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Case Report

Neurenteric cyst of the conus medullaris

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Abstract

Background: Neurenteric cysts (NECs) are rare developmental malformations of the central nervous system (CNS) which originate as benign congenital lesions. They originate from developmental foregut precursors, and are presumed to be the result of abnormal partitioning of the embryonic notochord plate. Such NECs predominantly arise in the cervical region in patients around 6 years of age or in their twenties or thirties. Notably, NECs of the conus medullaris are exceedingly rare, especially in patients of advanced age.

Case Description: A 70-year-old male presented with bilateral upper thigh and leg pain of over 20 years duration. His pain worsened over the past 3 years, and he sought surgical management. Although his neurological exam was normal, the lumbar magnetic resonance imaging revealed an intradural, nonenhancing, thin-walled, cystic lesion at L1/conus medullaris. The lesion was successfully resected without any adverse sequelae.

Conclusions: NECs are rare congenital legions that involve the spine. Here, an L1 intradural extramedullay neuroenteric cyst of the conus medullaris was resected without complications.

Key Words: Conus medullaris, neurenteric cysts, spinal cord tumor

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INTRODUCTION

Neurenteric cysts (NECs), known as enterogenous cysts or endodermal cysts, are rare developmental malformations of the central nervous system. As benign congenital lesions of foregut origin, NECs result from abnormal partitioning of the embryonic notochord plate and endoderm.[1]

Frequency

NECs represent 0.3–1.3% of all spinal cord tumors. They occur more frequently in males (66–73%), and are found most commonly in patients around 6 years of age or in those in their twenties or thirties.[2] NECs are typically associated with bone, soft tissue, and visceral abnormalities.[3] They are slow growing, and ultimately result in a progressive myelopathy.

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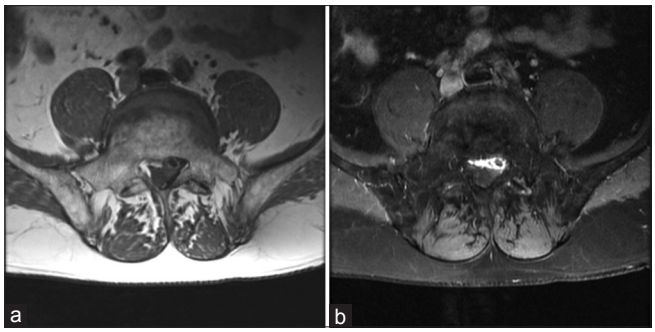

Figure 1:Axial MR images of a cystic lesion on the anterior portion of the spinal chord which demonstrates isolucency on T1-weighted MR images (a) and hyperintensity following gadolinium contrast on T2 weighted image (b)

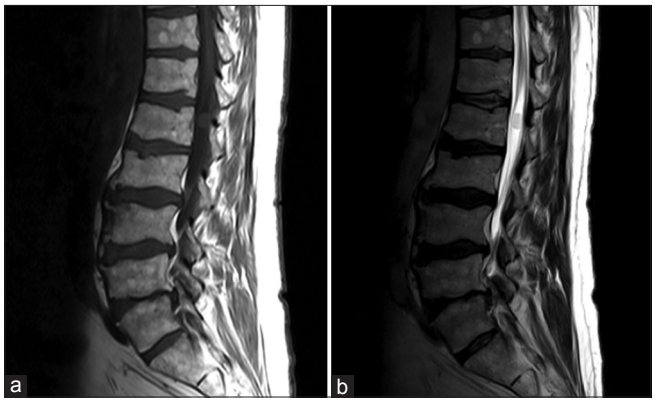

Figure 2: MR imaging revealing a non-enhancing cystic-appearing lesion growing off the conus medullaris at L1- L2 in the spinal with hyperintensity on a sagittal T1-weighted image (a) and hypointensity on a T2-weighted image (b)

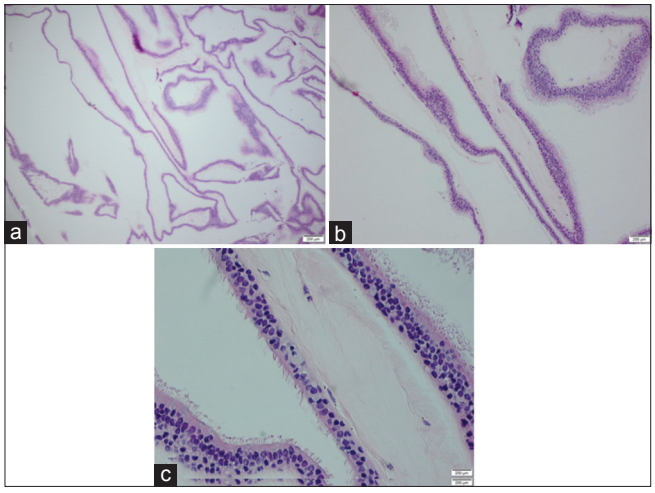

Figure 3: Microscopy images of the endodermal cyst revealing a simple ciliated columnar epithelium at 10x (a), 20x (b) and 40x (c) magnification

Localization

NECs predominantly occur in the lower cervical or cervicothoracic regions; 90% are intradural/extramedullary (IDEM), but 10% are either extradural or intramedullary. Here, we describe a very rare lumbar NEC (IDEM) involving the conus medullaris/L1 level in a 70-year-old male.

CASE REPORT

History, physical, and diagnostic work-up

A 70-year-old male presented with bilateral upper thigh pain of 20 years duration that progressed over the last 3 years. Although neurologically intact, the MRI documented an L1/conus medullaris intradural, nonenhancing, thin-walled, cystic lesion resulting in significant cauda equina compression. He underwent an L1–L2 laminectomy with intradural resection of two lesions that were adherent to the conus. Following careful resection/dissection, he remained neurologically intact, and was still asymptomatic 1 year later [Figures 1 and 2]. Pathologically, the lesions proved to be NECs typified by simple ciliated columnar epithelium [Figure 3].[4]

DISCUSSION

NECs typically occur in the cervical/cervicothoracic spine. Pathologically, they are lined by mucin secreting cuboidal or columnar epithelium of the intestinal or respiratory type. More than half of spinal NECs are associated with vertebral anomalies.[5] In the literature, there is only one case report of a true IDEM NEC involving the conus medullaris in that case there were no associated visceral or vertebral abnormalities.[3]

CONCLUSION

NECs are rare, congenital lesions of endodermal origin that most commonly occur in the cervical or cervicothoracic regions. Only rarely do they involve the conus medullaris where they are typically extradural/intramedullary in location. Here, a 70-year-old male presented with an IDEM NEC at the L1 level compressing the conus medullaris, which was successfully resected without complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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