

# True Intraspinal Neurenteric Cyst in the Lumbosacral Region

## —Case Report—

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### Abstract

A 56-year-old man presented with a very rare true neurenteric cyst in the conus medullaris without evidence of vertebral or visceral anomaly manifesting as a 6-month history of mild low back and bilateral inguinal pain. No motor weakness was found in the bilateral lower extremities. He had also suffered dysesthesia in the bilateral feet for several weeks before admission. Lumbar spine magnetic resonance (MR) imaging demonstrated a cystic intradural extramedullary mass at the L1-2 levels without enhancement after gadolinium injection. MR imaging, computed tomography, and radiography detected no vertebral anomaly. Lumbar laminectomy at the L1-2 levels was performed and the lesion was incompletely removed. Histological examination showed the cystic wall lined with ciliated columnar epithelium. Neurenteric cyst should be considered in the diagnosis of isolated cystic mass lesion at the lumbosacral region even in the absence of vertebral or visceral abnormality.

Key words: neurenteric cyst, conus medullaris, intraspinal cyst

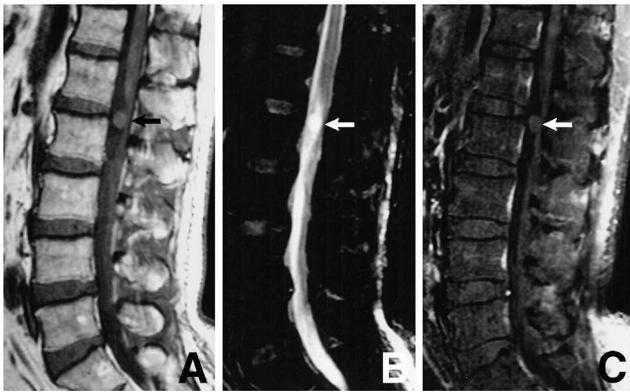
### Introduction

Neurenteric cyst is an unusual but well known developmental malformation occurring in the spinal canal or mediastinum.<sup>4)</sup> The embryological origin of neurenteric cyst is anomalous endodermal-neuroectodermal adhesion and abnormal separation of the endodermal cells in the 3rd week of embryonic life, leading to persistence of endodermal elements in the spinal canal.<sup>9,11)</sup> Persistence of the entire primitive neurenteric canal results in vertebral anomalies, whereas persistence of only part of the canal results in isolated intraspinal cyst.<sup>11)</sup> The most common location is the lower cervical and cervicothoracic regions.<sup>6)</sup> Neurenteric cyst is often associated with defects of vertebral bodies, allowing the spinal cord to interact with the alimentary canal.<sup>7)</sup> Isolated intraspinal enterogenous neurenteric cyst mainly occurs in the cervical region,<sup>6,7)</sup> and is usually associated with vertebral or visceral anomalies.<sup>6,8,9)</sup> We describe a case of true intraspinal neurenteric cyst located in the conus medullaris without evidence of vertebral or visceral anomalies.

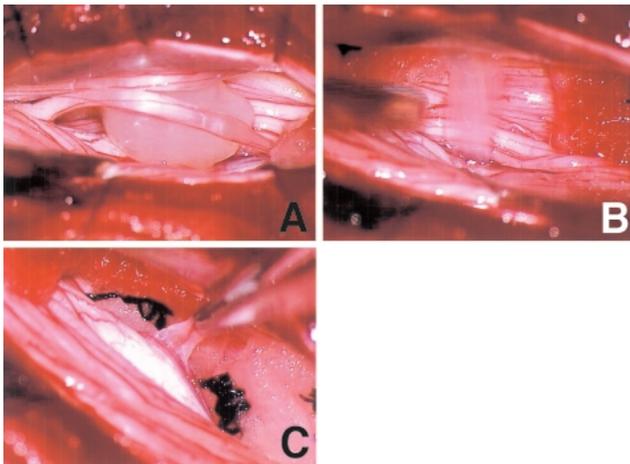
### Case Report

A 56-year-old man had been well without significant illness until 6 months before admission, when he noticed mild low back pain and bilateral inguinal pain. Neurological examination on admission found diminished bilateral lower extremity reflexes and dysesthesia in the bilateral feet, but no motor weakness in the upper or lower extremities. Physical examination found no abnormalities in the cardiovascular and respiratory systems. The abdomen was soft on palpation but no mass was found, and auscultation detected no abnormal sound. No subcutaneous mass or abnormal hair was detected in the lumbosacral region. Magnetic resonance (MR) imaging demonstrated an intradural mass in the ventral side of the conus medullaris at the L1-2 levels as a round and homogeneous isointense lesion without enhancement on T<sub>1</sub>-weighted images and as very hyperintense lesion on T<sub>2</sub>-weighted image (Fig. 1). MR imaging, computed tomography, and radiography detected no vertebral anomaly.

Total laminectomy was performed at the L-1 and L-2 levels, and the dura was opened. The cyst was identified in the cauda equina, with a thin membrane and filled with purulent-appearing material.



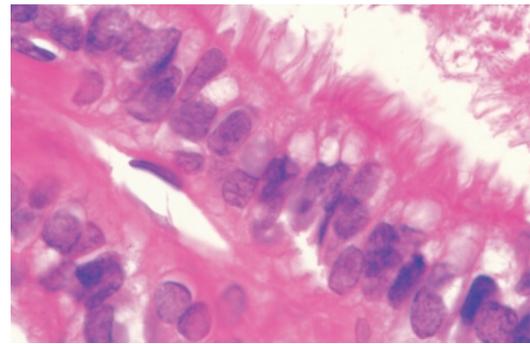
**Fig. 1** Preoperative sagittal magnetic resonance images showing a cystic intradural lesion in the ventral side of the conus medullaris at the L1-2 levels (arrow) as a round homogeneous isointense lesion (A) without enhancement (C) on the T<sub>1</sub>-weighted images, and as a very hyperintense lesion on the T<sub>2</sub>-weighted image (B).



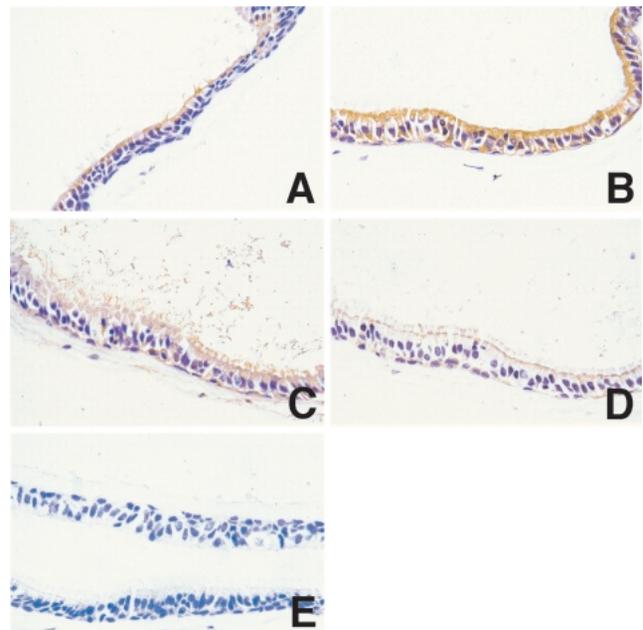
**Fig. 2** Operative photographs showing a cystic intradural extramedullary tumor with a whitish semitranslucent wall of the cyst before (A) and after (B) aspiration of the viscous content, and the cyst wall adherent to the conus medullaris (C).

The cyst was aspirated with a 21-gauge needle. The cyst wall was adherent to the nerve roots, so was removed incompletely to avoid possible nerve root damage (Fig. 2).

The patient recovered promptly from the operation, and his low back pain, bilateral inguinal pain, and dysesthesia in the bilateral feet improved rapidly. He had no neurological deficits at discharge.



**Fig. 3** Photomicrograph of the surgical specimen showing the cyst wall lined with ciliated columnar epithelium. Hematoxylin and eosin stain,  $\times 1000$ .



**Fig. 4** Photomicrographs of the cells lining the cyst wall showing positive immunohistochemical staining for epithelial membrane antigen (A), cytokeratin (B), carcinoembryonic antigen (C), and S-100 protein (D), but negative for glial fibrillary acidic protein (E).  $\times 200$ .

Histological examination showed that the cyst wall was lined with ciliated columnar epithelium (Fig. 3). Immunohistochemical examination revealed that the cells lining the cyst wall were positive for epithelial membrane antigen, cytokeratin, carcinoembryonic antigen, and S-100 protein, but negative for glial fibrillary acidic protein (Fig. 4). The final diagnosis was neurenteric cyst.

## Discussion

Only three cases of neurenteric cysts have been reported in the conus medullaris.<sup>6,8,9)</sup> One cyst was isolated and two were connected to presacral cyst or subcutaneous fistula. The case of isolated neurenteric cyst was associated with partial sacral agenesis.<sup>9)</sup> The present case showed no evidence of spinal or visceral abnormalities and no diverticulum. Therefore, neurenteric cyst should be considered in the diagnosis of isolated cystic mass lesion at the conus medullaris, even in the absence of concomitant vertebral or visceral abnormalities.

The natural history of untreated intraspinal neurenteric cyst is disappointing.<sup>10)</sup> Therefore, decompression of the cyst and excision of the membrane should be as complete as possible.<sup>2,10,11)</sup> Total resection is often difficult, as in our case, because of tight adhesion of the cyst to the neural tissue.<sup>1)</sup> However, even incomplete excision of the cyst wall usually produces excellent results.<sup>10)</sup>

Neurenteric cysts usually appear as non-homogeneous isointense to slightly hyperintense lesions without enhancement on T<sub>1</sub>-weighted images and hyperintense on T<sub>2</sub>-weighted images, although the T<sub>1</sub>-weighted signal intensity may vary depending on the protein concentration or hemorrhage within the cyst.<sup>1,11)</sup> The findings of MR imaging were consistent with these features in the present case, but the final diagnosis depends on the histological findings. The neurenteric cyst wall consists of ciliated or nonciliated columnar cells with secretory granules, containing mucin.<sup>3)</sup> Immunohistochemical staining of the wall is positive for cytokeratin, epithelial membrane antigen, and carcinoembryonic antigen, and negative for S-100 protein and glial fibrillary acidic protein, which confirms the endodermal origin.<sup>5)</sup> The findings in our case were consistent with these features except for positive immunostaining for S-100 protein, which has also been documented previously.<sup>5)</sup>

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