




An Unusual Case of Neurenteric Cyst in a Patient with Split Cord Malformation

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Abstract

Keywords

- ▶ neurenteric cyst
- ▶ enterogenous cyst
- ▶ split cord malformation
- ▶ diastematomyelia
- ▶ diplomyelia

Neurenteric cyst in a split cord malformation is a rare finding. We report an adult female becoming acutely symptomatic secondary to an expanding neurenteric cyst, though previous imaging had demonstrated stability. We discuss our workup and management with surgical resection and possible etiologies of her acute decline.

Introduction

Split cord malformations (SCMs) are rare congenital malformations. Two forms of SCMs have been described. Diastematomyelia (SCM type 1) is a malformation in which two hemicords in separate dural sacs are split by a bony or cartilaginous ridge. Diplomyelia (SCM type 2) is two cords within a single dural tube. Most patients are symptomatic, presenting with low back pain, weakness, scoliosis, and incontinence. Females are much more commonly affected than males.

Neurenteric cysts or enterogenous cysts are rare congenital abnormalities that may form from incomplete resorption of the neurenteric canal. A cyst may develop because of an abnormal connection between primitive endoderm and ectoderm in the third week of embryogenesis.¹ Males are much more commonly affected than females.

We present a case of SCM associated with the neurenteric cyst at the same level. To date, only nine cases have been reported.²

Case Report

A 63-year-old female presented to us with 5 days of progressive weakness, numbness, and urinary retention. Her history was remarkable for scoliosis with T8 butterfly vertebra, congenital fusion of T9–11, and a known cystic lesion in her thoracic spinal cord last imaged in 2015 and deemed stable from prior comparisons. She initially presented to an outside hospital with 1 day of urinary retention and was found to have a left ureteral stone for which a ureteral stent was placed. She was discharged home shortly after the procedure. Over the ensuing 3 days, she had progressive difficulty ambulating to and from the bathroom at home, prompting her return to the emergency room. She was flaccid in her right lower extremity with complete numbness. She demonstrated grade 4 power in her left lower extremity with decreased sensation to light touch. Imaging demonstrated a progression of her known cyst, which had grown from 4.5 to 5.5 cm in the craniocaudal dimension. The lesion now spanned T6–11 and measured 2.1 × 3.0 cm in its

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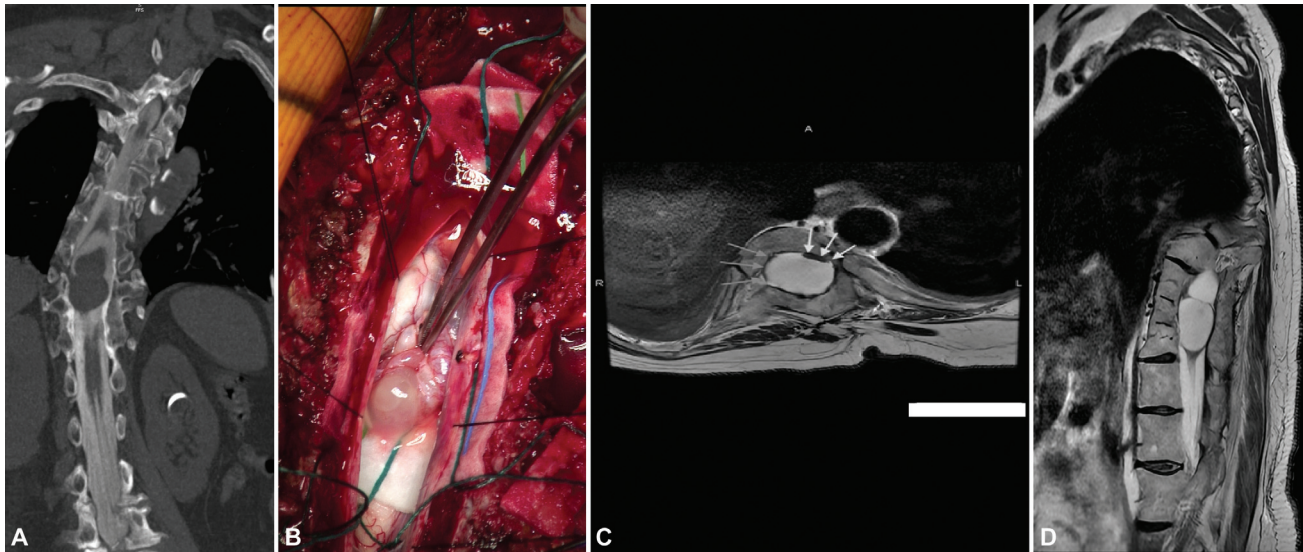


Fig. 1 (A) Maximum intensity projection of coronal computed tomographic myelogram demonstrating diastematomyelia of the lower thoracic spinal cord as well as expansile intradural multiloculated cyst, likely intramedullary in origin arising from the right hemicord and does not fill with contrast. (B) Intraoperative picture of intramedullary cyst. (C) Axial T2 magnetic resonance imaging (MRI) demonstrating compression of the two spinal cords, with right (gray arrows) more compressed than left (white arrows). (D) Sagittal T2 MRI of the spine showing compression of the left hemicord by the cyst and causing syrinx.

maximum anteroposterior and transverse dimensions with compression at T9–11, with no abnormal enhancement seen on postcontrast imaging (►Fig. 1).

After transfer to our institution for neurosurgical evaluation, she underwent cyst aspiration with improvement in her left lower extremity symptoms; however, she maintained significant right lower extremity weakness. She underwent T8–T12 laminectomies with intradural/intramedullary mass debulking and cyst fenestration. The cyst was found to be intradural and intramedullary with a very large extramedullary component. The cyst affecting the right hemicord was predominantly intramedullary in nature. Direct nerve root stimulation identified intercostal sensory nerves running along the dorsal wall of the cyst; no motor nerves were involved. The cyst wall structure and intramedullary component were resected freely and sent for permanent pathology and fluid analysis. Pathology returned consistent with a cystic lesion lined by a columnar epithelium with occasional mucin vacuoles and cilia. Focal squamous metaplasia was present. Immunostaining was performed, and the cells were positive for cytokeratin AE1/AE3 and negative for glial fibrillary acidic protein and S100 (ruling out an ependymal cyst). Mucicarmine stain was positive in the occasional mucin vacuoles. Overall, the findings were consistent with an enterogenous cyst.

Post procedurally her examination improved. She was full strength with appropriate coordination and intact sensation in her left lower extremity. Her right lower extremity improved to grade 3 power proximally with grade 2 power distally. She worked with physical and occupational therapy and was discharged to acute rehabilitation. At time of discharge, her right lower extremity had improved to grade 4 power proximally with grade 3 power distally. At 6 months follow-up, she endorsed persistent numbness in her right leg

though she was ambulatory with a cane with grade 4 power in right dorsiflexion but otherwise grade 5 power.

Discussion

SCM associated with a dorsally located neurenteric cyst in an adult female is very rare. To date, nine cases of neurenteric cysts associated with SCM have been reported, with three of the nine being in females, four being located dorsally, with only one other being in the thoracic region.² Further, less than 5% of neurenteric cysts have an intramedullary component,¹ with no other reports of intramedullary cyst associated with SCM. Her sudden onset worsening in adulthood also makes this case particularly unique. Neurenteric cyst volume instability has been linked to osmotic and hemodynamic factors such as production and resorption of mucin, venous stasis with thrombosis, and hemorrhage.^{1,3} Our patient had no signs of venous thrombosis or hemorrhage, and the time course of her symptoms makes the ebb and flow of mucin production less likely. A case of acutely enlarging enteric cyst has been described in pregnancy.⁴ The worsening was postulated to be secondary to diurnal increases in intraspinal pressure related to pregnancy and labor.

Our patient remained completely asymptomatic prior to her presentation for urinary retention to outside hospital. She walked three miles a day and denied any preceding abnormal sensation, or changes in bowel or bladder function up until the day prior to presentation to outside hospital. Her numbness and paraplegia developed over the course of 3 days following her initial presentation for urinary retention. Taken together this may suggest that another factor led the cyst to grow rather than the cyst being the initial inciting factor. Given the presentation of urinary retention with ureterolithiasis status post-stent placement at outside

hospital, this seems to be the inciting factor. Valsalva as might be predicted in urinary strain is known to increase intraspinal pressures,⁵ which may have led to the development of symptoms in the setting of mass lesion. However, it is possible that acute urinary retention from spinal cord compression led to stasis predisposing to ureterolithiasis.

Her acute neurological decline necessitating surgical intervention in the setting of SCM and neurenteric cyst with significant intramedullary component make this case particularly unique.

Conflict of Interest

None declared.

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