Adult Presentations/Outcomes of Occult Spinal Dysraphism

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Introduction

Occult spinal dysraphism (OSD) is defined as a group of congenital clinicopathological entities in the spinal cord. These pathological changes are underpinned by similar embryological causes and have multiple or single common forms of presentation. OSD generally appears in children, the most common OSD form in adults being tethered cord (TC) [1]. TC syndrome (TCS) is a neurological disorder caused by tissue attachments to the spinal cord at different levels. These attachments cause abnormal stretching of the spinal cord, limiting its movement. In some of these cases of congenital deterioration, a spinal cord with the conus in normal position can still be tethered by a thick filum or split cord. This limits its growth. As a result, patients can experience a wide spectrum of clinical presentations from asymptomatic to paraplegic [1–3]. TCS was described in 1976 by Hoffman et al. [4], who observed that the spinal cord was tethered via a thickened terminal filum to the sacral bones in 31 children and a noticeable neurological improvement followed release of the cord.

OSD syndromes are not rare pathological disorders, particularly in developing countries. The development of imaging technologies has increased the number of adults in whom OSDs such as congenital TCS are diagnosed. The incidence of OSD is not known. However, the incidence of neural tube defects is estimated at 0.17–6.39 per 1000 live births worldwide [5]. Occult spinal dysraphic lesions have been reduced in developed countries owing to folic acid supplementation during pregnancy and to prenatal diagnosis of dysraphic malformations, which often lead to termination of the pregnancy.

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Although the management of congenital OSD/TCS diagnosed in adulthood remains controversial, the results of recent clinical studies of surgical intervention in adults are encouraging [1, 6–11]. Herein, the presentation and surgical treatment outcomes of OSD diagnosed in adults are discussed.

Tethered Cord Syndrome in Adults

TCS is one of the most progressive forms of neurological deterioration resulting from spinal cord tethering by various dysraphic spinal abnormalities. Generally, the tethered cord entity is accompanied by one or more of OSD forms such as split cord malformations (bony septa or fibrous bands), intradural dermoid masses such as hamartomas or lipomas, tight or thick filum terminale, neurenteric cysts, meningocele manqué, scoliosis, syringohydromyelia, lipomyelomeningocele, dermal sinus tract, and diastematomyelia. These entities have a propensity for multiple expressions within the same patient (Case Study 1, Fig. 7.1) [1]. These malformations can be accompanied by malformations of other organs. Over our 12 years of experience,
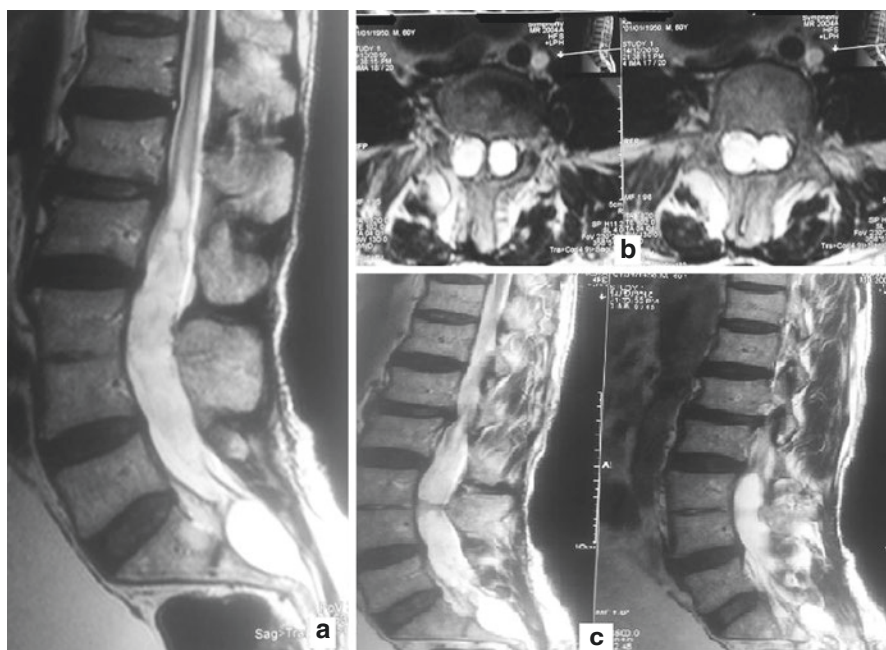

Fig. 7.1 A 61-year-old male patient presented with numbness and low back and leg pain: (a) T2-weighted sagittal MRI demonstrating cyst extends between L1 and S2. Note that L3 and L4 vertebral fusion anomaly (block vertebra) is apparent; (b) T2-weighted axial MRI showing that diastematomyelia extends between L2 and L4. Note split cord malformation as bony septum at L3 level and dermal sinus tract are apparent; (c) T2-weighted axial MRI demonstrating that filum terminale terminates at S1–S2 level (Case Study 1)

Table 7.1 Co-malformations detected in 31 adult TCS patients

Malformation	No. of patients	Percentage
Filum terminale (tethered cord)	31	100%
At L2 level	1	3.2%
At L3 level	4	12.9%
At L4 level	4	12.9%
At L5 level	10	32.3%
At S1 level	9	29%
At S2 level	3	9.7%
Diastematomyelia	13	41.9%
Vertebral fusion anomalies	13	41.9%
Split cord malformation	10	32.3%
As bony septum	6	19.4%
As fibrous bands	4	12.9%
Lipoma	8	25.8%
Scoliosis	8	25.8%
Syringohydromyelia	6	19.4%
Dermal sinus tract	6	19.4%
Foot anomalies (equinovarus, pes planus)	6	19.4%
Hypertrichosis	5	16.1%
Lipomeningocele	4	12.9%
Kidney malformations (hypoplastic, horseshoe)	4	12.9%
Skin hyperpigmentation	3	9.7%
Lipomyelomeningocele	2	6.5%
Chiari malformation	2	6.5%
Arachnoidal cyst	2	6.5%
Hydrocephalus	2	6.5%
Mitral valve prolapse	1	3.2%
Only tethered cord without co-malformation	3	9.7%

6 out of 31 operated adult TCS patients (25 patients were previously reported in 2 papers [1, 10]) had foot anomalies, and four had kidney malformations (Table 7.1).

The structure of the filum terminale is especially important in the development of TCS. Recently, Tehli et al. compared histopathological analyses of filum terminale between TCS patients and normal human fetuses. They observed adipose tissue, fibrosis, hyalinization, and meningothelial proliferation in filum terminale samples from TCS patients, but none of these in normal fetal samples. Elastic fibers were present in all TCS specimens and the adult cadaver, but not in the fetuses. Peripheral nerves, ganglion cells, and ependymal cells were observed in the normal human fetal filum terminale samples. Because these changes were not observed in fetuses, the authors suggested that this syndrome did not begin during intrauterine life [7, 10, 12]. According to this work, TC could be an acquired rather than a congenital syndrome.

Most adult patients experience TC during normal daily activity or mild static neurological deficits in childhood. The development of imaging technologies has increased the number of adults in whom congenital TCS is diagnosed. However,

unlike the pediatric population, adults may deny any significant symptoms and be unwilling to visit their doctors, so TCS, which is the most common form of OSD in adults, is usually diagnosed after progressive scoliosis, foot deformities, leg length discrepancy, or muscular atrophy, potentially increasing the chances of disability. In some cases, serious traumas such as traffic accidents or falling, as well as dynamic and static changes that occur in the spinal column (such as those during pregnancy), can aggravate the degenerative processes and induce back and leg pains.

Case Study 1 *A 61-year-old male was referred with low back and right leg pains. He had fallen 6 years earlier. His neurological examination was intact, except for weakness in his right leg distal muscles group of 4/5, right L2–L5 and S1 dermatomal hypoesthesia, neurogenic claudication, and the Babinski sign bilaterally evaluated as no response. MRI showed diastematomyelia at L2–L4, block vertebra at L3–L4, split cord malformation at L3 level as bony septum, and dermal sinus tract. A giant cyst extended between L1 and S2, while the filum terminale terminated at S1–S2 level* (Fig. 7.1). *Urodynamic study showed normocompliance normotonic bladder function. The patient underwent bony septum resection and duraplasty using total L3–L4 laminoplasty, followed by cystectomy using total S1 laminoplasty. Histopathological examinations revealed that the cyst was arachnoidal. The patient recovered quite well and was discharged after 3 days without complications. He was doing well on his postoperative 62nd-month doctor visit.*

Pathophysiology of TCS

TC coexists in open and occult forms of spinal dysraphisms [13]. The normal spinal cord is free in the spinal canal except for denticulate ligaments and nerve roots. As a result of TC, the spinal cord is tightly fixed. Therefore, there is no normal movement of the spinal cord when under pressure. During embryogenesis, the nascent spinal cord fills the spinal canal. Throughout the development of the fetus, the vertebral column grows faster than the spinal cord. Thus, the distal end of the spinal cord is located at the level of the first or second lumbar vertebral body in adults, but it can be located at the level of the third lumbar vertebral body in children. If an abnormality affects the nature of the spinal cord, such as split cord malformation, myelomeningocele, tight or thick filum terminale, diastematomyelia, or lipoma, the spinal cord is tethered [1, 14, 15]. This results in stretching of the spinal cord and causes neurological injury even in the fetus. At the time of birth, the spinal cord is normally located between the first and second lumbar vertebral bodies. After birth, continuing growth stretches the TC further; this injures the spinal cord both by directly stretching it and by interfering with its blood supply and oxidative metabolism [16]. If neurological findings are already presented, then further clinical deterioration can be anticipated. Adults with TC can show deterioration due to daily repetitive stretching and cumulative microtraumas during extension and flexion of the TC. A sudden flexion movement of the spine can also produce a symptomatic

onset of TCS [1, 10, 16]. Irreversible neuronal injury can result from sudden stretching of the already chronically stretched TC [16]. Yamada et al. demonstrated changes in spinal cord blood flow and oxidative metabolism following tethering of the spinal cord in both experimental animals and humans [16]. Usually a TC results in a low conus position. However, many cases of TCS have been reported with the conus at a normal level [1, 10, 13].

Sex and Age

The female preponderance in adult patients with TCS is well established in all published reports [1, 6, 9, 10, 13, 17, 18]. However, some authors have reported whole series of males, such as Akay et al., who reported nine young males with SCMs [19]. In agreement with previous publications, there was a female predominance of 18:13 (58%) in our patients. Klekamp reported in his series that 71.8% of his patients who underwent untethering were female [6]. Almost all congenital TCS is seen in children especially during the first year of life. The mean age of adult TCS patients in our experience is 32.1 ± 11.8 (18–68) years.

Clinical Presentation

Most adult patients with a TC have practiced their normal daily activities without complaint since childhood despite this congenital disorder. However, during adulthood, they become more aware of significant symptoms than the pediatric population. In some cases the symptoms of the TCS appear after a traffic accident or fall. Furthermore, physiological dynamic or static changes in the spinal column, such as those that happen during pregnancy, can aggravate the degenerative processes and induce back and leg pains.

Neurological deficits developed in adult TC patients are generally irreversible. Stretching of the conus medullaris and nerve roots can induce back pain, leg weakness, foot deformity, scoliosis, sensory loss, and/or bowel or bladder dysfunction [2, 3, 10, 13, 20]. Untreated TCS can progress in 27.5%, 40%, and 60% of cases at 1, 2, and 5 years after diagnosis, respectively [3]. In our series, the most common presenting symptom was pain, which was seen in 27 patients (87.1%); 20 patients experienced low back pain (64.5%), and 18 experienced leg pains (58.1%). The pain was followed by bladder dysfunction in 16 patients (51.6%) and muscular weakness in 16 (Table 7.2).

In TCS, stretching of the conus medullaris and nerve roots can induce back pain, leg weakness, foot deformity, scoliosis, sensory loss, and/or bowel or bladder dysfunction [2, 3, 13, 21]. In our experience during the last 12 years, 116 adult patients were diagnosed with TC after permanent or severe pain in their back and/or legs (incidentally) or in those experiencing serious urological complaints. Only 31 adult patients (26.7%) diagnosed as symptomatic TC were treated surgically after all investigations and full evaluation of their clinical status. Patients with tolerable

Table 7.2 The presenting symptoms of 31 adult TCS patients

Presenting symptoms	No. of patients	Percentage
Low back pain	20	64.5%
Leg pain	18	58.1%
Bladder dysfunction (incontinence or retention)	16	51.6%
Muscular weakness	16	51.6%
Numbness	8	25.8%
Unsteady gait	6	19.4%
Bowel dysfunction (fecal incontinence)	5	16.1%
Muscular atrophy	4	12.9%
Lumbar swelling	3	9.7%
Frequent lumbar skin infection	2	6.5%
Headache	1	3.2%
Vertigo	1	3.2%

Table 7.3 The clinical findings in neurological examinations of 31 adult TCS patients

Clinical findings	No. of patients	Percentage
Bladder dysfunction (incontinence or retention)	16	51.6%
Muscular weakness (motor deficit)	16	51.6%
Hypoesthesia	11	35.5%
Increased deep tendon reflex	8	25.8%
Extensor or indifferent Babinski	6/4	19.4%/12.9%
Loss of anal tone and sensation (fecal incontinence)	5	16.1%
Muscular atrophy	5	16.1%
Lumbar local tenderness	4	12.9%
Positive clonus	3	9.7%
Positive Hoffman	2	6.5%
Normal clinical findings	6	19.4%

pain, as seen in all of the adult patients with TCS in our series, were treated conservatively, with close follow-up (3-month periods) to preclude permanent new neurological deficits.

The patients have to undergo full neurological examination, especially those who experience low back and leg pains associated with urinary complaints. The most common clinical findings in our series were bladder dysfunction and muscular weakness, each seen in 16 patients (51.6%). Pathological (abnormal) responses to all of deep tendon reflexes, Babinski reflex, clonus, and Hoffman sign, noticed in 18 patients of our series (58.1%), are important indicators for checking the craniospinal MRI (Table 7.3). Because large numbers of new neurological deficits are expected in their age group, the surgical intervention rate was 100% among all children and adolescents with TCS referred to us (68 children and adolescents were diagnosed with TCS). Klekamp reported that 43 of his 85 (50.8%) adult patients with TCS underwent an untethering procedure [6].

Some authors suggest that the reversible symptoms associated with TCS are related to metabolic derangements and alterations in oxidative metabolism [16]. Not all neuronal injuries in TCS patients will necessarily be repaired after surgical intervention; the persistence of symptoms can vary depending on the severity of the

condition and the interval between presentation and initial treatment [1, 10]. If treated early, patients can recover from their neurological deficits. Chronic or excessive stretching of the spinal cord can lead to permanent disability. Although the period between initial symptoms and time of surgery ranged between 4 months and 25 years in our series, with an average of 5.2 years, early surgical intervention after increasing intensity of pain or severity of complaints gave good results; 64.5% ($n = 20$) of our patients recovered well.

Associated Malformations

Three of our 31 operated adult TCS patients were free of other malformations associated with TC (Case Study 2, Fig. 7.2). Therefore, malformations in patients with classic symptoms of TCS should alert physicians to refer such patients to neurosurgeons. Split cord malformations (SCMs) are mainly associated with TCS in adults [2]. Two types of SCM are distinguished on the basis of the state of the dural tube and the nature of the median septum (Table 7.1).

Some SCM cases show signs of TC but no spur is found in the malformations. This happens in type II SCM. The cleft is generally partial or incompletely split. Spina bifida is often present with the possible formation of hydromyelia and a thin fibrous band (FB) [15]. The management of TCS can change according to the associated SCM type [10]. For type I SCM, removal of the FB or bony septum (BS) can suffice (Case Study 1). If not, untethering is necessary. However, for type II SCM, removal of the FB should be followed directly by untethering.
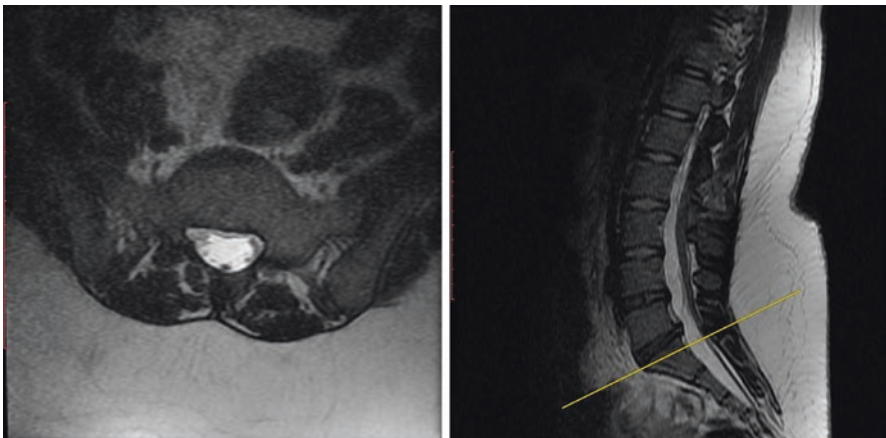

Fig. 7.2 A 21-year-old female presented with low back and leg pains for more than 5 years. Her neurological examination was intact, except rare loss of control of urination. T2-weighted MRI demonstrated that terminal filum terminates at L5 level. Right side: sagittal MRI demonstrated the tethered cord level; *yellow line* shows the cross-section at S1 level that axial MRI shows on the left side. Left side: axial MRI demonstrating tethered cord level (Case Study 2)

Case Study 2 *A 21-year-old female was referred to us with pain in her low back and both legs for more than 5 years. During the last month before the referral, she developed urinary incontinence. Her neurological examination was intact except for an infrequent loss of control of urination. MRI showed that the filum terminale terminated at L5 level* (Fig. 7.2). *The patient underwent untethering using L5 laminoplasty. She recovered well and was discharged after 2 days without complications. The patient was doing well on her postoperative 24th-month doctor visit.*

Diagnosis of Adults with TCS

The diagnosis of OSD or TCS in adult patients is not well established until back and/or leg pain manifests. Several pathological conditions including fatty and thickened filum terminale, lipomyelomeningocele, meningocele, split cord malformation, diastematomyelia, and post-repair myelomeningocele can be responsible for TCS [1, 21]. It is well established that early surgical intervention of congenital TCS in children, whether symptomatic or otherwise, prevents additional neurosurgical deficits [2, 13, 22]. However, despite early diagnosis of these conditions, there remains an ongoing debate as to whether asymptomatic adult patients with TCS should undergo prophylactic surgery [1, 13]. According to Rajpal et al., the risk of new neurological deterioration in patients with congenital TCS increases with age. Therefore, Rajpal et al. suggest surgery for adult patients with TCS to protect them from developing new neurological deficits that could be permanent [9].

Although low back pain is the cardinal symptom for TCS, there are other presenting symptoms such as leg pain, anorectal and perineal pain, fatigue, recurrent bladder infections, progressive muscular weakness of the lower extremities, unsteady gait, patchy sensory loss, sacral sensory loss, progressive deformities (foot size discrepancies, foot and leg deformities, or scoliosis), bladder and bowel dysfunction (incontinence/retention), and sexual dysfunction (Table 7.2).

Midline cutaneous abnormalities are possible markers for ODS/TCS. Most patients with a TC have a discoloration or a lesion of some type on their skin in the midline [1, 10]. These skin markers are mostly located in the lumbosacral area. Therefore, full physical examination of suspected TC patients is recommended. Hairy patches/dimples (hypertrichosis), subcutaneous lipomas, myelomeningocele sac over the back, cutaneous hemangioma, port-wine stain, pigmentary nevus, and dermal sinus are the most common midline cutaneous markers associated with OSDs/TCS. After physical examination, careful neurological examination is required. The neurological findings in our 31 adult TCS patients are given in Table 7.3.

As with all OSD, TC in child or adult patients is determined by MRI, which shows a low level of the conus medullaris and other pathological findings such as thickened filum terminale, split cord malformation, or diastematomyelia that can be associated with TC (Fig. 7.3). Patients presenting with suspicious symptoms (back pain, leg pain, and/or urinary-fecal incontinence) (Table 7.2) associated with
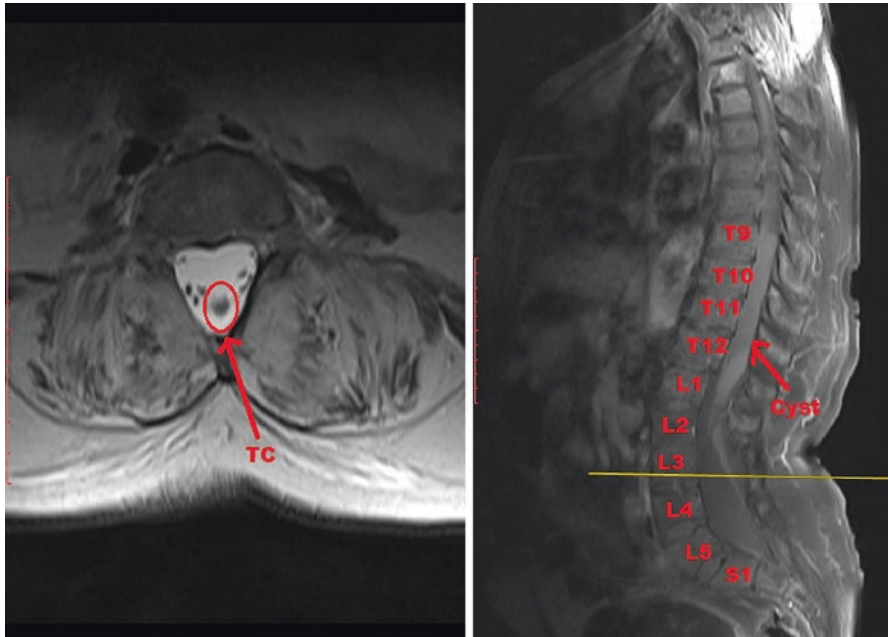
Fig. 7.3 A 68-year-old male patient presented with low back pain and progressive myelopathy. Right side: T1-weighted sagittal MRI with contrast enhancement demonstrating cyst extends between T9 and L2 and measures 12×86 mm. Note that L3 and L4 vertebral fusion anomaly (block vertebra) is apparent; *yellow line* shows the cross-section at L3 level that axial MRI shows on the left side. Left side: axial MRI demonstrated that filum terminale terminates inferior to L3 level (Case Study 3). *TC* tethered cord, *T* thoracic vertebral body, *L* lumbar vertebral body, *S* sacral vertebral body

any malformations or structural lesions such as lipomyelomeningocele, lipomeningocele, dermal sinus tract, cutaneous stigmata (hypertrichosis), scoliosis, or other system malformations (e.g., congenital heart disease, congenital kidney disease, or genetic syndrome) (Table 7.1) have to undergo detailed neurological and physical examinations, followed by craniospinal MRIs. Especially in patients with chronic low back pain that increases with static and dynamic movements, full physical and neurological examination has to be done first, and then plain X-rays of the spine have to be taken. After plain X-ray, lumbar MRI is the next least invasive imaging test for low back, back, or radiating leg pain. Lumbar MRI helps to reveal the level of the conus medullaris. If the TC is below the L2 level in an adult patient, craniospinal MRI is required before planning treatment.

The subtly differing properties of that signal from various tissues enable MRI to differentiate among organs and potentially to reveal contrasts between benign and malignant tissues (Fig. 7.3). MRI is excellent at demonstrating degenerative spinal changes such as arthritis, which can narrow the spaces through which the spinal nerves travel. In addition, it can determine herniation of the spinal discs between vertebral levels that can bulge and compress either the spinal cord or a nerve.
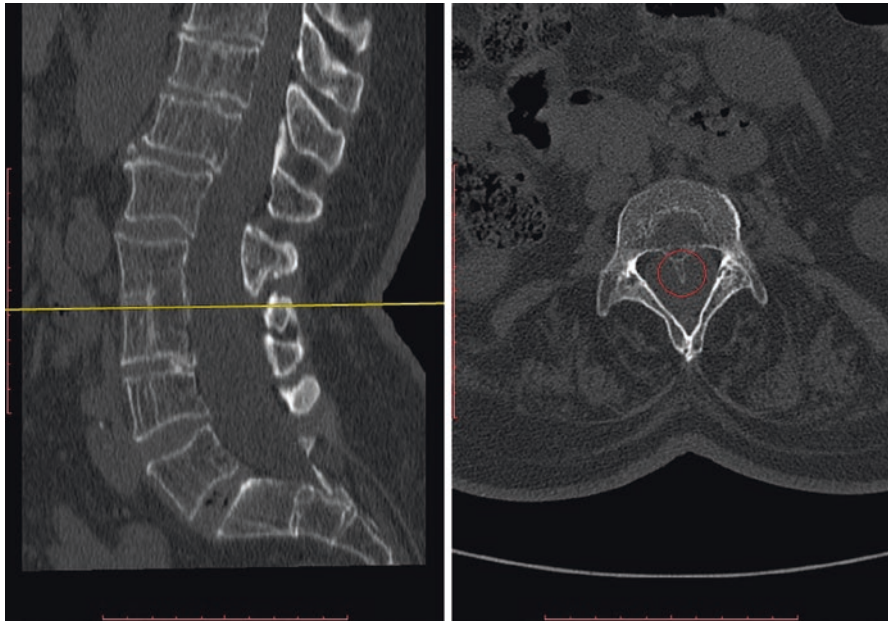
Fig. 7.4 CT imaging of Case Study 3. Right side: axial CT demonstrating posterior bony septum at L3 level. Left side: sagittal CT demonstrating block vertebra between L3 and L4 bodies; *yellow line* shows the cross-section at L3 level that axial CT shows on the right side

Computed tomography (CT) imaging can help define congenital bony anomalies better than MRI (Figs. 7.4 and 7.5) but has lost its prominence in the diagnostic assessment of TC and other OSD forms. High-resolution CT with thin slices and with reconstruction is useful in very complex OSD and to facilitate surgical planning. Three-dimensional CT can help to reveal scoliosis and bony markers to facilitate posterior instrumentation placement (Fig. 7.6).

Urodynamic study establishes the pressure-flow relationship between the bladder and urethra to define lower urinary tract function. This testing should assess the voiding phase of both bladder and urethral functions as well as the filling and storage phases. Simple urodynamic testing involves noninvasive uroflow study, obtaining a postvoid residual urine measurement, and performing single-channel cystometrography. In simple single-channel cystometrography, water is generally used as the fluid medium because it assesses the first sensations of filling, fullness, and urinary urge well. Bladder compliance and uninhibited detrusor phasic contractions can also be noted during filling cystometrography.

A video-urodynamic study is used to evaluate patients with incontinence. Radiographic contrast substance is the fluid medium for such studies. Deterioration of kidney function secondary to high bladder pressure transmitted to the upper urinary tract has been seen in the setting of neurogenic lower tract dysfunction, which can arise in OSD cases and other neurogenic conditions. Urodynamic studies can reveal low bladder capacity and overflow incontinence and serve as predictors for good surgical outcomes [1, 10] or as a baseline for postoperative follow-up [13].
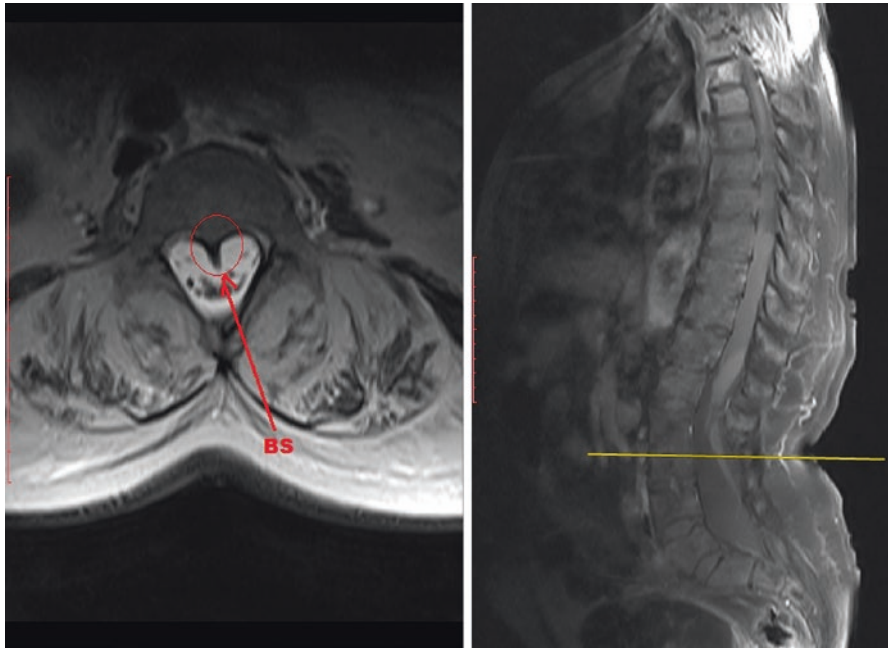
Fig. 7.5 MRI of Case Study 3. Right side: T1-weighted sagittal MRI with contrast enhancement; *yellow line* shows the cross-section at L3 level that axial MRI shows on the left side. Left side: T2-weighted axial MRI demonstrating posterior bony septum (BS) at L3 level (Case Study 3)

These urodynamic studies are useful because the therapeutic results are tied to understanding of the pathophysiological characteristics of a given case. Thus, surgeons can make a correct and complete diagnosis. Surgery on OSD cases presenting with urinary incontinence, if incorrectly diagnosed, can have substantial failure and high complication rates.

Unfortunately, urodynamic study is expensive and requires good specialized expertise and equipment, which can limit its availability. On the other hand, these studies are unphysiological in nature, and the reference ranges are wide. Therefore, the significance of the findings obtained must be assessed in association with the patient's symptoms. Studies that show abnormalities with no associated symptoms are not conclusive.

Case Study 3 *A 68-year-old male was referred to us with low back pain for 7 years and progressive myelopathy in both lower extremities for 4 months. He was operated on for inguinal hernia, cataract, and benign prostatic hyperplasia. His neurological examination was intact except for weakness in his right leg muscles group of 4/5 and weakness in his left leg muscles group of 2/5, bilateral L2–L5 and S1 dermatomal hypoesthesia, unsteady gait, joint position sense bilaterally evaluated as absent, bilateral positive clonus reflex, and the Babinski sign bilaterally evaluated as positive. MRI and CT imaging showed diastematomyelia at L1–L3, block vertebra at L3–L4, split cord malformation as an anterior fibrous band*

Fig. 7.6 Three-dimensional CT of Case Study 4 showing right curved thoracolumbar scoliosis

at level L2, and posterior bony septum at level L3. A giant cyst extended between T9 and L2 and measured 12x86 mm, while the filum terminale terminated at L4 level (Figs. 7.3, 7.4, 7.5, 7.7, and 7.8). *Urodynamic study showed normocompliance normotonic bladder function. The patient underwent laminotomy between T9 and L5 using a high-speed drill motor. The anterior fibrous band and posterior bony septum were resected at levels L2 and L3, respectively, followed by cystectomy, duraplasty, and total laminoplasty between T9 and L5 using nonabsorbable sutures* (Figs. 7.9, 7.10, and 7.11). *Histopathological examinations revealed that the cyst was arachnoidal. The patient was discharged after 7 days without complications. According to the neurological scoring system* (Table 7.4), *his neurological examination at discharge showed improvement of pain intensity (from 2 to 3), motor weakness (from 2 to 4: left leg distal muscles group still unchanged), sensory disturbance (from 1 to 3), and gait ataxia (from 1 to 3). He had no*
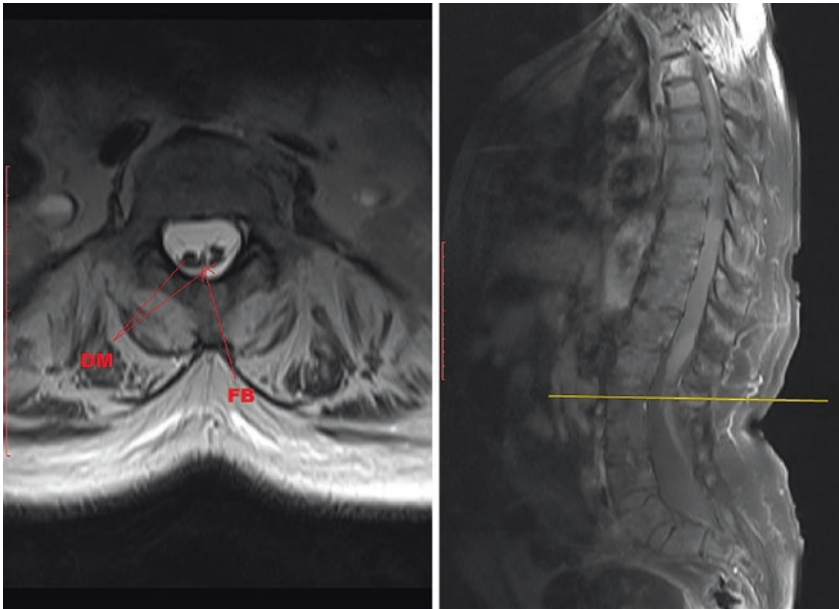
Fig. 7.7 MRI of Case Study 3. Right side: T1-weighted sagittal MRI with contrast enhancement; *yellow line* shows the cross-section at L2 level that axial MRI shows on the left side. Left side: T2-weighted axial MRI demonstrating anterior fibrous band (FB) at L2 level (Case Study 3). *DM* diastematomyelia
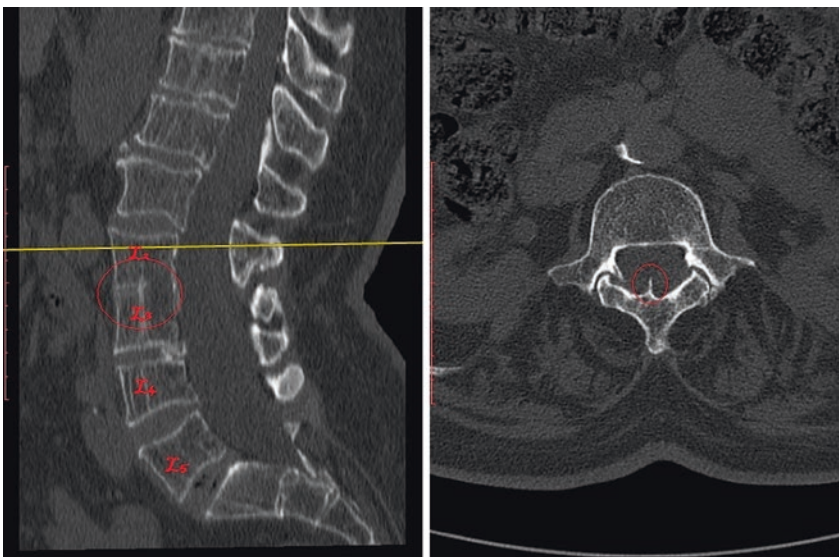

Fig. 7.8 CT imaging of Case Study 3. Right side: axial CT demonstrated anterior FB at L2 level. Left side: sagittal CT demonstrated block vertebra between L3 and L4 bodies; *yellow line* shows the cross-section at L2 level that axial CT shows on the right side. *L* lumbar vertebral body
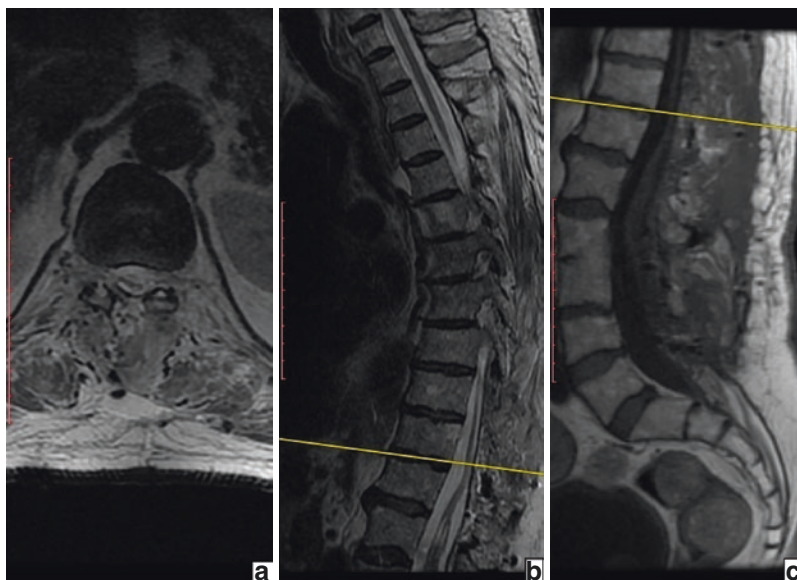
Fig. 7.9 Early postoperative MRI of Case Study 3: (**a**) T2-weighted axial MRI demonstrated axial cross-section at T11–T12 level; (**b**) T2-weighted sagittal thoracolumbar MRI demonstrated cyst was excised totally; (**c**) T1-weighted sagittal lumbar MRI showed cystectomy and postoperative changes; *yellow lines in* (**b**, **c**) show the cross-section at T11–T12 level that axial MRI shows in (**a**)
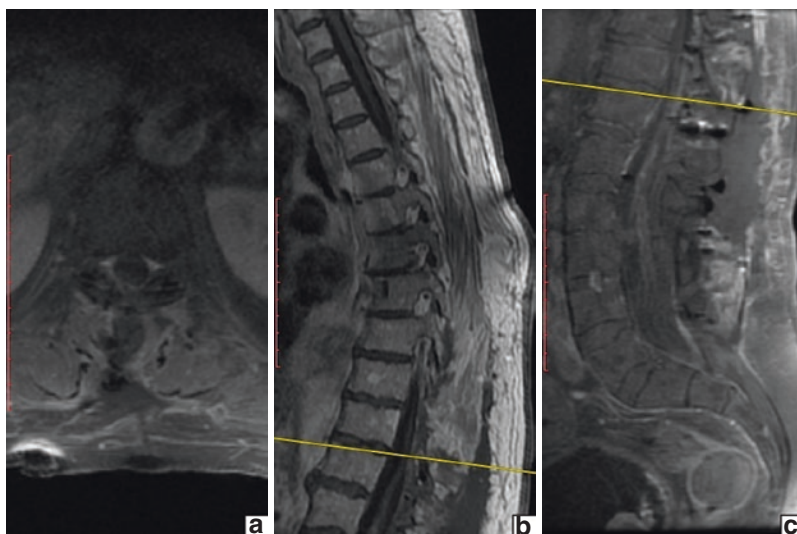

Fig. 7.10 Early postoperative MRI of Case Study 3; (**a**) T1-weighted axial MRI with contrast enhancement demonstrated axial cross-section superior to T12 level; (**b**) T1-weighted sagittal thoracolumbar MRI without contrast enhancement demonstrated cyst was excised totally; (**c**) T1-weighted sagittal lumbar MRI with contrast enhancement showing cystectomy and postoperative changes, *yellow lines in* (**b**, **c**) show the cross-section superior to T12 level that axial MRI shows in (**a**)
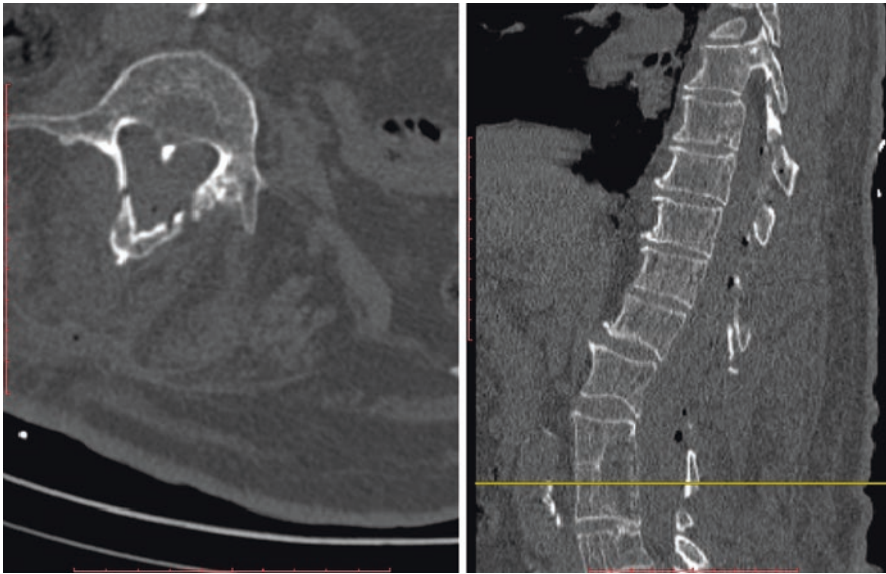
Fig. 7.11 Early postoperative CT imaging of Case Study 3. Right side: sagittal CT demonstrated total L3 laminoplasty using nonabsorbable sutures; *yellow line* shows the cross-section at L3 level that axial CT shows on the left side. Left side: axial CT demonstrating laminoplasty

Table 7.4 Neurological scoring system

Score	Pain intensity	Sensory disturbance, dysesthesias	Motor weakness	Gait ataxia	Sphincter function
5	None	Normal	Full power	Normal	Normal
4	Slight, no medication	Present, not significant	Movement against resistance	Unsteady, no aid	Slight disturbance, no catheter
3	Tolerable w/ medication	Significant, function not restricted	Movement against gravity	Mobile w/ aid	Residual in urodynamic studies, no catheter[a]
2	Insufficient control w/ medication	Some restriction of function	Movement w/o gravity	Few steps w/ aid	Rarely incontinent
1	Severe despite medication	Severe restriction of function	Contraction w/o movement	Standing w/ aid	Frequent catheter
0	Incapacitating	Incapacitated function	Paralysis	Paralysis	Permanent catheter

Data from Sofuoğlu et al. [10]
w/ with, *w/o* without
[a]Modified according to our protocol

complaints of sphincter dysfunction. The patient was sent to a physiotherapy center. His neurological examination was quite improved on his postoperative 6th-month doctor visit.

Other Adult OSD Forms

Spinal anomalies common to OSD include fatty and thickened filum terminale, split cord malformation, diastematomyelia, lipomyelomeningocele, meningocele, dermal sinus, tight filum terminale, neurenteric cysts, terminal myelocystocele, and meningocele manqué (Table 7.1).

Split Cord Malformation (SCM) and Diastematomyelia

Split cord malformations (SCMs) are mainly associated with TC in adults [2]. They are divided into two types on the basis of the state of the dural tube and the nature of the median septum: type I (i.e., diastematomyelia with septum) (Figs. 7.1 and 7.3) and type II (i.e., diastematomyelia without septum) (Fig. 7.12). The former type usually coexists with scoliosis and TCS. Cutaneous stigmata (hemangioma), skin discolorations, and hypertrichosis are characteristic features of type I SCM. Butterfly vertebrae, block vertebra, hemivertebrae, and spina bifida are the vertebral abnormalities that can be associated with this type [1, 14]. The BS (Figs. 7.4 and 7.5) or FB (Figs. 7.7 and 7.8) midline septum splits the spinal cord into two tubes each containing a hemicord. Beyond the BS, which can occur in either the thoracic or lumbar regions, the two hemicords adhere to each other and return to a normal anatomical position. Some cases of SCM show signs of TC though no spur is found in the malformation. This happens in type II SCM. The cleft is generally partial or incompletely split. Spina bifida is often present with the possible formation of hydromyelia, and a thin FB can form [15].

Diastematomyelia also accompanies SCM (Figs. 7.1 and 7.3). Diastematomyelia refers to the splitting of the spinal cord, conus medullaris, or filum terminale in the sagittal plane into two not necessarily equal hemicords [15, 19, 21]. A thick cutaneous hairy patch usually overlies the region of the diastematomyelia [1, 14, 15]. Diastematomyelia accounts for up to 44% of OSD cases [1]. Vertebral fusion anomalies and scoliosis are almost always associated with it. Tethering can result from dorsal or ventral tethering bands between the hemicords and the dura or from a thickened filum terminale.

In adult OSD, craniospinal MRI, particularly axial sections, axial sectioned CT, or CT myelography, is useful for demonstrating the hemicords and median septum (Fig. 7.1). As we previously explained, neurosurgeons have to decide about surgical intervention according to patients' complaints and neurological examination when confirmed by MRI and urodynamic study. Surgery can involve resecting the median septum and dividing a thickened filum and dorsal tethering bands or releasing the spinal cord if it is tethered. In our experience, resection of
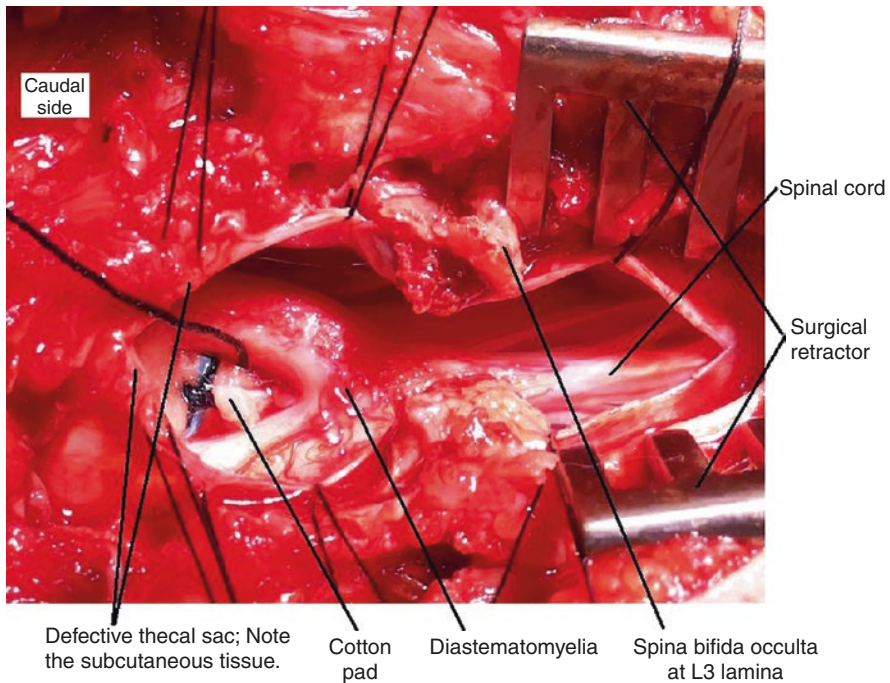
Fig. 7.12 Photo had been taken during the operation showing spina bifida occulta at L3–L4, diastematomyelia at L3, and the thecal sac terminated in the skin at the L3–L4 level. Note the subcutaneous fatty tissue in the caudal part of the dura (lipomeningocele). There was no bony septum or fibrous band between the hemispinal cords (SCM type II)

the median FB or BS in four patients (12.9% of the total) who presented with TCS without releasing the TC relieved or improved the neurological and urological symptoms.

Lipomyelomeningocele, Lipomeningocele, and Spinal Lipoma

Fatty accumulations within the spinal cord are common lesions associated with TC and take one of four forms: lipomyelomeningocele, lipomeningocele, spinal lipoma, and fatty filum. Meningocele is a type of spina bifida cystica characterized by the herniation of meninges through an abnormal opening in the spinal column. Myelomeningocele is another type of spina bifida cystica characterized by the herniation of spinal cord contents and meninges through an abnormal opening in the spinal column. When a lipoma covers the sites of meningocele and myelomeningocele, they are called lipomeningocele (Fig. 7.12) and lipomyelomeningocele, respectively.

Lipomyelomeningocele is a subcutaneous malformation within the spinal cord that extends through a defect of the lumbosacral fascia, lamina, dura, and pia into a

low-lying spinal cord [15]. Although it is the most common form of spinal lipoma, patients with lipomyelomeningoceles usually present to healthcare centers within the first few months to years of life. Therefore, these lesions are rarely seen in adult patients [1]. Lipomyelomeningocele is a lipoma of the conus medullaris [23].

The relative anatomy of the lipoma and neural tissues distinguishes three types of lipomyelomeningocele: dorsal, transitional, and caudal. In the dorsal type, lipomas have an area of attachment to the dorsal spinal cord at the splitting point in the lumbar or lumbosacral levels and are continuous with the subcutaneous tissue. In the transitional type, lipomas have attachments that extend beyond the splitting area down the conus medullaris, with a less distinct lipoma-cord interface. In this type, lipomas can extend through a dural defect. In the caudal type, lipomas arise predominantly from the caudal side of the conus medullaris.

Spinal cord (intradural) lipomas are rare intramedullary lesions almost always found within the thoracic spinal cord. They are not associated with cutaneous or bone malformations and often present with symptoms of spinal cord compression. The fatty thickened filum involves fatty infiltration into the whole length or part of the filum terminale. The fat within the thick filum can be diagnosed by MRI. The occurrence of incidental fat within the filum terminale in the normal adult population has been estimated to be 3.7% in cadaveric studies and 1.5–5% in MRI studies [23].

Fatty accumulations can be diagnosed by the associated subcutaneous lumbosacral masses found in almost all patients or by sagittal and coronal MRI. Surgery on the symptomatic patient has been advocated by many authors to prevent further decline in neurological status [1]. For lipomyelomeningoceles and lipomeningoceles, the aim of surgical intervention is to reconstruct the neural tube and then to repair the thecal sac to reformat the subarachnoid space (Fig. 7.13).

In our adult OSD cases, 2 out of 31 (6.5%) had lipomyelomeningocele, 4 (12.9%) had lipomeningocele, and lipomas were seen in 8 (25.8%). The operation for fatty accumulations has been significantly advanced by IONM and laminoplasty. Once

Fig. 7.13 Intraoperative photo shows duraplasty was performed using 5.0 absorbable sutures after exploring the spinal cord. Thus, the thecal sac was repaired
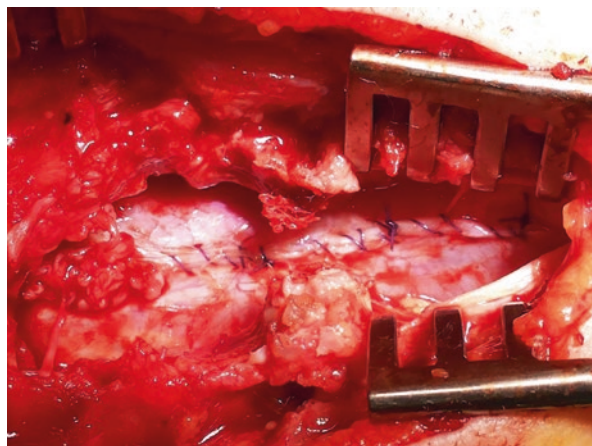
the spinal cord has been released, we recommend closure and enlargement of the dura with an allograft material to reduce the risk of re-tethering. Surgery for a lipoma of the filum terminale in the symptomatic patient has been recommended [1, 6, 10]. Surgery for an asymptomatic lipoma of the conus medullaris is still controversial [10].

Tight Filum Terminale Syndrome

This syndrome refers to TCS in a patient with a low-lying conus medullaris. In almost all such patients, the tip of the conus medullaris lies inferior to the L2 body. In this TCS, the filum terminale exceeds 2 mm in diameter, and there are no other tethered malformations. In 86% of patients, the tip of the conus medullaris lies inferior to L2. The tight thickened filum terminale and the low-lying conus can be visualized on MRI [23].

Terminal Myelocystoceles

These malformations are rare forms of OSD, the elements of which include expansion of the central canal of the caudal spinal cord by a cerebrospinal fluid (CSF)-containing terminal cyst, which is itself surrounded by an expanded dural sac. Patients harboring these lesions typically have no bowel or bladder control and possess poor lower-extremity function. Therefore, such patients are diagnosed in early childhood. These malformations are associated with a lipoma or multiple congenital genitourinary and orthopedic defects such as omphalocele, cloacal exstrophy, imperforate anus, renal abnormalities, ambiguous genitalia, pelvic deformity, and talipes equinovarus. In such OSDs, tethering results from the attachment of the myelocystocele to the inferior aspect of the spinal cord [23]. Terminal myelocystocele, like all OSDs, can be diagnosed by MRI. The aim of surgery is to separate the spinal cord from the fluid-filled terminal myelocystocele, reconstruct the neural tube, and then repair the dura to reformat the subarachnoid space.

Spinal Neurenteric Cysts

These cysts are rare congenital malformations formed by entrapment of endodermal tissue between a split notochord. Such cysts can be extraspinal with mediastinal or abdominal extension, or they can be intramedullary. They are frequently associated with spina bifida and can occur with no associated dysraphic lesions. Patients with these lesions can present with acute progressive signs as a result of spinal cord compression. A neurenteric cyst can be diagnosed by MRI. The aim of surgery is to remove the cysts totally. Subtotal resection of them increases the recurrence rate [23].

Dermal Sinus Tract

A dermal sinus tract appears as a midline dimple in the lower lumbar and lumbosacral region (Fig. 7.1). Patients with dermal sinus tract are usually referred to neurosurgeons or general surgery with recurrent infection in the affected area, which can present as meningitis. The dermal sinus tract can extend from the skin surface to the dura, subarachnoid space, or the spinal cord, thereby causing tethering [23]. It can be visualized by MRI or CT myelography.

Meningocele Manqué

Meningocele manqué refers to a dysraphic lesion of dorsal tethered bands composed of fibrotic or atretic neural tissue connecting the spinal cord to the dura or surrounding structures. These lesions are usually found incidentally during surgical exploration for other OSDs, and they can occur far from the site of obvious tethering. These tethered bands are almost always found at the site of diastematomyelia. The aim of surgical intervention is to remove these bands. Meningocele manqué can be visualized by MRI [23].

Case Study 4 *A 19-year-old female was referred to us with low back pain, uncontrolled leg contractions, and unsteady gait for 6 years and progressive myelopathy in the left leg and urinary-fecal incontinence for 4 months. She had had spinal malformations since her birth. She was referred to another healthcare center after urodynamic study that showed low bladder capacity and overflow incontinence. The urological surgeon recommended a Foley catheter three times and oxybutynin 5 mg twice a day. HerPlease check if edit to sentence starting "Her neurological examination…" is okay. neurological examination was intact except for weakness in her left leg muscles group of 2/5, ataxic gait, walking only a few steps with aid, left positive clonus reflex, and deep tendon reflexes increased in the left leg. MRI and CT imaging showed thoracolumbar scoliosis* (Figs. 7.6 *and* 7.14) *and a lipomyelomeningocele extending between T10 and L1. The filum terminale terminated at S2 level. The patient underwent total T9, T10, and S1 laminectomy (the condition of the bone was not suitable for laminoplasty) and repair of the lipomyelomeningocele* (Fig. 7.15)*; then the TC was released at S2. She was discharged after 14 days with no improvement in her neurological scoring system. After an anesthesiologist consultation, she was planned to undergo correction for her scoliosis but after a delay of 1 year.*

Standard Management of Adult OSD

Almost all authors recommend surgery as soon as neurological symptoms appear [1, 8, 10, 13, 17, 21, 22]. However, a few have recommended surgery for those with progressive neurological symptoms [6]. We described the standard management protocol that we use to treat adult TC in previous paper [10]. Although the aim of
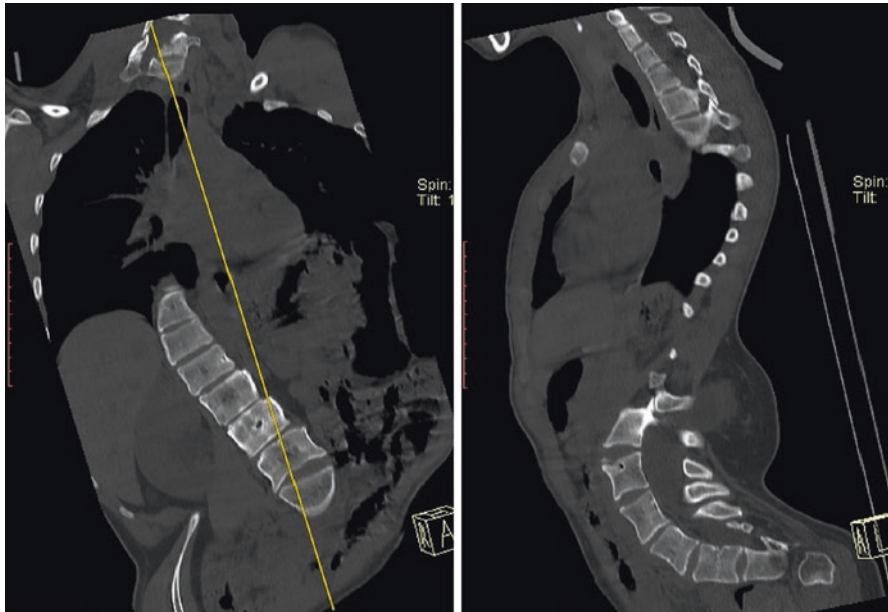
Fig. 7.14 CT of Case Study 4 showing right curved thoracolumbar scoliosis. Right side: sagittal CT. Left side: coronal CT

the initial approach of surgical intervention is to deal with the pathological conditions that cause the symptoms in symptomatic OSD patients, the surgical management of asymptomatic OSD remains controversial. Therefore, patients who present with suspicious symptoms (back pain, leg pain, and/or urinary-fecal incontinence) (Table 7.2) associated with any malformation or structural lesion such as lipomyelomeningocele, lipomeningocele, dermal sinus tract, cutaneous stigmata (hypertrichosis), scoliosis, or another system malformation (e.g., congenital heart diseases, congenital kidney diseases, or genetic syndromes) (Tables 7.1) have to undergo detailed neurological and physical examination followed by craniospinal MRIs. If any findings support OSD, the patients must undergo a urodynamic test. If all investigations lead us to diagnose the patient as symptomatic OSD, the neurosurgeons have to explain the risks, complications, and benefits of surgical intervention to the patients and their families in order to help them decide about the operative option.

If both the physicians and the patient's family opt for surgery, neurophysiological monitoring and laminoplasty rather than laminectomy have to be applied. Intraoperative surgical strategies depend on additional pathological entities associated with OSD. Klekamp recommended a complete resection, including the capsules in hamartomas (i.e., lipomas, dermoid cysts, epidermoid cysts, and neurenteric cysts), and using artificial materials for duraplasty [6]. In our series, eight adult OSD patients had lipomas, but the lipomas were resectioned in only three of those eight. One patient had recurrence after 6 years, even after complete resection of the lipoma with its capsule. To avoid serious complications and re-tethering, the lipoma
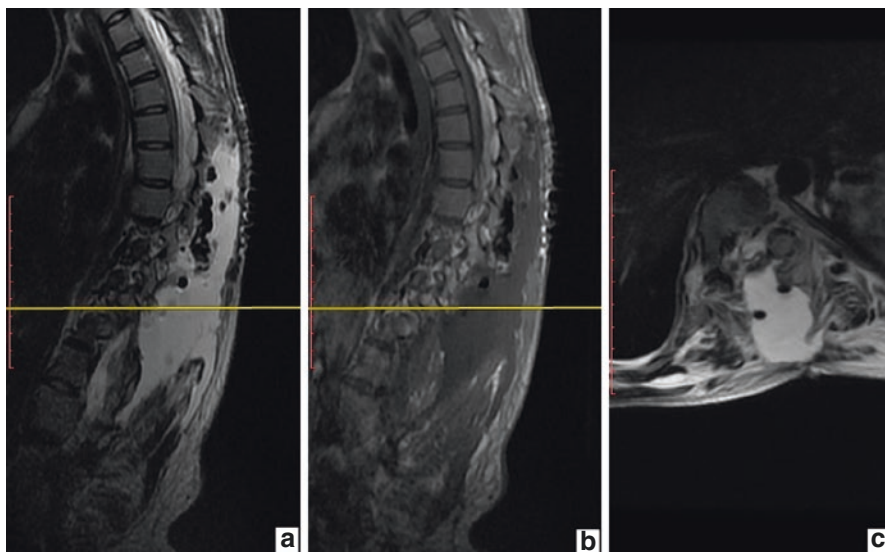
Fig. 7.15 Early postoperative MRI of Case Study 4 demonstrating reconstruction of thecal sac. (**a**) T2-weighted sagittal MRI; (**b**) T1-weighted sagittal MRI; (**c**) T2-weighted axial MRI: *yellow lines in* (**a**, **b**) show the cross-section at T10–T11 level that axial MRI shows in (**c**)

was left without resection after intraoperative evaluation showed that it did not press on nerve roots or the dural sac. However, asymptomatic patients have an increased risk of developing further neurological deterioration. Close patient follow-up and timely treatment for local pathologies after changes in urodynamic or manual motor testing are detected can protect patients from new deficits.

Adult OSD patients are less likely to show neurological deterioration because there has already been rapid spinal column growth in a spinal cord that is tethered caudally. Four of our patients did not undergo TC release; only bony septum resection had been applied. Two of those four also underwent cystectomy (Case Studies 1 and 3). The guide here is the consistency of clinical presentation and neurological examination with the urodynamic study. In these four cases, the presenting symptom was pain, while the urodynamic tests showed normocompliant normotonic bladder function with less than 50 ml postvoid residual urine (normal). Therefore, bony septum resection sufficed to resolve their complaints, rather than an untethering procedure that could result in new neurosurgical deficits. Thus, our management does not agree with the literature, in which most authors suggest that TC release is a necessary intervention in adult TC patients [6, 8, 13, 17].

Surgical Procedure

Under general anesthesia and using intraoperative neurophysiological monitoring (IONM), the patients are positioned prone using a supporting roll on each side. A paramedian vertical midline incision is made between the superior and

inferior laminas detected via MRI (e.g., if the conus medullaris is at L5 level, the incision would be extended between L4 and S1). The paraspinal muscles are dissected. If the aim is to perform partial or total (complete) laminectomy, the dissection is unilateral; if the surgeons plan a laminoplasty, the dissection is bilateral.

Hemilaminectomy, laminectomy, or laminotomy is completed (in cases of spina bifida occulta, defective laminotomy is used). If the purpose is laminoplasty, bilateral laminotomy is performed using high-speed drills or Kerrison rongeurs. Next, the ligamentum flavum and the adipose tissue are removed. The spinal cord can continue to the S1 or S2 levels by giving some sacral rootlets. Laminectomy or laminotomy should be performed up to this level. The operative microscope is brought in over the operation field. The thecal sac should be opened in the midline and tacked up bilaterally using strong sutures.

After all nerve roots, filum terminale, and arachnoid bands have been exposed, the neurosurgeons select the filum terminale using the microscope (generally senior neurosurgeons select it by its suspected structure) and IONM. Under the microscope, the filum terminale appears darker than the nerve rootlets. This darker color is related to its fibrovascular tag structure that contains a large vessel, which becomes smaller through thecal sac [7, 15, 18] (Fig. 7.16). However, this vessel is not a reliable landmark for the filum terminale since similar vessels can be found on the rootlets, and sometimes no vessel can be seen on the filum terminale [1, 10]. The use of the IONM probe is recommended to check whether the tissue is neural. This helps to avoid cutting one of the rootlets instead of the filum terminale to be tethered. The rootlets are retracted laterally, and the filum terminale is coagulated and cut after it is identified. All connective tissues attached to the caudal part of the spinal cord and conus medullaris should be released. After hemostasis using physiological saline, duraplasty is performed using 5.0 absorbable sutures (Fig. 7.13). To avoid a CSF fistula after tight closure of the dura, the surgeons use Fibrin Sealant Products. If BS or FB is present, it should be resected first, before the untethering
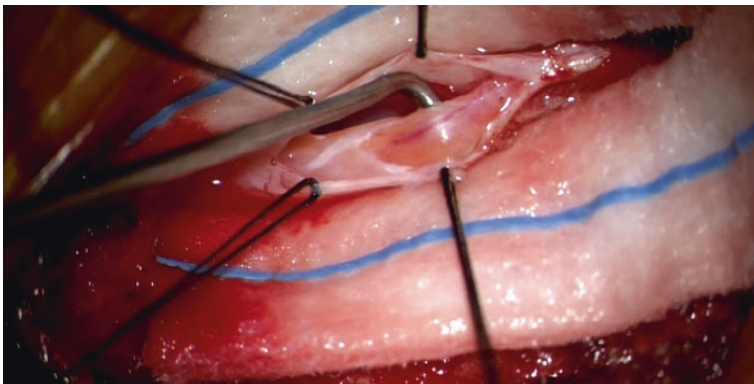

Fig. 7.16 Intraoperative photo shows TC at S2 level before untethering procedure. Note that the darker color of the terminal filum is related to its fibrovascular tag structure containing a large vessel, which becomes smaller through the thecal sac. Note the operation field is kept clean, and the CSF circulation between neural elements preserved using paddy cottons (Case Study 5)

procedure. In cases of dermal sinus, the tracts can be attached to the thecal sac. Therefore, these structures should also be removed and duraplasty should be done if necessary.

In cases of lipoma, cyst, or other hamartomas that press on the nerve rootlets and narrow the thecal sac, the hamartomas should be removed too. To avoid serious complications and prevent re-tethering, lipomas should not be removed aggressively. This decision should be taken after intraoperative evaluation, keeping the operation field clean and preserving CSF circulation between the neural elements.

Scoliosis Associated with Adult OSD

Scoliosis is defined as a lateral curvature of the spine of at least 10° with vertebral rotation. The most common type of scoliosis in the pediatric and young adult populations is idiopathic. After skeletal maturity is reached, a patient with adolescent idiopathic scoliosis is defined as having adult idiopathic scoliosis. Such patients have had scoliosis while growing into adulthood. Typically, there is a slow increase (about 0.5–2° per year) in the lateral curvature. The scoliosis associated with adult OSD generally coexists with type II SCM, has a single big curvature, and affects the thoracic and thoracolumbar spine [1, 10]. Such scoliosis is thought to be secondary to the OSD itself. It is essential to assess all scoliosis patients fully and perform craniospinal MRI before planning surgery to determine whether there is any co-malformation or OSD.

Symptoms

Shoulder asymmetry, a rib hump, or a prominence of the lower back on the side of the lateral curvature is the most physical symptoms of scoliosis. Both types of adult scoliosis can progress over time. Curves that reach 50° or more can progress more rapidly than those that are less than 50°. Neurological symptoms can include back pain, especially in adults with large curvatures, and shortness of breath with activity if the curvature in the thoracic spine exceeds 80°. Although adult scoliosis alone rarely causes paralysis or other severe neurological problems, it can be associated with OSDs such as TC or SCM, which can cause nerve irritation, leg pain, muscular weakness, and sphincter dysfunction. In our experience, two out of eight scoliosis patients presented because of their humps. Both patients were female. One of them had hemiparalysis (Case Study 4).

Diagnostic Work-Up

Scoliosis can first be recognized clinically with a physical examination. Neurological examination is mandatory for full assessment of the patient. Plain and scoliosis radiography (full-length, whole spine X-rays need to be performed) is necessary to

determine the magnitude and type of scoliosis fully. For a proper scoliosis assessment, lateral bending X-rays are used to assess the rigidity of the scoliosis. In our experience, 8 out of 31 adult OSD patients (25.8%) presented to us with scoliosis. Therefore, MRI and three-dimensional CT imaging are needed if there is OSD in a pediatric or young adult patient.

Treatment and Follow-Up

The treatment of adult scoliosis associated with OSD is very individualized and is based on the specific symptoms and age of the patient. Many adult patients with scoliosis have very minor symptoms, diagnosed incidentally, and they live with it without treatment. Patients with predominant symptoms of moderate back and leg pain are typically treated conservatively. Physical therapy exercises, swimming, and sometimes bracing can help to reduce pain intensity. Patients with severe back and leg pain can benefit from steroid injections to help relieve the leg pain. If the scoliosis curvature measures more than 50° or the patient asks to have his/her hump treated for cosmetic reasons, surgical correction can be useful. Decompressive surgery, fusion procedure, and osteotomies are the most commonly used surgical approaches. The aim of surgical intervention is to remove pressure on the nerves and spinal cord and to maintain stabilization and obtain sagittal balance of the spine. Thus, surgery stops the scoliosis from progressing. The length of the fusion, or the number of spine levels included, depends on the age of the patient, the type of scoliosis, and the spinal area involved. IONM and an expert spine surgeon team are essential for precluding serious complications. Early postoperative lung X-rays are essential for identifying lung complications.

Follow-up is mandatory after scoliosis surgery. It is important to have an adult scoliosis specialist monitor the curvature over time. These curvatures can worsen owing to disc degeneration, which can happen in elderly patients. This can also cause sagittal imbalance because it makes the patient lean progressively further forward. In severe cases, arthritis in the spine facets can lead to bone spurs, local back pain due to stiffness of the back, radiating leg pain, and numbness down the legs from pinched nerves.

Case Study 5 *An 18-year-old female was referred to us from another country with a hump in her back. Her mother said she had had spinal malformation since birth. Her hump was minimal up to the age of 12. Her neurological examination was intact. Full-spine X-ray showed left curved thoracolumbar scoliosis (T2–L3) of 105° (Fig. 7.17a). MRI and CT imaging showed left curved thoracolumbar scoliosis extending between T2 and L3 (Fig. 7.18a). The filum terminale terminated at S2 level (Figs. 7.18b, c). The patient underwent total S2 laminectomy, and then the TC was untethered at the S2 level (Fig. 7.16). After 2 days, posterior instrumentation was performed between the T3 and L2 vertebral bodies, followed by a right T8 hemicorporectomy (Fig. 7.17b). The follow-up X-rays demonstrated a curve*
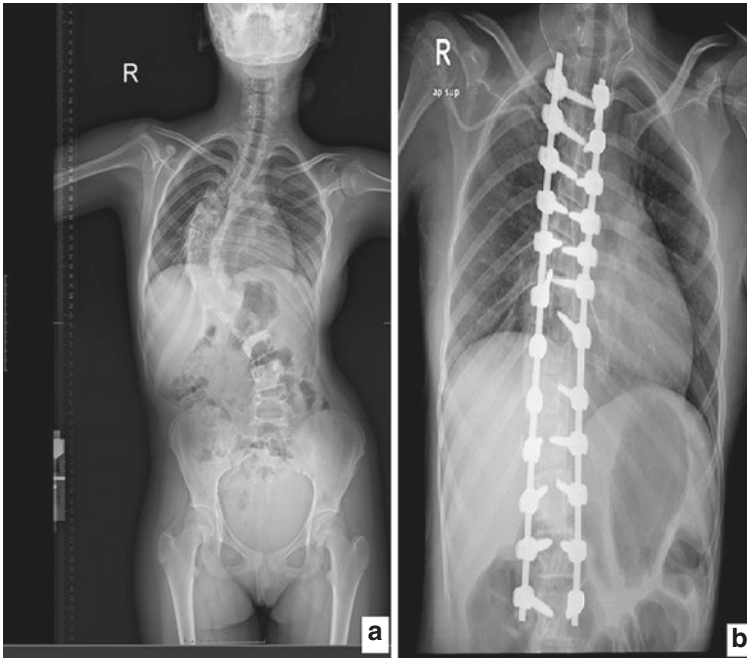
Fig. 7.17 An 18-year-old female patient presented with a hump in her back. Plain full-spine X-ray of Case Study 5: (**a**) preoperative X-rays showed left curved thoracolumbar scoliosis of 105°; (**b**) early postoperative X-rays
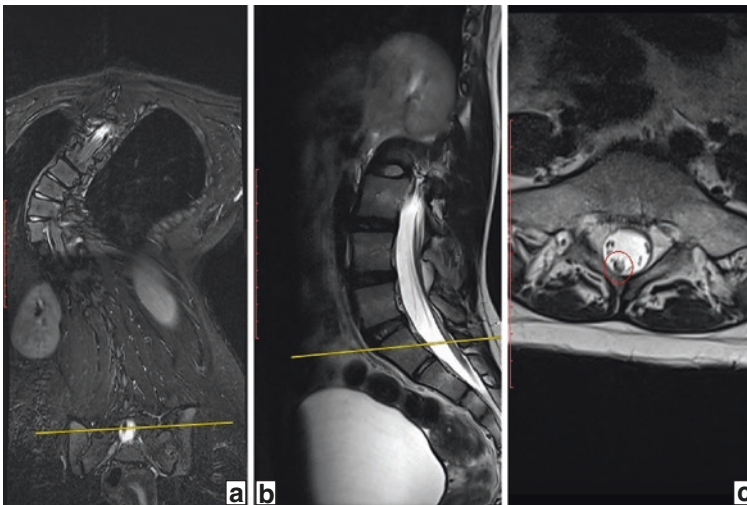

Fig. 7.18 Preoperative MRI of Case Study 5: (**a**) STIR coronal MRI demonstrated left curved scoliosis; (**b**) T2-weighted sagittal thoracolumbar MRI demonstrated cyst was excised totally; (**c**) T2-weighted axial lumbar MRI showed axial cross-sectional S2 level – note TC at S2 level; *yellow lines in* (**a**, **b**) show the cross-section at S2 level that axial MRI shows in (**c**)
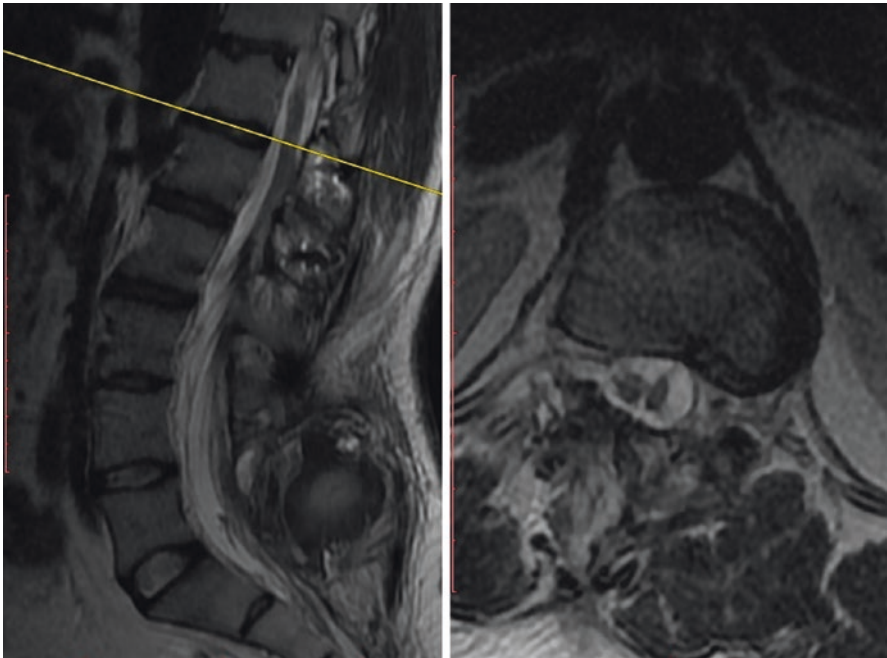
Fig. 7.19 A 31-year-old male presented with low back and left leg pain for 3 years and muscular weakness for 3 months. Early postoperative T2-weighted MRI demonstrated resection of FB at T12-L1 level. Left side: sagittal MRI; *yellow line* shows the cross-sectional T12-L1 level that axial MRI shows on the left side. Right side, axial MRI demonstrated diastematomyelia postoperatively (Case Study 6)

correction to 18 and a balanced spine. The patient was discharged after 7 days without complications. On her postoperative 15th-month doctor visit, she was doing well.

Case Study 6 *A 31-year-old male presented with low back and left leg pain for 3 years and muscular weakness for 3 months. The patient smoked and was on medication for hypertension. His neurological examination was intact except for weakness in his left leg distal muscles group of 2/5, left L2–L5 and S1 dermatomal hypoesthesia, unsteady gait, increased response of deep tendon reflexes in the left leg, and atrophy in his left leg (right and left pretibial circumferences were 28.1 and 24.2 cm, respectively). MRI showed diastematomyelia at T12-L1, and split cord malformation as a fibrous band at L3 level was apparent (Fig. 7.19). The filum terminale terminated at L5 level. CT showed a left curved thoracic scoliosis of 35° (Fig. 7.20). Urodynamic study showed normocompliance normotonic bladder function. The patient underwent FB resection and duraplasty using total T12-L1 laminoplasty via mini screws plaque (Fig. 7.21), followed by release of the TC at L5 level using total L5 laminectomy (Fig. 7.22). The patient's pain decreased but neurological status did not improve. He was discharged after 10 days without complications and was referred to the physiotherapy department.*
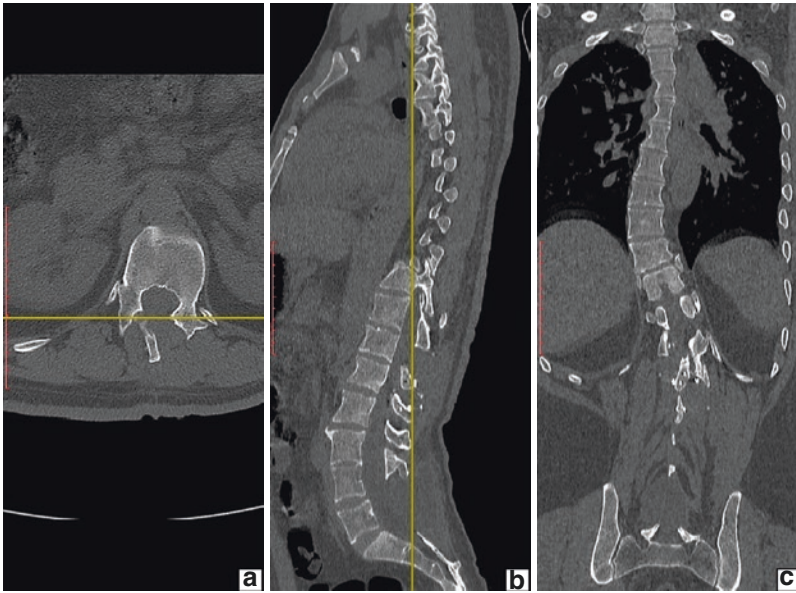
Fig. 7.20 CT of Case Study 6 showing thoracic scoliosis: (**a**) axial CT; (**b**) sagittal CT; (**c**) coronal CT
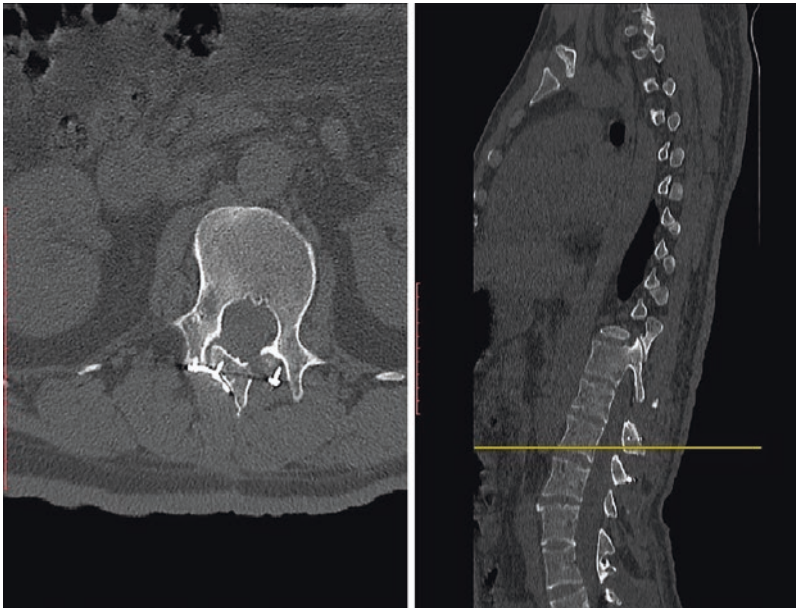

Fig. 7.21 Postoperative CT of Case Study 6 showing L1 laminoplasty using mini screws plaque. Left side: axial CT. Right side, sagittal CT
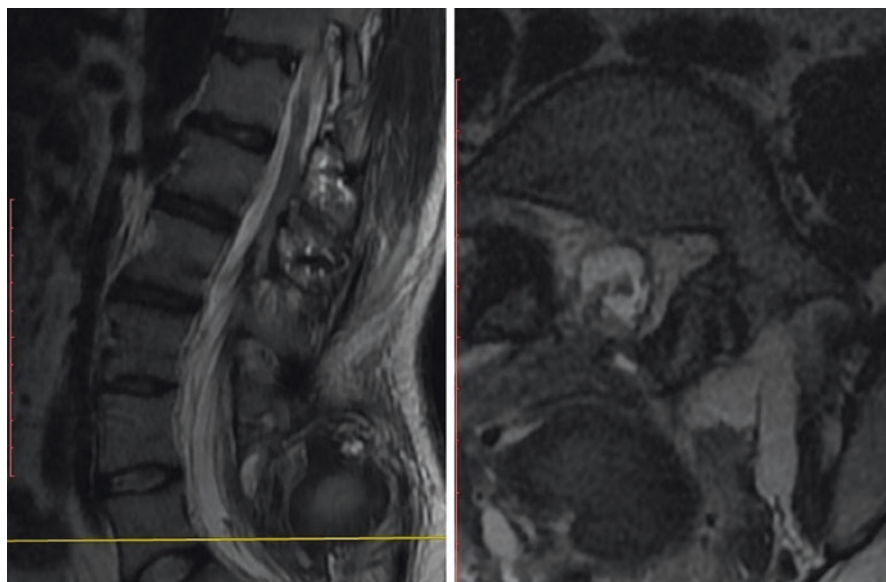
Fig. 7.22 Early postoperative T2-weighted MRI of Case Study 6 showed releasing TC at L5 level. Left side: sagittal MRI; *yellow line* shows the cross-sectional L5 level that axial MRI shows on the left side. Right side: axial MRI

Surgical Outcomes of Adult OSD Patients

Early surgical intervention after increasing intensity of pain or severity of complaints gave good results; good recovery was seen in 64.5% and improvement in 29.0% of our patients. Only two patients had worsened neurological deficits. Our good surgical outcomes were related to the full assessment of each patient separately. Those who were diagnosed incidentally with TC after moderate pain were closely followed up, and when their complaints started to be symptomatic and urological complaints were added to their clinical pictures, urodynamic studies were re-performed, and then a decision was made for surgical intervention.

Iskandar et al. [17] reported that untethering improved 22 of 27 patients (81.5%) presenting with pain, 13 of 27 (48.1%) with motor or sensory dysfunction, and 11 of 18 (61.1%) with bowel and bladder disturbances. Sofuoglu et al. [10] reported that TC release and/or BS resection in their patients improved and eliminated pain in 15.8% (3 out of 19 patients) and 84.6% (16 out of 19), respectively. Muscular weakness recovered in 8 of their 12 patients (66.7%), was unchanged in 2, and worsened and improved in 1 patient each. However, sensory disturbance remained unchanged in 50% (four out of eight patients) in their series. Bladder and bowel dysfunction improved, recovered, and remained unchanged in 50% (6 out of 12 patients), 25%, and 25%, respectively. These surgical outcomes are better than those

reported by Lee et al. [8], who found that surgical intervention improved back and leg pain in 78% and 83% of patients, respectively. Motor weakness stabilized or improved in only 27% and 64% of their patients, respectively. Sensory deficits remained unchanged in 50%. Urological abnormalities improved in 50% of patients undergoing untethering and remained stable in 45%.

Surgical outcomes of adult OSD patients are rated as recovered, unchanged, or worsened. Unless the surgical intervention leads to full recovery or resolution of the major symptoms (leg and/or back pain, motor weakness, sphincter dysfunction, or gait ataxia) that affect the patient's quality of life, patient satisfaction and quality of life continue to be poor. Therefore, we divide the surgical outcomes into two major groups. The first is the "fully recovered group" comprising patients who have good clinical outcomes with resolution of their major symptoms. The second is the "others group," which includes all cases with at least one major symptom unresolved. We investigate the factors that could have affected the outcomes in our 31 adult OSD patients.

Gender and Age

The mean age of our adult OSD patients was 32.1 ± 11.8 (18–68) years. At their final follow-up, after 62.9 months on average, 64.5% had good clinical outcomes. The 20 recovered patients comprised 13 females and 7 males. The gender factor was not statistically significant (OR 0.45, $p = 0.25$). The mean age of the recovered group was 31.2 ± 10.2 (20–61) years, while that of the "others group" was 33.5 ± 11.3 (18–68) years. Although the recovered group was younger on average, the age factor was not statistically significant (OR 0.93, $p = 0.88$).

Long-Term Symptoms and Neurological Examination on Presentation

The interval between the initial symptom and the time of surgical intervention in our sample ranged between 4 months and 25 years with an average of 5.2 years. Bladder dysfunction when associated with muscular weakness (OR 7.0, $p = 0.014$) and long-term (i.e., more than 6 months) symptoms (OR 24.5, $p = 0.0001$) are independent risk factors leading to poor, minimally improved, or almost unchanged surgical outcomes even after rehabilitation programs. Therefore, we recommend early surgical intervention in symptomatic patients irrespective of the fact that most OSD or TC cases diagnosed in adults are asymptomatic. During the last 12 years, our team diagnosed 116 adult patients with OSD after incidental permanent or severe pain in the back and/or leg or after serious urological complaints. Only 31 adult patients (26.7%) who were diagnosed as symptomatic OSD were treated surgically. Klekamp reported that only 50.8% of adult patients with TC underwent an untethering procedure [6]. Asymptomatic TC in our sample was identified in 85 patients (73.3%).

Patients have to undergo full neurological examinations, especially those who experience low back and leg pain associated with urination complaints and those with malformations associated with TC (Table 7.2). Surgical intervention is recommended in children and adolescents to reduce the expected risk of further neurological deficits [10].

Associated Malformations

Three patients (9.7%) in our operated 31 adult OSD were free of other malformations associated with TC (Table 7.1). In our experience, seven out of eight patients with SCM were treated by BS or FB resection. Only four of those seven needed a subsequent untethering procedure to relieve their pain or other complaints. In the remaining three patients, the untethering procedure was not applied, so new neurological deficits were precluded. Out of eight patients with lipoma, only two were treated by removal of the lipoma. A third patient was treated by cystectomy. Four out of five patients who presented with dermal sinus tract were treated by resection and surgical repair of the skin. Except for two young adult female patients with scoliosis (who wanted the operation for cosmetic reasons), patients with scoliosis and syringohydromyelia did not need additional surgical intervention, but 38% of our child TCS patients who presented with the same malformations underwent additional surgery to treat their complaints [10]. Surgical outcomes in cases without lipomas have better recovery chances than those with lipomas (OR 2.4, $p = 0.001$). Aggressive surgical treatment of lipoma was the reason behind the only recurrence case 6 years later. Lipoma was associated with motor weakness, atrophy, or bladder dysfunction, but only with trend-level significance (OR 1.49, $p = 0.09$).

Surgical Approaches

IONM and laminoplasty have been integral to surgery, especially in recent years. IONM is useful for distinguishing functional from nonfunctional nerve roots. Thus, neurosurgeons can reduce the serious complications that could result from surgical intervention in adult OSD cases. Laminoplasty reduces the adhesions that could facilitate recurrence of TCS in adults; even our recurrent patient underwent laminoplasty, but his recurrence was thought to be related to the aggressive removal of the lipoma.

Comparing laminoplasty, hemilaminectomy, and laminectomy in our sample, the authors found that laminoplasty leads to a mean hospital stay of 3.9 ± 2.2 (2–8) days, which is shorter than for the hemilaminectomy and laminectomy approaches, which lead to mean hospital stays of 4.0 ± 2.2 (2–7) and 6.4 ± 3.3 (3–12) days, respectively. However, the difference is not statistically significant ($p = 0.57$ and $p = 0.29$, respectively). None of the three approaches is superior to the others in regard to surgery-related complications.

A multivariate regression model showed that compared to laminoplasty (OR 2.05, $p = 0.047$) and hemilaminectomy (OR 1.875, $p = 0.049$), laminectomy is independent of other risk factors associated with poor or marginally improved (almost unchanged) surgical outcomes. This statistically significant difference could be related to IONM, not to the approach itself.

In cases of lipoma, cyst, or other hamartomas that press on the nerve rootlets and cause narrowing of the thecal sac, the hamartomas should be removed too. To avoid serious complications and to prevent re-tethering, lipomas should not be removed aggressively. This decision should be taken after intraoperative evaluation. The operation field should be kept clean, and the CSF circulation between the neural elements should be preserved.

In the adult population, the rate of re-tethering is reportedly as high as 29% [5, 6, 17, 19, 20, 24]. Re-tethering developed in one of our male patients 6 years postoperation, so the re-tethering rate in our patients was only 3.2%. This patient underwent an untethering procedure and partial surgical resection of his lipoma. His case shows that laminoplasty is effective in protecting the thecal sac and nerve roots from fibrosis and granulation tissues.

Pre- and Postoperative Urodynamic Studies

The urodynamic test has value in predicting most of the future deteriorations in adult OSD patients. Adults diagnosed incidentally with OSD after moderate pain were followed up closely. When their complaints started to become symptomatic and urological complaints were added to their clinical pictures, urodynamic studies were re-performed, and the surgical intervention decision was taken.

We recommend surgery for adult OSD patients whose urodynamic test shows an overactive detrusor muscle. Postoperative urodynamic tests show no improvement in those patients. The long-term symptoms can be held responsible for this. On the other hand, when the urodynamic tests showed postvoid residual urine >100 ml, especially in patients with short-term (under 6 months) symptoms, postoperative urodynamic tests showed improvement in all 12 patients (either normal or close to normal residual urine: 50–100 ml) ($p = 0.0001$).

Complications of Adult OSD

Despite the good results obtained in almost all surgical interventions for symptomatic OSD in adults, tethered cord releasing is a complex procedure and has serious complications. Therefore, it is suggested that surgery be planned according to the dominant symptoms, with full neurological examination, craniospinal MR imaging, and urodynamic tests.

Over the 12-year period of our experience, surgery-related complications included three out of 31 (9.7%) patients suffering CSF leakages, three (9.7%) suffering surgical site infections (SSI), and one (3.2%) suffering a late pseudomeningocele. This

pseudomeningocele developed in a female patient 4 years after untethering surgery. All of these patients were successfully treated either conservatively or surgically. Two of our 31 patients (6.5%) complained of worsening muscular weakness. Iskandar et al. [17] reported that one of their 34 patients (2.9%) suffered CSF leakage, 5 (14.7%) suffered pseudomeningocele, 2 (5.9%) complained of worsening bladder dysfunction, 4 (11.8%) experienced persistent pain, and 1 (2.9%) complained of worsening pain postoperatively.

In the adult population, the rate of re-tethering is reportedly as high as 29% [5, 6, 17, 20, 24]. Usually recurrent TCS can lead to the same significant complaints seen in patients before primary surgery. Re-tethering was observed in one (3.2%) of our patients, who underwent an untethering procedure and complete resection of his lipoma surgically, and then improved. The patient was doing well on his postoperative 38th-month doctor visit. In this case, the authors noticed that laminoplasty was effective for protecting the thecal sac and nerve roots from fibrosis and granulation tissues. Solmaz et al. [18] reported a high re-tethering rate of 24.5% among children with TC.

To avoid serious complications, neurosurgeons have to study the craniospinal MRI carefully. If necessary, CT must be performed to investigate the bone structures. IONM and laminoplasty are also needed. To prevent re-tethering, lipomas should not be removed aggressively; the operation field should be kept clean, and CSF circulation between the neural elements should be preserved. Fibrin Sealant Products can be required for avoiding CSF fistula formation after tight closure of the dura.

Follow-Up of Adult OSD Cases

Patients with tolerable pain, which was seen in all of the adult patients with OSD in our series, were treated conservatively, with close follow-up (3-month periods) to preclude permanent new neurological deficits. Operated adult TCS patients were subjected to full neurological examinations on the 1st-, 3rd-, 6th-, 12th-, and 24th-month control visits. Subsequently, each patient was called every 2 years if he/she had no new complaint. The patients must be evaluated pre- and postoperatively according to a neurological scoring system [10] (Table 7.4). If the patient experiences the same symptoms as those presenting before surgery, lumbar MRI has to be performed.

Conclusions

OSDs are congenital deteriorations that can lead to serious complaints. Despite the good results reported after surgical interventions for symptomatic OSD in adults, surgical intervention such as tethered cord releasing is a complex procedure and has serious complications. Therefore, it is suggested that surgery be planned according to the dominant symptoms, with full neurological examination, craniospinal

imaging, and urodynamic tests. Laminoplasty (or hemilaminectomy), short-term (less than 6 months) symptoms, patients without lipomas, and presentation with moderate or mild symptoms seem to be reliable predictors for good surgical outcomes. Urodynamic study can be used as a predictive tool for diagnosing TCS in asymptomatic adult patients, and it could be a good predictor for disease prognosis.

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