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Case Report

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Spinal Neurenteric Cyst of the Ventral Cervicothoracic Junction With Klippel-Feil Syndrome as a Symptom of Progressive Myelopathy: A Case Report

Jong Tae Lee [1,2], Myeong Jin Ko [1,2], Hee sung Kim [3], Seung Won Park [1,4], and Young-Seok Lee [1,2]

[1]Department of Neurosurgery, College of Medicine, Chung-Ang University, Seoul, Korea
[2]Department of Neurosurgery, College of Medicine, Chung-Ang University Hospital, Seoul, Korea
[3]Department of Pathology, College of Medicine, Chung-Ang University Hospital, Seoul, Korea
[4]Department of Neurosurgery, College of Medicine, Chung-Ang University Gwangmyeong Hospital, Gwangmyeong, Korea

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Address for correspondence:
Young-Seok Lee
Department of Neurosurgery, Chung Ang University College of Medicine, 84 Heukseok-ro, Dongjak-gu, Seoul 06974, Korea.
Email: leeys1026@cau.ac.kr

ORCID iDs
Jong Tae Lee
https://orcid.org/0009-0005-3865-5947
Myeong Jin Ko
https://orcid.org/0000-0002-1850-7753
Hee sung Kim
https://orcid.org/0000-0002-8154-2391
Seung Won Park
https://orcid.org/0000-0001-8305-7501
Young-Seok Lee
https://orcid.org/0000-0002-6881-812X

Conflict of Interest
The authors have no financial conflicts of interest.

ABSTRACT

Neurenteric cysts are rare and account for only 0.7%–1.3% of all spinal tumors. Spinal neurenteric cysts are associated with spina bifida, split-cord malformations, and Klippel-Feil syndrome, a rare congenital disorder characterized by fusion of two or more cervical vertebrae. Klippel-Feil syndrome is rarely accompanied by neurenteric cysts. In this case report, we describe a cervicothoracic junction neurenteric cyst associated with Klippel-Feil syndrome in a 30-year-old man who presented with a 2-month history of neck pain with radiation of pain into both arms and a 1-month history of weakness in the left arm. Magnetic resonance imaging (MRI) of the spine revealed an expansive intradural extramedullary cystic lesion anterior to the spinal cord at the cervicothoracic junction. The neurenteric cyst was removed using an anterior approach, accompanied by C5–C6 corpectomy. The patient's condition improved postoperatively, and he was discharged after postoperative MRI. Spinal neurenteric cysts should be considered in the differential diagnosis in cases of vertebral developmental abnormalities concurrent with intraspinal cysts.

Keywords: Neurenteric cyst; Klippel-Feil syndrome; Spinal neoplasm, Cervical vertebrae

INTRODUCTION

Neurenteric cyst represent a benign congenital anomalies, originating from endodermal cells, which are one of the three primary germ layers in early embryonic development. Predominantly situated in the spine, particularly in extramedullary and ventral locations, they account for a small percentage (0.7%–1.3%) of spinal cord tumors.[2,10,11,14,18] Neurenteric cyst may coexist with other spinal abnormalities like spina bifida or cleft vertebrae. The manifestations depend on the cyst's size and location and can range from mild back pain to more severe symptoms like weakness or paresthesia. In the context of cyst removal, anterior or posterior surgical approach is considered.

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In parallel, Kippel-Feil syndrome is characterized by the improper fusion of two or more vertebrae, usually in the cervical region. It is a rare condition, occurring in approximately 0.0025% of newborns. The syndrome typically presents with a triad of a low posterior hairline, a short-webbed neck, and restricted neck motion.[12] In the case of Klippel-Feil syndrome, the primary course of action revolves around providing symptomatic care. Notably, both neurenteric cyst and Kippel-Feil syndrome may exhibit associations with various additional anomalies, but the concurrent presentation of these two conditions is exceptionally uncommon.[1,5]

The concurrent occurrence of both neurenteric cyst and Klippel-Feil syndrome in the same patient is particularly rare and noteworthy. Each condition on its own presents diagnostic and management challenges, and their combination can lead to more complex clinical scenarios.

This report describes the successful removal of a symptomatic cervical neurenteric cyst in a 30-year-old male with Klippel-Feil syndrome. The procedure was performed using an anterior approach, improving the patient's clinical outcomes.

CASE REPORT

Clinical presentation

A 30-year-old man presented with worsening discomfort and weakening of his upper limbs that had persisted for 2 months. He frequently experienced the involuntary dropping of objects from his left hand. There were no noticeable signs of reduced leg strength, and bowel and bladder functions were normal. His medical history revealed no significant health issues.

Examination

The patient complained of neck and radiating pain from the elbow to the left upper limb hand. We observed decreased motor strength in the left biceps and triceps muscles, although sensory function appeared normal. Plain cervical spine radiography revealed congenital fusion of the cervical vertebrae spanning C5–T3, accompanied by scoliosis. Magnetic resonance imaging (MRI) confirmed the presence of a non-enhancing cystic lesion within the spinal canal but outside the spinal cord, extending from the level of the fused vertebrae at C5 to C7. Upon examination, there were no typical signs of Klippel-Feil syndrome, such as a shortened neck with limited mobility.

Preoperative sagittal (A) and axial (B) T2-weighted MRI illustrated the fusion of vertebral bodies from C5 to T3, along with a sizeable cystic lesion situated intradurally and extramedullary in front of the spinal cord and extending from C5 to C7. Cervical spine 3D computer tomography (C) and lateral radiography of the cervical spine (D) revealed the fusion and deformity of the cervical vertebral bodies (**FIGURE 1**).

Operation

General anesthesia was induced, and intraoperative neuromonitoring was implemented; a corpectomy at the C5–C6 level was performed, along with the removal of the cyst, while the patient was in a supine position. (A) During the procedure, the dura mater was opened, revealing a cyst that pushed the spinal cord towards the back. The cyst was partially excised, and a clear mucous-like fluid was drained. (B, C) The cyst and enclosing wall were completely

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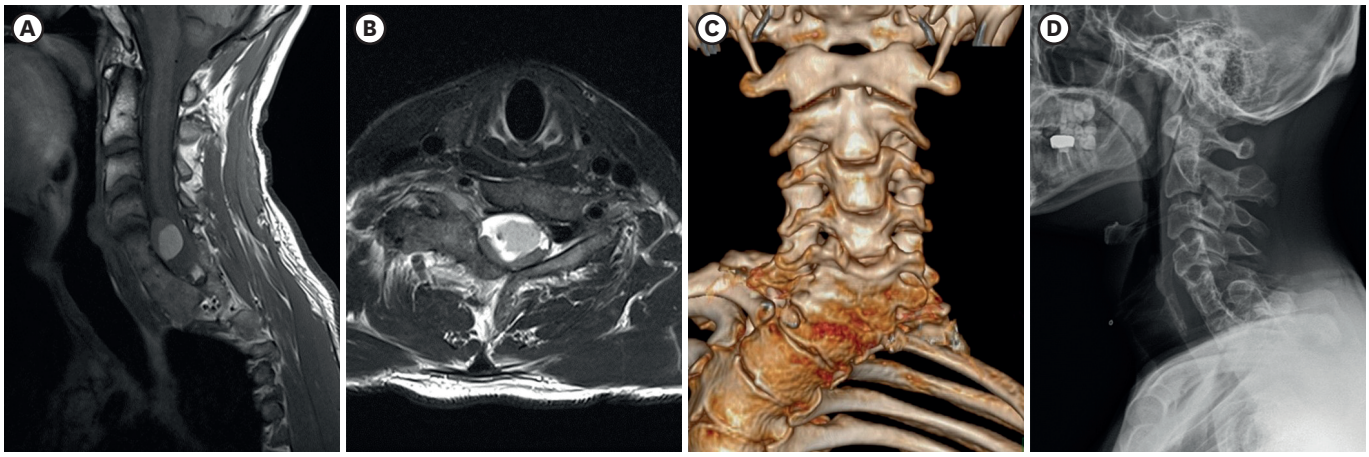

FIGURE 1. Preoperative sagittal (A) and axial (B) T2-weighted magnetic resonance images demonstrating expansive intradural-extramedullary cystic lesion anterior of the spinal cord, extending from C5 to C7. Preoperative cervical spine 3D computer tomography (C) and C-spine lateral radiograph of cervical spine (D) demonstrating fused and deformed cervical vertebrae spanning from C5 to T3.

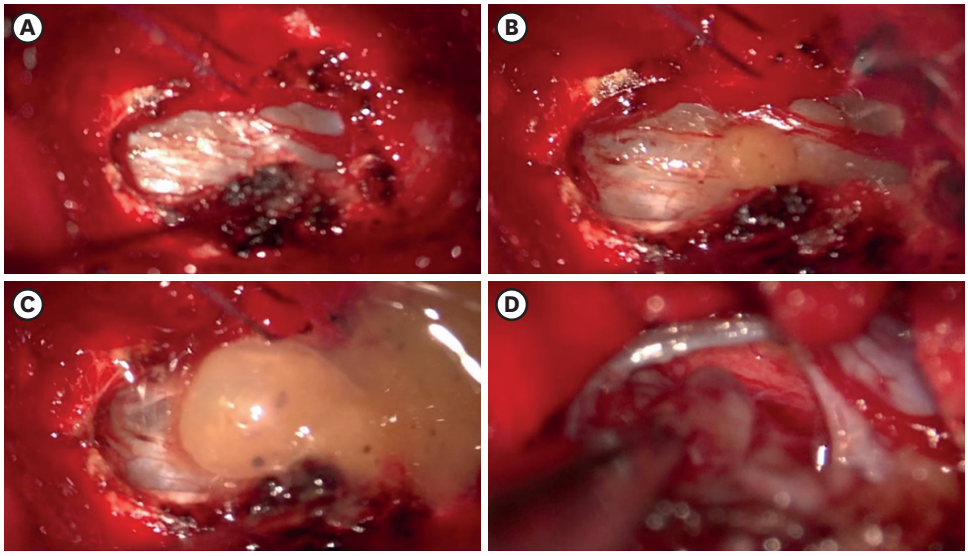

FIGURE 2. Intraoperative photograph of the neurenteric cyst. After the C56 corpectomy, the dura is exposed (A); Cyst rupture and cyst removal (B and C); Intradura cyst wall removal (D)

removed without complications. (D) Throughout surgery, monitoring of somatosensory- and motor-evoked potentials consistently demonstrated stable readings (**FIGURE 2**).

Postoperative course

The patient was released satisfactorily on postoperative day 1. Subsequent evaluations conducted at 2 weeks and 2 months postoperatively revealed noticeable enhancement in his neurological function. A follow-up cervical MRI performed 2 months postoperatively indicated no remaining lesions when viewed on T2-weighted magnetic resonance images (**FIGURE 3**).

Pathological examination

Hematoxylin-eosin-stained slides showed a lesion with columnar epithelium on the collagen layer and a cyst with gastric and respiratory epithelium, consistent with a neurenteric cyst (**FIGURE 4**).

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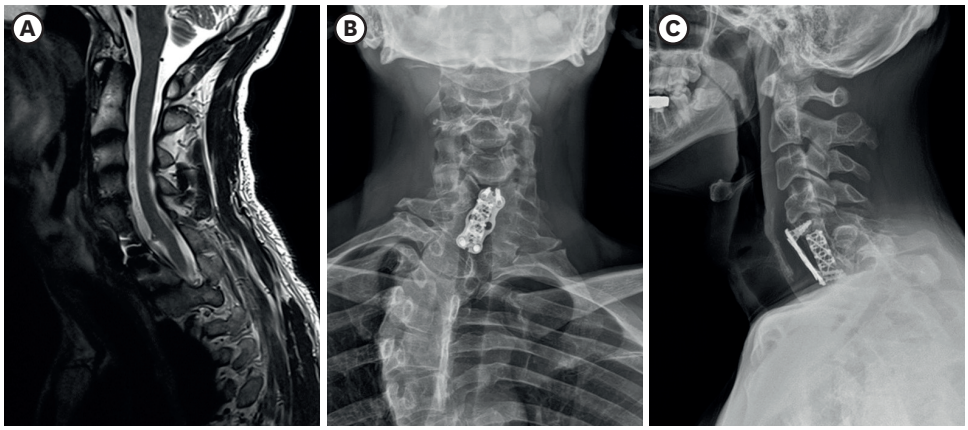

FIGURE 3. Two months follow up T2-weighted magnetic resonance images (A) demonstrate total neurenteric cyst removal; C-spine anterior (B) and lateral (C) radiograph of cervical spine demonstrating C56 corpectomy.

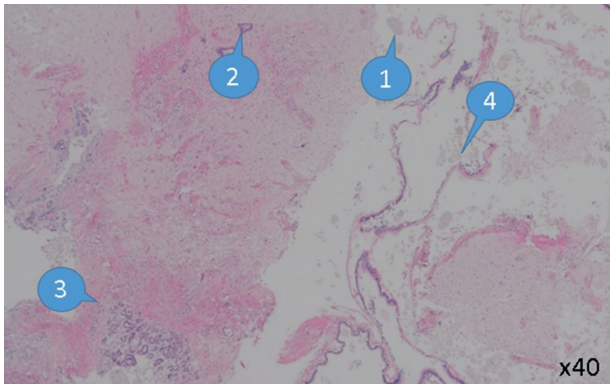

FIGURE 4. Histopathological findings of the cyst. Hematoxylin-eosin stained slide show (1) endodermal and glial tissue, (2, 3) columnar epithelium on the layer combined with gastric epithelium, and (4) respiratory epithelium (hematoxylin-eosin, ×40).

DISCUSSION

Neurenteric cyst is congenital anomalies arising from endodermal cells, more common in males during their second or third decades of life. Typically, location of neurentric cyst found in the ventral and extramedullary regions of the spine, especially at the cervicothoracic junction. Occurrence in the supratentorial part of the brain is rare.[15] The Associated abnormalities often accompanied by spinal abnormalities like cleft vertebrae, spina bifida, or diastematomyelia.[3] In the event of spinal intradural cyst, various diagnostic possibilities should be explored. It is essential to not only consider the presence of a neuroenteric cyst but also other cyst types, such as ependymal cysts or arachnoid cysts. Distinguishing between these cysts can be achieved through a comprehensive assessment of the patient's clinical history, the cyst's location, and the findings from contrast-enhanced MRI imaging. Clinical presentation of neurenteric cyst vary and can include localized pain, myelopathic symptoms, radicular issues, and can mimic conditions like multiple sclerosis, paraplegia, or meningitis. Characters of pathology are epithelial linings of these cysts range from cuboidal to columnar, resembling gastrointestinal or respiratory epithelia.[2,4,6,9-11,14,15,17,18] Radiological evaluation, especially through MRI, is crucial for early diagnosis, revealing various signal intensities depending on the content of the cyst. Importantly, these cysts do not typically enhance with

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contrast or contain mural nodules, distinguishing them from neoplastic lesions.[11] Complete surgical removal is recommended, with the anterior approach being preferable for ventrally located cysts due to better visibility and higher chances of complete removal.[1,5]

The symptom of Klippel-Feil syndrome Characterized by a low posterior hairline, short-webbed neck, and limited neck motion, with less than 40% of patients exhibiting the classic triad.[7,12] The associated conditions of Klippel-Feil syndrome can be accompanied by Sprengel deformity, congenital scoliosis, and other musculoskeletal, cardiovascular, renal, and neural anomalies. The association of neurenteric cyst with vertebral abnormalities suggests an embryological development failure, likely due to failed separation of the ectoderm from the endoderm.[12]

Despite their benign nature, untreated neurenteric cysts follow an unfavorable natural course.[18,20] Complete surgical removal is the recommended treatment, resulting in neurological symptom improvement in most cases.[9,11,13,14] Incomplete removal of the cyst is the main factor associated with recurrence.[16] Extramedullary cysts allow for a clear dissection plane, facilitating complete resection and expected neurological function stabilization or improvement.[1,8] However, intramedullary cysts present challenges due to the adherence of the cyst wall to the spinal cord.

There are two primary methods for removing the cyst. The first, as outlined in this study, is the anterior approach. Due to the typical ventral location of the cyst in relation to the spinal cord, the anterior approach offers superior visibility during surgery and a higher likelihood of complete removal.[19] The second method, the posterior approach, is the more commonly employed approach. Although it is associated with fewer complications than other methods, there is a risk of cyst leakage and meningitis resulting from cyst aspiration.[5,19]

CONCLUSION

When examining patients with intraspinal cystic masses alongside vertebral developmental irregularities, such as Klippel-Feil syndrome, it is crucial to include neurenteric cyst in the list of potential diagnoses. Neurenteric cyst can manifest differently depending on their size and location. The objective is the thorough elimination of the cyst, and any shortcomings in achieving this goal pose a considerable risk of recurrence. To accomplish this objective, it is crucial to meticulously select the surgical approach.

REFERENCES

1. Agnoli AL, Laun A, Schönmayr R. Enterogenous intraspinal cysts. **J Neurosurg** 61:834-840, 1984 PUBMED | CROSSREF
2. Agrawal D, Suri A, Mahapatra AK, Sharma MC. Intramedullary neurenteric cyst presenting as infantile paraplegia: a case and review. **Pediatr Neurosurg** 37:93-96, 2002 PUBMED | CROSSREF
3. Cai C, Shen C, Yang W, Zhang Q, Hu X. Intraspinal neurenteric cysts in children. **Can J Neurol Sci** 35:609-615, 2008 PUBMED | CROSSREF
4. Chavda SV, Davies AM, Cassar-Pullicino VN. Enterogenous cysts of the central nervous system: a report of eight cases. **Clin Radiol** 36:245-251, 1985 PUBMED | CROSSREF
5. 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** 103 Suppl:512-523, 2005 PUBMED
6. Fortuna A, Mercuri S. Intradural spinal cysts. **Acta Neurochir (Wien)** 68:289-314, 1983 PUBMED | CROSSREF

https://doi.org/10.13004/kjnt.2024.20.e8

7. Hinojosa M, Tatagiba M, Harada K, Samii M. Dermoid cyst in the posterior fossa accompanied by Klippel-Feil syndrome. **Childs Nerv Syst** 17:97-100, 2001 PUBMED | CROSSREF
8. Knight G, Griffiths T, Williams I. Gastrocystoma of the spinal cord. **Br J Surg** 42:635-638, 1955 PUBMED | CROSSREF
9. Kumar R, Jain R, Rao KM, Hussain N. Intraspinal neurenteric cysts--report of three paediatric cases. **Childs Nerv Syst** 17:584-588, 2001 PUBMED | CROSSREF
10. Kumar R, Nayak SR. Unusual neuroenteric cysts: diagnosis and management. **Pediatr Neurosurg** 37:321-330, 2002 PUBMED | CROSSREF
11. 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** 94 Suppl:305-309, 2001 PUBMED
12. Litrenta J, Bi AS, Dryer JW. Klippel-feil syndrome: Pathogenesis, diagnosis, and management. **J Am Acad Orthop Surg** 29:951-960, 2021 PUBMED | CROSSREF
13. Menezes AH, Traynelis VC. Spinal neurenteric cysts in the magnetic resonance imaging era. **Neurosurgery** 58:97-105, 2006 PUBMED | CROSSREF
14. Muzumdar D, Bhatt Y, Sheth J. Intramedullary cervical neurenteric cyst mimicking an abscess. **Pediatr Neurosurg** 44:55-61, 2008 PUBMED | CROSSREF
15. Osborn AG, Preece MT. Intracranial cysts: radiologic-pathologic correlation and imaging approach. **Radiology** 239:650-664, 2006 PUBMED | CROSSREF
16. Paleologos TS, Thom M, Thomas DG. Spinal neurenteric cysts without associated malformations. Are they the same as those presenting in spinal dysraphism? **Br J Neurosurg** 14:185-194, 2000 PUBMED | CROSSREF
17. Riviérez M, Buisson G, Kujas M, Ridarch A, Mignon E, Jouannelle A, et al. Intramedullary neurenteric cyst without any associated malformation. One case evaluated by RMI and electron microscopic study. **Acta Neurochir (Wien)** 139:887-890, 1997 PUBMED | CROSSREF
18. 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** 2:372-376, 2005 PUBMED | CROSSREF
19. Sasani M, Ozer AF, Oktenoglu BT, Peker K, Bozkus MH, Sarioglu AC. Excision of an asymptomatic cervical intradural neurenteric cyst through the anterior approach: a study of two cases and a review of the literature. **Spine J** 7:720-727, 2007 PUBMED | CROSSREF
20. 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)** 47:237-239, 2007 PUBMED | CROSSREF