

### 2. Congenital dermal sinus with a tethered cord

Dermal sinus tracts (DSTs) are remnants of incomplete neural tube closure during embryologic development. The incidence is reported to be 1 in 2500 live births. Dermal sinus tracts can be associated with spinal fluid drainage, intradural dermoid or epidermoid cysts, and spinal cord tethering.<sup>1</sup>

Tethered cord syndrome (TCS) is characterized by low fixation of the spinal cord, with a conus situated below the L1–L2 intervertebral space.<sup>2</sup> Fixation might be caused by an intradural lipoma, a thickened filum, spinal cord tumour, diastematomyelia, or a dermal sinus, and creates traction on the spinal cord. In adults, accidents, deliveries, and sporting activities might also cause or precipitate symptoms of TCS. Stretching of the caudal end of the spinal cord can alter mitochondrial oxidative metabolism; this has been suggested as a possible pathogenesis for progressive neurologic, urologic, or orthopedic deficits.<sup>3</sup>

Several cutaneous lesions along the midline neuroaxis, such as hemangiomas, hypertrichosis, dermal sinus, subcutaneous lipoma, and aplasia cutis, are associated with occult spinal dysraphism and TCS.<sup>4</sup> They are mostly seen in lumbar regions (41%), followed by lumbosacral (23%), sacrococcygeal junction (13%), thoracic (10%), and cervical (1%) areas.<sup>5</sup>

In most patients with congenital dermal sinus (CDS), infectious complications such as recurrent meningitis, soft-tissue or subdural empyema, and infected dermal or epidermoid cysts prompt early diagnosis. Sometimes cells proliferating within the dermal sinus contain excess cholesterol crystals, predisposing patients to aseptic meningitis.<sup>6</sup> Inclusion tumours or cysts can also lead to neurologic signs of compression in patients with CDS.<sup>7</sup> Perilesional hypertrichosis and scarlike areas (as in the case of our patient), along with capillary malformations, often coexist and assist diagnosis. They can, however, exist separately.

### Diagnosis

In the differential diagnosis of CDS, coccygeal pits should come to mind first. They are located within the gluteal cleft and have no accompanying intradural pathology; deep tendon reflexes located above the gluteal cleft are often associated with intradural lesions.<sup>2</sup>

Radiologic imaging is the main diagnostic tool for all similar lesions in the midline lumbar and lumbosacral region. Magnetic resonance imaging best demonstrates the extent and depth of the sinus, reveals additional intradural pathologies, and can assist with surgical interventions if they are necessary.<sup>8</sup> In our case, a thick adipose tissue was revealed by the magnetic



resonance image, although it was not lipomatous and did not involve the spinal canal.

### Management

Management of CDS depends on the neurologic deficit. An asymptomatic CDS-related tethered cord by itself is not necessarily an indication for surgery, but risk of infection and possible further neurologic complications should be addressed.<sup>9</sup> In our patient, there was no association between the spinal cord and risk of intradural infection, so surgery was not performed. But most cases become symptomatic during the rapid-growth period of childhood, or after accidents, labour, and sporting activities in adults.<sup>8</sup> Children who have congenital tethered cords benefit from surgical untethering, as it prevents neurologic deterioration. In such circumstances, surgical treatment is recommended.<sup>9</sup>

All midline skin lesions in children should be carefully examined. Radiologic imaging and early diagnosis of lower back lesions facilitate swift surgical treatment and rehabilitation, so that any possible progression to neurologic deficit and bladder or bowel dysfunction can be avoided.<sup>6</sup>

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**Competing interests**  
None declared

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