Spinal Dysraphism

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31.1 Definition

Spinal dysraphism (SD) is an umbrella term that includes congenital midline neural tube defects. Midline closing of bone, neural, or other mesenchymal tissue is defective. Spinal dysraphism can be classified as open type (spina bifida aperta; SBA) and closed type (spina bifida occulta; SBO) dysraphism. Open-type dysraphism includes hemimyelocele, meningocele, myelomeningocele, and hemimyelomeningocele. Closed dysraphism includes lipomyelomeningocele, dermal sinus, diastematomyelia, slit notochord, tight filum terminale, myelocystocele, neurenteric cyst, and developmental tumors such as spinal lipomas (Table 31.1).

31.2 Physical Examination

Clinical symptoms of SBO are often due to tethering of the spinal cord. Tethering of the spinal cord may cause back pain, perineal sensation loss, myelopathy of lower extremities, incontinence, and neurogenic bladder (Video 31.4).

Spinal bifida occulta can be associated with a cutaneous stigma (hypertrichosis, dimple, capillary hemangiomas, or sinus tract; Fig. 31.1).

SD is a congenital spinal malformation, so its occurrence accompanied by other congenital system anomalies is not rare. The most common accompanying

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Table 31.1 Classification of spinal dysraphism

	Spina bifida occulta	
Spina bifida aperta	*With subcutaneous mass*	*Without subcutaneous mass*
• Myelomeningocele	• Lipomyelomeningocele	*Simple*
• Meningocele	• Lipomyelocele	• Spinal lipoma
• Hemimyelocele	• Terminal myelocystocele	• Tight filum terminale
• Hemimyelomeningocele	• Meningocele	• Dermal sinus
		• Persistent terminal ventricle
		Complex
		Disorders of midline notochordal integration
		• Diastematomyelia
		• Neurenteric cysts
		Disorders of segmental notochordal formation
		• Caudal agenesis
		• Segmental spinal dysgenesis

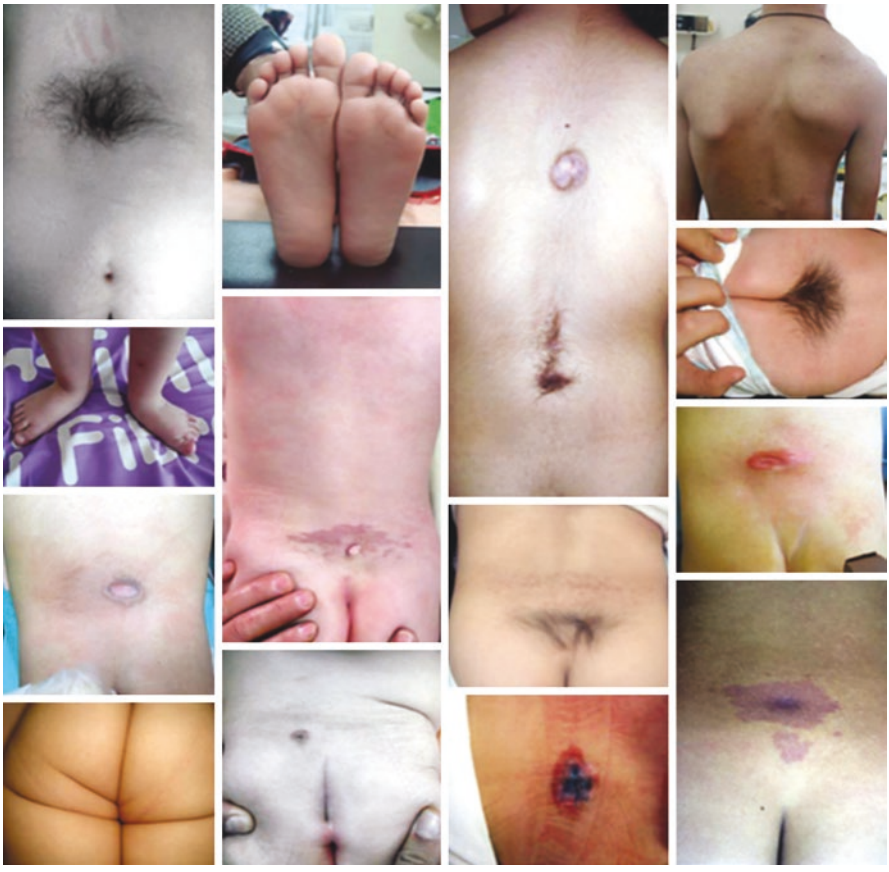

Fig. 31.1 Patients' photographs demonstrate different types of cutaneous stigmata and orthopedic anomalies associated with spinal dysraphism. (GUFM Division of Pediatric Neurosurgery archive)

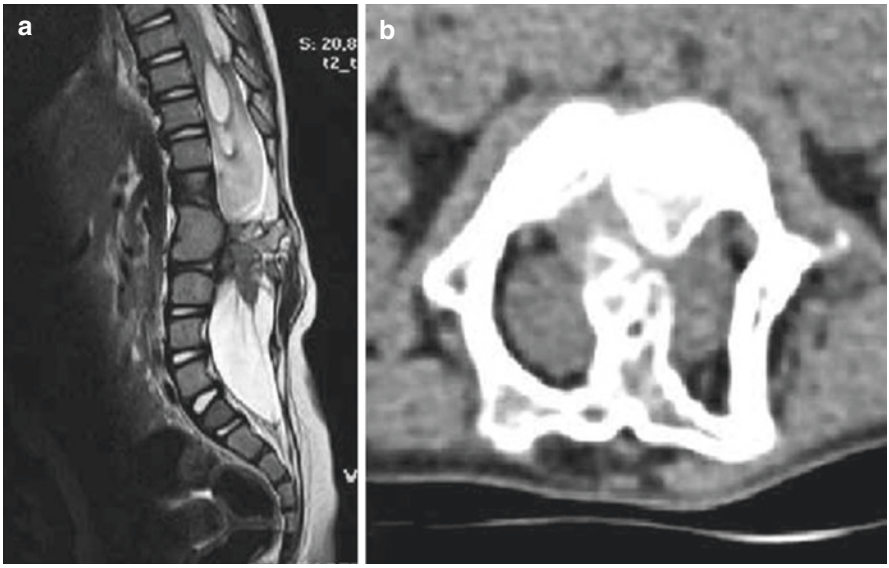

Fig. 31.2 (**a**) T2-weighted sagittal MRI images of an SCM patient. (**b**) Axial CT scan shows a midline osseous spur

congenital anomalies with SD are urologic problems. Studies showed that nearly 25% of SD patients have urologic pathologies. Also, different types of cardiovascular, renal, and skeletal (particularly in lower extremities) malformations are reported.

31.3 Imaging

Computerized tomography is helpful in the assessment of bony malformations (Fig. 31.2). Magnetic resonance imagining (MRI) is the gold standard radiological modality. Complete spinal and cranial MRI scanning is mandatory for these patients, because multiple congenital malformations are not rare in these patients.

31.4 Treatment Options

Surgical treatment is indicated as soon as possible for open-type spinal dysraphism. Local infection and central nervous system infection are the main potential risks for these patients. The main surgical aim is to close the spinal opening which exposes the neural tissue to infectious risks.

Clinical symptoms of occult spinal dysraphism are usually due to tethering of the spinal cord. Neurological deficits usually progress slowly and coincide with body growth. Surgical untethering is indicated as soon as possible for patients with neurological findings. Surgical treatment is aimed to release the tethering of the spinal cord.

There is a debate in the literature about the management of asymptomatic SBO as spinal lipomas, including lipomyelomeningocele. Some studies advocate early prophylactic untethering surgery. However, Kulkarni et al. reported follow-up results of conservative management of asymptomatic spinal lipomas. This study showed no statistical difference in the follow-up result of neurological deterioration between early surgery and conservative management.

Since most of the closed-type patients have subtle neurological problems, surgery in those patients puts them at great risk for further deterioration, so every precaution should be exercised during the surgery of those patients. Intraoperative neuromonitorisation (IONM) is the most important of those measures. Whenever possible, surgeons should rely on IONM both for their (for medicolegal aspects) and patients' safety.

31.5 Expected Outcomes

The most important prognostic parameter for symptomatic SBO is early diagnosis and treatment. Because severe neurological deficits may not be reversible after surgery. On the other hand, the most important prognostic parameter for SBA is the location of the defect and related level of the neurological deficit. Low-level lesion location and good spontaneous movement of lower extremities have a good functional outcome.

31.6 Potential Complications

Hydrocephalus is usually seen in SBA patients. CSF shunting is usually necessary. Other systemic disorders associated with SBA are vertebral deformities, genitourinary and gastrointestinal dysfunctions.

Spinal deformities, such as scoliosis, can be seen with spinal dysraphism. Scoliosis is the result of spinal cord tethering. So surgical correction of scoliosis without untethering the spinal cord will be a disaster.

Latex hypersensitivity is commonly seen in spina bifida aperta, especially in myelomeningocele patients. Frequent and early exposure to latex products is suspicious. Latex-free surgical instruments should be used to avoid latex allergy for these patients.

31.7 What Should Patient and Family Know?

Although surgical closure of SBA and surgical untethering of symptomatic SBO is usually necessary, spinal dysraphism is a congenital embryological disorder. No single intervention is expected to correct all of the problems in a particular patient. Anatomical, urological, cosmetic, and neurological problems require different management techniques and lifelong medical follow-up with a multidisciplinary team is

mandatory. Prevention plays an important role; in particular, folic acid and folates supplementation before conception have been proven useful to prevent congenital midline neural tube defects.

Further Readings

Guggisberg D, Hadj-Rabia S, Viney C, Bodemer C, Brunelle F, Zerah M, et al. Skin markers of occult spinal dysraphism in children—a review of 54 cases. Arch Dermatol. 2004;140(9):1109–14.

Pang D. Perspectives on spinal dysraphism: past, present, and future. J Korean Neurosurg Soc. 2020;63(3):366–72.

Copp AJ, Stanier P, Greene NDE. Neural tube defects: recent advances, unsolved questions, and controversies. Lancet Neurol. 2013;12(8):799–810.