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Lumbar Intradural Neurenteric Cyst: A Rare Pathology in an Unusual Location

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A previously healthy 48-year-old female presented to the emergency department with a 2-week history of low back pain, progressive lower extremities weakness and right leg numbness. There were no bowel or bladder dysfunction symptoms. Spine MRI showed an intradural cystic lesion dorsal to the spinal cord at the level of L1 measuring 1.6 x 2.1 x 4.1 cm, which was T1 hypointense and T2 hyperintense, with a small soft tissue component and no gadolinium enhancement (Figure 1). A small lipomatous component was also noted. There were no associated vertebral anomalies. The patient underwent a T12-L2 laminectomy and cyst resection, which was subtotal due to the cyst adherence to the conus medullaris. Histopathology showed characteristic features of a neurenteric cyst, with respiratory-type epithelium in the cyst wall (Figure 2). Eight months later, follow-up MRI showed no evidence of recurrence. The patient reported improved sensation in the lower extremities, however there was some residual weakness predominantly in the proximal hip flexors bilaterally.

Spinal intradural extramedullary neurenteric cysts are rare, making up 0.7-1.3% of all spinal tumours, with a propensity for cervical and upper thoracic regions.[1,2] Here we present a case of a neurenteric cyst in an uncommon location, at the L1 vertebral level. Neurenteric cysts are hypothesized to result from abnormal partitioning of the foregut endoderm from the notochordal plate during early embryogenesis.[1] Histopathologically, a neurenteric cyst is lined by cuboidal or columnar epithelial cells which may demonstrate features of the respiratory or gastrointestinal tract, such as cilia, mucous glands, and/or goblet cells.[2] This accounts for alternative terminologies in the literature including 'endodermal' cyst, or 'bronchogenic' cyst when lined by respiratory epithelium, such as in our case.

The differential for intradural extramedullary lesions is broad, with meningioma, schwannoma, and neurofibroma representing the most common lesions.[3] Non-neoplastic intradural extramedullary spinal mass lesions include lipoma, dermoid, epidermoid, and

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arachnoid cysts, and rarely neurenteric cysts.[2] The differential can sometimes be narrowed based on characteristic features on MRI, which is the modality of choice for distinguishing such lesions. The most common MRI finding for spinal neurenteric cyst is a homogeneous lesion that appears hypointense on T1 and hyperintense on T2, and does not enhance following administration of gadolinium contrast agent.[1,2,4] However, variable T1 and T2 intensity have been described depending on the protein content of the cyst. Cases reported in the literature also occasionally demonstrate a lipomatous component on MRI, as seen in our case.[4] Given the variable appearance and non-specific clinical presentation, histopathology is required for definitive diagnosis. Spinal neurenteric cysts may be associated with congenital vertebral anomalies such as Klippel-Feil, anterior spina bifida, butterfly vertebrae, and hemivertebrae.[5] While they are often indolent, spinal neurenteric cysts may become symptomatic with cord compression.[2,4] When possible, total surgical resection is the most effective management option to prevent recurrence.[1,6]

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Figure Legends

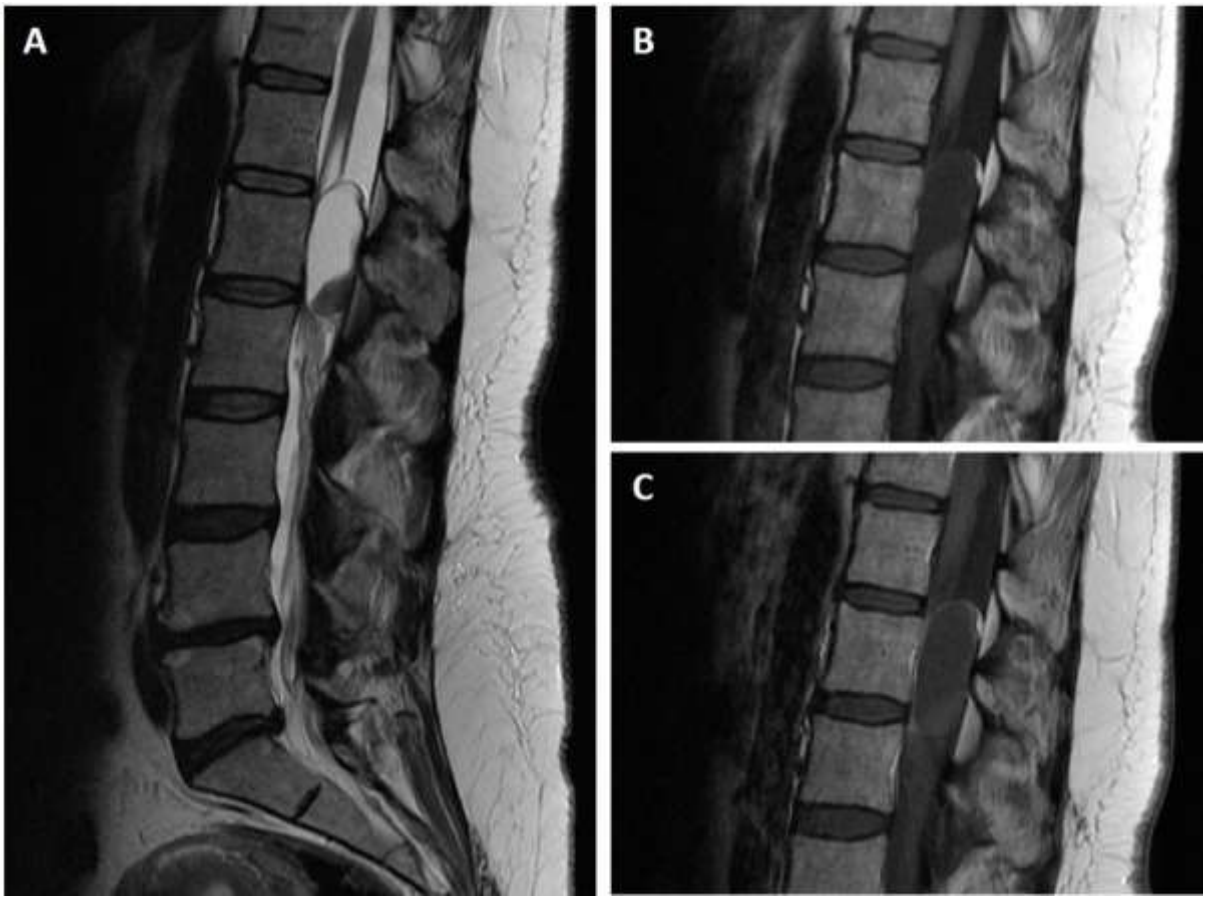

Figure 1. MRI images revealed an intradural lesion measuring 1.6 x 2.1 x 4.1 cm (AP x TR x CC) dorsal to the L1 vertebral body, demonstrating hyperintense signal on T2-weighted imaging shown in sagittal plane **(A)**. Comparison of pre-gadolinium T1-weighted imaging **(B)** and post-gadolinium T1-weighted imaging **(C)** demonstrated no enhancement of the lesion. A small soft tissue component is seen posteroinferiorly, and a tiny focus of fat with chemical shift artifact is noted on T2 at the postero-superior corner of the lesion **(A, B).**

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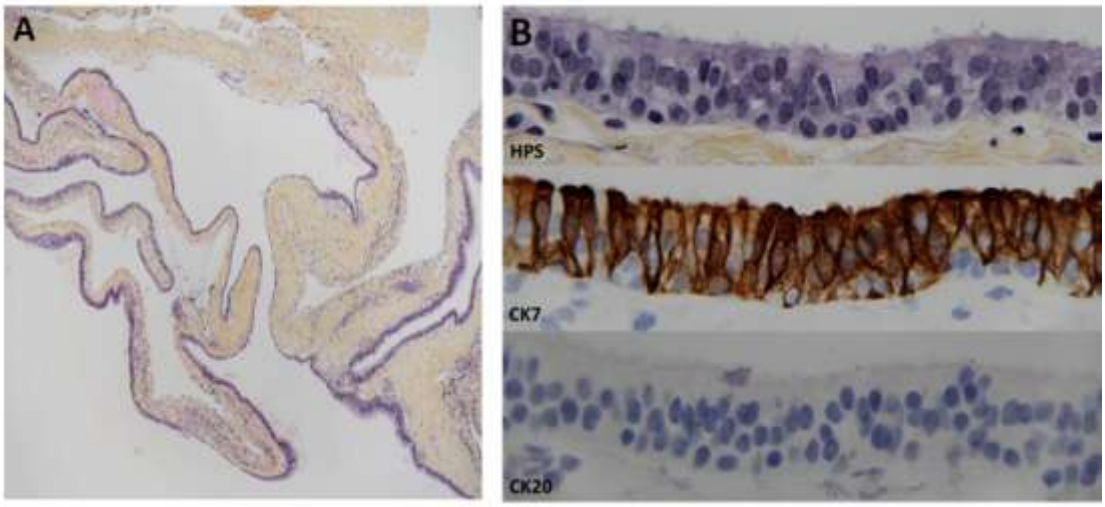

Figure 2. (A) Medium-power view of the partially resected cyst wall stained with Hematoxylin Phloxine Saffron (HPS). **(B)** Cyst wall demonstrating pseudostratified ciliated columnar epithelial cells (respiratory type), that are strongly immunoreactive for intermediate filament cytokeratin CK7 and immuno-negative for CK20. Given the respiratory type epithelium, the cyst could be termed a bronchogenic cyst.

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Conflict of Interest

The authors have no conflicts of interest or financial support to disclose.

Statement of Authorship

We confirm that each author has participated sufficiently in this submission, taking public responsibility for its content, and has approved this submission.

SB organized the images, performed the literature review, and prepared the manuscript. JPR reviewed the pathology and revised the manuscript for intellectual content. MCW revised the manuscript for intellectual content. OI reviewed the radiological findings and revised the manuscript for intellectual content. DT reviewed the radiological findings and revised the manuscript for intellectual content. BYMK designed and conceptualized study, reviewed the radiological images, and revised the manuscript for intellectual content.

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