SUPRATENTORIAL NEURENTERIC CYST CASE REPORT AND LITERATURE REVIEW

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RÉSUMÉ : Le kyste neurentérique est une lésion rare du système nerveux central considéré comme un défaut de séparation entre le feuillet ectodermal et endodermal durant la 3e semaine de l'embryogenèse. Au niveau du système nerveux central, la majorité de ces kystes siège habituellement dans la moelle cervico-dorsale. La localisation intracrânienne sus-tentorielle est exceptionnelle. Nous décrivons un kyste neurentérique frontal révélé par des crises convulsives chez un homme de 63 ans ayant une séropositivité HIV. A travers ce cas clinique, nous avons fait une revue de la littérature autour de cette entité pathologique dont l'étiologie reste imprécise.

Mots clés : *Kyste neurentérique supratentoriel, Kyste endodermique, Epilepsie, Système nerveux central.*

ABSTRACT : Neurenteric cysts are uncommon lesion of the central nervous system and are considered to result from failure of separation of ectodermal and endodermal layers during the 3rd week of embryogenesis. The majority of these cysts occur in the cervico-thoracic spine with a predilection for the ventral cord. The intracranial supratentorial location is exceptional. We describe an right frontal neurenteric cyst revealed by seizures in a 63 year old man. We report this case and discuss the relevant literature.

Key words *: Supratentorial neurenteric cyst, Endodermal cyst, Epilepsy, Central nervous system.*

INTRODUCTION

The neurenteric cyst is a benign cystic lesion sometimes covered by a ciliated mucus-secreting epithelium, simple or pseudostratified and resembling the epithelium of the respiratory or digestive tract. These cysts are generally considered to be lesions of endodermic origin, secondary to a fault in the partition between the neuroectodermic layer and the endodermic layer. They are most often described in the medullary canal, typically at the level of the anterior part of the spinal cord between the lower section of the cervical column and the upper section of the dorsal column. The intracranial localization is rare and is most often situated in the posterior indent.

CLINICAL CASE

The authors report on a rare case of the intracranial and supratentorial localization of a neurenteric cyst with a literature review spanning from 2007 to 2017 on the Pub Med and EM-Consulte servers, with the following keywords : supratentorial neurenteric cysts, endodermal cyst, central nervous system, which permitted the discovery of 9 articles. 5 were on PubMed and 4 were on EM-Consulte. A Google and Google Scholar search revealed an additional 4 articles which provide a non-specific coverage of the topic.

A 63-year-old man was hospitalized in 2001 after a seizure. The cerebral scanner showed a large right frontal cyst similar to an arachnoid cyst but with a fleshy posterior component.

Faced with these radiological and clinical elements, no surgical procedure was performed. 9 years after the first seizure, the patient had another. The cerebral MRI showed an augmentation in the volume of the cyst with a massive effect on the median line (Fig1: A, B, C, D).

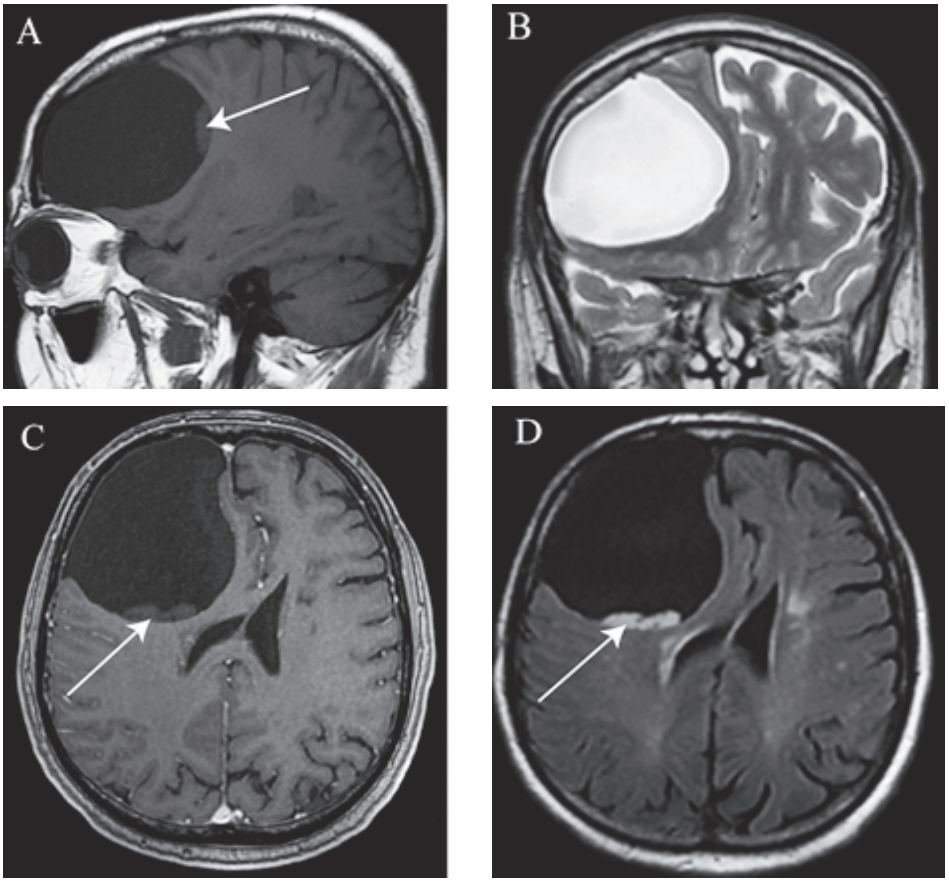

Fig. 1 (a, b, c, d) : MRI T1 axial cross-section with gadolinium (a), sagittal cross-section T1 without gadolinium (b), coronal cross-section T2 (d) and FLAIR sequence axial cross-section (c) : frontal right cystic image with a posterior tissue component with no contrast. Important mass effect on the lateral ventricle and the median line. Hypersignal on the fleshy posterior section in the FLAIR sequence.

An anticonvulsive treatment with a base of Levetiracetam and Clobazam was established. Faced with the onset of a new seizure and the augmentation of the volume of the cyst as shown on the MRI with a mass effect, a surgical treatment was proposed.

With a frontal right craniotomy, the cyst was punctured to reveal an off-white liquid deposit. With the opening of the dura mater, the lining of the cyst was found, also off-white and with gelatinous contents. The content of the cyst was evacuated and its lining was removed. The preoperatory aspect did not resemble an arachnoid cyst, but instead a parasitic cyst.

The content of the cyst was sent to the laboratory for bacteriological and parasitic analysis. These results proved negative.

The histological analysis concluded with an endodermic cyst with a lining bordered with cubocylindrical cells, certain cilia, and other mucus-secreting cells.

This coating was peeled off and worn down in certain areas. In certain zones, a metaplasia was found (Fig 2 a, b).

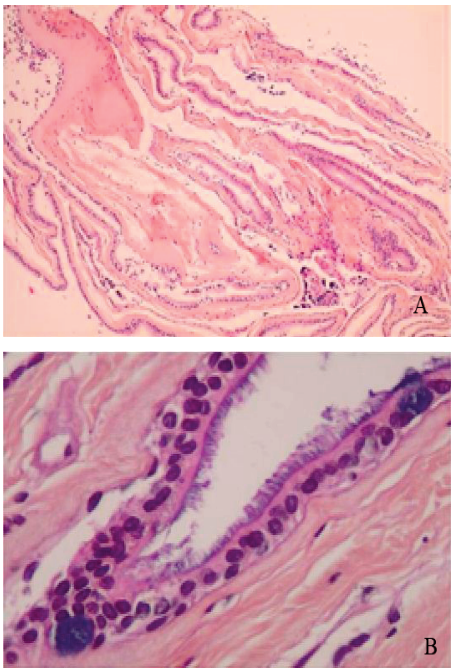

Fig. 2 (A, B) : histological cross-section of the cyst's lining, HES coloration. A (4X), B (20X). Fibrous cyst lining with a cylindrical coating. The borders of the cyst are covered by ciliated cylindrical cells and caliciform cells.

The postoperatory procedures were simple, as the J4 postoperatory control scanner confirmed the total exeresis of the cyst with pneumocephalus and CSF in the operating room.

The patient left the hospital at J5 postoperatory under anticonvulsant treatment.

The follow-up at 3 months found no evidence of further seizures.

DISCUSSION

The neurenteric cyst, also called endodermic cyst, enterogenic cyst, anterior intestinal cyst, bronchogenic cyst or even epithelial cyst, is generally found in the posterior mediastinum, the abdomen, or the pelvis and can contain endodermic and neuroectodermic elements [1]

Localization in the central nervous system is rare, and, in this case, they are most often found in the medullary canal. Touching the cervicodorsal region, they are intradural and extramedullary and are most often seated at the level of the anterior section of the medullary belt [2]

There are reported cases of intracranial neurenteric cysts which are generally located at the level of the brain stem, in the posterior indent. The supratentorial localization is much less frequent. Only 5 cases have been reported on PubMed in the past 10 years. The cyst may induce 2 types of symptoms: inflammatory symptoms in reaction with the content of the cyst or symptoms due to the effect of the mass [1]

In our case, the clinical manifestations of a convex neurenteric cyst are secondary to the compression or the irritation of neurological structures adjacent to the cyst. The revelatory symptomatology of the cyst was that of generalized seizures in the patient.

The neurenteric cyst appeared on the scanner as a well-defined hypodense image which did not take the contrast. The MRI also showed the characteristics of a cyst with a hyposignal T1 image without augmentation after the contrast. In the T2 sequences, it appeared hyperintense [2, 3]. In our patient's case, a posterior tissue component was found which did not augment after the gadolinium.

The neurenteric cyst poses a differential diagnostic problem with intracranial cystic lesions [4]. This is particularly true for arachnoid cysts, which was considered as a diagnostic but ruled out because of the associated component tissue. The differential diagnostic also rests with dermal and epidermal cysts as well as parasitic cysts. A diagnostic problem may also arise with tumors like the xantoastrocytome pleomorphe, the cystic metastases.

The treatment consists of a complete surgical exeresis, emptying of the cystic content and removal of the lining. The complete exeresis is the guarantor of a clinical and conditional improvement in order to avoid a relapse. However, in certain cases, because the lining of the cyst adheres closely to the parenchyma and a complete removal is impossible in certain zones, the risk of a return or dissemination becomes elevated [5]

The evolution towards malignity was also reported for repeated partial exeresis on the same cyst or with multiple relapses [6, 7].

CONCLUSION

The neurenteric cyst is a benign lesion. Its development in the CNS is rare and its supratentorial localization is even rarer. The prognostic after surgical exeresis is favorable but relapses can be observed in the case of partial exeresis.

ETHICAL STATEMENT

• Funding: This study was not funded
• Conflict of Interest: The authors declare that they have no competing interest.
• Ethical approval: Yes
• Informed consent: Informed consent was obtained from all individual participants included in the study.

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