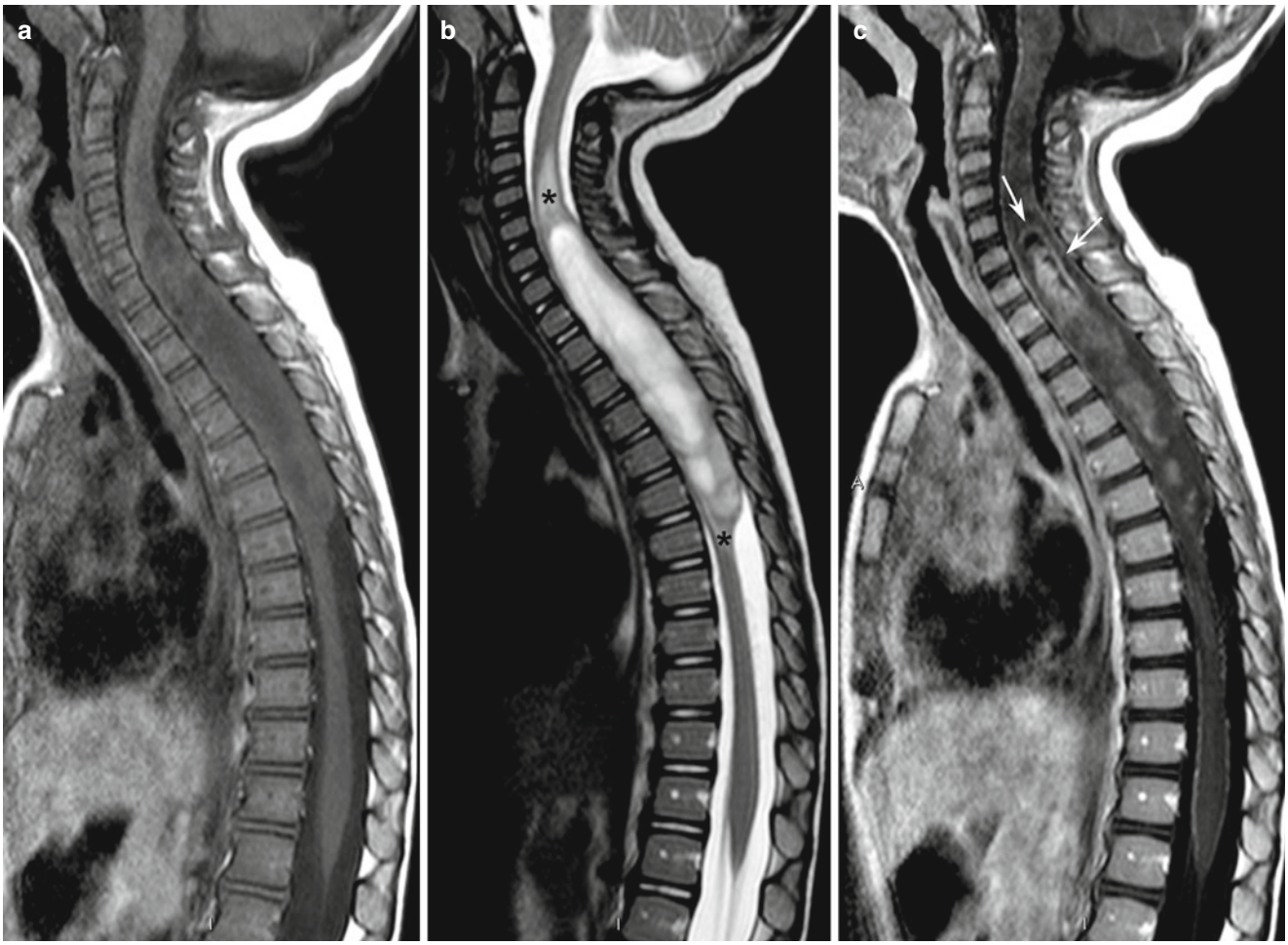

Fig. 13 Intramedullary pilocytic astrocytoma in a 2-year-old girl. Sagittal T1-weighted (**a**) and T2-weighted (**b**) images show intrinsic spinal cord lesion at the C6–T7 level, causing significant expansion of the cord and eliciting perifocal edema (*asterisks*, **b**). Gd-enhanced sagittal T1-weighted image (**c**) shows patchy enhancement, consistent with a mostly solid lesion, although a small peripheral cystic component with enhancing neoplastic walls (*arrows*) is seen at the cranial margin of the lesion. Aspecific venous engorgement is also seen along the pial surface of the thoracolumbar cord

bone marrow infiltration, and compressive myelopathy compression from the tumor, whereas CT detects the osteolytic or osteosclerotic nature of the lesion and the degree of involvement of bone [24].

Among primary bone tumors, *osteoid osteomas* are characterized by an acute presentation with nocturnal pain that recedes with nonsteroid anti-inflammatory medications and/or with painful scoliosis. CT shows the nidus of the osteoma as a rounded hypodense lesion surrounded by a hyperdense sclerotic ring and containing a calcified spot resulting in a "target" appearance. Because of the small size of the lesion, accuracy of MRI in identification of osteoid osteomas is not high; however, MRI clearly detects the extensive reactive soft-tissue masses that are frequently associated with the osteoma (Fig. 14) [25].

Extrinsic compression of the spinal cord and/or nerve roots is the presenting sign of paravertebral *neuroblastoma.* These tumors are the most common non-CNS solid tumors in the pediatric age group and typically affect children younger than 5 years. The tumor mass is typically huge and may show extensive necrotic-hemorrhagic areas and calcification. The clinical presentation is variable depending on the variable location and size of the mass; however, neurologic signs typically ensue with intraspinal extension and thecal sac compression. Neuroblastomas originating from the paravertebral sympathetic chains typically display a "dumbbell" growth through one or more neural foramina, extending a variably sized component into the spinal canal that compresses and displaces the thecal sac and spinal cord (Fig. 15). Other extradural tumors causing spinal cord compression

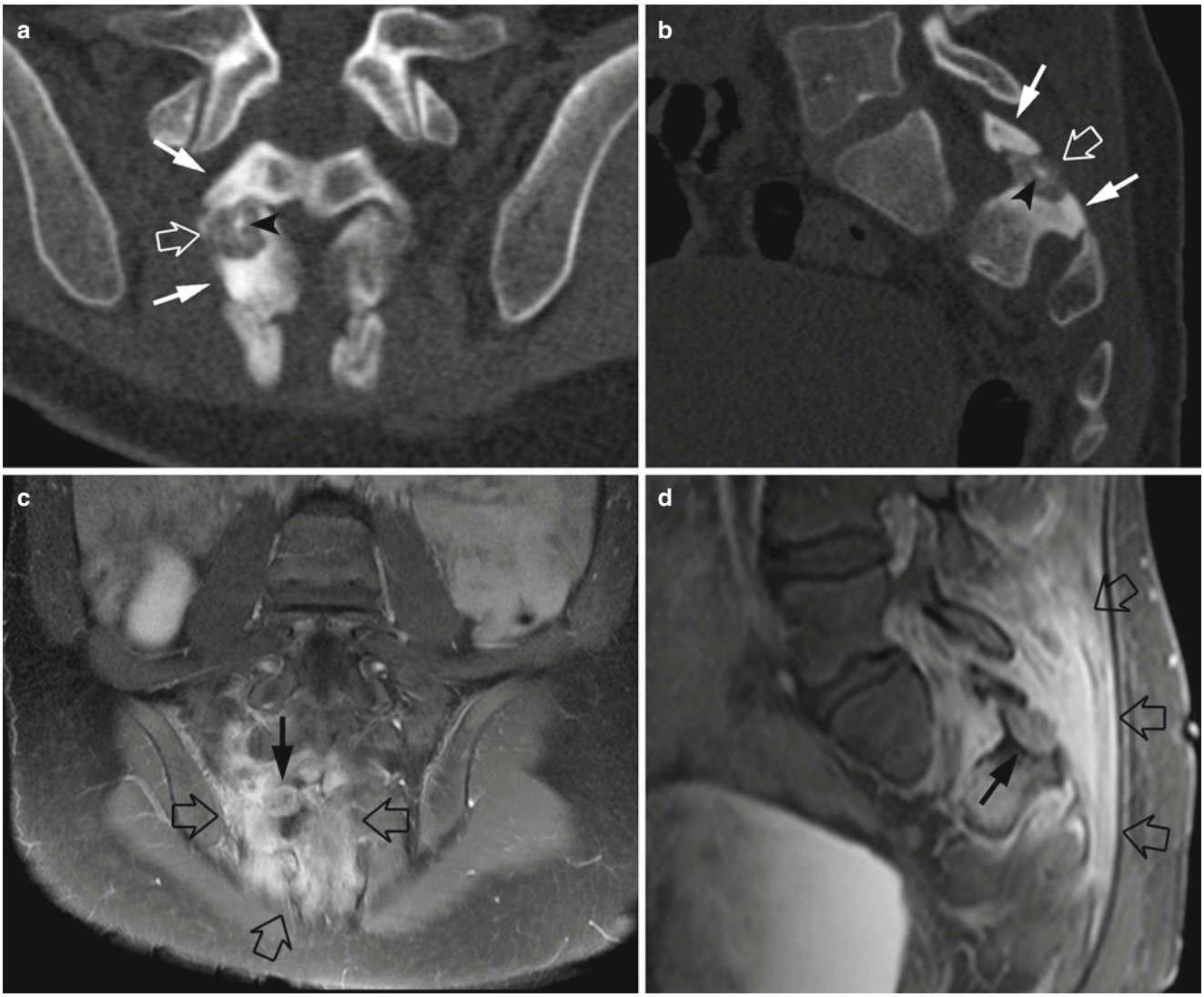

Fig. 14 Osteoid osteoma in a 12-year-old boy with intense low back pain. Coronal (**a**) and sagittal (**b**) reformatted CT images show radiolucent lesion (*empty arrow*) with central sclerotic spot (*arrowhead*, **a**) involving the right S2 lamina and inducing a perifocal sclerotic reaction (*thin arrows*). Contrast-enhanced coronal (**c**) and sagittal (**d**) MR images acquired with a fat suppression technique barely show the lesion (*arrow*), engulfed by a marked inflammatory reaction of the surrounding soft tissues (*empty arrows*)

include Ewing's sarcoma, hematological malignancies, and nerve sheath tumors [24].

Musculoskeletal Disorders

A large host of musculoskeletal conditions may present with acute or chronic pain, often resulting in an emergency room presentation. Only the most common entities will be briefly discussed here.

Juvenile Idiopathic Arthritis Arthritis is a heterogeneous group of musculoskeletal disorders, comprising a large host of different diseases or conditions that affect the joints, bones, muscles, cartilage, and other connective tissues, hampering or halting physical movement. The most common form in children is juvenile idiopathic arthritis (JIA). In the spine, JIA most commonly involves the cervical region; symptomatic patients present with pain, stiffness, torticollis, and limited range motion of the head.

MRI reveals synovitis and joint effusion, bone marrow edema, and bone erosions generally involving the C2–C3 level (Fig. 16). There can be malalignment of the articular surfaces of atlanto-occipital, atlanto-axial or cervical facet joints, or of two adjacent vertebrae. Interruption of the osseous joint

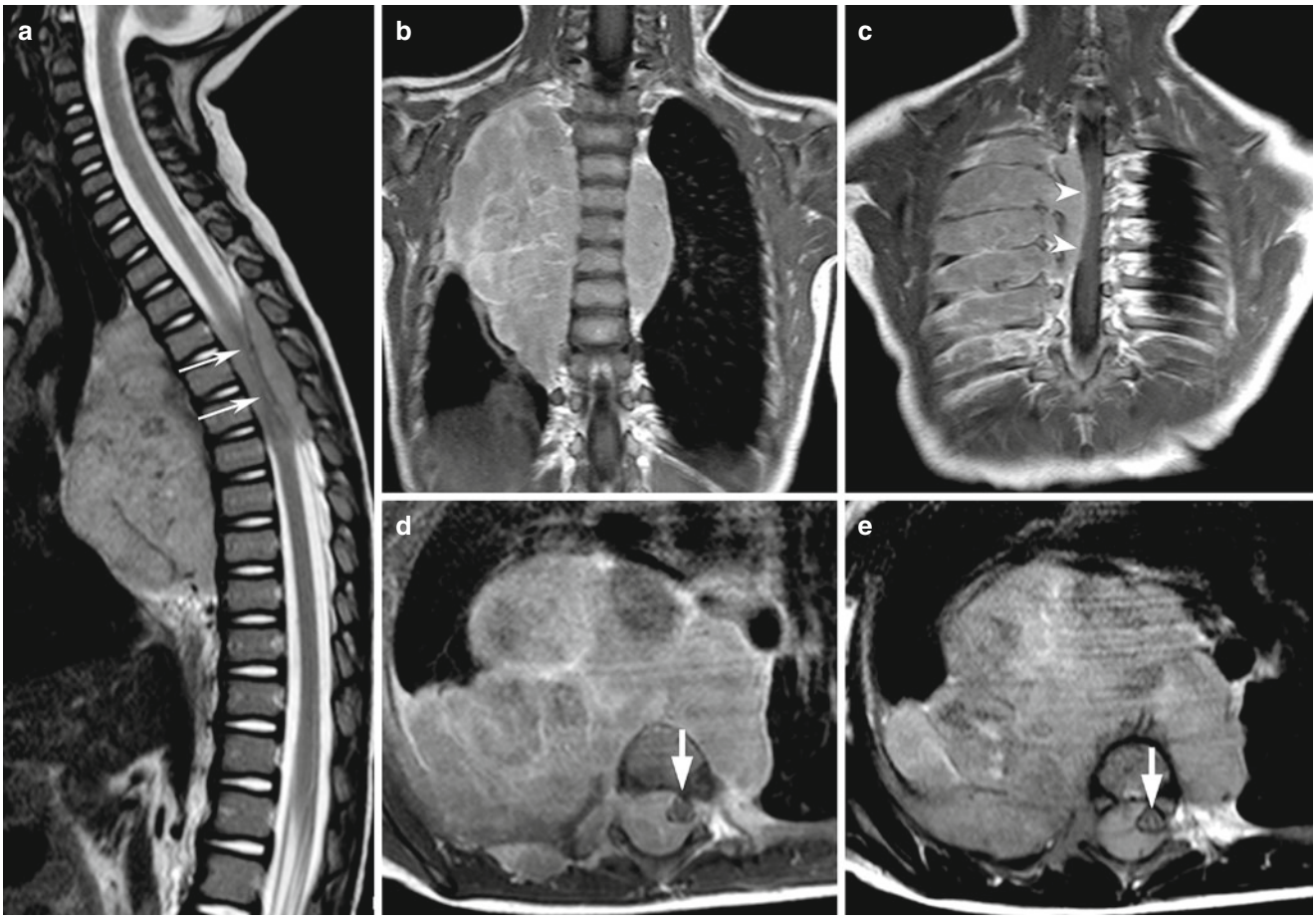

Fig. 15 Thoracic neuroblastoma causing spinal cord compression in a 1-year-old boy. Sagittal T2-weighted image shows huge prevertebral mass that has extended intraspinally, causing spinal cord compression (*arrows*). Contrast-enhanced coronal T1-weighted images (**b**, **c**) show intraspinal extension (*arrows*) has occurred through multiple neural foramina. Notice the absence of scoliosis despite the huge size of the mass, a typical feature of neuroblastoma. On contrast-enhanced axial T1-weighted image (**d**) and axial T2-weighted image (**e**) the thecal sac containing the cord is seen to be markedly displaced (*arrow*, **d**); central spinal cord T2 hyperintensity (*arrow*, **e**) is consistent with edema due to mechanical compression

surface with signs of inflammation, including hyperintense signal of the interarticular space, synovia, and subjacent bone in STIR images with corresponding enhancement on post-contrast T1-weighted images, is classically found [26]. The dens becomes eroded, first anteriorly and then posteriorly, and may become hypertrophic causing narrowing of the spinal canal at the craniocervical junction with possible neurological impairment due to spinal cord compression.

Disc Space Calcification Disc space calcification (DSC), or intervertebral disc calcification, is a poorly understood, uncommon condition characterized by calcification of the intervertebral disc, usually in the cervical spine. Most patients are boys aged 6–10 years who complain with local pain or torticollis; intraspinal herniation of a calcified disc fragment may cause acute neurologic signs such as radiculopathy or sensorimotor signs, similar to other forms of disc herniation. The etiology of DSC is unknown, and the entity is considered idiopathic in the pediatric age group; it is typically self-limiting, and conservative treatment is advocated [27].

Conventional X-rays may show an ovoid calcification in the intervertebral disc space. CT scan confirms the presence of the calcification in the disc space as well as any extruded disc fragment that may impinge on the thecal sac and spinal cord [28]. The vertebral end plates may appear irregular, with areas of subchondral sclerosis or Schmorl nodes. MRI is especially useful to depict associated herniations and the effects on the spinal cord (Fig. 17).

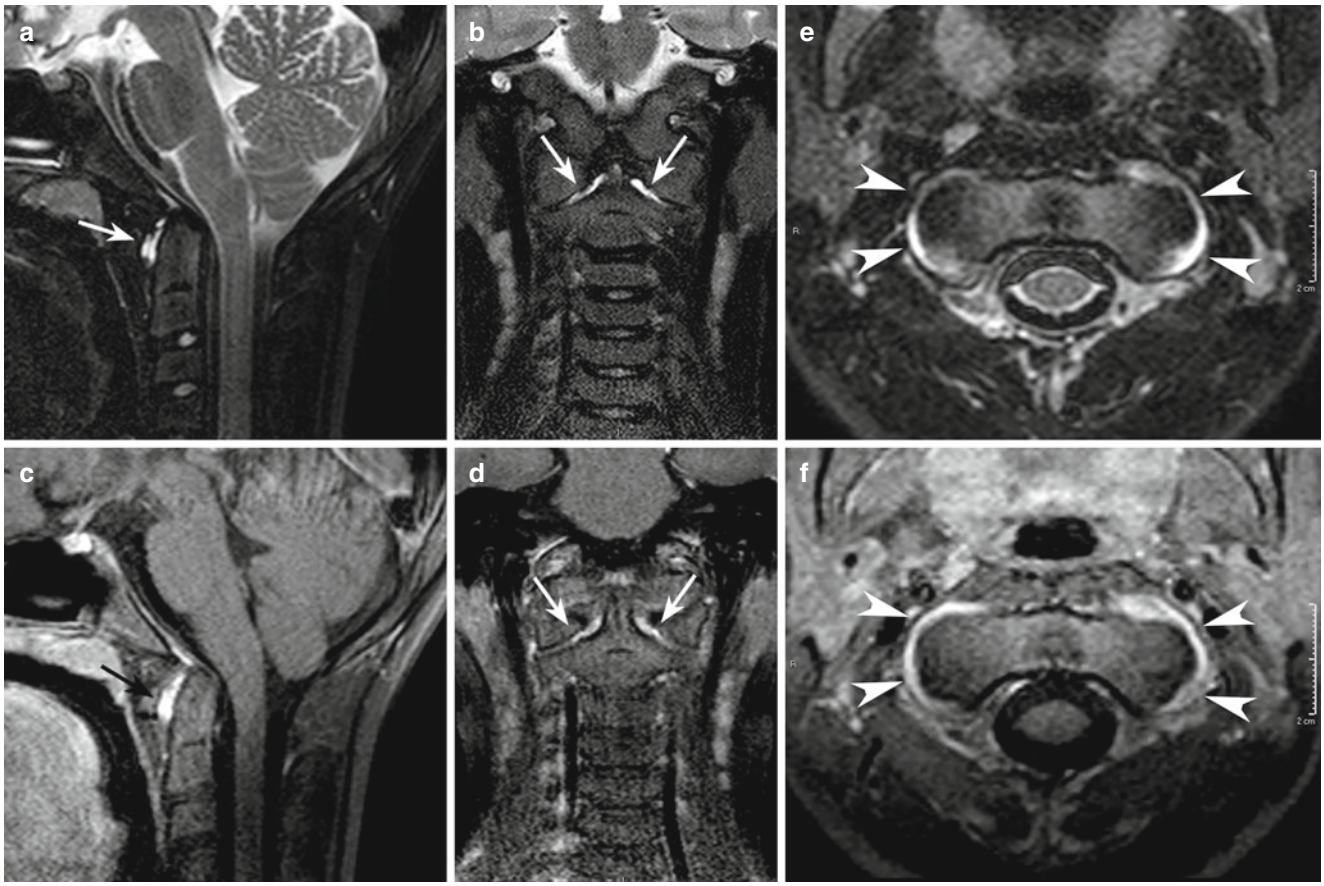

Fig. 16 Juvenile idiopathic arthritis in a 9-year-old girl presenting with acute cervical pain. (**a**) Sagittal and (**b**) coronal STIR images show fluid collection in the atlanto-axial interarticular space bilaterally (*arrows*). (**c**) Sagittal and (**d**) coronal Gd-enhanced T1-weighted images with fat suppression show the collections enhance (*arrows*), consistent with synovitis. (**e**) Axial STIR image and (**f**) Gd-enhanced, fat-suppressed T1-weighted image show enhancing collection surrounding the lateral masses of the atlas bilaterally (*arrows*)

Chronic Recurrent Multifocal Osteomyelitis Chronic recurrent multifocal osteomyelitis (CRMO) is a sterile skeletal inflammation occurring primarily in childhood and adolescence, predominantly in girls. The cause is unknown; autoimmune mechanisms and genetic susceptibility have been implicated. The disease has a long, fluctuating course with exacerbations and remissions. Pain, rigidity, and malaise are the most common complaints. The diagnosis is often one of exclusion in a patient with multiple localized skeletal lesions. Vertebral involvement is often multifocal, with the thoracic spine being involved most commonly. When present, vertebral collapse may progress to kyphosis and vertebra plana.

On CT, the involved vertebrae show a mottled lytic-sclerotic appearance and wedge-like deformation. MRI shows osseous edema of the involved vertebrae, which is exquisitely depicted by STIR images, while the intervertebral discs are typically spared. On post-contrast T1-weighted images, enhancement of the involved vertebrae is best appreciated when fat-suppressed techniques are used [29] (Fig. 18).

Spondylolysis and Spondylolisthesis Spondylolysis is a bone defect of the neural arch, usually at level of the pars interarticularis. Its cause may be traumatic (stress fracture) or dysplastic. Spondylolysis accounts for the majority of cases of low back pain in children older than 7–8 years and is commonly seen in individuals who practice sports [30]. It is more often bilateral than unilateral and involves L5 in the vast majority of cases. This condition allows for excessive motility, and therefore potential instability, of the lumbar spine with respect to the sacrum, often leading to spondylolisthesis, i.e., anterior dislocation, of L5 with respect to S1.

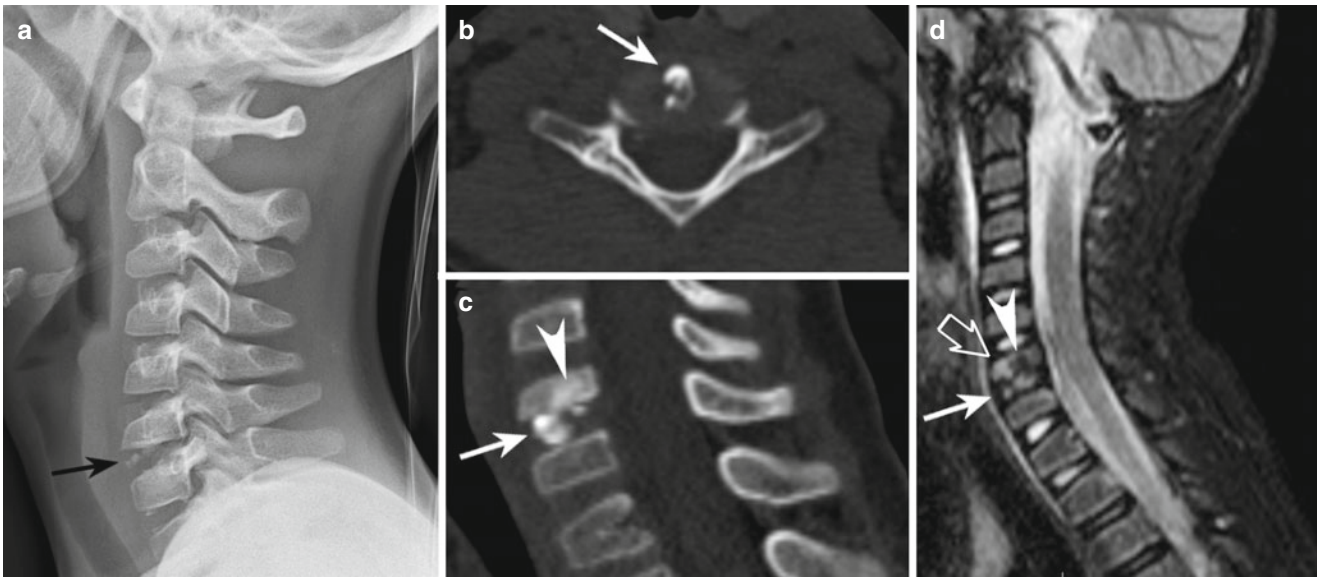

Fig. 17 Disc space calcification in a 7-year-old girl. (**a**) Conventional X-ray of the cervical spine in lateral projection shows densities in the C6–C7 intervertebral space (*arrow*). (**b**) Axial CT scan and (**c**) sagittal reformatted CT image shows calcified clusters involving the C6–C7 intervertebral disc. The C6 vertebral body is slightly reduced in height and shows a subchondral hyperdensity (*arrowhead*). (**d**) Sagittal STIR image shows the C6–C7 disc (*arrow*) is slightly hypointense compared to unaffected discs; the subchondral calcification also appears hypointense (*arrowhead*) and stands out on a diffuse background of mild hyperintensity (*empty arrow*) indicative of bone marrow edema

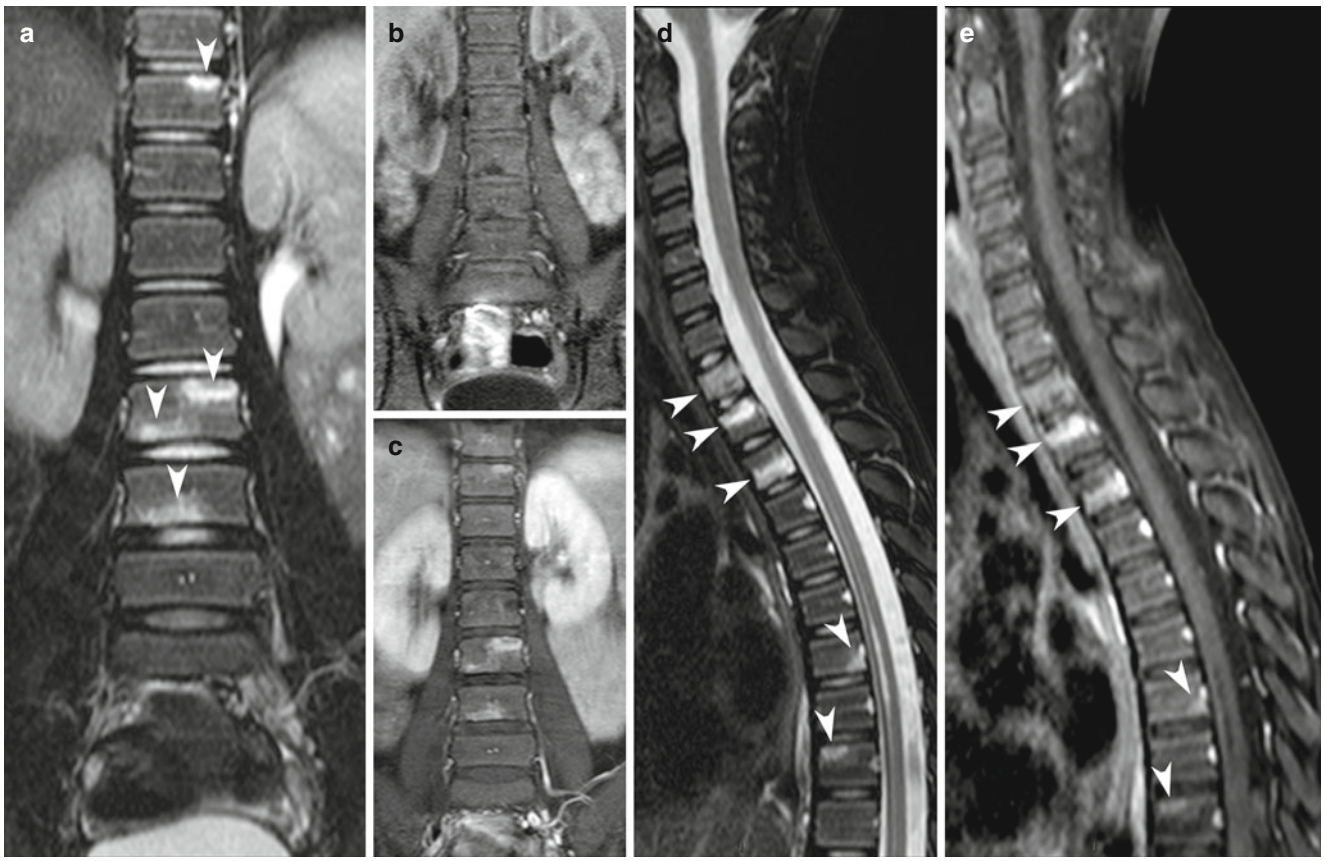

Fig. 18 Chronic recurrent multifocal osteomyelitis in a 12-year-old girl. (**a**) Coronal and (**d**) sagittal STIR images show multiple ill-defined areas of abnormal signal intensity involving several vertebral bodies in the whole spine (*arrowheads*). (**b**) Coronal fat-suppressed T1-weighted image is inconspicuous; (**c**) coronal and (**e**) sagittal Gd-enhanced fat-suppressed T1-weighted images show enhancement of the vertebral abnormalities (*arrowheads*). Notice there is no vertebral collapse in this case, and the intervertebral discs are spared. There is incidental hydrosyringomyelia

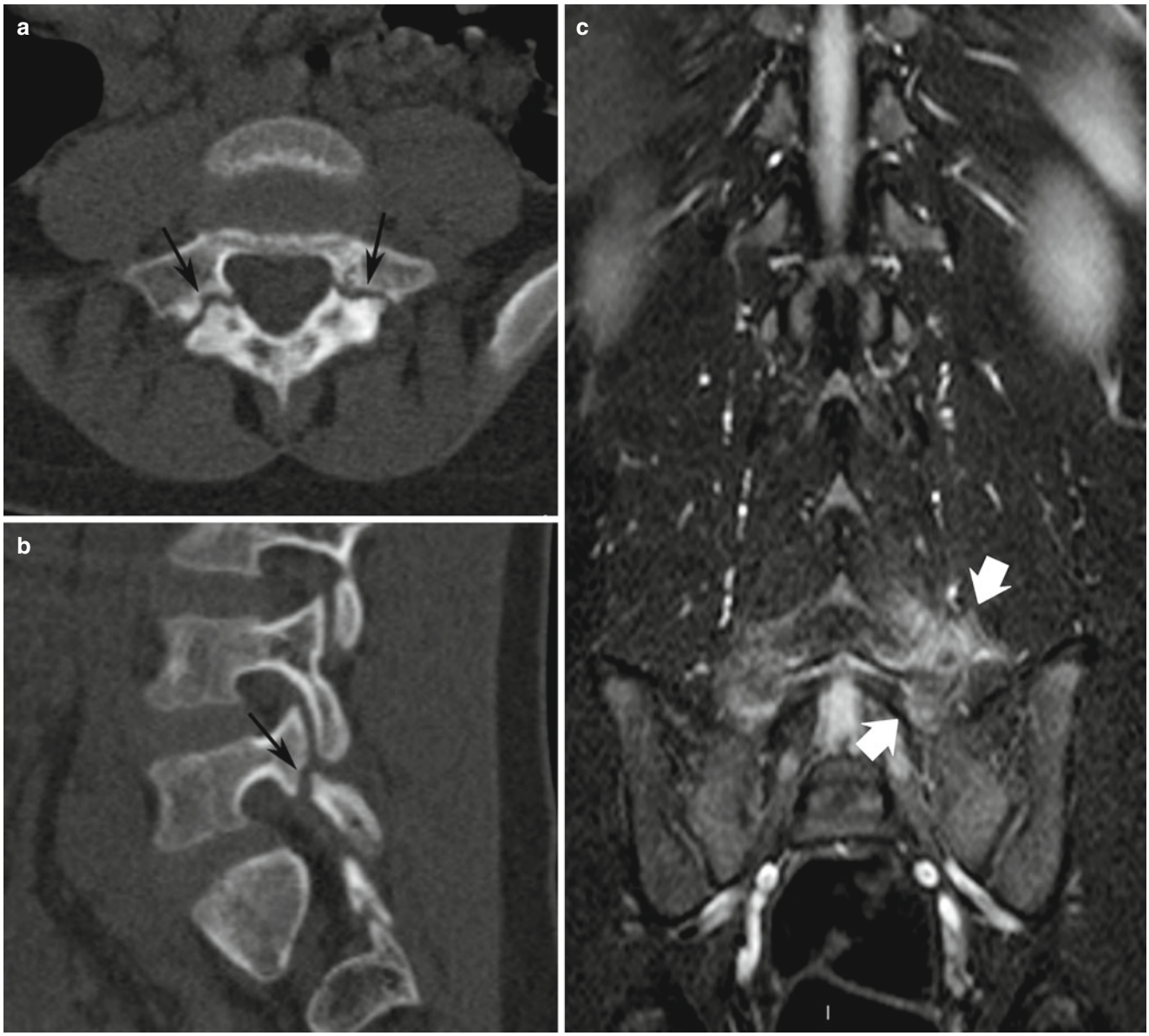

Fig. 19 Isthmic spondylolysis of L5 in a 9-year-old boy presenting with low back pain. CT scan on axial (**a**) and parasagittal (**b**) projections shows radiolucent line crossing the isthmus (or pars interarticularis) bilaterally (*arrows*), surrounded by reactive sclerosis. There is not associated spondylolisthesis in this case; however, note that images were acquired in the supine position, i.e., without the normal axial load which could exacerbate listhesis. Coronal STIR MR image (**c**) does not directly show the lysis, but reveals inflammatory reaction involving the surrounding soft tissues (*arrows*)

CT shows a thin, irregular interruption of the neural arch at level of the isthmus; sagittal reformats perfectly display the possible associated listhesis. On MRI, the defect may not be seen equally well; however, MRI is more useful to evaluate associated features, such as disc bulging and foraminal stenosis. MRI also shows redundant masses of inflammatory tissue surrounding the pars defect as well as edema of the spongious bone, well seen on STIR images (Fig. 19).

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