

CASE REPORT / CAS CLINIQUES

CERVICAL INTRADURAL AND EXTRAMEDULLARY EPIDERMOID CYST: A RARE CAUSE OF CERVICAL SPINE COMPRESSION- A CASE REPORT**KYSTE ÉPIDERMOÏDE CERVICAL INTRADURAL EXTRAMÉDULAIRE : UNE CAUSE RARE DE COMPRESSION MEDULLAIRE CERVICALE**

Haidara Aderehime ¹
 SQUARE Ibrahima Sory ²
 TOPKA André ¹
 DEROU Keabléon Louis ¹
 YAO Konan Serges ¹

1. Service de Neurochirurgie CHU de Bouaké (Côte d'Ivoire)
2. Service de Neurochirurgie, Hôpital de l'amitié sino-guinéenne de Kipé (Guinée)

E-Mail Contact - HAIDARA Aderehime: adermedic@gmail.com

Mots-clés: imagerie par résonance magnétique, intrarachidien, kyste épidermoïde.

Keywords: epidermoid cyst, intraspinal, magnetic resonance imaging

ABSTRACT**Background:**

Epidermoid cysts of the central nervous system are benign congenital tumors resulting from the abnormal inclusion of ectodermal tissues during the neural tube closure. Although intracranial location is commonly observed, spinal epidermoid cysts are rare representing 1% of adult spinal cord tumors.

Case report:

Herein, we report the case of a cervical intradural extramedullary epidermoid cyst in a 22-years-old male with previous history of cerebellar astrocytoma. The cyst was surgically removed with favorable outcome.

Conclusion:

Furthermore, we discussed the clinical, radiological and therapeutic aspects of an unusual location of epidermoid cyst.

RESUME**Introduction :**

Les kystes épidermoïdes du système nerveux central sont des lésions bénignes d'origine congénitale, résultant de l'inclusion anormale de tissus ectodermiques lors de la fermeture du tube neural. Bien que la localisation intracrânienne soit inhabituelle, les kystes épidermoïdes rachidiens sont rares et représentent 1% des tumeurs de la moelle épinière chez l'adulte.

Cas clinique :

Nous rapportons ici le cas d'un kyste épidermoïde extramédullaire intradural cervical chez un homme de 22 ans ayant des antécédents d'astrocytome cérébelleux opéré quelques années auparavant. Le kyste a été enlevé chirurgicalement avec un résultat favorable.

Conclusion :

Nous avons discuté à travers une revue de littérature, les aspects cliniques, radiologiques et thérapeutiques d'un emplacement inhabituel du kyste épidermoïde.

INTRODUCTION

Epidermoid cysts (EC) are rare benign neoplasms which account for less than 1% of all adult intraspinal tumors (6,10). They are congenital condition due to an abnormal implantation of epidermal cells during neural tube closure (2,13,14,18). However, EC can be acquired in patients who had previous lumbar puncture, spinal trauma or surgery (8,9,12,20,24,25,26). Moreover, EC can be extra/intra dural or extra /intra medullary (6,7,15,27). When located in the spine, EC commonly occurs at the lumbosacral region. Therefore, cervical and thoracic spinal cord locations are rare (1,2,4,12,14,17,19). Clinically, spinal EC leads to an insidious onset of spinal cord compression in which the severity is correlated with the cyst size and location (2,7,10,19). Spinal MRI is the imaging study of choice in the diagnosis as it shows an iso/hypo-intense image in T1-weighted (T1W) and an hyperintense image in T2-weighted (2,13,15,16,21). The treatment of epidermoid cysts is essentially surgical, and when possible, complete removal must be the goal because of the recurrence risk (17,27).

CASE

A 22-year-old male presented with a 5 months' history of gradual gait disorders and right sided weakness. He had a history of a posterior fossa surgery 5 years ago for a cerebellar astrocytoma. This had left him with cerebellar sequelae syndrome.

His clinical examination showed a cervical spinal cord compression syndrome with right hemiparesis grade 3/5, hyperreflexia, and bilateral Babinski sign. An occipito-cervical scar related to his previous surgery was noticed upon skin examination.

The MRI of the cervical spine revealed a well-defined intradural extramedullary lesion extending from C5 to C6 level and markedly compressing the cord. The lesion was hypointense on T1W images, hyperintense on T2W with no enhancement after contrast injection suggesting at first a spinal arachnoid cyst. The brain MRI, requested in relation to the history of cerebellar astrocytoma, was normal except for the cavity left by the previous tumor resection (**Figure 1**).

The patient underwent surgery. A laminectomy was performed from C3 to C7. After opening the dura, we identified a well-encapsulated tumor mass (**Figure 2**). The lesion was oval, pearly white, soft with a cleavage plane allowing easy removal at once without damaging the spinal cord tissue. The histopathology exam showed a squamous epithelium, stratified, keratinized, with desquamated epithelial cells rich in keratin and cholesterol consistent with an epidermoid cyst. The postoperative outcome was good with complete motor recovery after physiotherapy. Control MRI of the cervical spine at 6 and 12 months did not show any recurrence.

DISCUSSION

Epidermoid cyst (EC), also called "pearly tumor" has been described for the first time in 1835 by Cruveilhier (6). The intraspinal location of the epidermoid cysts account for less than 1% of all intraspinal tumors in adults and are mostly intradural extramedullary (6,7,10,15,25,27). Their origin can be congenital or acquired. The congenital form takes its origin from anomalous inclusion of the ectoderm tissue during the closure of the neural tube in early fetal life and possibly may be associated with a defective closure of the dural tube (2,13,14,18). Acquired epidermoid cysts usually occur years after iatrogenic penetration of the skin fragments due to trauma including lumbar spinal punctures (2,4,12,14,17,19). Reports of this ectopic ectodermal migrating to sites more rostral have also been reported (1,3,16). EC commonly occurs at the lumbosacral region. Therefore, cervical and thoracic spinal cord locations are rare (1,2,4,12,14,17,19). In our clinical case, the localization was cervical intradural extramedullary. Indeed cervical localizations reported in the literature would be related to an extension of the cyst from an intracranial starting point, most often in the posterior cerebral fossa (1,16). It should be noted that our patient was operated on for 5 years for a

cerebellar astrocytoma. The clinical picture is generally progressive medullary compression whose severity is a function of the size and the location of the lesion. Acute forms have been reported and may be due to rupture of cyst contents resulting in chemical meningitis (1,3,16). MRI is the gold standard for the radiological diagnosis of the epidermoid cyst (13,15). The lesion appears, as in our case, most often in a typical homogeneous or heterogeneous hypointense signal on a T1-weighted image and a hyperintense signal on a T2-weighted image. No enhancement is observed in most cases after injection of contrast products. However, atypical signal intensity variations have been reported in epidermoid tumors. The lesion can appear as a hyperintense signal on a T1-weighted image and a hypo-intense signal on a T2-weighted image (2,13,15,16,21). This variability of the signal makes the preoperative diagnosis difficult and could be related to the chemical state of cholesterol or the relative composition of cholesterol and keratin (5). When the differential diagnosis arises with an epidermoid cyst, the diffusion sequences make it possible to make the difference. The epidermoid cyst has a heterogeneous appearance on the FLAIR sequence, with a high signal on the diffusion and restriction sequence on the ADC map. Celik (5) reported an unusual case of intraspinal epidermoid cyst associated with an arachnoid cyst in the same patient where the diffusion sequences were useful to differentiate the two lesions.

The treatment of the intraspinal epidermoid cyst is essentially surgical (2,6,12,27). The removal can be easy and complete, as in our case, with a cleavable arachnoidal plane between the capsule of the lesion and the medullary tissue (27). The lesion appears most often macroscopically rounded, pearly white color, firm consistency, encapsulated in monobloc and non-haemorrhagic. However, total excision may not always be possible when the tumor is strongly adherent making the procedure dangerous with a risk of neural damage (10). In the case of partial resection, rigorous postoperative monitoring of the residual tumor is required because recurrences, between 10 and 30%, have been reported in the literature (8,27). Radiotherapy is not established in the treatment of EC; it should be considered as an alternative to palliative surgery and in patients who cannot undergo surgery (3).

CONCLUSIONS

The intraspinal epidermoid cyst is a rare disease which pathogenesis is not clearly elucidated. Its prognosis is generously favorable. The intraspinal epidermoid cyst must be considered in spinal cord compression with a hyperintense image in T1 and hypertense in T2 on the MRI. Total excision with microsurgical technic may lead to complete recovery for patients with symptomatic lesions.

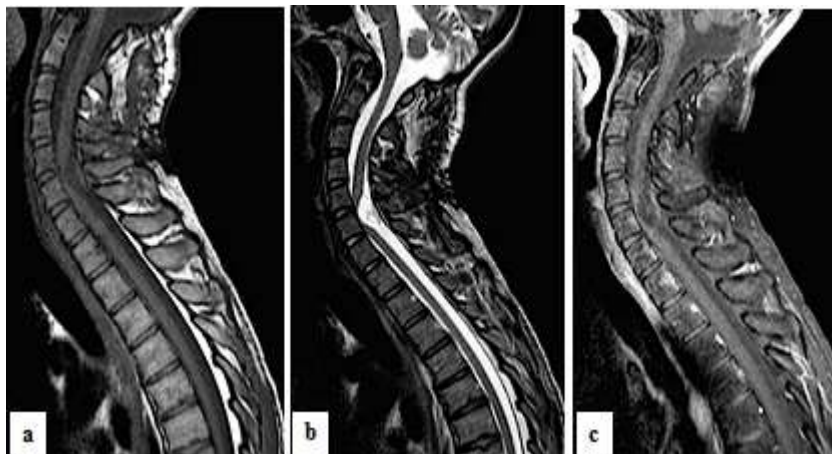


Figure 1 (a) Sagittal T1-weighted image shows hypointense mass, extending from C5 to C6. (b) On the T2-weighted image the tumor mass is hyperintense and heterogeneous. (c) No enhancement of the lesion after contrast administration.

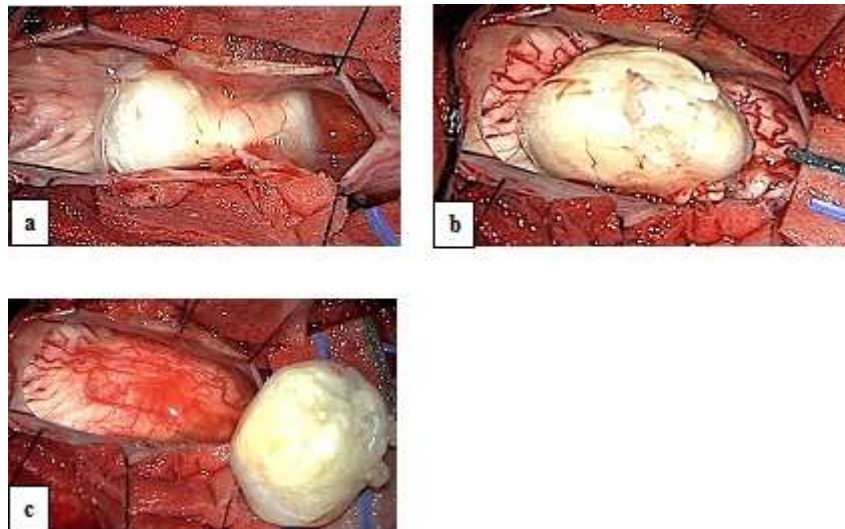


Figure 2 : (a) Subarachnoid encapsulated oval mass after opening the dura. (b) Pearl white color appears with a cleavable arachnoidal plane. (c) Complete excision of the lesion without neural tissue damage.

REFERENCES

1. ARGAWALL A. A cervical intramedullary epidermoid cyst with liquid contents. *Asian Spine J.* 2011;5(1):59-63.
2. BEECHAR VB, ZINN PO, HECK KA, FULLER GN, HAN I, PATEL AJ, ROOPER AE. Spinal epidermoid tumors: case report and review of the literature. *Neurospine.* 2018;15(2): 117-22.
3. BRETZ A, VAN DEN BERGE D, STORME G. Intraspinal epidermoid cyst successfully treated with radiotherapy : case report. *Neurosurgery* 2003;53:1429-32.
4. CARRE S, SANOUSSI S, DIETEMANN JL, SALATINO S, GUESSOUM M. Kyste épidermoïde J. *neuroradiol* 1997;24:65-6.
5. CELIK AO, BARIS MM, DEMIRTAS H. Unusual case of spinal epidermoid cyst and a concomitant spinal arachnoid cyst. *BMJ Case Rep.* 2016: doi:10.1136/ bcr-2015-214002.
6. CHANDRA PS, MANDJARI T, DEVI BI, CHANDRAMOULI BI, SRIKANTH SG, SHANKAR SK. Intramedullary spinal epidermoid cyst. *Neurol India.* 2000;48(1):75-7.
7. FERRARA P, COSTA S, RIGANTE D, MULE A, D'ALEO C, PULITANÒ S. Intramedullary epidermoid cyst presenting with abnormal urological manifestations. *Spinal Cord.* 2003;41(11):645-8.
8. FUNAO H, ISOGAI N, DAIMON K, MIMA Y, SUGIRA H, KOYANAGI T, NAKAMURA M, H MATSUMOTO, MATSUMOTO M, ISHII K. A rare case of intradural and extramedullary epidermoid cyst after repetitive epidural anesthesia: case report and review of the literature. *World J Surg Oncol.* 2017;15(1):131.
9. GARDNER DJ, O'GORMAN AM, BLUNDEL JE. Intraspinal epidermoid tumor: late complication of lumbar picture. *CMAJ* 1989;141(3): 223-5.
10. GOTECHA S, RANADE D, SHARMA S, PUNIA P, KOTECHA M. Giant intradural intramedullary epidermoid cyst. Report of two cases with varied presentations. *Asian J Neurosurg.* 2014;9(4):244.
11. JANG JS, LEE SH. Multiple intramedullary and intradural epidermoid cysts in the conus medullaris and the lumbar spine lipomyelomeningocele repair. *J Korean Neurosurg Soc.* 2003;33:512-3.
12. JEONG IH, LEE JK, MOON KS, KWAK HJ, JOO SP, KIM TS, KIM JH, KIM SH. Iatrogenic intraspinal epidermoid tumor: case report. *Pediatr Neurosurg.* 2006; 42(6):395-8.
13. KIKUCHI K, MIKI H, H NAKAGAWA The Utility of diffusion-weighted imaging with navigator-echo technique for the diagnosis of spinal epidermoid. *Am J Neuroradiol.*2002;1:1164-6.
14. KUMAR A, SINGH P, JAIN P, BADOLE CM. Intramedullary spinal epidermoid cyst of the cervicodorsal region: a rare entity. *J Pediatr Neurosci.* 2010;5(1):49-51.
15. LAI SW, CHAN WP, CHEN CY, CHIEN JC, CHU JS, CHIU WT. MRI of epidermoid cyst of the conus medullaris. *Spinal Cord.* 2005;43(5):320-3.
16. LIM J, CHO K. Epidermoid cyst with unusual magnetic resonance characteristics and spinal extension. *World J Surg Oncol.* 2015;13:240.
17. LIN YP, LI YJ, CHEN BL, GUO YH. Lumbar laminotomy and replantation for the treatment of adulte

- spinal epidermoid cyst: A case report. *Medicine (Baltimore)* 2018;97(1):e9334.
18. MANNO NJ, UIHLEIN A, KERNOHAN JW. Intraspinal epidermoids. *J Neurosurg.* 1962;19: 754-6.
 19. MUNSHI A, TALAPATRA K, RAMADWAR M, JALALI R. Spinal epidermoid cyst with sudden onset of paraplegia. *J Can Res Ther.* 2009; 5:290-2.
 20. PARK JC, CHUNG CK, KIM HJ. Iatrogenic spinal epidermoid tumor. A complication of spinal puncture in an adult. *Korean J spine.* 2014;11(3):195-7.
 21. POTE U, PARASNIS R, PATHAK S, BISHT A. Intradural epidermoid cyst with atypical magnetic resonance imaging and clinical presentation. *J Orthop Allied Sci.* 2016;4(2):91-4.
 22. 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(3):528-33.
 23. SWAMY MN A. Case of intramedullar epidermoid. *Med J Armed Forces India.* 2008;64(1):72-3.
 24. SHENG HS, LIN J, HWANG HO, YIN B, ZHANG N. Spinal epidermoid cyst formation after spinal fracture operation: A case report. *Turk Neurosurg.* 2013;23(6):800-2.
 25. SINGH K, PANDEY S, GUPTA PK, SHARMA V, SANTHOSH D, GHOSH A Acquired dorsal intraspinal epidermoid cyst in an adult female. *Surg Neurol Int.* 2016;7(Suppl 3):S67-9.
 26. SONG JH, KIM MH, SHIN KM. Intraspinal epidermoid cyst occurring 15 years after lipomyelomeningocele repair. Case report. *J Neurosurg.* 1999;90(2 Suppl):252-4.
 27. YIN H, ZHANG D, WU Z, ZHOU W, XIAO J. Surgery and outcomes of six patients with intradural epidermoid cysts in the lumbar spine. *World J Surg Oncol.* 2014;12:50.