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# Infant Rudimentary Meningocele with Tethering of the Cervical Cord: A Case Report

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## Abstract

Rudimentary meningoceles of the spine with dural extension are very rare and warrant surgical excision to prevent infection and long-term neurological deficits in pediatric patients. We present the case of a 5-month-old infant with a tethered spinal cord secondary to a rudimentary meningocele. The patient presented shortly after birth with a midline cervical dimple that was evaluated for a suspected dermal sinus tract. Magnetic resonance imaging scan of the spine showed a sinus tract with intradural extension to C2-3 and external opening at the level of spinous process C5. En bloc surgical excision and spinal cord release were successfully performed. Histological analysis of the specimen confirmed the presence of two blunt sinus tracts and staining was consistent with a rudimentary meningocele. Intradural rudimentary meningoceles in infants can successfully be managed with surgical intervention. Surgery is indicated to prevent future motor complications from spinal cord tethering and neoplastic growth from the rudimentary meningocele.

**Keywords**

- cervical dermal sinus tract
- rudimentary meningoceles
- tethered spinal cord

## Introduction

Rudimentary meningoceles arise from involution and scarring from incomplete neural tube closure during pregnancy. In the setting of spinal cord tethering, surgery is indicated to prevent future neurologic sequelae. Rudimentary meningoceles are rare congenital spinal malformations that typically present with solid cutaneous masses and carry the risk of future neoplastic growth.[1,2] This entity is typically present at birth with cutaneous deformities and is characteristically on the scalp or midline spine.[3] Intradural rudimentary meningoceles are infrequently reported and carry operative challenges that are not well established in the literature. To date, there have been three reported cases of intradural rudimentary meningoceles in the spine.[2–4] Here we present the surgical management of an asymptomatic infant with a rudimentary meningocele and tethered spinal cord.

## Case Report

### History and Preoperative MRI

The patient presented at birth with a midline cervical and posterior right shoulder dimple. The patient was born at 40 weeks, 4 days to a 29-year-old primigravida whose pregnancy was complicated by a prenatal ultrasound showing possible amniotic band syndrome. The patient underwent a total spinal ultrasound that was inconclusive for a dermal sinus tract.

At the age of 4 months, the patient underwent a total spine magnetic resonance imaging (MRI). The MRI revealed

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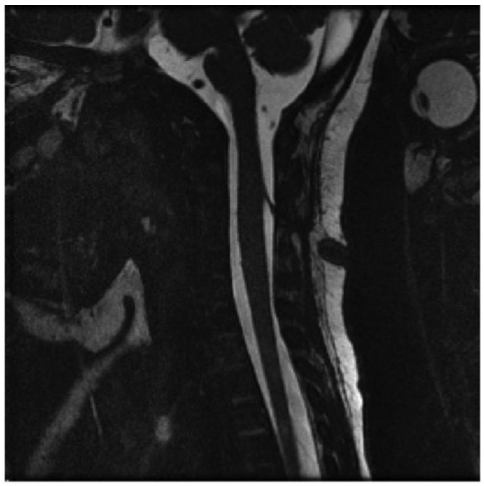

**Fig. 1** Preoperative magnetic resonance imaging scan. T2 sagittal scan shows dorsal dermal sinus tract and external opening at the level of spinous process C5.

what appeared to be a dorsal dermal sinus tract with intraspinal extension to C2-3 and external opening at level of spinous process C5 (►**Fig. 1**). There was a small ($5 \times 8$ mm) subcutaneous nodule in the soft tissue at the C5 level felt to represent an epidermoid cyst. The MRI also showed a hypoplastic C3-4 intervertebral disc with vertebral segmentation abnormality and spinal laminar nonfusion of C4 vertebra (►**Fig. 1**). The scan was negative for hydromyelia. We recommended surgical resection of the dermal sinus tract and suspected dermoid cyst and tethered cord release.

### Surgical Management

At the time of surgery, the patient was 5 months old and had a normal neurological examination. She underwent general anesthesia for excision of dermal sinus tract, en bloc resection of the presumed subcutaneous epidermoid and associated tract, and release of the tethered cervical cord via intradural exploration. At the time of surgery, the patient was placed in the prone position and somatosensory-evoked potential and motor-evoked potential neuromonitoring were initiated. An ellipse incision allowed for en bloc removal of the dermal sinus tract and exposed the intradural attachment (►**Fig. 2A**). The team performed C2-4 laminectomies to obtain adequate dural exposure and complete dermal sinus tract excision. A small tract remnant at the level of the dura was left and was tied off with a silk suture. The excised tract appeared bubbled in nature with a distinct white cutaneous nodule and was sent to pathology for evaluation (►**Fig. 2B**).

An operating microscope was used to facilitate the intradural microdissection for the remainder of the tethered cord release. The dura was opened caudally to the insertion point of the tract and extended cranially around this point to visualize the fibrous stalk that penetrated the dura and attached to the spinal cord (►**Fig. 2C**). The spinal cord was completely detethered and the dura was closed in standard running fashion. Neuromonitoring signals remained stable throughout the duration of the case. 11 days postoperatively, the patient underwent revision surgery for wound dehiscence and presumed cerebrospinal fluid leak, although no discrete leak site was observed. Cultures from the surgical site were negative. A lumbar drain was placed, and plastic surgery assisted with a two-layer muscle closure over the dura. The patient has had an uneventful recovery since.

### Histological Diagnosis

Histologically, the specimen had two epidermal openings that formed two blunt sinus tracts lined with squamous epithelium. The specimen had dilated irregular vessels forming a cord around fibrous tissue and epithelial membrane antigen immunostaining was positive for meningothelial cells. The findings were consistent with rudimentary meningocele.

## Discussion

Dermal sinus tracts are a rare spinal dysraphism that typically presents in approximately 0.04% of neonates with skin abnormalities, neurologic deficits, or infection.[5] Cervical dermal sinus tracts are rare (<1% of dermal sinus tracts) and surgical outcomes are widely absent from present literature.[5] Rudimentary meningoceles of the spine are exceedingly rare and only 18 cases have been reported; of these 18 patients, only three cases had intradural rudimentary meningocele extension.[1–4] Given the rarity of both cervical dermal sinus tracts and rudimentary meningoceles, there is a need to report surgical outcomes to inform future management of these conditions.

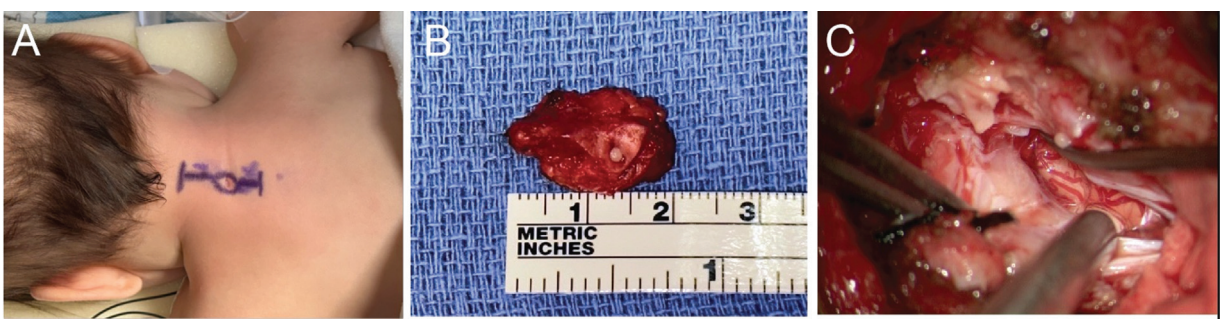


**Fig. 2** Intraoperative pictures. (**A**) Preoperative image of skin lesion. (**B**) Excised dermal sinus tract with prominent white nodule on skin surface. (**C**) Microsurgical intradural exploration and detethering.

 

Dermal sinus tracts and rudimentary meningoceles are thought to be of similar embryogenic origin and require surgical intervention.[3,5] The presence of dermal sinus tracts raises the risk of hydrocephalus, meningitis, and other infections.[5] Rudimentary meningoceles carry the risk of nerve damage and can manifest bladder and bowel dysfunction and muscle paralysis.[3] The rare nature of rudimentary meningoceles has complicated a definitive understanding of the origin and pathogenicity of this condition. Recent reports have questioned if rudimentary meningoceles carry the risk of neoplastic growth. A recent study has shown that a neoplastic cutaneous meningioma can grow within a rudimentary meningocele. Histopathological analysis in these cases ($n = 4$) reported meningeal whorl formations, psammoma bodies, and collagen bundles in addition to the meningothelial cells of the rudimentary meningocele.[2]

Current understanding defines rudimentary meningoceles as spinal dysraphisms and type I cutaneous meningiomas as neoplasms. Rudimentary meningoceles do not always have features of meningioma and are not synonymous with type I cutaneous meningiomas.[2] Our patient's histopathological analysis was negative for indications of cutaneous meningioma. Nevertheless, the risk of neoplastic growth from rudimentary meningoceles warrants surgical intervention to prevent neurological dysfunction and potential meningioma growth.

Tethered spinal cords have been reported in up to 63% of dermal sinus tract cases.[1,5] Tethered spinal cords are associated with sensory loss, motor weakness, and urinary tract dysfunction.[2] Although some cases may present asymptomatically, diagnosis of a tethered spinal cord requires prompt treatment to prevent future sequelae. In total, the risk of future infection, neoplastic growth, and sensory and motor deficits call for prompt surgical treatment of intradural rudimentary meningoceles in infants.

Presently, there are three reported cases of intradural rudimentary meningoceles in the spine that were surgically managed.[2–4] In a case similar to our patient, a 14-month-old female presented with a cutaneous skin dimple and MRI confirmed a cervical dermal sinus tract with spinal cord tethering.[3] In other related cases, a 9-month-old male presented with a congenital skin-covered hump in the thoracolumbar region with intradural attachment via vascular stalk and a 15-year-old male presented with a protuberant skin lesion that penetrated the dura and attached to the phylum terminale.[2,4] The second two cases were not associated with an identifiable tract and at presentation had outward skin protrusions from the meningocele. No cases of recurrence or neurological deficit after surgery have been reported. Taken together, these cases suggest surgical excision is an appropriate treatment option for intradural rudimentary meningoceles in the spine.

## Conclusion

Cervical dermal sinus tracts and rudimentary meningoceles are extremely rare spinal conditions. Surgical intervention can prevent future neurologic deficits. Here we present the case of an asymptomatic infant with a midline cervical dimple at birth. MRI confirmed the presence of a cutaneous tract with intraspinal extension. Our study supplements the current literature on intradural rudimentary meningoceles and reports the successful surgical management of this condition in an infant presenting with a tethered cervical cord.

### Authors' Contributions

RMS and JSH conceptualized and designed the study. RMS drafted the manuscript. JSH, SH, AL, DPF, HWP, and RGK critically evaluated the manuscript. RGK approved the final version to be published. All the authors agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

### Ethics

*Study approval statement*: This study protocol was reviewed and approved by the University of Pittsburgh Institutional Review Board.

### Consent

Informed consent was obtained for the publication of this case and associated images.

### Conflict of Interest

None declared.

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