

Surgical Nuances of Intramedullary White Epidermoid Cyst in the Conus Medullaris: A Rare Entity

Abstract

Spinal epidermoid cysts (ECs) are benign slow-growing spinal tumors. They account for <1% of spinal tumors and are usually found intradural extramedullary. This report is regarding two rare cases of intramedullary white ECs present at the conus medullaris. In the first case, a 32-year-old male presented with a complaint of lower backache for 5 years, which progressively increased in intensity, radiating to the left leg. The patient had left lower limb weakness in the form of difficulty in walking. On examination, power of left knee and ankle was 4/5. Left extensor hallucis longus power was 3/5. Left Babinski sign was extensor. In the second case, a 42-year-old male, presented with a complaint of numbness over the left foot for 5–6 months. On examination, the power of the left ankle was 3/5, left extensor hallucis longus was 3/5. Both patients had EC in conus medullaris, which was hyperintense on T1-weighted magnetic resonance imaging and underwent laminectomy with the evacuation of the cyst with electrocoagulation of cyst epithelial lining. White ECs are extremely rare in the conus medullaris. Electrocoagulation of the cyst wall is like walking on a tight rope. Liberal electrocoagulation can lead to the neurological deficit but decreases the chances of recurrence. On the other hand, conservative electrocoagulation can lead to recurrence but decreases the chance of a new deficit. Recurrence should also lead to suspicion of atypical changes in the cyst wall, which may require adjuvant treatment such as radiotherapy and chemotherapy.

Keywords: *Conus medullaris, dermoid cyst, intramedullary spinal lesions, white epidermoid cysts*

Introduction

Spinal epidermoid cysts (ECs) are rare and account for <1% of the spinal tumors.^[1,2] They can be congenital or acquired. Acquired are mostly found at the L1 level, which is associated with spinal procedures such as lumbar punctures. Congenital EC is due to the presence of ectodermal tissue in the spinal cord. EC is usually intradural extramedullary but intramedullary EC is very rare. In the current literature, white ECs are usually described intracranially, but intramedullary white epidermoid has not been mentioned in the conus medullaris to the best of our knowledge, though few atypical ECs have been described which bear radiological resemblance to white ECs. In this article, we discuss two cases of intradural intramedullary white ECs in the conus medullaris.

Case Reports

Case 1

In the first case, a 32-year-old male presented with a complaint of

lower backache for 5 years, which progressively increased in intensity, radiating to the left leg. The patient had left lower limb weakness in the form of difficulty in walking. There was no sphincter involvement. On examination, the power of the left knee 4/5, ankle in dorsiflexion and plantar flexion 4/5, left extensor hallucis longus 3/5, and the left Babinski sign was positive. No history of previous spine intervention was present. Magnetic resonance imaging (MRI) of the spine revealed intramedullary lesion in the conus at L1–2. Lesion was hyperintense with areas of isointense on T1-weighted image and heterogenous on T2 weighted image. Margins were enhanced on contrast T1 weighted MRI [Figure 1]. The patient underwent D12–L2 laminectomy. The dura was opened and intramedullary cystic lesion was found at the conus. The lesion was thin-walled with visualization of intrinsic yellowish content. Posterior midline myelotomy was performed

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

How to cite this article: Aggarwal V, Narang A, Jain R, Maheshwari C, Kavita D. Surgical nuances of intramedullary white epidermoid cyst in the conus medullaris: A rare entity. *Asian J Neurosurg* 2021;16:614-7.

Submitted: 29-Jun-2020

Accepted: 24-Nov-2020

Published: 14-Sep-2021

**Varun Aggarwal,
Amit Narang,
Rahul Jain,
Chandni
Maheshwari,
Divya Kavita**

Neurosurgery, Guru Gobind Singh Medical College and Hospital, Baba Farid University of Health Sciences, Faridkot, Punjab, India

Address for correspondence:

*Dr. Amit Narang,
Department of Neurosurgery
Guru Gobind Singh Medical
College and Hospital, Baba
Farid University of Health
Sciences, Faridkot - 151 203,
Punjab, India.
E-mail: drnarangamit@gmail.
com*

Access this article online

Website: www.asianjns.org

DOI: 10.4103/ajns.AJNS_321_20

Quick Response Code:



followed by evacuation of dirty yellowish pultaceous material suggestive of EC. The cyst wall was firmly adhered to the conus medullaris proximally and nerve roots distally. The cyst was evacuated, but the wall could not be excised to prevent injury to neural tissue; hence, epithelial lining of the cyst wall was biopsied and extensively electrocoagulated to prevent recurrence. On histopathology, the cyst was lined by keratinizing stratified squamous epithelium and contained degenerated lamellated keratinous material [Figure 2]. All these features were characteristic of intramedullary white EC. Immediately after the surgery, the patient worsened to Grade 3/5 power in the right lower limb with foot drop, which partially recovered over a period of the next 6 months. Left-sided weakness totally recovered after surgery.

Case 2

A 42-year-old male, presented with a complaint of numbness over the left foot for 5–6 months. On

examination, the power of the left ankle was 3/5, left extensor hallucis longus was 3/5, and 50% loss of sensation of touch, pain, and temperature at L3–L5 dermatome. Sphincter involvement was absent. History of previous spine surgery or lumbar puncture was absent. MRI spine revealed intramedullary lesion in the conus at D12–L1. Lesion was hyperintense with areas of isointense on T1-weighted image and heterogenous on T2 weighted image [Figure 3]. The patient was operated with D12–L1 laminectomy. The dura was opened in the midline and intramedullary cystic lesion was found at the conus. Posterior midline myelotomy was performed followed by evacuation of yellowish pultaceous material. The cyst wall was firmly adhered to the neural tissue of conus medullaris and could not be excised to prevent injury to neural tissue. The cyst was evacuated and the epithelial lining of the cyst wall was electrocoagulated although not extensively to prevent any new deficit, based on previous experience. On histopathology, the cyst was lined by stratified squamous



Figure 1: (a) Sagittal T1W image, the lesion appears hyperintense with few areas of isointense areas within, in the region of the conus medullaris. (b) Sagittal T2W image shows well defined, mixed-signal intensity lesion in the region of the conus medullaris. (c) Sagittal postcontrast T1W shows enhancement of capsule or wall. (d) Coronal postcontrast T1W, the lesion shows heterogeneous enhancement. (e) Axial T1W image shows well defined, centrally placed, and hyperintense intramedullary lesion. (f and g) Postoperative Sagittal T1W and T2W images revealed no recurrence of cyst content

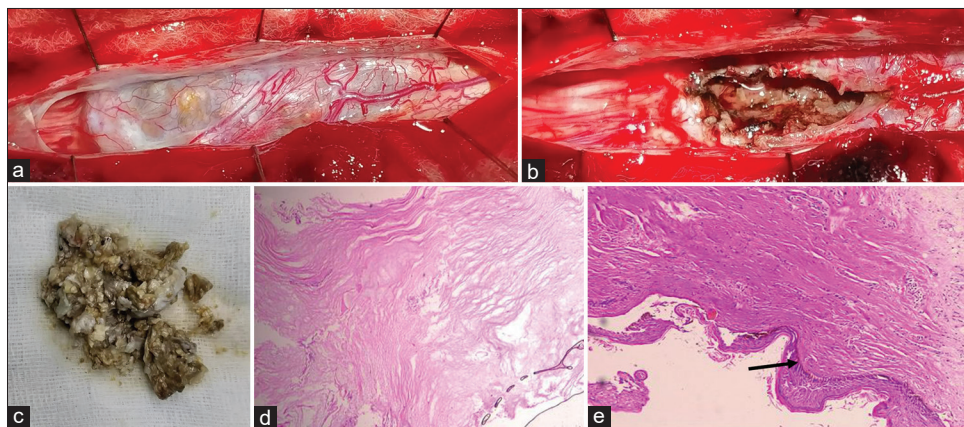


Figure 2: (a) Visualization of thin-walled cyst with intrinsic yellowish content at the conus. (b) Cyst cavity after the evacuation of content and electrocoagulated epithelial lining of cyst wall. (c) Dirty yellowish pultaceous material evacuated from the cyst. (d) Histopathology of cyst contents showing lamellated keratinous debris. (e) Histopathology of the cyst wall showed keratinizing stratified squamous epithelium

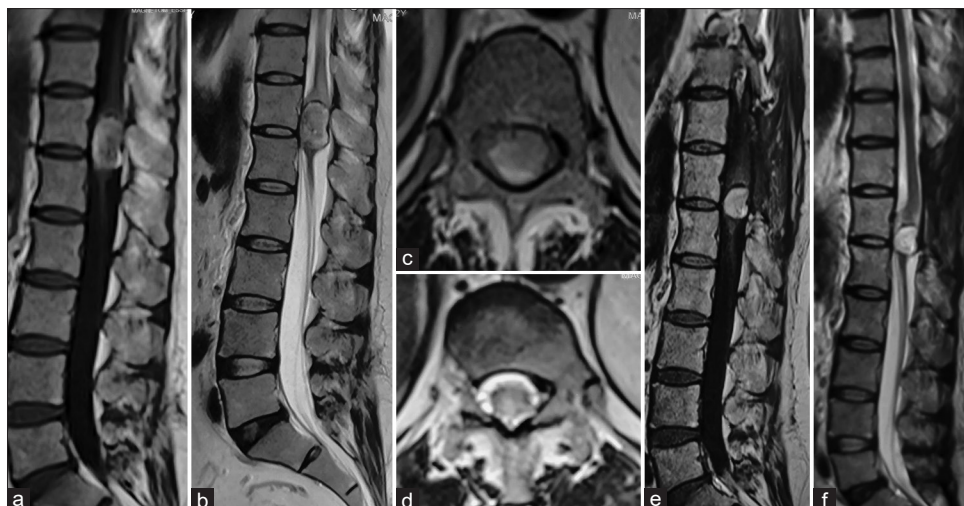


Figure 3: (a) Sagittal T1W image, the lesion appears hyperintense with few areas of isointense areas in the center. (b) Sagittal T2W image shows well defined, mixed-signal intensity lesion in the region of the conus medullaris. (c) Axial T1W image shows centrally placed hyperintense intramedullary lesion (d) Axial T2W image showed centrally placed heterogenous intramedullary lesion (e and f) Postoperative Sagittal T1W and T2W showed recurrence of cyst contents

epithelium and underlying fibro collagenous tissue showed hemorrhage and lymphomononuclear infiltrate [Figure 4]. All these features were characteristic of intramedullary white EC. Immediately after the surgery, the patient recovered completely with no new deficit. On 5 months, follow-up patient developed numbness with recurrence of the cyst at the same site. The patient was advised to repeat surgery, which he refused.

Discussion

Cruveilhier was the first to describe spinal ECs in the year 1829, naming them *tumeurs perlées*, which means pearly tumors. Spinal ECs are congenital or acquired. Congenital is most commonly reported in the conus medullaris region and arises due to the accidental inclusion of the ectoderm during the closure of the neural tube. The congenital ECs are frequently associated with hemivertebra, dermal sinus, spina bifida, and syringomyelia.^[3,4] Acquired spinal ECs are mostly because of spinal trauma or any invasive procedure such as lumbar puncture or spine surgery, which cause deposition of ectopic epidermal cells in the intradural space and lead to cyst formation. Intramedullary EC are very rare. The most common site of involvement is the thoracic cord (usually at D4–6 and D11–12 regions), followed by the lumbar and cervical cord.^[5-7] The diagnosis of intramedullary EC is often based on operative and histological findings.^[5]

White EC is usually described in the brain and the brainstem. Radiologically white epidermoid is hyperintense on T1 and heterogenous on T2-weighted image in contrast to regular EC which is hypointense on T1 and hyperintense on T2.^[8] White EC is difficult to diagnose radiologically from dermoid cyst because of similar features on radiology.^[9] White EC appears white on T1-weighted MRI because of high protein content or hemorrhage inside the cyst. Table 1 shows how various features on MRI correlate

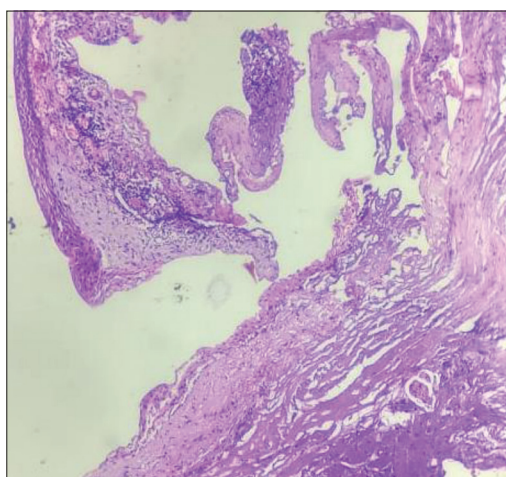


Figure 4: Histopathology of cyst showing lining of stratified squamous epithelium and underlying fibro collagenous tissue with hemorrhage and lymphomononuclear infiltrate

with the biochemical content of the cyst.^[10,11] Recurrent hemorrhage is because of granulation tissue bleed, which leads to chronic inflammation of cyst wall with firm adhesion to neural tissue. As a result, the cyst wall is tightly adhered to the neural tissue, which results in the inability to excise the wall intraoperatively. Hence, cyst evacuation and electrocoagulation of the epithelium of the wall is done to prevent recurrence. The coagulation of the wall should be meticulous in order to prevent recurrence and postoperative new deficits. Both the patients were managed with the evacuation of the cyst and electrocoagulation epithelium of the wall. The electrocoagulation in the first patient was extensive, which prevented recurrence but it leads to right-sided foot drop. We suspected thermal injury to neural tissue due to electrocautery as the cause of a new deficit; hence in the second patient, conservative electrocoagulation was done to prevent a deficit, but this patient had recurrence at 5 months follow-up. Atypical

Table 1: Prediction of biochemical contents based on MRI features of white epidermoid cyst^[10,11]

MRI		Biochemical analysis		
T1	T2	Protein	Lipid	Viscosity
Hyper	Hyper	High protein	High lipids	Semi viscous
Hyper	Hypo	High protein	Not mentioned	High viscous
Heterogenous	Heterogenous	Inflammation or recurrent bleed of different chronology		

hyperplasia of the epithelium and malignant transformation should be suspected in recurrent lesions and adjuvant treatment such as chemotherapy and radiotherapy can be considered in such cases.^[12] Chemical meningitis can be prevented by avoiding spillage of content and using steroids postoperatively. None of our patients had chemical meningitis in the postoperative period. Neurophysiological monitoring should be recommended to avoid neurological deficit because of thermal injury and dissection. In our institute, neurophysiological monitoring was not available.

Conclusion

Intramedullary, conus medullaris white ECs are extremely rare spinal ECs which occur due to hemorrhage resulting in inflammation in tumor. We recommend tumor decompression with meticulous electrocoagulation of the cyst wall to avoid recurrence and postoperative complications. The cyst wall is tightly adhered in white ECs and removal can lead to neurological deficit, so it should be avoided.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

- Amato VG, Assietti R, Arienta C. Intramedullary epidermoid cyst: Preoperative diagnosis and surgical management after MRI introduction. Case report and updating of the literature. *J Neurosurg Sci* 2002;46:122-6.
- Roux A, Mercier C, Larbrisseau A, Dube LJ, Dupuis C, Del Carpio R. Intramedullary epidermoid cysts of the spinal cord. Case report. *J Neurosurg* 1992;76:528-33.
- Manno NJ, Uihlein A, Kernohan JW. Intraspinial epidermoids. *J Neurosurg* 1962;19:754-65.
- Tekkök IH, Palaoglu S, Erbeni A, Onol B. Intramedullary epidermoid cyst of the cervical spinal cord associated with an extraspinal neuroenteric cyst: Case report. *Neurosurgery* 1992;31:121-5.
- Chandra PS, Manjari T, Devi BI, Chandramouli BA, Srikanth SG, Shankar SK. Intramedullary spinal epidermoid cyst. *Neurol India* 2000;48:75-7.
- Gonzalvo A, Hall N, McMahon JH, Fabinyi GC. Intramedullary spinal epidermoid cyst of the upper thoracic region. *J Clin Neurosci* 2009;16:142-4.
- Ogden AT, Khandji AG, McCormick PC, Kaiser MG. Intramedullary inclusion cysts of the cervicothoracic junction. Report of two cases in adults and review of the literature. *J Neurosurg Spine* 2007;7:236-42.
- Horowitz BL, Chari MV, James R, Bryan RN. MR of intracranial epidermoid tumors: Correlation of *in vivo* imaging with *in vitro* 13C spectroscopy. *AJNR Am J Neuroradiol* 1990;11:299-302.
- Gosal J, Joseph J, Khatri D, Das KK, Jaiswal A, Gupta A. White epidermoid of the sylvian fissure masquerading as a dermoid cyst: An extremely rare occurrence. *Asian J Neurosurg* 2019;14:553-6.
- Chen CY, Wong JS, Hsieh SC, Chu JS, Chan WP. Intracranial epidermoid cyst with hemorrhage: MR imaging findings. *AJNR Am J Neuroradiol* 2006;27:427-9.
- Timmer FA, Sluzewski M, Treskes M, van Rooij WJ, Teepen JL, Wijnalda D. Chemical analysis of an epidermoid cyst with unusual CT and MR characteristics. *AJNR Am J Neuroradiol* 1998;19:1111-2.
- Li J, Qian M, Huang X, Zhao L, Yang X, Xiao J. Repeated recurrent epidermoid cyst with atypical hyperplasia: A case report and literature review. *Medicine (Baltimore)* 2017;96:e8950.