Dumbbell Tumors of the Spine

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22.1 Definition

The term "dumbbell tumor" was initially introduced by Heuer in 1929 to describe spinal tumors that acquire an hourglass shape as they encounter an anatomic barrier, such as dura mater, a nerve-root foramen, or other bony elements, as they grow [1–3]. Spinal tumors with significant intraspinal and paravertebral involvement are classified into four types based on the location of the tumor: intramedullary, intradural extramedullary, epidural, and dumbbell-shaped [4]. Dumbbell tumors can be assigned to various groups according to the constricting structure and other details of tumor location [5]. These days, the term "dumbbell tumor" does not refer to the hourglass shape, but rather acts as a stand-in conceptual term referring to separate tumors that connect and comprise two or more separate regions, such as the intradural or epidural space, or locations outside the spinal canal [6].

Schwannoma and meningioma are the two most common intradural spinal tumors. Ninety percent of spinal dumbbell tumors are schwannomas [4], and up to 33% of schwannomas have a dumbbell form [7]. Multiple schwannomas more frequently represent a clinical manifestation of neurofibromatosis type 2 [8]. Non-schwannoma non-neurofibroma dumbbell tumors of the spinal cord include 28 different pathological entities: hemangioma [9, 10]; meningioma [11]; malignant peripheral nerve sheath tumors [12]; neurogenic paravertebral tumors with origin from neurogenic elements within the thorax [13], including neuroblastoma [14], ganglioneuroblastoma [14], and ganglioneuroma [15]; hemangioblastoma [16];

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liposarcoma [17]; lipoblastoma [18]; angiomatosis [19]; angiolipoma [20]; rhabdomyosarcoma [6]; spine extraosseous chordoma (SEC) [21]; mesenchymal chondrosarcoma [22]; soft tissue chondroma [23]; osteochondroma [24]; malignant glomus tumor [25]; malignant solitary fibrous tumor [26]; plasmacytoma [27]; metastasis [28]; Ewing sarcoma [29]; atypical teratoid rhabdoid tumor [30]; lymphoma [31]; lymphangioma [32]; meningeal melanocytoma [33]; small cell malignant tumor [34]; and peripheral primitive neuroectodermal tumor (PNET) [35]. In addition, malignant dumbbell tumors accounted for 64% of cases in pediatric patients and 2.8% in adult patients [6].

22.2 Epidemiology

In large series of spinal cord tumors, the incidence of dumbbell shaped lesions varies between 6% [3] and 24% [2, 6–8, 36, 37]. There is a significantly higher rate in the cervical spine of up to 44% [6].

22.3 Classification

The first classification of dumbbell tumors was the Eden Classification, which served as the gold standard for decades [2]. Eden designated four cross-sectional configurations for dumbbell tumors in 1958, a time at which neither computed tomography (CT) nor magnetic resonance imaging (MRI) had been developed (Fig. 22.1). Although this is a morphological as opposed to a surgical classification, it was postulated that Type I, II and III tumors can be operated on through a posterior route, and that the anterior approach is appropriate for Eden Type IV tumors. In cases of Type II and III tumors (i.e., the extraspinal component of the tumor compresses and shifts the vertebral artery and extends anteriorly beyond the vertebral artery), combined anterior and posterior approaches can be considered [38]. One series of 118 cases showed that Type III tumors were the most frequent type (53%) followed by Type II (33%), Type I (9%), and Type IV (5%) [6].

There are seven other classifications of dumbbell tumors, in addition to the Eden Classification. Liu et al. introduced an anatomic classification of dumbbell tumors in 2017 [34]. The largest transverse section of the tumor was divided into four areas; each area needed different surgical procedures. A modification of the Eden classification due to CT and MRI advances was provided by Toyama et al. [39]. In addition to an axial configuration, Toyama et al. categorized dumbbell tumors in an imaging-based anatomic 3-dimensional classification according to the number of intervertebral and transverse foramina involved for each tumor, which was better suited for surgical planning [5]**.** Special sub-classifications for cervical dumbbell tumors of the spine were developed by Hiramatsu [40], which classified both the horizontal and craniocaudal spread of dumbbell tumors. Jiang et al. developed the so-called Peking University Third Hospital (PUTH) anatomical classification of the cervical dumbbell tumors in 2009 [41], which differentiated between erosive or compressive

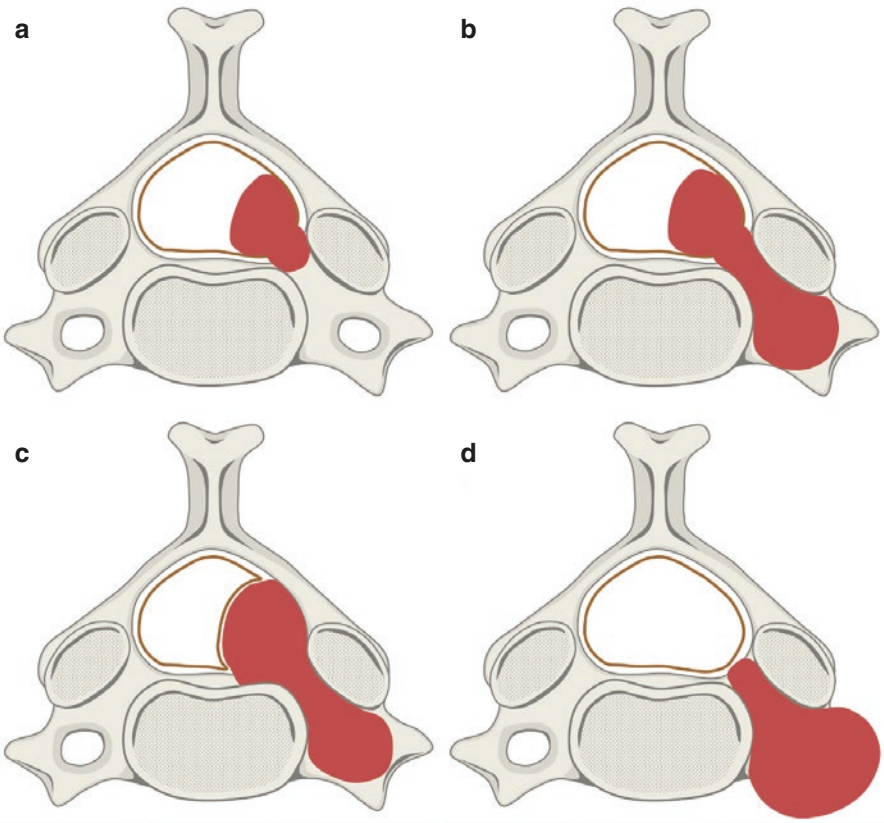


Fig. 22.1 Eden Classification of the dumbbell tumors of the spine. (**a**) Type 1: intradural and extradural. (**b**) Type 2: intradural, extradural, and paravertebral. (**c**) Type 3: intradural and paravertebral. (**d**) Type 4: Foraminal and Paravertebral

bony change. This classification included intraspinal lesions in front of the spinal cord, and standardized surgical procedures according to category. Sridhar et al. proposed a 5-type classification system that was limited to giant invasive dumbbell spinal schwannomas [42]. Modifications of the Shridhar et al. classification were provided by Park et al. [43] and Kotil et al. [44].

22.4 Differential Diagnosis

22.4.1 Symptoms

Presentation of spinal dumbbell tumors depends on the size and location of the tumor. Most patients with spinal dumbbell tumors present with similar symptoms, regardless of the underlying pathology. Non-radicular pain is a common symptom,

followed by sensory deficits and gait disturbances, radiculopathy, motor weakness, ambulation, and bowel and bladder-function impairment [45, 46]. Non-radicular pain can persist through the follow-up, while radiculopathy tends to completely resolve following surgery [46]. Extraforaminal thoracic giant dumbbell tumors (e.g., giant schwannomas) can compress the lungs and vascular structures, and giant lumbosacral tumors can compress abdominal and visceral structures, resulting in urination problems and constipation [47]. Furthermore, tumors that are located in the cauda equina can cause vertebral erosion, resulting in instability and pain [47]. Vascular lesions can present with bleeding that mimics spinal subarachnoid hemorrhage or leads to progressive neurological deficits [48, 49]. Known primary tumor in the patient history could indicate the presence of metastasis. In pediatric patients, rare childhood tumors—like neuroblastoma, ganglioneuroma, atypical rhabdoid theratoid tumor, Ewing sarcoma, or lymphoma—should be taken into consideration. Multiple schwannomas can indicate the presence of neurofibromatosis type 2. Dumbbell tumors among pediatric aged patients are more likely to be malignant than among adult patients [6].

22.4.2 Radiological Presentation

More than half of dumbbell tumors are completely restricted to the extradural space, although preoperative MRI in some cases suggests the presence of intradural/extradural tumors [50, 51]. A wide variety of unusual lesions, which can cause neural foraminal widening, need to be taken into consideration, including the following: neoplastic lesions, such as benign/malign peripheral nerve sheath tumors (PNSTs), solitary bone plasmacytoma (SBP), chondroid chordoma, superior sulcus tumor, and metastasis; and non-neoplastic lesions, such as infectious process (tuberculosis, hydatid cyst), aneurysmal bone cyst (ABC), synovial cyst, traumatic pseudomeningocele, arachnoid cyst, or vertebral artery tortuosity [52].

It is often impossible to differentiate between dumbbell-shaped schwannoma, meningioma, or vascular lesion. MRI yields no definitive findings—both neurogenic tumors and hemangiomas are isointense-to-hypointense in T1 imaging, and homogeneously hyperintense in T2 imaging [53]. Regular enhancement is seen upon gadolinium administration. As a general rule, a schwannoma diagnosis should be made when a spinal intradural extramedullary tumor shows hyperintensity on T2-weighted imaging or intense enhancement without dural tail sign; otherwise meningioma is a more probable diagnosis [54]. Positive predictors of meningiomas are the dural tail-sign on contrast-enhanced T1-weighted imaging and calcification on CT scans [55]. For schwannomas, fluid signal intensity of the tumor on T2-weigthed imaging, rim enhancement on T1-weigthed imaging with gadolinium-diethylenetriamine pentaacetic acid (Gd-DTPA), and bone scalloping on CT scans with bone windows are predictive factors [55]. One characteristic of spinal meningiomas is the so called “gingko leaf sign” [56] seen on axial post-contrast T1 imaging: the distorted spinal cord pushed to one side of the theca by the meningioma depicts the “leaf”-shape, and the stretched dentate ligament depicts the p “stem” of

the leaf (seen as a non-enhancing "streak") [56]. In large tumors, cystic and necrotic zones can be observed [57]. Intracranial schwannomas are more likely to accompany spinal schwannomas [57], which is one reason for why we recommend MRI of the head and total spine prior to surgery.

Vascular lesions are particularly important when making a differential diagnosis since they can present with bleeding [48]. It is also important to take vascular tumor in consideration when planning surgery. A cavernous hemangioma should be included in the differential diagnosis of dumbbell-shaped spinal tumors when the p intervertebral foramina is not highly dilated and a non-enhanced nerve root is identified in the tumor with a lobular contour [58]. One of the signs most indicative of hemangioblastoma is the presence of blood flow voids on T2-weighted MRI [59].

Differentiation between benign and malignant lesions on MRI could be made using dumbbell scoring system [27]. Tumors larger than 5 cm or that increase in size on follow-up, irregular boundaries with perilesional edema, irregularly lobulated shaped, and osteolytic p bone destruction could point to a malignant lesion [27] and could be important in decision-making and for indications for a biopsy. The occurrence of paraspinal infiltration along the muscle fascicles suggests the possibility of lymphoma, which can be decisive in treatment since lymphomas are extremely chemo-sensitive and radiosensitive [60].

We recommend CT studies in all cases in which bony erosions are suspected. The extent of widening and erosion of one or more neuroforamina can provide insight into the instability of the spine and the need for stabilization following resection. Bony erosions can result from large benign tumors due to the compression effect of the tumor, but also due to aggressive malignant neoplasms. In most cases, the bony change is simply compressive with the widened intervertebral foramen [41]. However, in some special cases, the tumor erodes the p vertebral body, the facet joint, or—less commonly—the lamina. Sometimes the tumor makes small but deep nidus in the bone. In these two circumstances, radical bony lesion resection is mandatory; a less aggressive bone lesion resection could lead to tumor recurrence [41]. Standard X-ray images, including anterior-posterior (AP), lateral, and flexion and extension dynamic X-ray evaluation should always be performed prior to surgery in order to a) help localize the segment where the surgery is going to be performed, and b) assess instability p with functional X-ray, especially in cervical spine.

22.5 Specific Pathological Entities in the Spine which can Present as Dumbbell Tumors

22.5.1 Schwannomas and Neurofibromas

Approximately 90% of schwannomas are solitary and sporadic; 4% arise in patients with neurofibromatosis type 2, and another 5% are multiple but unassociated with neurofibromatosis type 2 [61]. The vast majority of spinal schwannomas develops from the dorsal roots and is intradural. Dumbbell schwannomas have been reported

as a separate group of spinal tumors, which characteristically involve both the intradural and the extradural compartments, occupying the intervertebral foramen, and more frequently located in the cervical spine [6, 62]. Giant schwannomas always have an extraspinal part with widening of the neuroforamen, which makes them dumbbell-shaped tumors. The surgical method should be tailored to each individual case (Figs. 22.2, 22.3, 22.4 and 22.5 demonstrate two cases of dumbbell schwannomas of the cervical spine operated on by the senior author of this chapter [KIA]. Details of the surgical resection of cervical dumbbell schwannoma are contained in the surgical video section of the online edition of this manuscript.) Spinal nerve sheath tumors showing an intradural location can be resected via a posterior standard or via a unilateral approach with modifications, such as partial facetectomy or a spinal cord rotation technique with resection of the dentate ligament [63]. Extradural dumbbell extension exposure of the tumor along the surgical plane of the dural or perineural boundary (particularly originating from the C1 or C2 level) is the key procedure required to accomplish radical and safe resection of the tumor with use of anterior access in selected cases [63].

Spinal extradural schwannomas are a distinct pathological entity of dumbbell schwannomas and can be distinguished from other nerve sheath tumors growing inside the spinal canal by their clinicoradiological features, including large tumor size with erosion of the vertebral body and an unlikely nerve root origin. Schwannomas tend to occur in thoracic and lumbar areas, and neurofibromas are usually associated with neurofibromatosis type 1 and tend to occur in the cervical area [45, 64]. In dumbbell neurofibroma cases, the majority of the nerve fibers are entrapped within tumoral tissue. It is frequently impossible to remove the tumor without sacrificing the nerve root; aggressive surgery may, therefore, result in severe neurological deficits [65, 66]. Neurofibromas were associated with higher rates of recurrence and lower rates of gross total resection than other tumor types, particularly in patients with neurofibromatosis types 1 or 2 [45].

22.5.2 Meningioma

Meningiomas compose up to 5% of spinal dumbbell tumors and are an important entity in differential diagnosis [6]. Dumbbell meningiomas probably originate from the arachnoid villi at the nerve root exits [4]. They are mostly located intradurally but occasionally exhibit an extravertebral extension with a dumbbell-shaped form [4, 67, 68]. The meningioma's extravertebral extension could include tumor growth through the intervertebral foramina, but usually only to a minor extent [69]. Because there is little intraspinal space for tumor growth, a meningioma at this location is prone to grow through the dura and, subsequently, to the extradural/extravertebral space [70]. The most common location seems to be the thoracic spine as there are several reports of thoracic dumbbell meningiomas resected with a combined posterior and thoracoscopic approach [11] or with additional thoracotomy [67]. Cases of dumbbell meningiomas in the cervical spine were also described [69, 71]. (Fig. 22.6 demonstrates a case of dumbbell meningioma of cervical spine operated by the senior author [KIA]).

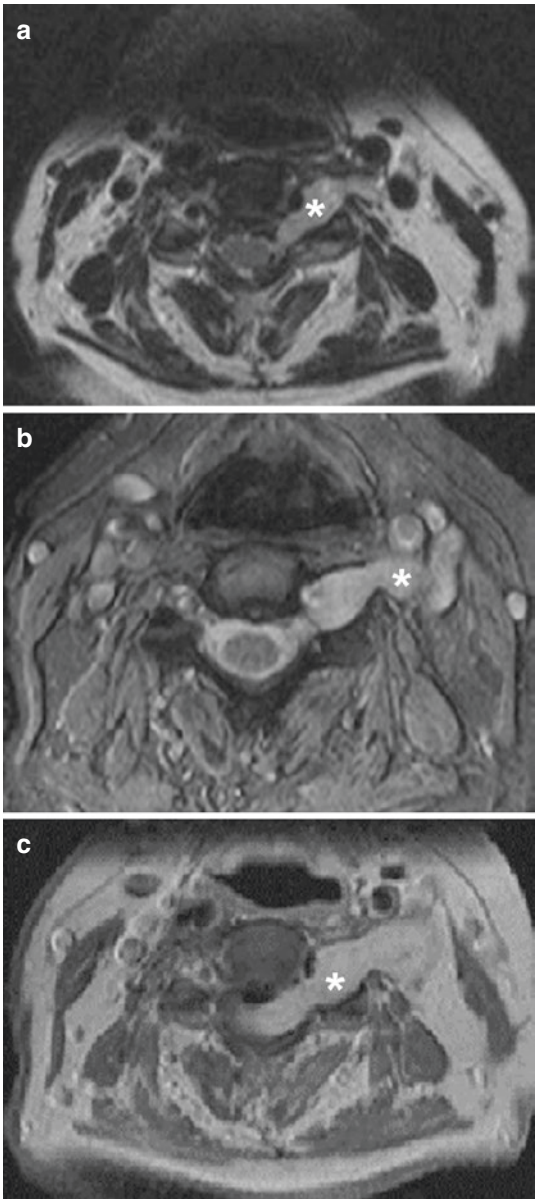


Fig. 22.2 Imaging of an 84-year-old female patient with left-sided radiculopathy and mild quadriparesis. She was diagnosed with a left-sided C3/4 dumbbell tumor (asterisk). The tumor was followed for 3 years where it showed progressive growth. (**a**) Initial T2-weighted axial MRI of the cervical spine showing homogeneously hyperintense dumbbell lesion in C3/4 foramen on the left side. (**b**) T2-weighted axial MRI of the cervical spine at 1-year follow up showing progressive growth of the extraspinal tumor component. Since the patient was clinically stable, surgery was not indicated. (**c**) Axial T1-postcontrast MRI of the cervical spine at 3-year follow-up showing dumbbell lesion with intense contrast enhancement and progressive intraspinal tumor growth with compression of the spinal cord and myelopathy. At this point, the patient had worsening quadriparesis

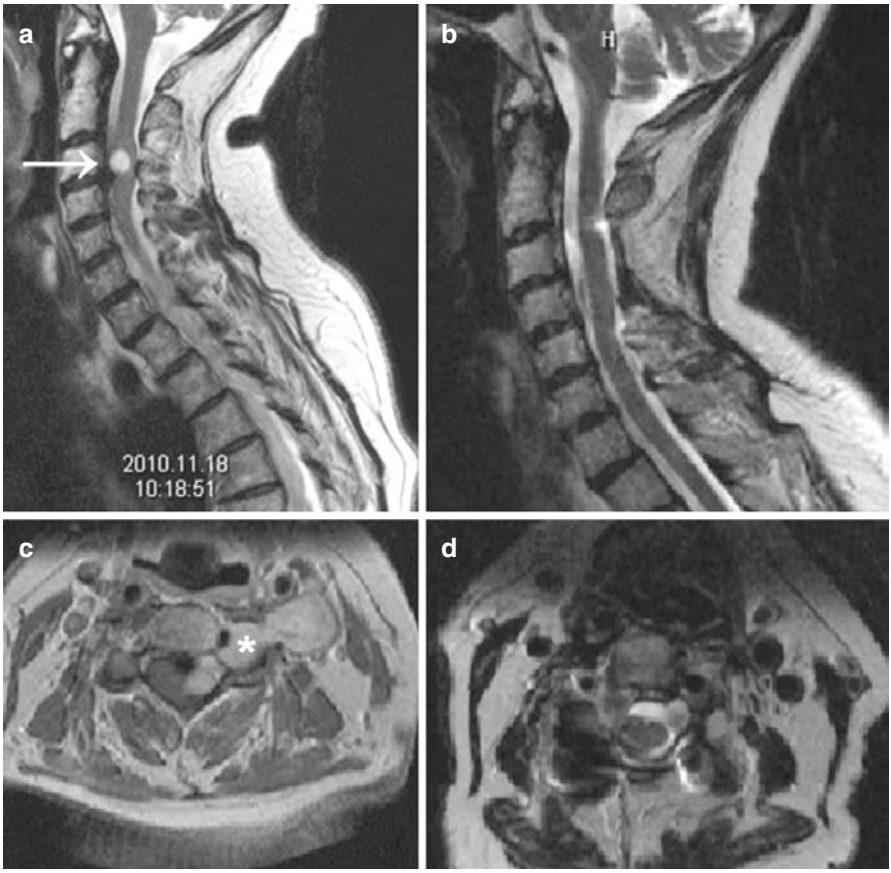


Fig. 22.3 Imaging of gross total resection of dumbbell lesion in left C3/4 neuroforamen via the posterior approach with laminectomy and unilateral facetectomy. The pathohistological diagnosis confirmed the diagnosis of schwannoma WHO I. Additional stabilization with lateral mass screws C2–5 was performed. (**a**) Preoperative sagittal T2-weighted MRI of the cervical spine showing an intradural homogenously hyperintense extramedullary tumor at C3/4 (arrow). (**b**) Postoperative sagittal T2-weighted MRI of the cervical spine showing complete resection of the tumor without any signs of myelopathy. (**c**) Preoperative axial T1-weighted post-contrast MRI of the C3/4 level showing the intraspinal and extraspinal expansion of the schwannoma with homogenous contrast enhancement (asterisk). (**d**) Postoperative axial T2-weighted MRI of the C3/4 level showing no sign of remaining tumor

22.5.3 Vascular Lesions

Capillary hemangiomas are benign vascular malformations most often found in the skin or soft tissue throughout the body of younger patients. They are histologically characterized by nodules of capillary-sized vessels lined with flattened endothelium, and often regress spontaneously. There have been more than 40 intradural

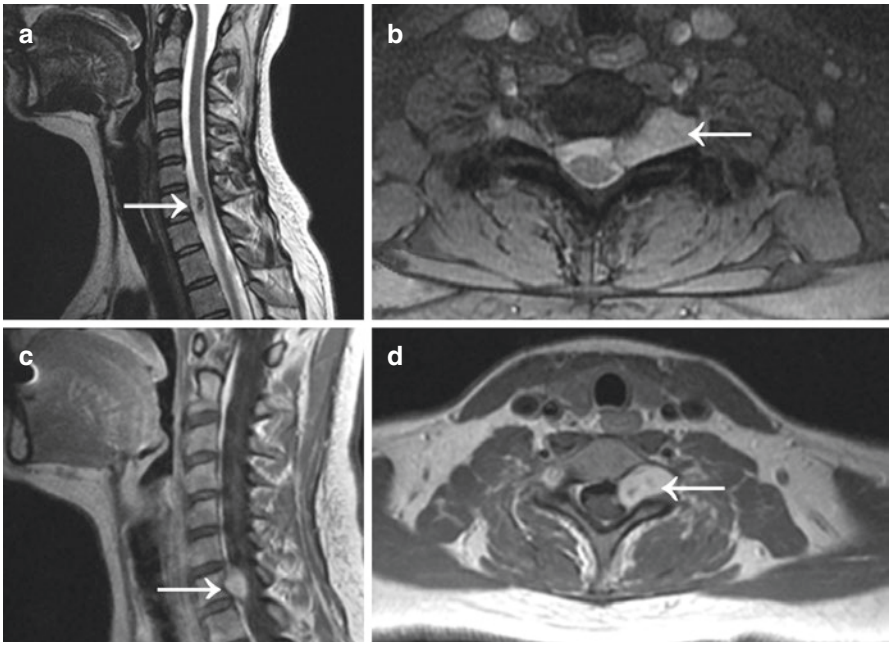


Fig. 22.4 Imaging of a 33-year-old patient with left arm numbness and weakness. (**a**) Preoperative sagittal and (**b**) axial T2-weighted MRI of the cervical spine showing hyperintense dumbbell tumor with (**c**) sagittal and (**d**) axial T1-weighted post-contrast MRI of the cervical spine demonstrating dumbbell tumor (arrow) with homogenous contrast enhancement at C7/T1 on the left side

spinal capillary hemangiomas described in the literature [72]. Extradural-only cases with spinal cord compression have also been described [10, 73], as well as cavernous hemangiomas with both intradural and extradural growth [9]. The majority of extradural cavernous hemangiomas are composed of an extension from a vertebral hemangioma into the spinal canal with extradural-only locations representing 1–2% of spinal hemangiomas [74]. These tumors commonly affect the vertebral bodies and extend into the epidural space, and are less commonly found within the thoracic cavity [75, 76]. Typical CT findings for hemangioma are lobulation, heterogeneous enhancement with contrast media, multiple ring-like calcifications, and an intact intervertebral foramen when the tumor extends to the spinal canal; however, these findings are not always observed [77]. On MRI, both neurogenic tumors and hemangiomas are isointense-to-hypointense on T1 images, and homogeneously hyperintense in T2 images [53]. They are usually presented as a progressive myelopathy, so early treatment may prevent permanent neurological deficits. Endovascular embolization has been recently used to remove a hemangioma, successfully minimizing blood loss during the operation. However, this management carries a risk of spinal infarction [78].

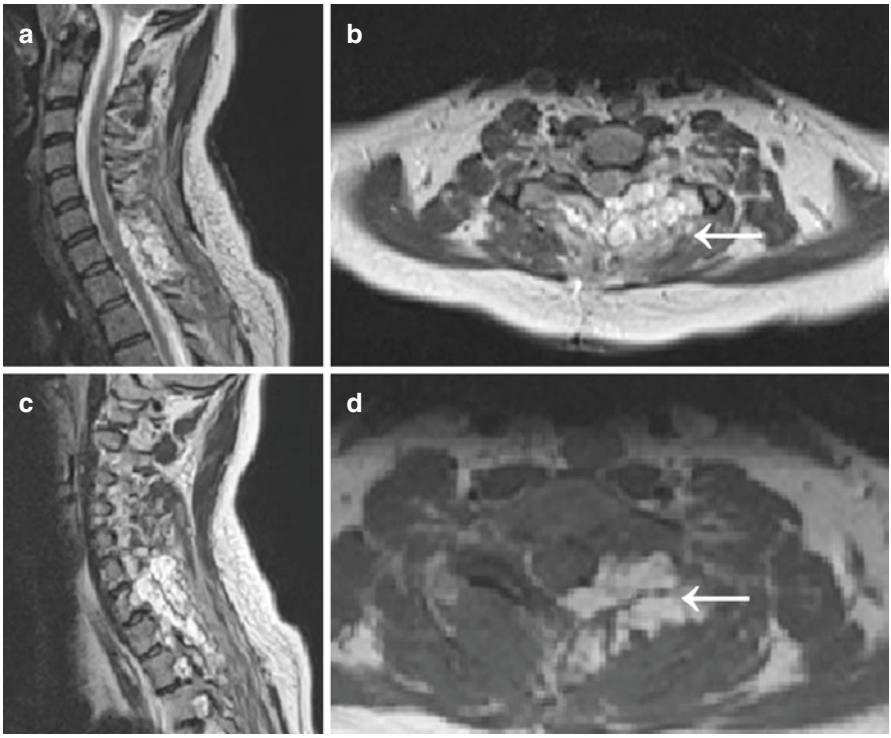


Fig. 22.5 Imaging of tumor gross total resection via posterior approach with unilateral facetectomy. The pathohistological diagnosis was schwannoma WHO I. (**a**) Postoperative sagittal and (**b**) axial T2-weighted MRI of the cervical spine with (**c**) sagittal and (**d**) axial T1-weighted post-contrast MRI of the cervical spine demonstrating complete resection of the tumor. Additional stabilization was unnecessary. Arrow shows abdominally harvested fat graft applied to dural suture in order to prevent cerebrospinal fluid leak

Gross total resection is the preferred treatment for these lesions. In the thoracic spine there are several approaches: a posterior approach, a posterior approach with thoracotomy, a thoracoscopic approach, and a combined posterior approach with thoracoscopy. Some authors suggest performing thoracotomy before posterior surgery because it allows for ligation of the involved arteries and prevents bleeding in the spinal canal [79]. Management of intraoperative bleeding and sufficient hemostasis are the pitfalls of successful surgery of these lesions. Details of the surgical resection of dumbbell hemangiomas are described in the surgical video section of the online edition of this manuscript. (Figs. 22.7 and 22.8. demonstrate the case of dumbbell hemangioma of the thoracic spine depicted in our surgical video in which the patient underwent a 2-stage procedure of intraspinal resection by the senior author (KIA) followed by thoracoscopic resection of the intrathoracic part of the tumor.

Hemangioblastomas located extradurally account for 8–12% of all spinal hemangioblastomas. They are more prevalent in patients with Von-Hippel Lindau disease [49]. There are several case reports describing these lesions. It is

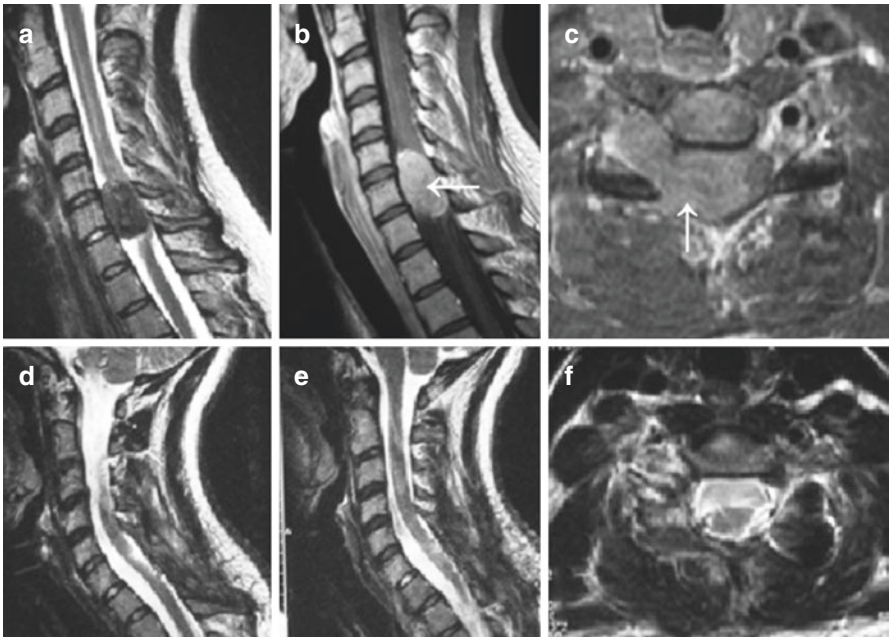


Fig. 22.6 Imaging of a 24-year-old female with quadriparesis and T1 sensory level, inability to walk, and intermittent urinary incontinence. (**a**) Preoperative T2-weighted sagittal MRI of the cervical spine demonstrating intradural extramedullary tumor with (**b**) homogenous contrast enhancement in sagittal T1-weighted post-contrast series and (**c**). axial T1-weighted post-contrast MRI of the cervical spine showing the dumbbell form of the tumor (arrows). The tumor underwent gross total resection via the posterior approach with laminectomy at C6/7 with unilateral facetectomy. The pathohistological diagnosis was meningioma. Additional stabilization of the C6/7 segment was performed. Postoperative (**d**, **e**) sagittal and (**f**) axial T2-weighted MRI of the cervical spine showing complete resection of meningioma. The patient fully recovered

recommended that resection of hemangioblastomas arising in the cervical spine be done via the lateral approach, which provides control of the vascular feeders [16]. Hemangioblastomas can also manifest subarachnoid hemorrhages in the cauda equine [49]. To avoid excessive blood loss, preoperative embolization and angiography to confirm the location of main feeder should be considered when hemangioblastoma is suspected. To prevent massive hemorrhage during surgery, early identification of feeders and blood flow direction with extracapsular resection needs to be carried out [49].

22.5.4 Paravertebral Neurogenic Tumors with Intraspinal Extension

Because the paravertebral space is rich in neurogenic tissue, primary neurogenic tumors originating here are the most common types of tumors, representing 90% of primary tumors of the posterior mediastinum, 63% of the retroperitoneal space, and

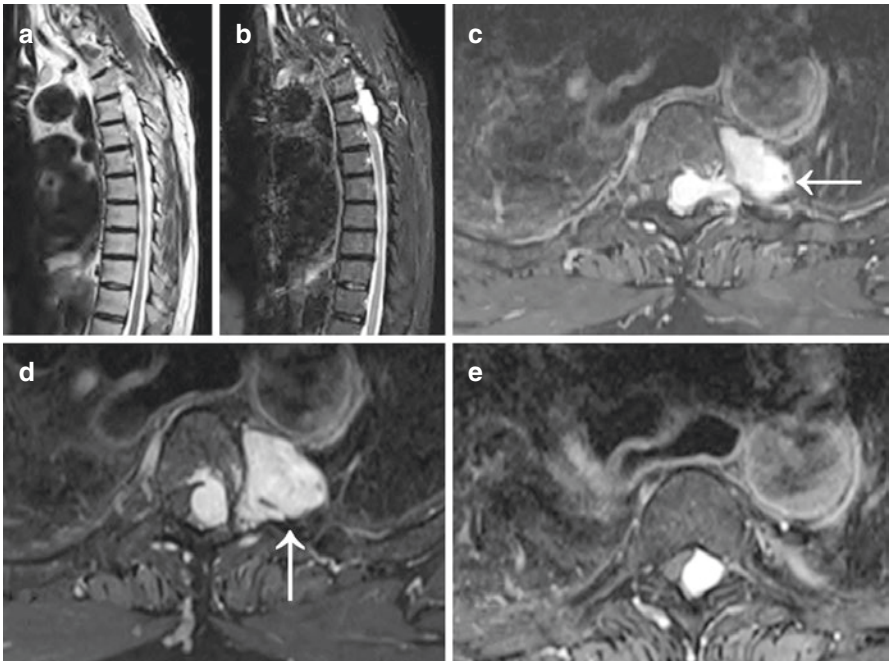
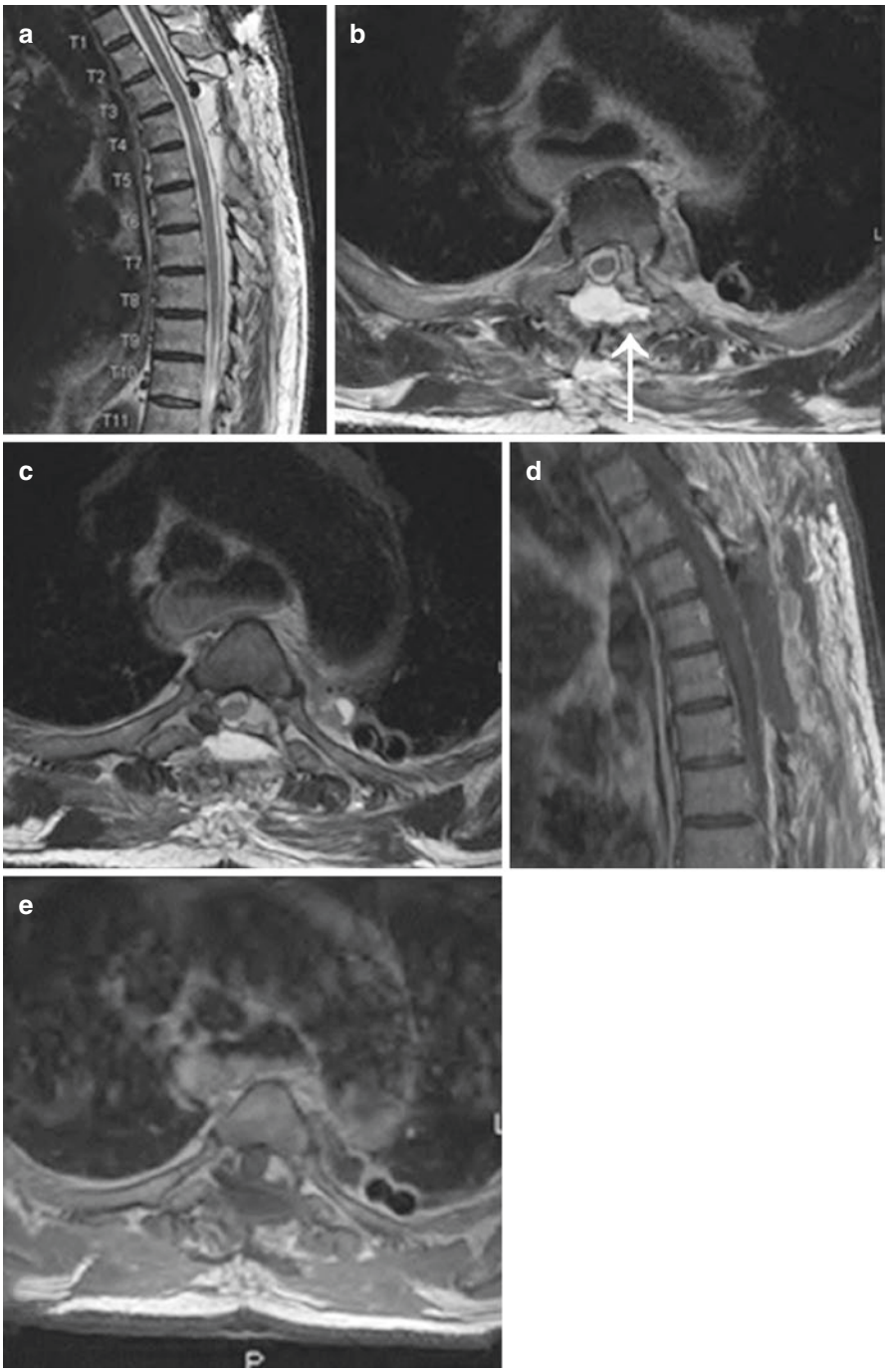


Fig. 22.7 Imaging of a 78-year-old patient with progressive paraparesis and ataxia of the thoracic spine. (**a**) T2 weighted sagittal sequence showing the hyperintense intraspinal tumor T3–5; (**b**) postcontrast T1-weighted sagittal sequences showing homogenous enhancement of the tumor; (**c, d, e**) postcontrast T1-weighted axial sequences revealing dumbbell-shaped tumor extending intraspinally with displacement of the spinal cord to the right and growth through the intervertebral foramen of T4 into the left middle thorax. Note the lobular contour of the mass, which can be a clue to the diagnosis of hemangioma (arrows)

Fig. 22.8 Further imaging of the 78-year-old patient undergoing a single 2-stage surgery, which included posterior microsurgical and transthoracic endoscopic resection of the tumor. For the posterior approach, a partial hemilaminectomy and facetectomy with partial costotransversectomy was performed, which revealed a highly vascular epidural tumor with spinal cord compression extending into the neuroforamen. Gelfoam powder (Pfizer, New York, NY) and extensive coagulation were used for hemostasis of this vascular tumor. The intraspinal epidural part of the tumor was completely resected with foraminotomy. A fat pad (arrow) was placed in the foramen as a marker for the extent of the thoracoscopic procedure. Second stage surgery included thoracoscopic resection of the extraforaminal part of the tumor. The fat pad was visualized endoscopically in the neuroforamen at the end of the resection providing an important orientation. Histopathological evaluation revealed capillary hemangioma. Postoperative MRI of the thoracic spine: (**a**) T2-weighted sagittal sequence showing the complete resection of the tumor; (**b, c**) T2-weighted axial sequences; (**d**) postcontrast T1-weighted axial sequence showing no signs of contrast enhancement; and (**e**) postcontrast T1-weighted axial sequence



10% of the neck [80]. The most common pathological type for patients age 2 year or younger is neuroblastoma of the suprarenal gland or paraganglionic retroperitoneal sympathetic tissue, while the most common type for adults is schwannoma in the retroperitoneal space or posterior mediastinum [8, 81]. Neurogenic tumors, especially neuroblastoma and extradural schwannoma [82] in the paravertebral gutter, have a predilection to spread through the intervertebral foramen to inside the spinal canal (i.e., forming a dumbbell-shaped tumor), which causes myelopathy due to either direct or indirect compression on the spinal cord [83, 84]. **Ganglioneuroma** is a rare, differentiated, benign, and slow-growing tumor that commonly arises from sympathetic ganglion cells. In rare cases, they can grow through the intervertebral foramina and present with a dumbbell shape. Most of these tumors are retroperitoneal and are more common in children and young adults [85]. On CT, these tumors show punctuate calcifications in pre-sacral region and sometimes present with extensive osteolytic bone destruction of the sacrum [86].

22.6 Malignant Spinal Dumbbell Tumors

22.6.1 Metastasis

A differential diagnosis of leptomeningeal metastasis includes ruling-out a wide range of malignant and benign conditions, such as congenital and degenerative lesions, infectious and autoimmune diseases, and neurinoma [8]. The radiologic distinction between metastases and neurinomas is based primarily on definite neuroimaging features, particularly the number of lesions, size and growth pattern [87]. Whereas metastases are often encountered as multiple small nodules at lower spinal structures (e.g., the cauda equina)—presumably—due to gravity, neurinomas appear as single lesions in the neuroforamen and might present at any height [8]. The clinical presentation of leptomeningeal metastases depends on the location and growth-pattern often resulting in general symptoms, such as nausea and headaches due to interruption of the cerebrospinal fluid (CSF) flow and, later signs of myelopathy due to compression of the spinal cord [87]. In patients with breast cancer, overall survival with current treatments remains limited to less than 6 months on average [88]. Surgical treatment combined with adjuvant or neoadjuvant therapy can improve neurologic function and lessen pain [28].

22.6.2 Malignant Peripheral Nerve Sheath Tumors (MPNSTs)

MPNSTs account for 3–10% of all soft tissue sarcomas, and are commonly located in the trunk, limbs, head, and neck, although there are some rare spinal cases [89]. MPNSTs have high metastatic potential and surgical resection is the preferred treatment of choice, if the tumor is resectable; however, there is no effective systemic therapy currently available. Surgical treatment of these lesions is defined by Enneking criteria as either Enneking appropriate (i.e., en bloc resection with wide

or marginal margins) or Enneking inappropriate (i.e., piecemeal or an intralesional resection) [90, 91], although a multicenter study showed similar rates of recurrence and survival for the two groups [90]. Prognosis of unresectable or metastatic MPNSTs is extremely poor, particularly in the spinal region, where the associated mortality rates are as high as 80%; larger lesions are also more likely to be related to higher malignancy [12, 92, 93]. Adjuvant photon beam therapy showed better local control, but carbon ion radiotherapy leads to better local control and increases in overall survival and progression-free survival [94].

Spinal extraosseous chordoma (SEC) is usually located in the cervical and epidural region and is extremely rare. SECs are less aggressive, have a lower rate of recurrence and metastasis, and have a better prognosis than those of the osseous origin [21].

Mesenchymal chondrosarcoma is a rare malignant tumor arising from bone or soft tissues. Calcification can be seen in tumors, which may influence or reflect the growth of tumor and disease progression. Gross total resection should be followed with adjuvant radiotherapy and close follow-ups due to the possibility of recurrence [22].

Myxoid liposarcoma (MLS) is a soft tissue sarcoma usually located in extremities. One-third of patients develop distant metastases and there are several reports that present as dumbbell-shaped spinal lesions [17, 95]. Treatment consists of surgical resection followed by adjuvant chemotherapy [17].

22.7 Surgical Technique

The best surgical approach for these tumors is dictated by the location and size of the tumor. Tumors located entirely or partially within the spinal canal can be accessed through the midline-posterior approach. In a large series reported in the literature, up to 80% of dumbbell tumors—predominantly schwannomas—were resected using only a posterior approach [6]. When the posterior approach is used, every effort should be made to maximize the excision of the paraspinal tumor through the same approach [46]. In cases in which an anterior approach is employed, the operation should be performed with surgeons who specialize in the region-specific approaches, such as head and neck, thoracic, and abdominal surgeons.

In the **cervical spine**, the posterior-midline approach is the standard common approach for intraspinal lesions [3]. McCormick (1996) has described a posterior-midline approach with partial laminectomy and complete unilateral facetectomy [65]. Further possibilities include a combined posterior and anterior approach [40], an anterior approach with corpectomy [38], a lateral approach with oblique corpectomy [96], an extensive posterolateral approach involving total lateral mass resection and laminectomy [97], and an anterolateral-transuncodiscal approach [68]. The location of the vertebral artery (VA) is important during resection of cervical dumbbell tumors [98]. When the VA is encased by the tumor, a posterior approach carries high risks [34]; the anterior approach may be more adequate in these cases.

McCormick [4] has also described a modified version of the lateral-extracavitary approach for removal of dumbbell and paraspinal tumors of both the thoracic and lumbar spine. For the **thoracic spine,** there are several surgical techniques available including the following: a combined posterior microsurgical approach for resection of the intraspinal part followed by subsequent video-assisted thoracoscopic surgery for intrathoracic part of the tumor [99, 100], a posterior-only approach [101], a single posterolateral approach [102], a posterior approach combined with thoracotomy [67, 79, 81, 103], a 1-step removal via posterolateral thoracotomy and extended foraminectomy [104], a transclavicular approach for tumors of the cervicothoracic junction [105], an extended lateral cavitary approach [4], and a thoracoscopic-only approach [106].

Posterior approaches for resection of dumbbell tumors of the **lumbar spine** include a posterolateral-transforaminal approach [57], a transparaspinal approach [107], a posterior approach with laminoplasty [108], a posterior approach with hemilaminectomy, facetectomy and stabilization [6], a posterior dual approach [109], and a mini-open [44] minimally invasive technique with [110] or without stabilization [111] (i.e., using tubular retractor). Giant lumbar schwannomas eroding the vertebral body and expanding into the retroperitoneal space requires a 2-stage surgery—a decompression and partial resection with root transection posteriorly, followed by an en bloc resection through the retroperitoneal approach by vascular surgeon [112].

Gross total resection should be performed whenever possible as long as there is no risk of vascular or neurologic injury since the risk of recurrence rises with subtotal resection [45, 113]. Subtotal resection is usually performed in cases when risk of vascular injury or neurological deficits is high as suggested by intraoperative nerve monitoring. Preservation of the involved nerve root should always be attempted when removing neurogenic dumbbell tumors [41]. In a large series in the literature, complete resection has been achieved in 86 to 100% of patients [4, 5, 8, 96, 98].

Spinal reconstruction surgery using instrumentation should be considered when the spinal column is weakened structurally during tumor removal when the tumor invades at multiple levels, such as with malignant neoplasms, neurofibromas, extradural-intravertebral tumors, and multidirectionally eroding tumors [5]. The cervical spine seems to be more prone to the development of the postoperative instability [5, 41]. Facetectomy with costotransversectomy in the thoracic spine may require additional stabilization. If bilateral laminectomy on more than two levels is performed or more than half of the lumbar vertebral body is compromised by the tumor, instrumentation and reconstruction may be considered [34].

22.8 Complications

Dumbbell lesions are associated with higher rates of CSF leakage, pseudomeningocoele, and wound infection compared with non-dumbbell spinal nerve sheath tumors [114]. Complications include surgery site infections [57], CSF leak, injury

to the surrounding structures in the neck, thorax and abdomen when using a combined approach (i.e., injury to the carotid artery, esophagus, recurrent laryngeal nerves, chylothorax, pneumothorax, colon perforation, ureter injury, retroperitoneal hematoma, injury of the aorta and iliac arteries), spinal cord edema, spinal cord ischemia due to injury of Adamkiewicz artery [115], pseudomeningocoele [116], and extensive bleeding in the case of vascular lesions. Complication rates increase as the size of the total excised tumor increases [57].

One of the important questions in surgery of dumbbell spinal tumors is how to deal with dural defect in intradural-extradural tumors (Eden Type I and II). Suturing autologous fascia over the dural defect, and then applying of several layers of dural graft and fibrin glue is one possible method [98]. A "separate-dural-incision method" is another type of dural incision and closure procedure for preventing postoperative CSF leakage during the surgical removal of dumbbell-shaped spinal tumors [117]. Adequate visualization of the intradural and extradural components of the tumor is achieved with the use of separate dural incisions. First, the dura mater is opened along the dural theca to provide adequate visualization of the intradural portion of the mass; then, a second incision is made along the nerve root to remove the extradural component. Meticulous suturing is essential in intradural lesion cases; however, the dura mater is usually thin and fragile in such cases. During suturing with a needle and thread, the dura mater can become lacerated proximal to the needle holes and result in CSF leakage. Instead of using a needle and thread for this technique, non-penetrating vascular clips were used to close the dural incisions [117]. A third technique that showed good results in prevention of the CSF leak in spinal surgery is the application of a previously harvested abdominal fat graft onto the dural suture [118]. After tumor resection, it may be very difficult or impossible to achieve watertight dural closure. Application of the fat graft application, along with fibrin glue, incorporated in dural closure may eliminate the risk of CSF leak.

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