Clinical Presentations of the Occult Spinal Dysraphisms

6

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Introduction

Occult spinal dysraphism (OSD) is comprised of a wide spectrum of closed congenital spinal anomalies, including lipomyelomeningocele, split cord malformation, neurenteric cyst, dermal sinus tract (DST), and tight filum terminale. These entities can occur in association with each other or even with open dysraphisms (myelomeningocele); that is, patients can have more than one expression of OSD. It should therefore come as no surprise that patients can present with an equally broad range of signs and symptoms. While some patients present at birth, others do not come to medical attention until adulthood. In this chapter, we will review the clinical presentation of patients with OSD (Table 6.1).

Table 6.1 Clinical presentation of OSD

Cutaneous stigmata
Motor weakness
Sensory disturbances
Orthopedic deformities
Urologic dysfunction
Infection
Pain

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Cutaneous Stigmata

Cutaneous abnormalities are present in 50–80% of patients with OSD and are frequently the reason for presentation for medical evaluation and investigation [1–6]. These stigmata are usually located in the midline and often occur near the level of the underlying intraspinal abnormality, which is usually in the lumbosacral region [2, 5]. It is not uncommon for a patient to have more than one cutaneous finding.

Focal hypertrichosis, commonly referred to as a hairy patch, is one of the most common cutaneous findings in OSD and is pathognomonic for split cord malformation. If hair is present, there is always some expression of split cord malformation. It may be mild, such as a duplicated central canal, but much more commonly, there is a true split cord. Focal hirsutism typically presents in one of two patterns, as a faun tail or silky down. A faun tail is a wide, usually triangular patch of coarse, terminal hair that can be several inches in length. Silky down refers to the presence of tufts of fine, soft, nonterminal hair or lanugo, often in a whorled pattern, which is usually limited to more discrete area in the midline (Fig. 6.1) [5, 7, 8].

A soft, non-tender, well-circumscribed subcutaneous mass is another common cutaneous finding (Fig. 6.2). The skin overlying these lesions may be normal or may exhibit another abnormality, such as hypertrichosis, hemangioma, or DST [5, 9, 10]. The vast majority of these subcutaneous masses are comprised of fat, which is seen in lipomyelomeningoceles, the most common type of spinal lipoma [9, 11]. In some cases, it can be quite subtle and go unnoticed without careful, deliberate inspection. In low-lying lipomyelomeningoceles, the mass may cause deviation or obscuration of the gluteal cleft. The majority is in the midline, but approximately one-third is paramedian in location. Asymmetric lesions are associated with a higher risk of neurologic deficit, typically in the lower extremity ipsilateral to the mass [2, 9, 11].

A small minority of subcutaneous masses are more fluctuant, which is seen in terminal myelocystoceles. These lesions are predominantly comprised of cerebrospinal fluid (CSF) but can also contain fat and neural elements and may swell with Valsalva maneuvers such as crying [10, 12]. Neuroenteric cysts can also occasionally present as a subcutaneous mass (Fig. 6.3).

Infantile hemangiomas are well-demarcated, vascular, typically raised lesions (Fig. 6.4). Flat capillary hemangiomas include port-wine stains, which are darker, purplish-red maculae with well-defined borders that can get darker over time, and nevus flammeus simplex, which are reddish-pink lesions with ill-defined borders [13]. A prospective study found that infantile hemangiomas >2.5 cm had a positive predictive value of 51.2% and a relative risk of 438 for OSD [14]. The association between flat capillary hemangiomas and OSD is less reliable than that between infantile hemangiomas and OSD [5, 13, 15, 16].

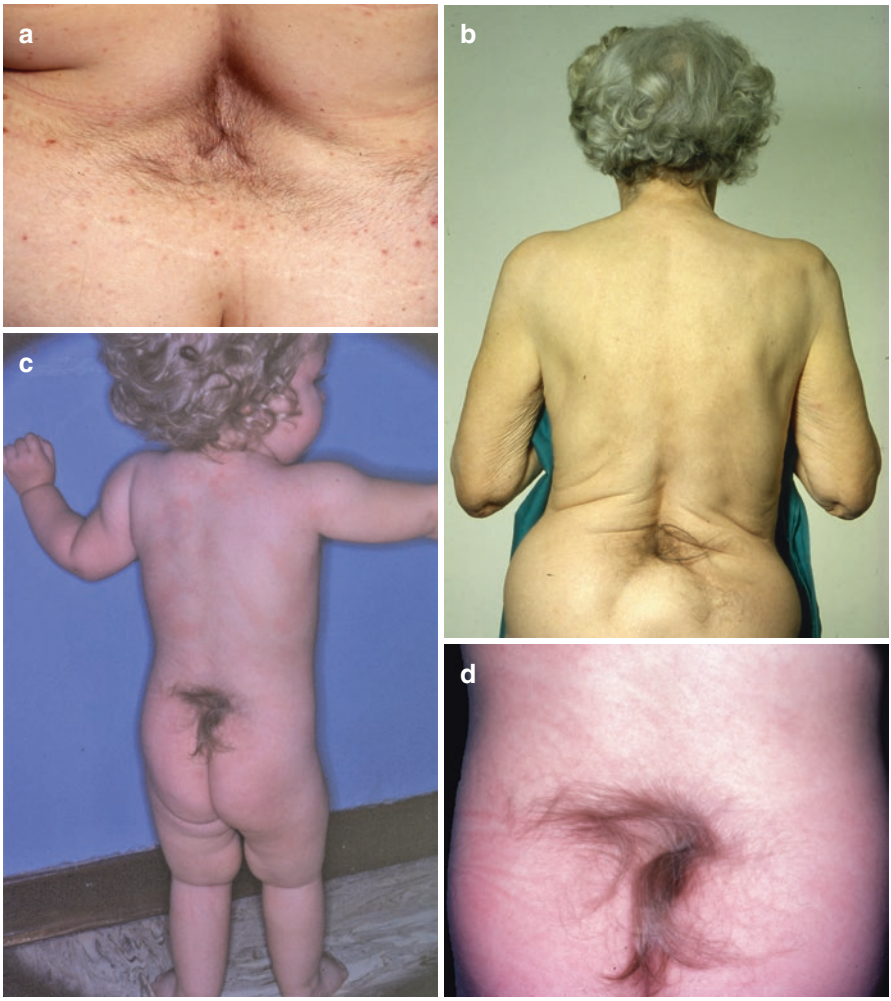
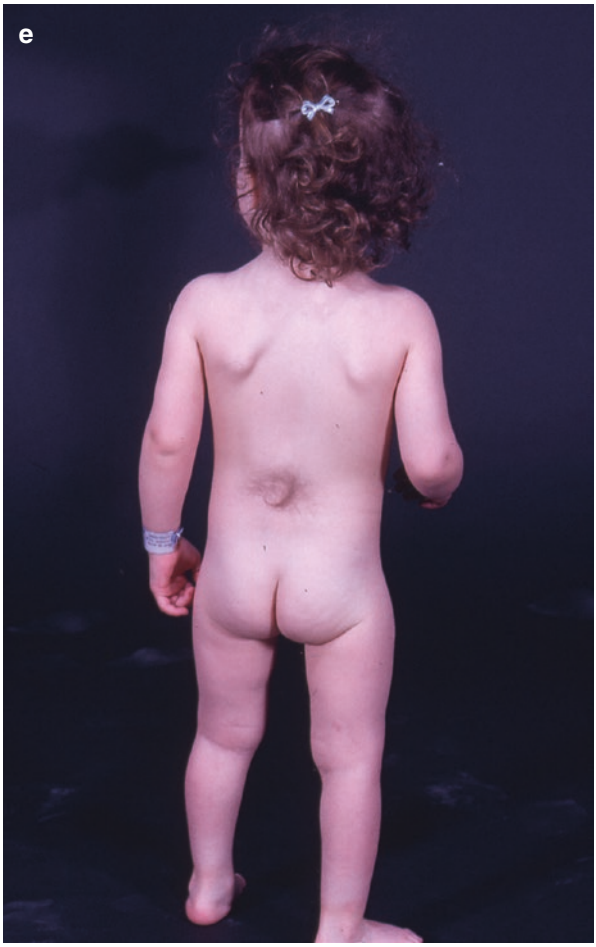


Fig. 6.1 (**a**) Adult with focal hirsutism in the lumbar region primarily in the midline. (**b**) Older adult female with evidence of a midline subcutaneous lipoma and focal hirsutism. This woman had draining osteomyelitis of her foot for more than four decades but had normal bowel and bladder function due to a split cord malformation with the lipoma involving only one hemicord. The other uninvolved hemicord maintained the neurological function of her bowel and bladder as well as of her other normally functioning leg. (**c**) Young child with typical focal hirsutism (faun tail) over the lower lumbar and sacral region. (**d**) Close-up view of the same child. (**e**) Another child with focal hirsutism at the thoracolumbar junction and normal neurological function

Fig. 6.1 (continued)



Hyperpigmented nevi and hypopigmented maculae have both been reported as being associated with OSD, but usually without further description of the lesions or histopathology [8, 17]. Aplasia cutis congenita is the congenital absence of the skin. It is typically found as a small, circular lesion in the vertex of the scalp, but several cases of aplasia cutis congenita in the midline in the lumbosacral region have been reported in association with OSD [18]. An atretic meningocele is a small area of dysplastic, thinned, scarified skin (Fig. 6.5). Sometimes referred to as a "cigarette burn" mark, it is often associated with an underlying meningocele manqué [13, 19, 20].

Caudal appendages, including skin tags, true tails, and pseudotails (Fig. 6.6), can also be seen in the presence of OSD [5, 17, 19]. A true tail, also called a persistent vestigial tail, is a midline appendage composed of skin, muscle, connective tissue, adipose tissue, blood vessels, and nerves. Usually located in sacrococcygeal region, it lacks vertebrae and is capable of spontaneous or reflexive movement.

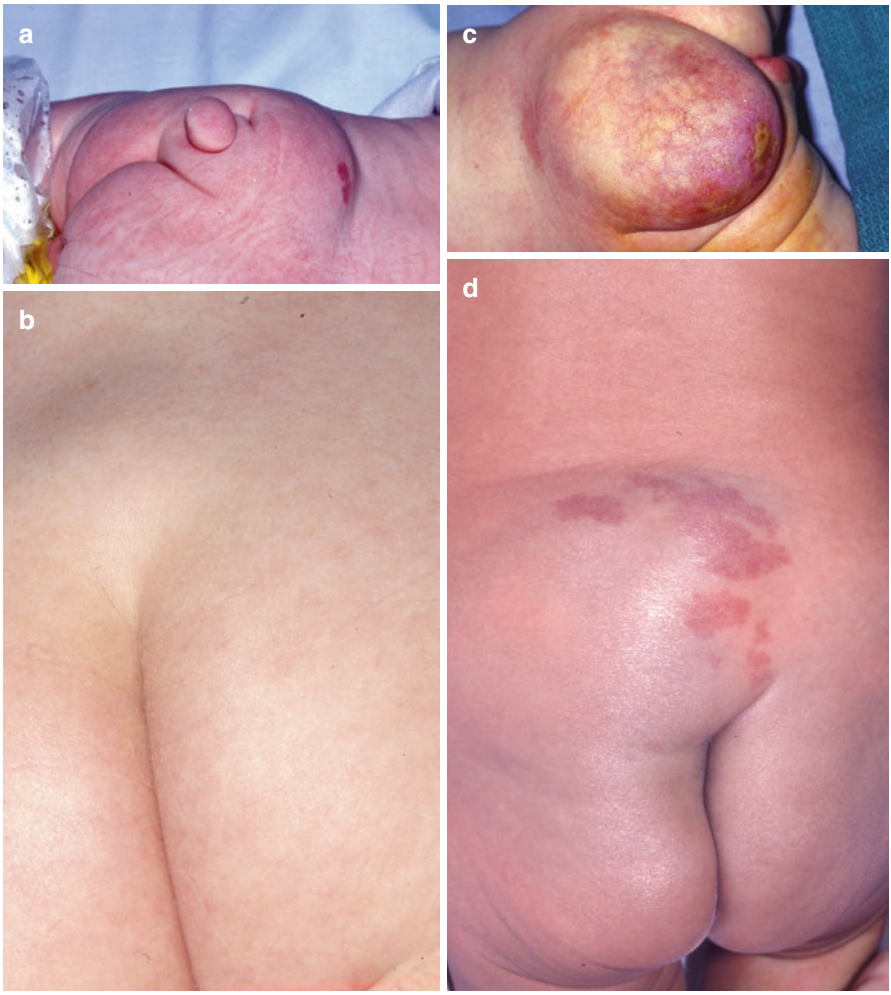


Fig. 6.2 (**a**) Neonate with a large lumbosacral lipoma and small capillary hemangioma on the superior right portion of the lipoma. Patient had normal neurologic and urologic function. (**b**) Subtle lipoma in an infant, again with normal neurologic function. (**c**) Subcutaneous midline lumbosacral mass which could be confused for a lipoma but in fact was a true myelomeningocele. There was little fatty involvement of either the neural elements or the surrounding dura. Shortly after this photograph was taken, the child developed a spontaneous CSF leak from the inferior left portion of the mass. The child was initially referred to the pediatric surgical service for a coccygeal teratoma. (**d**) Asymmetric lipoma of the lumbosacral region with superficial flat capillary hemangiomas and deviation of the gluteal crease because of the lipoma. The clinical exam in this child showed progressive loss of neurologic function of the left leg

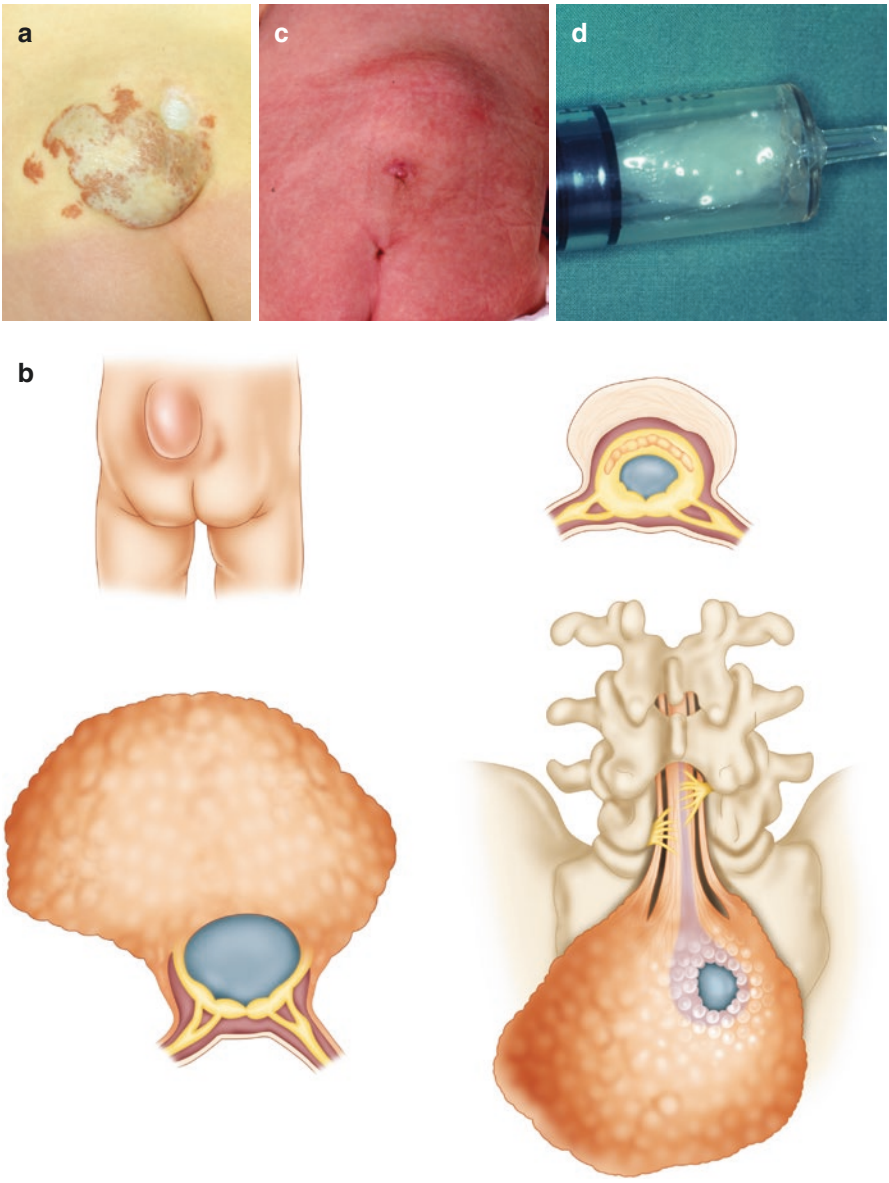


Fig. 6.3 (**a**) Newborn with subtle lumbosacral mass, superficial hemangioma, and a cleft over the dome of the lipoma. The cleft represents an area of dysmorphic skin overlying a CSF collection within the distal spinal cord typical of a lipomyelocystocele. (**b**) Illustration of the pathological changes of a lipomyelocystocele. (**c**) Infant with a midline lumbar mass superior to a dermal sinus tract. Although similar in appearance to a lipoma, this proved to be a subcutaneous neurenteric cyst. (**d**) Mucus obtained from the neuroenteric cyst at time of surgery

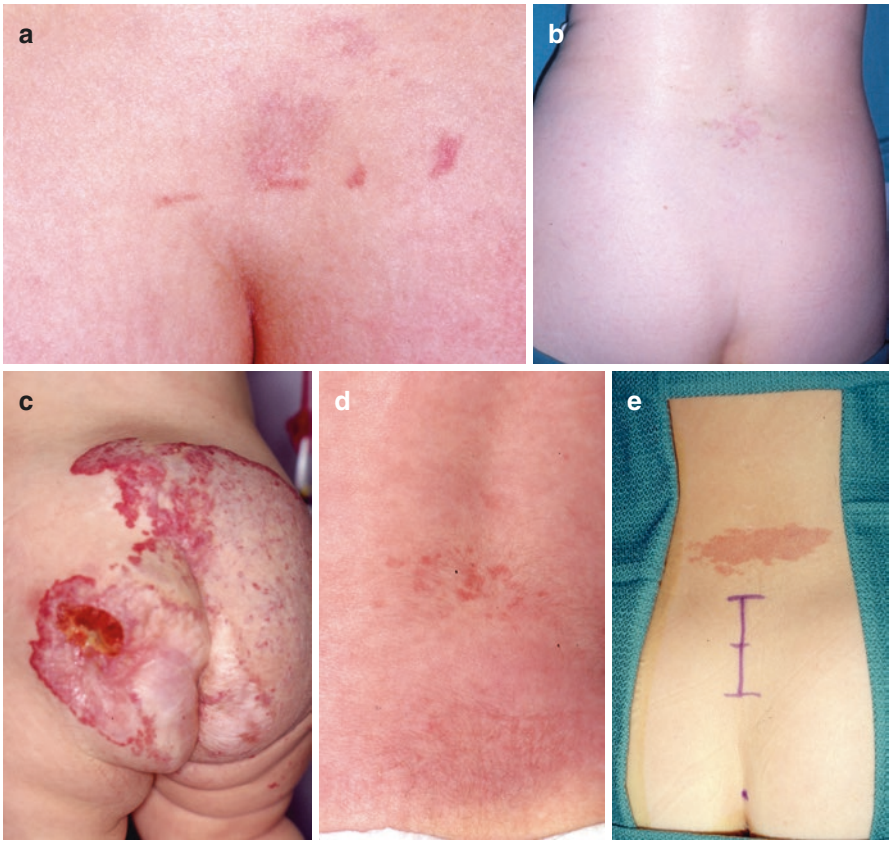


Fig. 6.4 (**a**) Subtle asymmetric flat capillary hemangioma in the lumbosacral region above the gluteal cleft. (**b**) More subtle hemangioma in the upper lumbar region in an adult with a thickened, taut filum terminale causing fixation of the distal spinal cord (tethered spinal cord syndrome). (**c**) Infant with a large lumbosacral lipoma and an overlying hemangioma in the process of involution. The superficial dermal layer has broken down in the inferior left quadrant of the hemangioma. Much of the velvety raised vascular aspect of the lesion on the right has involuted with time. (**d**) Spotty flat capillary hemangioma in an otherwise asymptomatic infant. (**e**) Broader flat capillary hemangioma in a symptomatic adult with tethered spinal cord syndrome

A pseudotail is composed of normal and abnormal tissues and is usually found in the coccygeal region. Often described as short or stump-like, it can include cartilage, fat, and teratomatous components. True tails occur more frequently in males, while pseudotails occur more frequently in females [5, 21].

A skin dimple or pit may indicate the presence of an underlying DST (Fig. 6.7). It is most often located in the midline anywhere along the spinal axis above the intergluteal crease, most commonly in the lumbosacral region. The dimple may be difficult to identify due to its small size, and intermittent drainage from the ostium is not uncommon. The underlying DST is composed of epithelial and fibrous tissue and typically courses rostrally as it goes deeper through the tissues [5, 13, 19, 22].

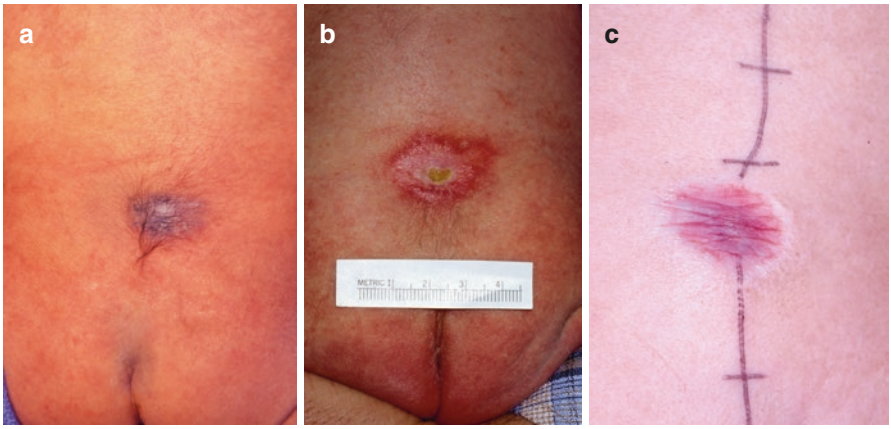


Fig. 6.5 (**a**) Neonate with a so-called cigarette burn over the mid-lumbar region. An area of hypertrichosis can be seen associated with the incomplete formation of the dermis. This lesion was so exquisitely sensitive to pressure that the infant was very uncomfortable lying on his back. (**b**) Newborn with a flat capillary hemangioma surrounding an area of incomplete dermis formation over the mid-lumbar region. This infant, too, had evidence of a thickened filum terminale and dural dehiscence directly under the central portion of the incomplete dermis. (**c**) Older infant with no capillary hemangioma but an area of distinct incomplete dermis formation, again associated with a thickened filum terminale. CSF is visible immediately under the thinned area of epithelium

At least 60% of DSTs terminate in the subarachnoid space. About half of all DSTs are associated with a dermoid or epidermoid cyst or inclusion tumor [23, 24].

It is important to distinguish the skin dimple associated with a DST from a simple coccygeal pit (Fig. 6.8). These congenital dermal sinuses are located below the level of the intergluteal crease. Present in 2–4% of the population, they are benign and do not require any further imaging or workup as they are not associated with OSD [3, 25].

Please refer to Chap. 4 for more detail and discussion on the cutaneous stigmata of OSD.

Infection

Infection may be the most serious and life-threatening clinical presentation in OSD and can be seen in patients with DST. The dermal sinus is a portal of entry for bacterial pathogens, which can result in bacterial meningitis. Aseptic chemical meningitis has also been reported secondary to spillage of epidermoid and dermoid cyst contents (desquamation of epithelium) into CSF spaces. In addition to meningitis, patients with DST can also present with subcutaneous, epidural, subdural, or intramedullary abscess formation or even infection of an associated inclusion tumor [22, 23, 26]. There has even been a reported case of patient who presented with a brain abscess as a manifestation of DST [27]. There should be a high level

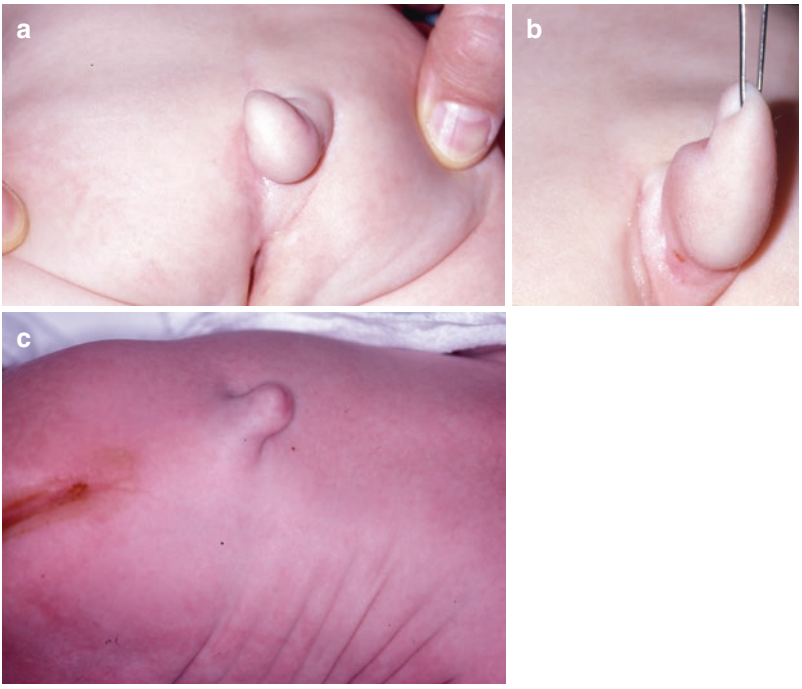


Fig. 6.6 (**a**) Asymmetric human tail in an infant with normal neurologic function but a low-lying conus secondary to a thickened and taut filum terminale. (**b**) A close-up view of the appendage. (**c**) Somewhat similar looking appendage from the lumbar region. However, this represents a skin-covered myelomeningocele rather than a human tail. Simple amputation of this will result in an increase in the neurologic deficit and CSF leak. A more formal myelomeningocele closure is necessary to address this lesion

of suspicion for DST in any young child who presents with aseptic meningitis or bacterial meningitis with an atypical organism, especially recurrent bouts of bacterial meningitis.

Reported organisms cultured from infected DSTs include *Staphylococcus aureus*, *Staphylococcus epidermidis*, *Staphylococcus albus*, *Escherichia coli*, *Klebsiella*, *Proteus mirabilis*, *Proteus vulgaris*, *Bacteroides fragilis*, *Bacteroides vulgatus*, *Bacteroides melaninogenicus*, *Peptostreptococcus*, and *Peptococcus magnus*. Multiple organisms may be identified in as many as 20% of patients [24].

Patients typically present with recurrent or persistent fever after appropriate treatment of other potential sources (such as otitis media, sinusitis, bronchitis, etc.), neck or back pain, purulent drainage from the tract, erythema and induration surrounding the ostium, and other meningeal signs [22, 24]. Rarely, CSF leakage has been reported [28]. Although all patients with OSD can develop progressive neurologic dysfunction secondary to tethering of the spinal cord (see section "Neurologic Symptoms" below), neurologic deterioration secondary to an infected DST, most commonly paraplegia, is usually relatively rapid [22].

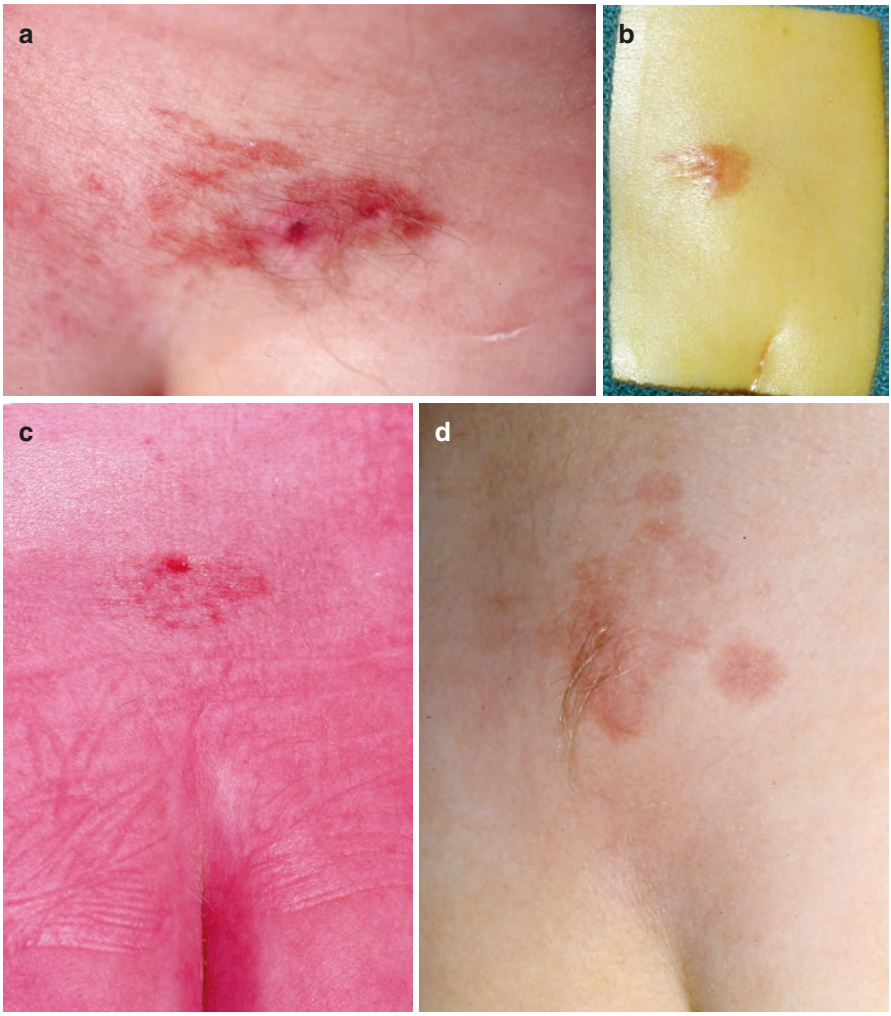


Fig. 6.7 (**a**) Dermal sinus tract with a paramedian opening to the right of the midline associated with a flat capillary hemangioma and limited but pathological hirsutism. (**b**) Asymmetrical dermal sinus tract to the left, again with a flat capillary hemangioma. (**c**) Midline dermal sinus tract in a child presenting with meningitis. (**d**) Flat capillary hemangioma and focal hirsutism in a midline dermal sinus tract. Note that all four of these lesions are above the gluteal crease

Recent series have indicated lower infection rates than older series, perhaps due to increased awareness of healthcare providers, especially primary care providers, about the cutaneous stigmata and sequelae of OSD. Although one recent series reported a 37.1% rate of meningitis, several other recent series report infection rates less than 15% [22, 23, 26, 29].

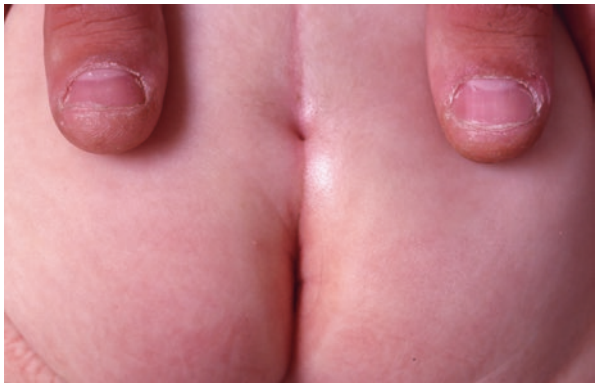


Fig. 6.8 Typical coccygeal pit located directly over the tip of the coccyx. The lesion is clearly within the gluteal crease and because of its location needs no further evaluation or consideration of surgical intervention. Lesions over the coccyx are not associated with surgically significant intradural pathology

Neurologic Symptoms

The natural history of OSD is one of progressive neurologic deteriorations. Upper motor neuron symptoms are due to tethering of the spinal cord. Pathologic fixation of the cord may result in repeated microtrauma to the spinal cord secondary to normal flexion and extension of the spinal column. It may also compromise the blood supply and oxidative neural metabolism to the spinal cord [2]. Lower motor neuron symptoms may be secondary to nerve root dysgenesis or local nerve root injury due to compression from a lumbosacral mass, such as a spinal lipoma, epidermoid or dermoid cyst, or abscess.

Neurologic deterioration tends to occur and progress during periods of growth as well as with repetitive flexion and extension movements of the spine. Symptoms are variable but can include progressive motor weakness, delayed gait development or gait deterioration, muscular atrophy, numbness, paresthesias, spasticity, and pain [1, 5].

Motor weakness is the most common symptom in the pediatric population, particularly in the lower extremities, and is often asymmetric. It may be accompanied by progressive muscle atrophy, which may be obscured by subcutaneous fat in infants. Patients may also develop increased tone and spasticity. Deep tendon reflexes can be hyperreflexic, normoreflexic, or hyporeflexic [1, 2, 5].

Sensory loss is less common and usually involves the buttocks, perineal region, and feet in a patchy, non-dermatomal distribution. Determination of altered or decreased pain appreciation in young children can be difficult, especially in the perineal area. Even experienced clinicians may misinterpret examination findings. Painless, trophic ulcerations can develop in these insensate areas. Nonhealing ulcers can lead to significant morbidity from additional complications such as osteomyelitis and toe amputations [1, 5, 30, 31].

Pain is much more prevalent in the adult population. It may be spontaneous, or it may be triggered by direct contact, activity, or trauma. Patients may experience

generalized low back pain, which may be exacerbated by progression of a scoliotic deformity. Tenderness of the subcutaneous mass is atypical in infant and children but can be seen in adolescents and adults. Pain in the lower extremities may be secondary to radiculopathy or may occur in a non-dermatomal distribution. Lhermitte sign has also been reported [9, 30, 32–34].

Hydrocephalus is a well-established sequela of infectious processes such as meningitis, ventriculitis, and/or arachnoiditis. It can also develop in association with intradural spinal tumors. Hydrocephalus in association with DST and/or dermoid tumors has rarely been reported. The mechanism for the development of hydrocephalus is not fully established but may favor a post-infectious or post-inflammatory phenomenon [22, 35].

Urologic Dysfunction

The progressive neurologic deterioration in OSD usually includes progressive bladder dysfunction. Urologic abnormalities tend to be more subtle than other symptoms but can range from a spastic, hypertonic bladder to a flaccid, hypotonic, overstretched bladder. Symptoms can include urinary frequency, urinary hesitancy, incomplete voiding, urinary incontinence, delay or regression in toilet training, and recurrent urinary tract infections. Subclinical findings on formal urodynamic testing can be an early indication of neurogenic bladder.

From a urologic standpoint, a flaccid bladder usually has a better prognosis than a spastic bladder, with less likelihood of upper tract damage. Detrusor hyperactivity leads to spasticity and hypertonia, causing elevated bladder filling pressures. Detrusor sphincter dyssynergia leads to elevated bladder-emptying pressures. This can result in a functional bladder outlet obstruction which can lead to vesicoureteral reflux, hydronephrosis, and, in severe cases, renal failure.

Chronic constipation is very common in patients with OSD. Bowel incontinence and sexual dysfunction are unusual and typically develop late in the progressive deterioration [1, 2, 5, 31, 36–38].

Please refer to Chap. 16 for more detail on the urologic aspects of OSD.

Orthopedic Deformities

The clinical presentation of OSD includes a wide spectrum of musculoskeletal abnormalities, predominantly involving the spinal column and lower extremities (Fig. 6.9). Sometimes referred to as the neuro-orthopedic syndrome, these deformities can be congenital, acquired, or iatrogenic. Congenital deformities include vertebral malformations such as hemivertebrae or congenital talipes equinovarus (CTEV), commonly known as club foot. Acquired deformities depend on the level of the neurological lesion but are frequently caused by muscular imbalance (i.e., flexors versus extensors). They may also develop as a result of habitual posturing or positioning. Some deformities may be iatrogenic, due to over or under correction of bony malformations or muscular imbalance [39, 40].

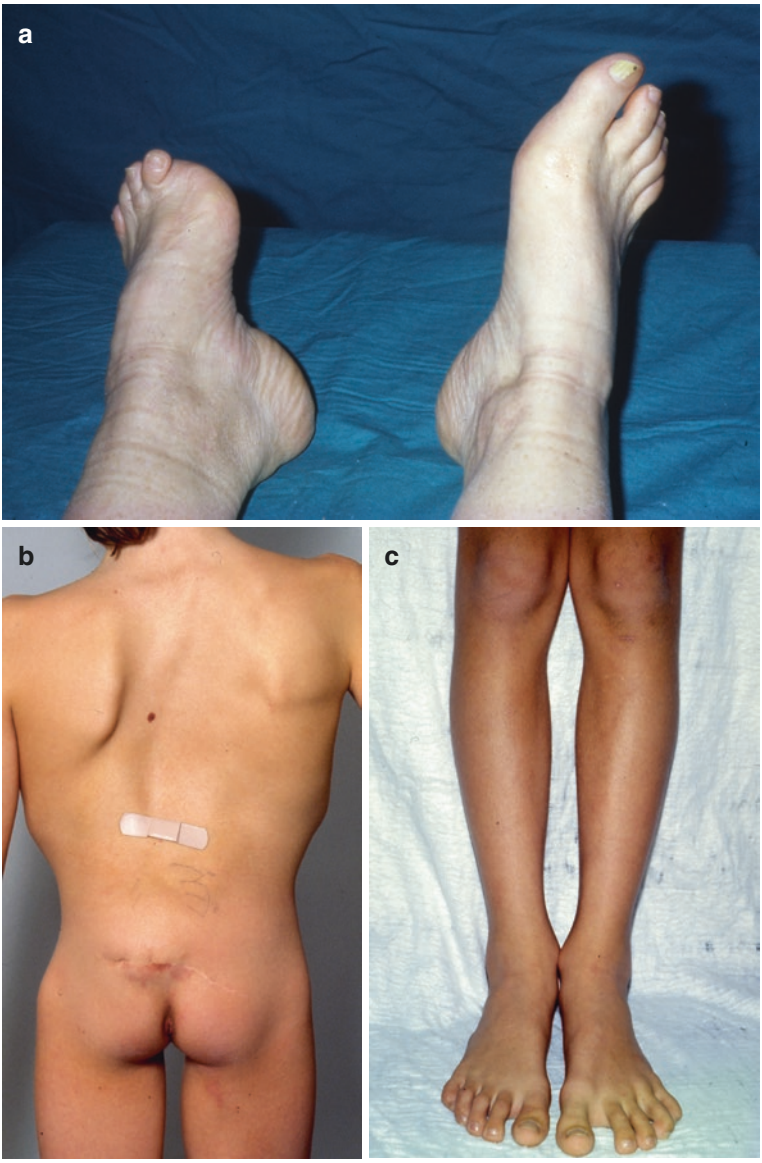


Fig. 6.9 (**a**) Elderly woman with split cord malformation who lost her left great toe from osteomyelitis because it was insensate. Note the high arch on the left compared to the right. Only one of the filum terminale was taut enough to produce a loss of sensory function causing the insensate left foot and the motor change resulting in the deformity of the foot. (**b**) Teenage male who had myelomeningocele repair elsewhere and had been followed in our spina bifida clinic for years. Note the well-healed small horizontal incision with loss of gluteal musculature. Over time, he progressively lost function in an asymmetric fashion. In point of fact, the cause of his decline was simple fixation of the distal cord from a tight filum terminale, not a myelomeningocele. (**c**) Minimally symptomatic child with split cord malformation with increased web space on the left foot as the only indication of a neurologic deficit. The patient also had no evidence of a neurogenic bladder at this point

Leg length discrepancy is one of the more commonly seen abnormalities. The abnormal, shorter lower extremity is usually weaker and may have an associated deformity. Foot deformities, including CTEV, pes cavovarus, pes cavus, calcaneus deformities, and vertical tali, are also frequently seen [39, 41, 42].

Scoliosis in OSD is multifactorial. While it is a manifestation of spinal cord tethering, other factors such as paraspinal muscular imbalance and vertebral anomalies such as segmentation anomalies, hemivertebrae, and butterfly vertebrae also contribute to the development and progression of scoliosis. Cord untethering may stabilize or even reverse the scoliotic deformity, particularly in patients whose scoliotic curves are <40° [13, 42–44].

Please refer to Chap. 16 for more detail on the orthopedic aspects of OSD.

Associated Syndromes

OSD has been associated with other congenital malformations and syndromes including anorectal malformations, Currarino triad, VACTERL (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities) association, and OEIS (omphalocele, exstrophy, imperforate anus, and spinal defects) complex [4, 13, 45–47]. Please refer to Chap. 19 for more details on syndromes associated with OSD.

Conclusions

The clinical presentation of patients with OSD is immensely variable. There is consensus in the published literature that the natural history of diastematomyelia and lipomyelomeningocele, while somewhat unpredictable, is that of progressive neurologic deterioration [48–53]. By analogy, this is thought to extend to all forms of OSD, a fact which is somewhat bolstered by the preoperative clinical histories of patients with these other entities. It is critically important to be able to recognize the clinical syndrome of OSD, however subtle or obvious, as timely surgical intervention can prevent or, in some cases, reverse the natural history of neurologic decline.

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