Case Report

Benign coccygeal dimple or dermal sinus tract: a big difference

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Abstract

Dermal sinus tracts are a type of closed spinal dysraphism occurring with an estimated incidence of 1/2500. They are often misdiagnosed as a benign coccygeal dimple, thereby increasing the risk of meningitis and tethering of the spinal cord. This case report describes a 3-month old infant with bacterial meningitis due to a dermal sinus tract in the lumbosacral region. After appropriate treatment with antibiotics she underwent surgical excision of the tract with intradural exploration and untethering of the cord.

Introduction

Dermal sinus tracts are a form of spinal dysraphism present at birth at the dorsal midline with an estimated incidence of 1/2500 and most frequently occurring in the lumbosacral region. Failure to separate the neural ectoderm and cutaneous ectoderm (non-disjunction) between the 3rd and 5th week of embryonic life creates an epithelial-lined tract between the skin and underlying spinal cord. Dermal sinus tracts can end intradurally (60%) or in the epidural space (10-20%). When extending into the dura, they are frequently associated with thickened fatty filum (40%), intradural inclusion tumors like dermoid or epidermoid tumors (50%), tethered cord (79%), myelomeningocele, lipomyelomeningocele or diastematomyelia. This is in contrast to benign coccygeal dimples that occur in 4% of the population and are not associated with dysraphisms. Children with dermal sinus tracts are at risk of infection. Older children can present with tethered cord syndrome (1–4).

Case presentation

A 3-month old girl was referred to our emergency department with suspected meningitis. She presented with acute-onset irritability, fever up to 38.5°C, vomiting and subsequent lethargy. Clinical investigation revealed a pale, moaning and hypertonic infant, with a sacral dimple. The mother mentioned that the sacral dimple was noticed at birth but no further investigations were undertaken. Vital parameters were stable.

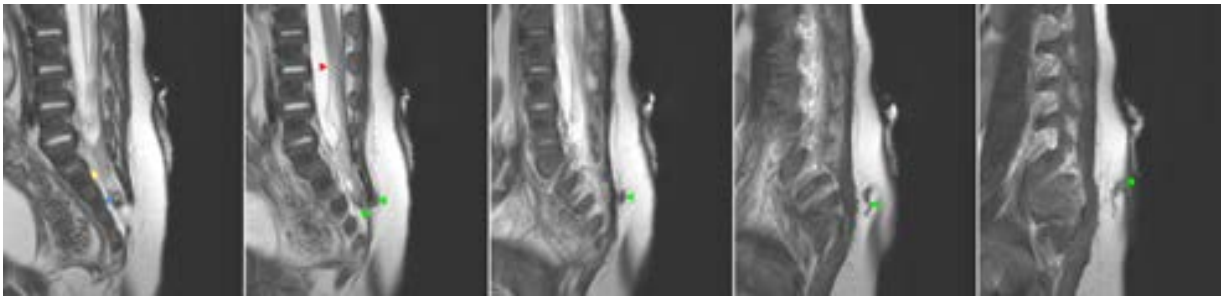
White blood cell count (WBC) was elevated to 27,6000/µL but C-reactive protein was low (1.6 mg/dL). Hemoculture was positive for Escherichia coli. Lumbar puncture showed WBC of 4371/µL, protein 1374 mg/L and decreased glucose (<2mg/dL). Cerebrospinal fluid cultures were positive for Escherichia coli, Bacteroides ovatus and Bifidobacterium breve (all commensals of the gut).

The diagnosis of a bacterial sepsis and meningitis was made. Broad spectrum antibiotics were started empirically. During her stay at the hospital clinical neurological examination was normal, except for initial but self-limiting axial hypotonia and head lag with pull-to-sit, likely secondary to the sepsis/meningitis. Neurological deficit of the lower limbs was never observed. The sacral dimple was located above the gluteal cleft, with no visible base. At presentation there was erythema around the sacral dimple with a small tuft of hair (Figure 1).

Imaging of the sacral dimple was performed because the clinical appearance suggested a dermal sinus tract. Ultrasound revealed a hypoechogenic trajectory reaching the spinal canal and suspicion of a low-reaching conus

Figure 1 : *Photograph of the lumbosacral region of the patient showing the ostium of the dermal sinus tract lying just above the gluteal cleft. This dermal sinus tract was associated with cutaneous stigmata; erythema and a small amount of hair. The bottom could not be clearly seen.*

Figure 2 : *Radiological findings. Adjacent T2-weighted (para)median sagittal images of the lumbosacral spine reveal the cutaneous opening (green arrowhead), which leads to a dermal sinus tract (green arrowheads), initially following a caudal course through the spina bifida of S3-5 just underneath the intact lamina of S2. There appears to be a dermoid tumor (blue arrowhead) at the junction of the dermal sinus tract with the thickened filum terminale (yellow arrowhead). There is a low-lying conus medullaris (L3-4 disc space; red arrowhead).*



medullaris with tethered cord. Additional magnetic resonance imaging (MRI) confirmed spina bifida of S3, S4 and S5, a dorsal dermal sinus with a cutaneous entry at the level of S3, with a typical U-shape on sagittal images, ending intradurally at level S3-4, likely on a thickened filum terminale. Along the intradural course of the tract, there was a dermal inclusion cyst. There was radiological tethering of the spinal cord, ending at level L3-4, but otherwise no central nervous system abnormalities (Figure 2). Further, there was a duplicated ureter with small kidneys.

After antibiotic treatment for the meningitis, she underwent surgical excision of the dermal sinus tract with intradural exploration, excision of the inclusion cyst, sectioning of the thickened filum and untethering of the cord (Figure 3). This procedure was performed under neuromonitoring (direct electromyogram with monitoring of the m. gastrocnemius (S1), abductor hallucis brevis (S2) and sphincter ani (S3-4)).

The abnormal skin opening, along with the dermal sinus tract, dermoid tumor and the thickened filum were excised. Pathologically, an epithelial lined lumen was detected, confirming the diagnosis of a dermal sinus tract (Figure 4).

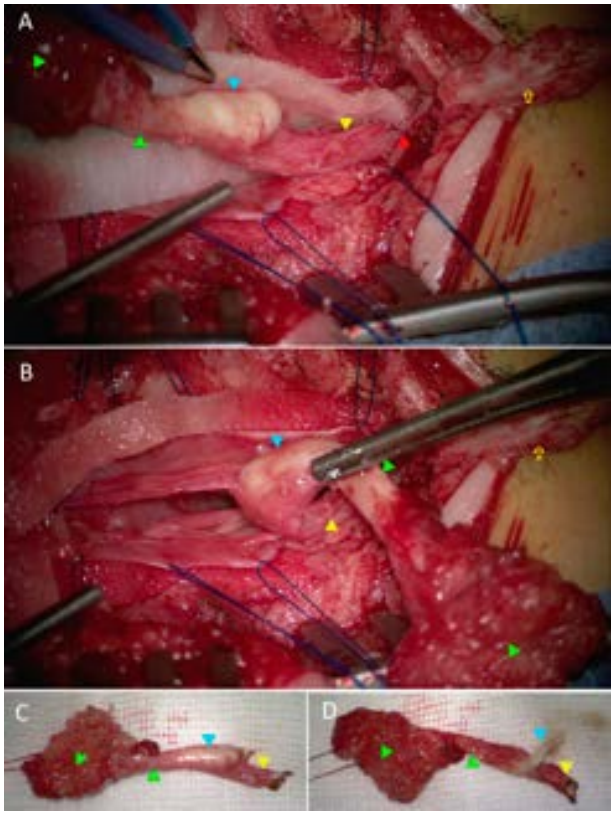
Discussion

Dermal sinus tracts can usually be distinguished from a benign coccygeal dimple on pure clinical grounds. Figure 5 show a benign coccygeal dimple of a newborn. Table 1 lists the differences. Benign coccygeal dimples are located within the gluteal cleft (1,2,4). Only atypical dimples (i.e. larger than 5 mm diameter, located more than 2.5 cm from the anus, not located on the midline and those whose base is not visible) are associated with a high risk of spinal dysraphism (5,6). Dermal sinus tracts are located above the gluteal cleft and are often associated with cutaneous stigmata such as erythema, skin tags, a small skin opening, pigmentation changes, hair or hypertrichosis, angiomata, subcutaneous masses, a human tail, local infection or inflammation (1,2,4).

Non-saccular limited dorsal myeloschizis (LDM) resemble dermal sinus tracts and they must therefore be distinguished from each other. A nonsaccular LDM has a tract without an epithelial lined lumen, consisting of mesodermal tissues. In contrast to dermal sinus tracts, non-saccular LDM are not associated with dermoid or epidermoid tumors. The patients are mostly older at diagnosis and do not have a history of meningitis. Non-saccular LDM are also associated with cutaneous stigmata different from those seen with dermal sinus tracts. The most common are pori and craters. Non-saccular LDM have a risk of tethering and in principle one could postpone surgery until these children are older. With a dermal sinus tract there is a risk of infection and later of tethering, so these children should be treated as soon as possible. Clinically and, radiologically, it is difficult to make a definitive distinction between both entities. Hence, we and others propose immediate surgery upon diagnosis (7–9).

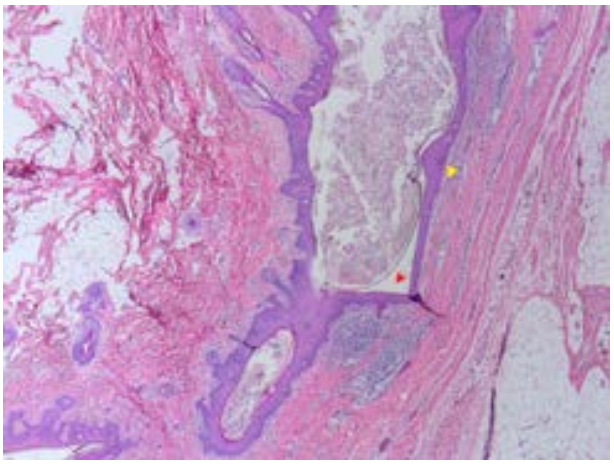
Definitive diagnosis of a dermal sinus tract or non-saccular LDM requires spinal MRI. When these are suspected, an ultrasound can already be performed. Ultrasound has a low sensitivity but high specificity (1,2). A retrospective study of 74 patients, who underwent an MRI preoperatively followed by surgery with intradural exploration, found that MRI even underreported intradural tracts and intraspinal inclusion cysts. MRI is therefore not sensitive enough to exclude these entities (10).

Figure 3 : *Intra-operative photographs. (A) and (B): The skin, subcutis and muscle are spread. The upper sacral and lowest lumbar laminae are deflected upwards (yellow open arrow). The dura is opened with blue-coloured tack-up sutures. The dermal sinus tract (green arrowheads) connects the skin (green arrowhead) with the thickened filum terminale (yellow arrowhead). At the junction between the latter, a small dermoid tumor can be seen as a pale cyst (blue arrowhead). The filum connects to the conus medullaris (red arrowhead). Several lumbar and sacral nerve roots are tightly attached to the thickened filum, likely due to preoperative infection. (C): The resected specimen includes the skin, dermal sinus tract, pale dermoid tumor and thickened filum terminale (same annotations as in (A) and (B)). (D): Upon incision of the dermoid tumor, keratin and a dark hair evacuate.*



Spinal dermal sinus tracts are noticed due to the presence of cutaneous abnormalities, an infection, a neurological deficit and/or orthopedic abnormalities. Children can be asymptomatic or may present with an infection for example meningitis, spinal or brain abscess. Patients can also present with recurrent meningitis or aseptic meningitis. Leakage of cerebrospinal fluid can also occur, increasing the risk of meningitis (11). The median age of infectious presentation is 2.5 years. The most frequent non-infectious presentation are neurological deficits (30%) such as abnormal reflexes, sensory changes, gait changes, altered bladder or bowel function or motor weakness of the lower

Figure 4 : *Photograph of a transverse section through the dermal sinus tract. The lumen of the dermal sinus tract (red arrowhead) surrounded by squamous epithelium (yellow arrowhead) is shown.*



limbs. Almost all children have normal neurological function at birth. With aging, neurological deterioration is more frequent due to increased tethering (1,2,4). Approximately 60-70% of adult patients with closed spinal dysraphism have urological symptoms (12). Orthopedic abnormalities including pes cavus, pes planus, clubfoot or scoliosis are also described later in life.

An infection, neurological deficits and orthopedic abnormalities can be prevented by referring children early for further investigation and treatment (1,2,4). Therefore it is important to make a correct diagnosis as soon as possible to avoid morbidity.

The most important risk associated with a dermal sinus tract is infection. When the diagnosis of a dermal sinus tract is made, urgent surgery is needed because of the increased risk of infection (2). In this case, the infant presented with a bacterial meningitis due to Escherichia coli, Bacteroides ovatus and Bifidobacterium breve. These are all commensals of the gut. Therefore there was a strong suspicion that the lumbosacral dimple was responsible for the infection.

Dermal sinus tracts can be associated with tethered cord syndrome. Traction induced neurological dysfunction is caused by mechanical tension or vascular stress on the spinal cord through growth and repetitive strain. This is the second most common presentation of a dermal sinus tract (1,2,4). In this case, the infant showed no symptoms of the tethered cord syndrome. A lumbar puncture before MRI should be avoided because of the risk of injuring the spinal cord.

Children with a dermal sinus tract have an increased risk of neurogenic bladder dysfunction. Therefore urological evaluation should be performed in all children. A retrospective study of 35 children who were treated for spinal dermal sinus tract reported that 77% had abnormal urological results. Urodynamic studies can detect urinary tract dysfunction before structural damage has occurred and is recommended (2).

We strongly recommend to always consider the possibility of a dermal sinus tract during postnatal clinical examination, during clinical examination of a child with recurrent meningitis, aseptic meningitis and a child with a neurological deficit of the lower limbs or orthopedic abnormalities. When characteristics are consistent with a dermal sinus tract (Table 1), the infant should be referred for further investigation.

Table I: *Distinction between dermal sinus tracts and benign coccygeal dimples*

Dermal sinus tract	Benign coccygeal dimple
Above gluteal cleft	Within gluteal cleft
Base not visible	Base visible
Associated cutaneous stigmata	No associated cutaneous stigmata
Can be associated with intradural pathology	Not associated with intradural pathology

Conclusion

Dermal sinus tracts are a type of closed spinal dysraphism involving significant morbidity if not diagnosed early. Often they are misdiagnosed as a benign coccygeal dimple. They occur most frequently in the lumbosacral region and are often associated with anomalies of the spinal cord. Dermal sinus tracts are located above the gluteal cleft, associated with cutaneous stigmata, the base of the tract is not visible and can be associated with intradural pathology. They can present as an infection, neurological deficit of the lower limbs or orthopedic abnormalities and should therefore be diagnosed and treated early.

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Figure 5 : *Photograph of the lumbosacral region of a newborn showing a benign coccygeal dimple. The dimple is located in the gluteal cleft and is not associated with cutaneous stigmata.*