International Journal of Surgery Case Reports 82 (2021) 105932

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

Microsurgery for intradural epidermoid cyst at cauda equina level in a 9-year-old child: A case report

Ha Dai Duong [a,b], Anh Hoang Pham [a,b], Hung Thanh Chu [a,*], Tam Duc Le [a,c], Dung Tuan Pham [b], He Van Dong [b]

[a] Department of Surgery, Hanoi Medical University, Hanoi, Viet Nam
[b] Department of Neurosurgery I, Viet Duc University Hospital, Hanoi, Viet Nam
[c] Department of Neurosurgery and Spine Surgery, Hanoi Medical University Hospital, Hanoi, Viet Nam

ARTICLE INFO

Keywords:
Intradural epidermoid cyst
Spinal epidermoid cyst
Pediatric
Laminectomy
Durotomy
Total resection

ABSTRACT

Introduction and importance: Epidermoid cysts are rare benign tumors. Here, we present a case of spontaneous intradural epidermoid cyst at cauda equina level in a 9-year-old patient, which we believed the first case to be reported in Vietnam.

Case presentation: A 9-year-old boy presented with 4 months of spontaneous left lower extremity muscle weakness and paresthesia. The MRI images suggested the diagnosis of intradural epidermoid cyst at cauda equina level. The patient underwent L5–S1 laminectomy and durotomy for tumor resection. The histology confirmed the diagnosis of epidermoid cyst. Post-operative images demonstrated total cyst removal.

Clinical discussion: The epidermiology, presentation and diagnosis and strategy of treatments as well as their outcomes were discussed.

Conclusion: Diagnosis of spinal epidermoid cyst is often delayed for its obscure presentation. Microsurgical dissection along with intra-operative mobile C-Arms enable total tumor resection while preserving spinal stability and neurological function. Follow-up with post-operative magnetic resonance imaging and tumor marker are helpful.

1. Introduction and importance

Epidermoid cysts are rare benign tumors. It made up for less than 1% of all intraspinal tumors [1]. Since the first intramedullary epidermoid cyst case reported by Chiari in 1883, there are more than 100 cases published [2]. Etiology of spinal epidermoid cyst may be spontaneous or iatrogenic [2]. During the fetal period, the epidermal elements integrate into deeper tissue led to spontaneous epidermoid cyst. Iatrogenic spinal epidermoid cysts following lumbar puncture have been reported in the literature [3]. The diagnosis and treatment of intraspinal epidermoid cyst are often delayed, for its slow growth and obscure clinical manifestations [1].

2. Case presentation

We report an original case of a 9-year-old. He presented with 4 months of spontaneous left lower extremity muscle weakness and paresthesia. His parents denied any history of trauma, lumbar puncture or previous surgery related to the spine. Past medical history revealed no drug use. No family history was observed.

On physical examination, he had no back pain. He had left dorsiflexion and plantarflexion weakness with muscle strength of $^4/_5$ [4,5], difficulty walking on his left heel or on his left toes, Achilles tendon hyporeflexia, decreased pin/touch appreciation in the L5–S1 dermatomes [5]. No muscle atrophy, no evidence of spinal dysraphism was observed.

The magnetic resonance imaging of the lumbosacral spine showed a 26 × 12 mm well-circumscribed intradural mass at the L5–S1 level,

Abbreviations: C-Arm, medical imaging device, which is based on X-ray technology and frequently used in various operation rooms; CA 19-9, cancer antigen 19-9; DWI, diffusion weighted imaging; FLAIR, fluid-attenuated inversion recovery; L5, lumbar vertebra 5; MRI, magnetic resonance imaging; S1, sacrum vertebra 1; STIR, short-TI inversion recovery; T1, T2, sequences in MRI protocol.

* Corresponding author.
E-mail addresses: duongdaiha@hmu.edu.vn (H.D. Duong), hungchuthanh@gmail.com (H.T. Chu).

https://doi.org/10.1016/j.ijscr.2021.105932
Received 3 April 2021; Received in revised form 20 April 2021; Accepted 21 April 2021
Available online 29 April 2021

which was slightly hyperintense on T1, hetero-intense on T2 and STIR, and no perilesional edema (Fig. 1). The findings were coherent with a diagnosis of intradural epidermoid cyst at the cauda equina level [6], resulting in L5–S1 nerve roots compression.

The patient was operated on with posterior lumbar laminectomy for tumor removal with affirmation from the patient and his parents. We utilized intra-operation imaging with mobile C-Arm to identify the accurate level of the tumor and skin incision. We performed a laminectomy and then, a durotomy at the level of L5–S1. The lesion capsule was well-defined and closely attached to surrounding nerve roots and vessels of the cauda equina region. Its content was pearly, soft, and friable. The cyst with its capsule was meticulously dissected and totally removed under the microscope. The surrounding nerve roots and vessels were preserved. The procedure was performed by Dr. H.D.D. and his team.

Macroscopically, a well circumscribed and unilocular soft cyst with friable contents was observed (Fig. 2). The histopathology showed a fibrous capsule with stratified squamous epithelium supported by an outer layer of collagenous tissue, without any skin appendages (Fig. 3). These findings were coherent with an epidermoid cyst [6].

Post-operation, the patient was treated with antibiotics and rehabilitation. After 5 days, he was dismissed from the hospital. On 1-month and 2-month follow-up, the neurological deficits subsided. The patient was able to walk steadily, he also denied any numbness or tingling in his lower extremities. The 2-month post-operative imaging demonstrated a complete resection of the tumor on MRI images (Fig. 4) [7] [8]. The CA 19-9 level 2-month post-operative was 12.25 U/ml which lines in normal interval [9].

This paper has been reported in line with the SCARE 2020 criteria [10].

3. Clinical discussion

Congenital epidermoid cysts are rare. Together with its slow growth and vague clinical manifestations, its diagnoses were often delayed [11] [8]. Our patient had been symptomatic for 4 months, with his gait were affected by weakness and paresthesia in his left lower extremity. However, there was no indication for his lumbosacral MRI scan until in our hospital.

Epidermoid cysts are usually iso- or slightly hyperintense compared with CSF in T1- and T2- weighted images, present of slight heterogeneity in signal intensity was possible. They restrict on DWI, rarely suppress on FLAIR, and may peripherally enhance [12]. In our case, MRI demonstrates a typical pattern of epidermoid cysts, which is an intradural well-circumscribed mass, slightly hyperintense in T1-weighted images, hetero-intense in T2-weighted and FLAIR images.

Diagnosis of an epidermoid cyst could be based on macroscopic inspection [8]. Histologically, epidermoid cysts are lined by stratified squamous epithelium braced up by a layer of collagenous tissue; progressive shedding of keratin from epithelial cells toward the interior of the cyst produces a soft white and friable material content. Unlike dermoid cysts, there are no skin appendages in epidermoid cysts. [8]

The ideal treatment of intradural epidermoid cyst is gross total resection [13]. Emptying of the cyst content is normally performed with ease, but the thin tumor capsule is usually tightly adherent to the surrounding nerve roots and vessels. Its complete resection can cause neurological deficits [8]. On the contrary, residual capsules might lead to the recurrence of the tumor [14] [7]. In our case report, microsurgical dissection of the capsule allowed us to accomplish total resection without compromising neurological functions.

With the utilization of mobile C-Arms intra-operative imaging, we

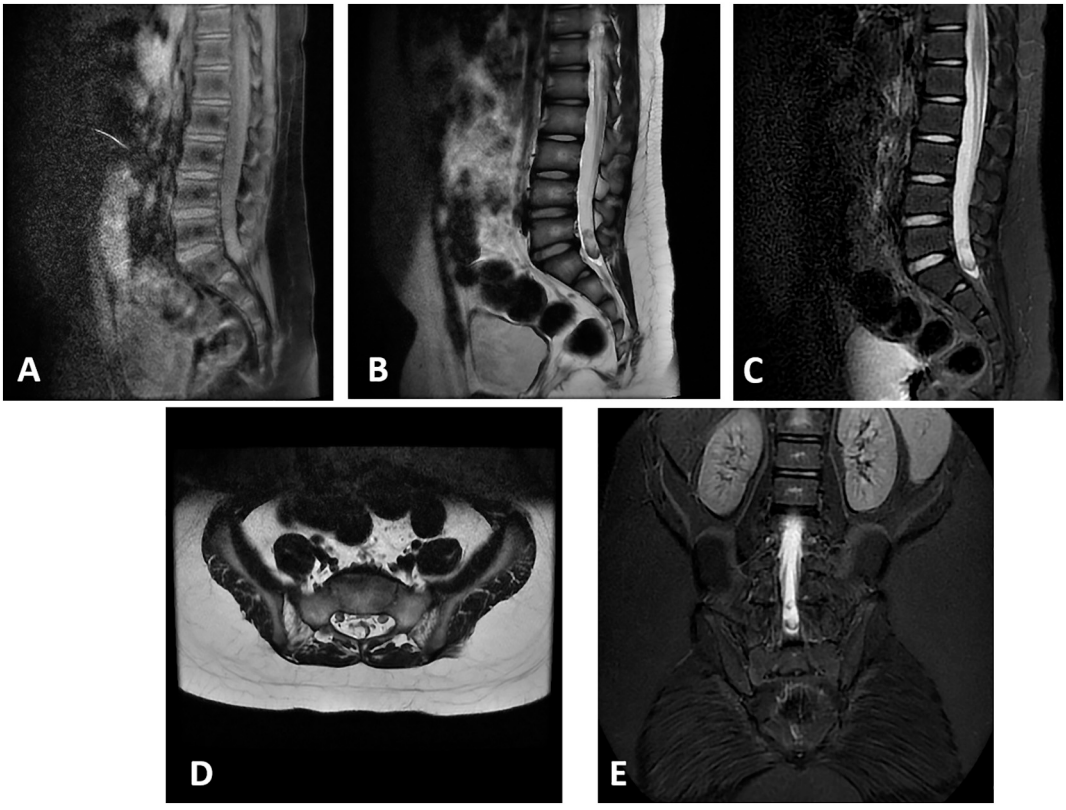

Fig. 1. A: T1 - weighted sagittal image shows a well-circumscribed intradural lesion at L5–S1 level, which is slightly hyperintense to CSF.
B: T2-weighted sagittal image shows a hetero-intense to CSF at L5–S1 level.
C: STIR sagittal image shows a hetero-intense to CSF at L5–S1 level.
D. T2-weighted axial image shows a well-circumscribed lesion with hetero-intense to CSF.
E: STIR coronal image shows a hetero-intense lesion.

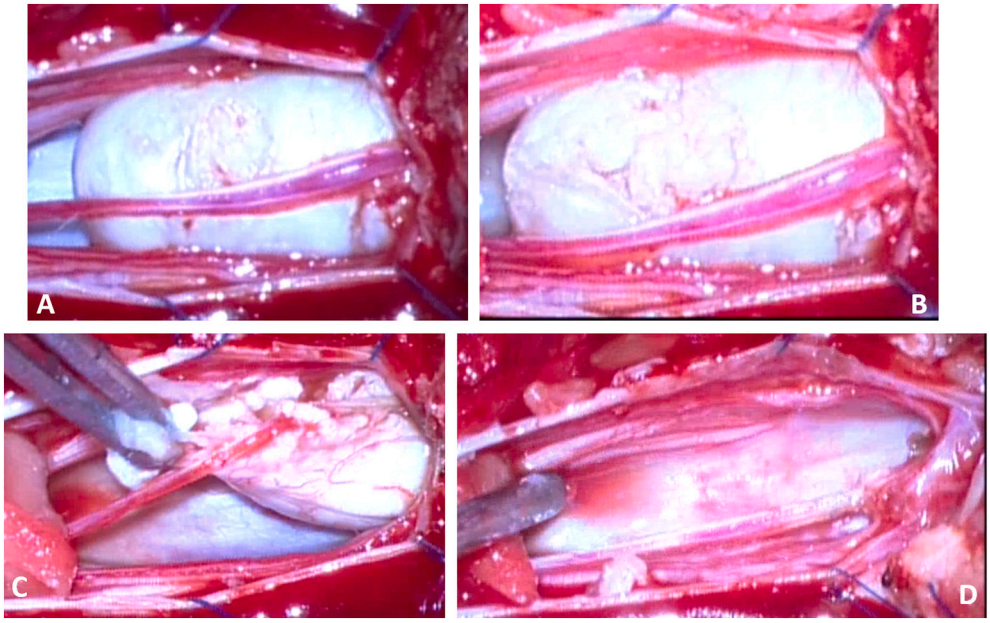

Fig. 2. A: Intra-operative image shows a well-circumscribed lesion with a white non-vascularized capsule.
B: Intra-operative image demonstrates that the cyst content was soft and friable.
C: The cyst capsule was closely attached to nerve roots and vessels.
D: After we completely resected the cyst capsule, we used normal saline to wash off all the cyst fragments.

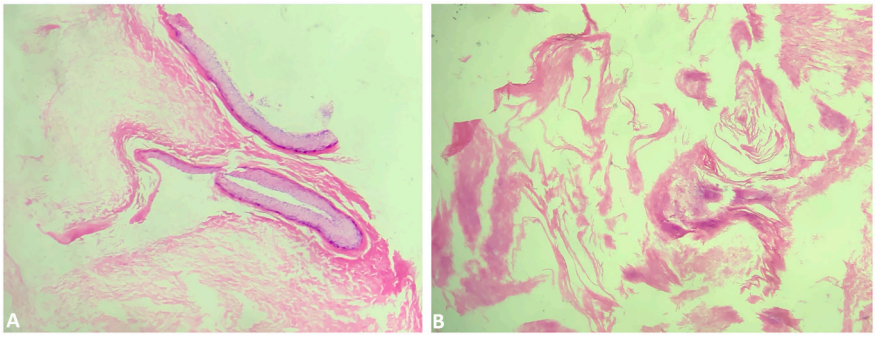

Fig. 3. A: The histology image shows stratified squamous epithelium supported by an outer layer of collagenous tissue.
B: The fibrous capsule was shown.

accurately decided the level L5–S1 for skin incision, laminectomy and durotomy, which facilitated us in the process of tumor resection. Excessive laminectomy leads to instability of the spine and requires fusion instrument [15] [16], which were complicated in our 9-year-old child case. Planned 3 or more levels of laminectomy or facetectomy equal or higher than 50% of the width of the joint on either side or both sides are decision criteria in favor for spinal fusion [17]. In our case, the patient has no spinal deformity pre-operatively, a L5–S1 laminectomy without facetectomy was performed intra-operation, and patient denies any back pain at one-month and two-months follow-up.

Postoperative follow-up mainly based on MRI images [9,13]. In our cases, MRI images show complete removal of the tumor, including in DWI images [8]. However, some reports have demonstrated the correlation between epidermoid cyst and the level of CA 19-9 [9]. Our patient's CA 19-9 level at two-month follow-up was inside normal interval.

4. Conclusion

Diagnosis of spinal epidermoid cyst is often delayed for its obscure presentation. Microsurgical dissection along with intra-operative mobile C-Arms enables total tumor resection while preserving spinal stability and neurological function. Follow-up with post-operative magnetic resonance imaging and tumor marker are helpful.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Sources of funding

The authors declared no funding for this research.

Ethical approval

The study was approved by the Research Ethics Committee of Hanoi Medical University. The procedures used in this study adhere to the tenets of the Declarations of Helsinki.

Consent

Written informed consent was obtained from the patient and his parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

CRediT authorship contribution statement

Ha Dai Duong: Conceptualization, Methodology, Investigation,

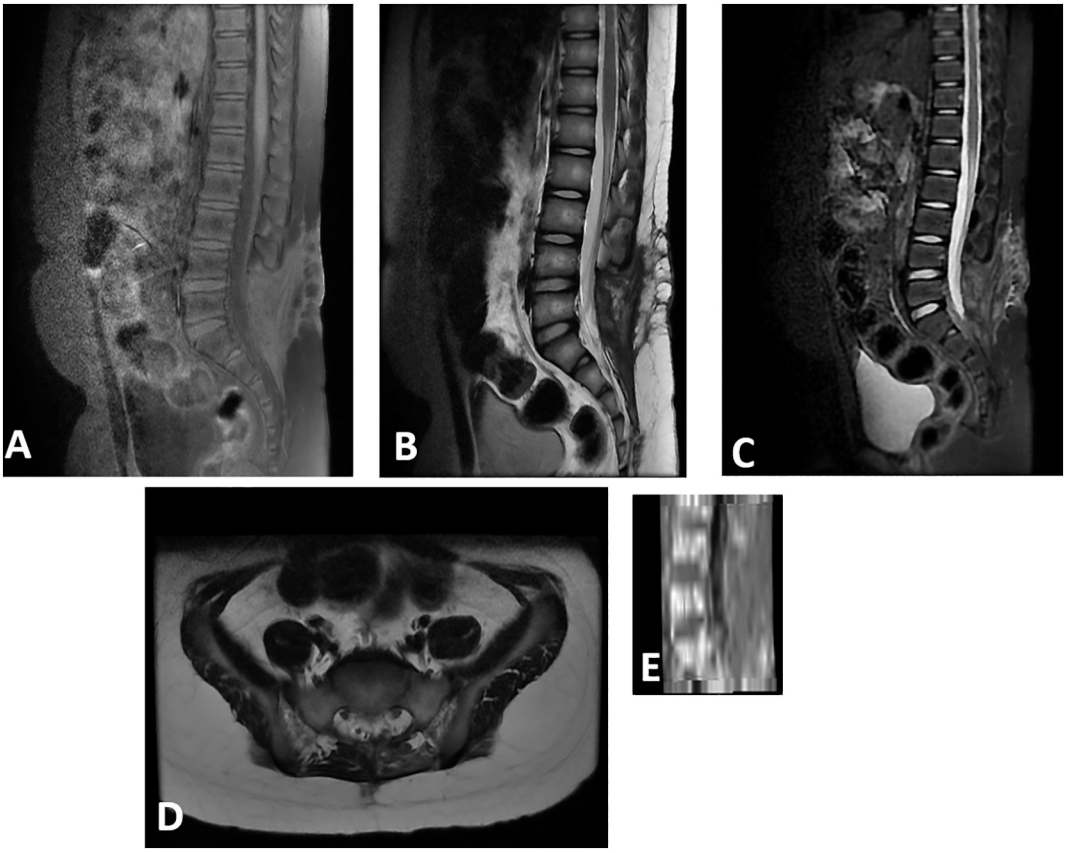

Fig. 4. A: T1-weighted image shows no lesion at the cauda equina level.
B: T2-weighted sagittal image shows no lesion at the cauda equina level.
C: STIR sagittal image shows no lesion at the cauda equina level.
D: T2- weighted axial image shows no lesion at cauda equina level.
E: DWI image shows no lesion at cauda equina level.

Writing - review & editing, Supervision.

Hung Thanh Chu: Conceptualization, Methodology, Investigation, Writing - original draft, Writing - review & editing, Visualization.

Anh Hoang Pham: Conceptualization, Methodology, Investigation, Writing - original draft, Writing - review & editing, Visualization.

Tam Duc Le: Visualization, Writing - original draft, Writing - review & editing.

Dung Tuan Pham: Visualization, Writing - original draft, Writing - review & editing.

He Van Dong: Conceptualization, Resources, Supervision.

Research registration (for case reports detailing a new surgical technique or new equipment/technology)

Not applied. This was not a first time a new surgical technique or new equipment/technology was used.

Guarantor

Hung Thanh Chu, MD
Hanoi Medical University, Hanoi, Vietnam
Email: hungchuthanh@gmail.com
ORCID ID: https://orcid.org/0000-0002-3698-5855

Declaration of competing interest

The authors declared no conflict of interest.

References

[1] O.M. Sîrbu, A.V. Chirteş, M. Mitricã, C.A. Sîrbu, Spinal intramedullary epidermoid cyst: case report and updated literature review, World Neurosurg 139 (Jul 2020) 39–50, https://doi.org/10.1016/j.wneu.2020.03.207.
[2] J.J.C. Hernandez, M. Anokwute, S.J.H. Martinez, J.R. Olivas, Intradural iatrogenic epidermoid cyst at cauda equina: a case report, Surg. Neurol. Int. 11 (2020) 299, https://doi.org/10.25259/sni_417_2020.
[3] H. Funao, N. Isogai, K. Daimon, et al., A rare case of intradural and extramedullary epidermoid cyst after repetitive epidural anesthesia: case report and review of the literature, World J Surg Oncol 15 (1) (2017) 131, https://doi.org/10.1186/s12957-017-1186-4. Jul 17.
[4] U.N. Sherman, Andrew l. Muscle Strength Grading. Text, 2020/09/03, https://www.ncbi.nlm.nih.gov/books/NBK436008/, 2020.
[5] S.C. Kirshblum, S.P. Burns, F. Biering-Sorensen, et al., International standards for neurological classification of spinal cord injury (revised 2011), J Spinal Cord Med 34 (6) (Nov 2011) 535–546, https://doi.org/10.1179/204577211x13207446293695.
[6] P. Balasundaram, A. Garg, A. Prabhakar, L.S. Joseph Devarajan, S.B. Gaikwad, G. Khanna, Evolution of epidermoid cyst into dermoid cyst: Embryological explanation and radiological-pathological correlation, Neuroradiol. J. 32 (2) (2019) 92–97, https://doi.org/10.1177/1971400918821086, 2019/04/01.
[7] A. Emad, A. Mohammad, P. Svetlana, G. Abdulkerim, G. Murat, A.-M. Ossama, Giant intracranial epidermoids: is total removal feasible? J. Neurosurg. 122 (4) (2015) 743–756, https://doi.org/10.3171/2014.11.JNS1481, 01 Apr. 2015.
[8] P. Ferrara, S. Costa, D. Rigante, et al., Intramedullary epidermoid cyst presenting with abnormal urological manifestations, Spinal Cord 41 (11) (2003) 645–648, https://doi.org/10.1038/sj.sc.3101482, 2003/11/01.
[9] Y. Wang, W. Yan, Q. Wu, G. Chen, J. Zhang, The implication of tumor biomarker CA19–9 in the diagnosis of intracranial epidermoid cyst, Oncotarget 8 (2) (2017) 2164–2170, https://doi.org/10.18632/oncotarget.12934. Jan 10.
[10] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (Dec 2020) 226–230, https://doi.org/10.1016/j.ijsu.2020.10.034.
[11] H.Y. Xiao, Z. Dan, W. Zhipeng, Z. Wang, Jianru, Surgery and outcomes of six patients with intradural epidermoid cysts in the lumbar spine. Original paper,

World J Surg Oncol 12 (1) (2014) 1–7, https://doi.org/10.1186/1477-7819-12-50, 2014-03-04.
[12] A.G. Osborn, G.L. Hedlund, K.L. Salzman, Osborn's Brain: Imaging, Pathology, and Anatomy, 2018.
[13] E. Aboud, M. Abolfotoh, S. Pravdenkova, A. Gokoglu, M. Gokden, O. Al-Mefty, Giant intracranial epidermoids: is total removal feasible? J. Neurosurg. 122 (4) (Apr 2015) 743–756, https://doi.org/10.3171/2014.11.Jns1481.
[14] M. Morita, A. Miyauchi, S. Okuda, T. Oda, H. Aono, M. Iwasaki, Intraspinal epidermoid tumor of the cauda equina region: seven cases and a review of the literature, J. Spinal Disord. Tech. 25 (5) (Jul 2012) 292–298, https://doi.org/10.1097/BSD.0b013e31821e2464.
[15] A. Bisschop, B.J. van Royen, M.G. Mullender, et al., Which factors prognosticate spinal instability following lumbar laminectomy? Eur. Spine J. 21 (12) (Dec 2012) 2640–2648, https://doi.org/10.1007/s00586-012-2250-y.
[16] R. Ahmed, A.H. Menezes, O.O. Awe, K.B. Mahaney, J.C. Torner, S.L. Weinstein, Long-term incidence and risk factors for development of spinal deformity following resection of pediatric intramedullary spinal cord tumors: clinical article. Article, J Neurosurg Pediatr 13 (6) (2014) 613–621, https://doi.org/10.3171/2014.1.PEDS13317.
[17] M.J. Avila, C.M. Walter, J. Skoch, et al., Fusion after intradural spine tumor resection in adults: a review of evidence and practices, Clin. Neurol. Neurosurg. 138 (Nov 2015) 169–173, https://doi.org/10.1016/j.clineuro.2015.08.020.