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Case Report

Intramedullary enterogenous cyst of the conus medullaris presenting as lower limb pain

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Abstract

Enterogenous cysts account for 0.7-1.3% of spinal axis tumors. Cervical and thoracic segments are most often affected and they are rare in the lumbar region. Intramedullary variant which comprises less than 5% of enterogenous cysts are densely adherent to the surrounding tissue and preclude total excision. Partial excision is associated with recurrence and is the most common unfavorable outcome in these cysts. Hence, such patients need follow-up with serial imaging. We describe a case of conus medullaris enterogenous cyst presenting as lower limb pain. Due to dense adhesion of the cyst to the surrounding neural tissue, subtotal excision was done. The patient is symptom and tumor free at one year interval. We describe our case, discuss its uniqueness and review the literature on this rare but difficult to cure tumor.

Key words: Conus medullaris tumor, enterogenous cyst, intramedullary tumor, lower limb pain, neurenteric cyst

INTRODUCTION

The term enterogenous cyst was first used by Harriman to describe cysts previously known as neuroenteric, endodermal or respiratory cysts.[1] These cysts account for 0.7-1.3% of spinal axis tumors.[2] Enterogenous cysts are typically non neoplastic, intradural mass lesions occurring in childhood or early adulthood presenting with spinal cord or cranial nerve compression.[3] In the spinal canal, cervical and thoracic segments are most often affected and these cysts are generally very rare within the lumbar spinal canal.[4] Even though total surgical excision is the treatment of choice, it may be limited by adhesion to surrounding neural structures.[4] Partial excision carries a high risk of recurrence.[5]

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CASE REPORT

A 21-year-old male student presented to us with bilateral lower limb pain radiating from the gluteal region to the thigh and below (L1-S2) segment. The pain was of radicular nature and not associated with limb weakness. On examination the power in both lower limbs were grade 5/5 and there were no sensory deficits. Reflexes were normal. He had no autonomic disturbances (bowel/ bladder/sexual). Magnetic resonance imaging (MRI) showed a cystic lesion at D12- L1 with contrast enhancement of the wall. [Figures 1 and 2] Pre operative differential diagnosis included cystic variant of astroctytoma, ependymoma, neurofibroma, abscess or parasitic cyst.

A decision to operate the patient was taken. In prone position, D12- L1 laminectomy was performed. Midline durotomy revealed a partially cystic mass densely adherent to nerve rootlets. [Figure 3] The cyst was first aspirated with syringe and needle, following which the walls were dissected off the nerve rootlets. The cyst fluid was mucinous in consistency. At places, the mass was very densely adherent to rootlets and hence could not be separated. Near total excision of the cyst wall was performed. Hemostasis followed by water tight dural closure

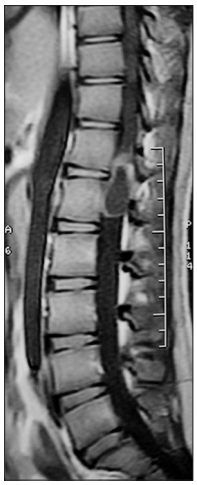

Figure 1: Post contrast sagittal image of lumbo sacral spine showing a cystic lesion at D12-L1 level with enhancement of cyst wall

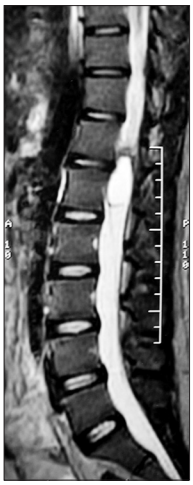

Figure 2: T2W sagittal image of spine showing a hyperintense cystic lesion at D12 – L1 level with hypointense cyst wall

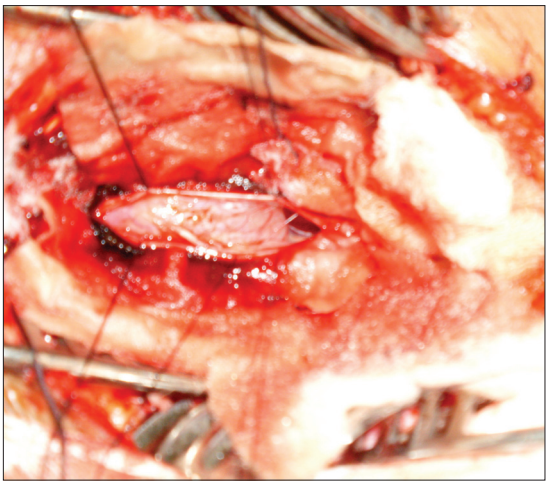

Figure 3: Intra-operative image after performing laminectomy at D12- L1 level. The lesion appears predominantly cystic but the walls are densely adherent to the surrounding nerve rootlets

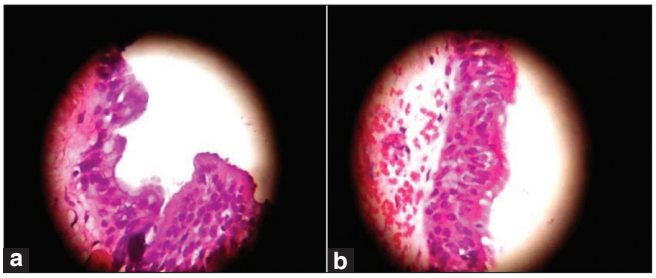

Figure 4: Histopathology image of cyst wall showing pseudostratified lining with goblet cells (a) and cilia (b)

was achieved. Patient was extubated on table and had no added neurological deficits. In the immediate post operative period, he had urinary retention which gradually improved with steroid cover. Histopathology revealed tissue lined by ciliated columnar cells and goblet cells with focal mucin secretion resting on fibro-collagenous base suggestive of enterogenous cyst. [Figure 4] At post operative period of one year, he is totally asymptomatic. Follow up MRI showed no recurrent tumor mass. [Figure 5]

DISCUSSION

Enterogenous cysts are considered congenital anomalies of endodermal origin generally localized in the mediastinum, abdominal cavity, and rarely the spinal canal or skull. They are also known as endodermal, neuroenteric, epithelial, bronchogenic, respiratory and foregut cysts. The general location for the intraspinal enterogenous cysts is in the subarachnoid space anterior to the spinal cord.[6] Approximately 90% are located in the intradural/extramedullary compartment, while the remaining 10% are divided between an intradural-intramedullary or extradural location. Less than 5% of lesions are located within the intramedullary compartment and that too mainly in the upper cervical and lower thoracic spine.[7]

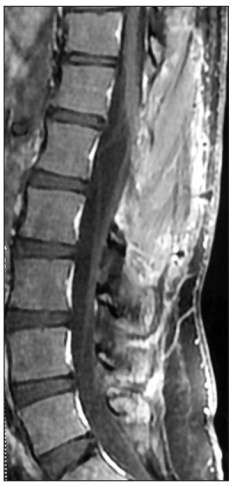

Figure 5: One year post operative, post contrast sagittal image of lumbo sacral spine showing no residual or recurrent contrast enhancing tumor

Individuals diagnosed with neurenteric cysts most frequently present in the second and third decades of life with a male-to-female ratio of approximately 2:1.[8] The majority of adult patients with neurenteric cysts present with progressive focal pain at the level of spinal axis pathology, fluctuating myelopathic signs, or radicular symptoms, depending on the size and location of lesion. The fluctuating nature of these symptoms have been attributed to cyst volumetric flux associated with periodic leakage of fluid content secondary to osmotic and hemodynamic factors.[5,9] Our case was unique in having radicular sciatica like pain without back pain.

Neurenteric cysts may be associated with bony abnormalities of the spine in approximately 50% of cases like spinal dysraphism, scoliosis, spina bifida, split cord malformation, hemivertebra, and Kippel-Fiel syndrome. In addition to disruption of the normal osseous architecture of the spine, neurenteric cysts can

be associated with malformations of the gastrointestinal tract, anal atresia, renal defects, cardiac abnormalities, and overlying cutaneous changes.[5] This was again absent in our case.

The most common MRI finding associated with neurenteric cysts are noncontrast-enhancing lesions that are isointense on T1-weighted sequences and hyperintense on T2-weighted imaging.[10] Similar to our case, Muzumdar *et al.* have described a neurenteric cyst with an abscess or granuloma-like presentation associated with peripheral enhancement of the cyst wall on MRI.[11] In a study by Kimura *et al.*, fluid-attenuated inversion recovery (FLAIR) sequence may show increased sensitivity for characterizing neurenteric cysts (lesion hyperintense to Cerebrospinal fluid).[10]

The diagnostic histopathology of neurenteric cysts has been described classically on hematoxylin and eosin (H and E) stained samples as a collection of mucin producing simple columnar or cuboidal ciliated and nonciliated goblet cells surrounding a central cystic cavity.[5] The mucus-secreting columnar epithelium of the neurenteric cyst wall has been described to be similar to that of the intestinal or respiratory epithelium. In most cases of enterogenous cysts like in our case, the wall often contains ciliated epithelial cells (unlike intestinal or respiratory epithelium). Langman postulated that ciliated cells may be displaced toward the evolving ventral spinal cord by the budding respiratory primordium.[12] Fabinyi and Adams suggested the origin for these cells from ciliated epithelium within the immature esophagus.[13]

Surgical resection is the first line of treatment for neurenteric cysts with the goal of neural decompression and prevention of cyst refilling.[4,5] When possible, total excision is the ideal outcome given the association between partial resection and cyst recurrence.[4] However, total resection of the intramedullary variant is generally not advocated as there is a high risk of damaging the spinal cord.[4,7] Operative interventions have ranged from cyst aspiration, subtotal resection with cyst marsupialization to cystosubarachnoid shunting.[4,7] Aspiration alone has been linked to recurrence and is, therefore, the least desirable intervention.[5] The utility of marsupilization and cystosubarachnoid shunting remains controversial. Several investigators cite cyst content leakage and associated central nervous system (CNS) irritation as evidence to avoid these techniques.[14]

The risk of surgical morbidity must be considered when assessing the goals of surgical resection.[5] Worsening of symptoms is documented in 11% of cases and failure to regain premorbid neurological function is cited in 18%.[15] Our patient had improvement of symptoms with no added neurological deficits. The most frequently reported suboptimal outcome is cyst recurrence. Reports of postsurgical recurrence have ranged between 0 and 37%. Our patient has been tumor free for a post operative period of more than one year. Partial resection remains the primary risk factor for the recurrence in neurenteric cyst management.[5]

Our case is unique in several respects: 1. intramedullary location (less than 5%); 2. conus medullaris (usual location is upper cervical or lower thoracic); 3. no vertebral anomalies (more than 50% are associated with anomalies); 4. absence of back pain (pain originated from gluteal region to the lower limb); and 5. MRI showed a cyst with contrast enhancing walls (usually no enhancement is seen) Our literature search revealed only one such similar case of lumbosacral/conus medullaris enterogenous cyst reported by Yunoki *et al.*[16]

CONCLUSIONS

Enterogenous cysts should be considered as a differential diagnosis in all cystic lesions of the spine. These lesions, when they appear in the intramedullary space, are densely adherent to the surrounding neural structures. Considering the possibility that the cystic lesion might be an enterogenous cyst, partial to subtotal excision should be kept as an option to be exercised after seeing the intra-operative condition. With a recurrence rate as high as 37%, patients with partial resection in particular should be followed with serial imaging to assess for recurrence of cyst.

REFERENCES

1. Harriman DG. An intraspinal enterogenous cyst. J Pathol Bacteriol 1958;75:413-9.
2. Fortuna A, Mercuri S. Intradural spinal cysts. Acta Neurochir (Wien) 1983;68:289-314.
3. D'Almeida AC, Stewart DH Jr. Neurenteric cyst. Case report and literature review. Neurosurgery 1981;8:596-9.
4. Arslan E, Cakir E, Kuzeyli K, Guvercin AR, Gazioglu G, Arslan EA, *et al.* Recurrent lumbar spinal intradural enterogenous cyst. A Case report. Turk Neurosurg 2010;20:402-5.
5. Savage JJ, Casey JN, McNeill IT, Sherman JH. Neuroenteric cysts of the spine. J Craniovertebr Junction Spine 2010;1:58-63.
6. Reinders JW, Wesseling P, Hilkens PH. Intramedullary enterogenous cyst presenting with spastic paraparesis during two consecutive pregnancies. A case report. J Neurol Neurosurg Psychiatry. 2001;71:528-30.
7. Lippman CR, Arginteanu M, Purohit D, Naidich TP, Camins MB. Intramedullary neurenteric cysts of the spine. Case report and review of the literature. J Neurosurg 2001;94:305-9.
8. Rao MB, Rout D, Misra BK, Radhakrishnan VV. Craniospinal and spinal enterogenous cysts. Report of three cases. Clin Neurol Neurosurg 1996;98:32-6.
9. Garg N, Sampath S, Yasha TC, Chandramouli BA, Devi BI, Kovoor JM. Is total excision of spinal neurenteric cysts possible? Br J Neurosurg 2008;22:241-51.
10. Kimura H, Nagatomi A, Ochi M, Kurisu K. Intracranial neurenteric cyst with recurrence and extensive craniospinal dissemination. Acta Neurochir (Wien) 2006;148:347-52.
11. Muzumdar D, Bhatt Y, Sheth J. Intramedullary cervical neurenteric cyst mimicking an abscess. Pediatr Neurosurg 2008;44:55-61.
12. Langman J. Medical embryology; human development, normal and abnormal. 2nd ed. Baltimore: Williams and Wilkins; 1969.
13. Fabinyi GC, Adams JE. High cervical spinal cord compression by an enterogenous cyst. Case report. J Neurosurg 1979;51:556-9.
14. Rotondo M, D'Avanzo R, Natale M, Pasqualetto L, Bocchetti A, Agozzino L, *et al.* Intramedullary neurenteric cysts of the spine. Report of three cases. J Neurosurg Spine 2005;2:372-6.
15. de Oliveira RS, Cinalli G, Roujeau T, Sainte-Rose C, Pierre-Kahn A, Zerah M. Neurenteric cysts in children. 16 consecutive cases and review of the literature. J Neurosurg 2005;103:512-23.
16. Yunoki M, Hirashita K, Gohda Y, Yoshino K, Fujimoto S, Mizobuchi K. True intraspinal neurenteric cyst in the lumbosacral region: Case report. Neurol Med Chir (Tokyo) 2007;47(5):237-9.

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