A RARE CASE OF NEURENTERIC CYST INVOLVING THE ANTERIOR FOSSA

Anca Buliman1,3, Tabita Cazac1,2, Nicholas Marandici1,2, M. Gorgan1,2

1“Carol Davila” University of Medicine and Pharmacy, Bucharest, Romania
2Department of Neurosurgery, “Bagdasar-Arseni” Emergency Hospital, Bucharest, Romania
3“Titu Maiorescu” University, Faculty of Medicine, Bucharest, Romania

Corresponding author: Tabita Cazac
E-mail: tabi_cazac@yahoo.com

Abstract

Neuroenteric cysts, are rare benign endodermal lesions which mostly occur in the central nervous system. We report a case of a neuroenteric cyst in a 30-year-old man who presented with rhinoliquorrhea at the ENT department. After clinical examination, a semisolid mass was revealed in the left nostril. The Computer Tomography Scan revealed a frontal ethmoidal nasal meningoencephalocele with inferior extension into the left nostril. Gadolinium-enhanced T1-weighted MR images showed a well-defined frontal mass with ring-like enhancement and extension into the cribriform plate of the ethmoid bone and into the left nostril. The lesion measured 10/10/20 mm. The tumor was totally resected using a unilateral subfrontal approach. At five months’ follow-up, the patient showed significant amelioration of symptoms and remission of cerebrospinal fluid leakage. Native and Contrast-enhanced Cerebral Computer Tomography, as well as Magnetic Resonance Imaging showed total surgical resection of the cyst. Supratentorial neurenteric cysts involving the anterior fossa are rare. Intracranial neuroenteric cysts should be differentiated by any well-demarcated cystic tumors. The gold standard treatment remains complete surgical resection with favorable outcome.

Keywords: neuroenteric cysts, benign endodermal lesions, rhinoliquorrhea, frontal mass

Introduction

In the following, we will be presenting the case of a patient diagnosed with a frontal ethmoidal nasal neuroenteric cyst, treated in our Neurosurgical Department. The tumor was totally resected using a unilateral subfrontal approach. We reviewed medical records, imaging, treatment and follow-up of the case. At the same time, we made a review of the present literature.

Case presentation

A 30-year-old man from an urban area presented with rhinoliquorrhea at the ENT department. After clinical examination, a semisolid mass was revealed in the left nostril, therefore further investigation was needed. A Computer Tomography Scan was performed (Figure 1, Figure 2), rising the suspicion of a frontal ethmoidal nasal meningoencephalocele with inferior extension into the left nostril. Due to these findings, the patient was referred to the neurosurgery department at “Bagdasar-Arseni” Emergency Hospital from Bucharest.
A RARE CASE OF NEUROENTERIC CYST INVOLVING THE ANTERIOR FOSSA

Figure 1 – CT scan with bone window. Left frontal ethmoidal nasal meningoencephalocele with inferior extension into the left nostril

Figure 2 – Native CT scan. Left frontal ethmoidal nasal meningoencephalocele with inferior extension into the left nostril

Figure 3 – Contrast SE IRM – coronal view. Left frontal ethmoidal nasal meningoencephalocele with inferior extension into the left nostril

Figure 4 – Contrast and T2 IRM. Left frontal ethmoidal nasal meningoencephalocele with inferior extension into the left nostril
Even though the patient’s history showed an acute episode of meningitis at the age of 14 y/o, no other focal, motor or sensory deficits, no cranial nerves deficits and no signs of intracranial hypertension were documented.

The general clinical systemic examination revealed normal values. The patient was conscious, cooperative, with a good general state, blood pressure = 130/80 mmHg, and the heart rate = 88 beats/min.

Paraclinical investigations: the EKG and the pulmonary X-ray revealed normal values.

Gadolinium-enhanced T1-weighted MR images (Figures 3, 4, 5, 6) demonstrated a well-defined frontal mass with ring-like enhancement and extension into the cribriform plate of the ethmoid bone and into the left nostril. The lesion measured 10/10/20 mm. No significant vasogenic edema was identified. The MR images also revealed a cortical malformation – frontal polymicrogyria – previously undiagnosed.

Based on the patient's symptoms, the clinical and paraclinical investigations, the preoperative diagnosis made was “Frontal ethmoidal nasal cystic tumor (probably dermoid cyst”).

An elective surgical procedure was proposed for the patient, namely the surgical treatment of the cyst by means of a unilateral subfrontal approach.

Surgical procedure

Before surgery, the patient received Dexamethasone 16 mg/d, Furosemide 40 mg/d, antiepileptic drugs, analgesics and intravenous hydration.

After the patient received a general anesthetic, he was positioned supine on the operating room table, with the neck flexed on the chest and the head extended on the neck. A standard bicoronal skin incision was made: above the zygomatic arch, one cm in front of the tragus (to avoid the frontal branch of the facial nerve and the frontal branch of the superficial temporal artery), curving anteriorly and beyond the midline. Then, the periosteum was incised behind the skin incision and preserved to cover the frontal air sinus and floor of frontal fossa at the end of surgery. A basal left frontal craniotomy was performed with frontal sinus opening, in order to avoid brain retraction. Dural opening was low and parallel with the skull base.

An extranevraxial mass was easily identified as a thin-walled, grey-white cyst with dimensions of about 10/10/20 mm. The cyst was inserted on the cribriform plate of the ethmoid bone, with extension in the left nostril. No feeding arteries were noted. The membrane was punctured and the viscous fluid was aspirated from the cyst. Minimal adhesions between the cyst wall and surrounding structures were identified, therefore the wall was peeled away easily and the cribriform plate was resected in order to complete the evacuation of the nasal extension of the cyst. The attachments to the cribriform plate were entirely coagulated.

Dura mater was anchored and closed in a watertight manner. A frontal sinus cranialization was performed, packing the sinus with muscle and fat, removing the intrasinusal mucosa and applying antibiotic solutions and Gel foam. Sinus defect was covered with pericranium, which was then sutured to the dura, tangent to the craniotomy, being excluded in this way the communication between frontal sinus and intracranial space, avoiding infections. An epidural drain was placed and reconstruction of
the bone defect was performed using acrylic cement. Finally, skin closure was realized in a two-layers suture technique.

The histological examination revealed a neuroenteric cyst with its wall formed by pseudostratified columnar epithelium and a basement membrane with intestinal-like structure.

Postoperative evolution
The patient was discharged ten days after surgery (Figure 8).

At five months’ follow-up, neurosurgical visit, the patient was in good condition, with significant amelioration of symptoms and remission of cerebrospinal fluid leakage (Figure 9).

He experienced no other cranial nerve abnormalities, no motor paresis or sensory impairments, no seizure or episodes of loss of consciousness.

Native and Contrast-enhanced Cerebral Computer Tomography, as well as Magnetic Resonance Imaging showed total surgical resection of the cyst (Figure 10).

Figure 7 – Histological examination. Neuroenteric cyst with a pseudostratified columnar epithelium. A – H&E, magnification x 10. B – H&E, magnification x 40. C – Van Gieson, magnification x 10

Figure 8 – Postoperative day 10 – Patient’s status

Figure 9 – Patient’s status - at two months’ follow-up, neurosurgical visit

Figure 10 – Cranial 3D reconstruction
Discussions

First described by Puuseep in 1934, neuroenteric cysts, are rare benign endodermal lesions which mostly occur in the central nervous system [1, 2, 3]. To date, about 80 intracranial neuroenteric cysts have been published in the literature [4, 11, 13].

Neurenteric cysts can originate anywhere along the central nervous system axis, being known as endodermal, enteric, enterogenous, respiratory, gastroenterogenous, and archenteric cysts. These lesions have also been defined as gastrocytomas or intestinomas [4-9].

Based on their origin cell, epithelial cysts are divided into two subtypes: neuroepithelial cysts, arising from primitive ependyma, and endodermal cysts, originating from the transient neurenteric canal, due to the notochord dysgenesis, around the third or fourth week of gestation [2, 14- 18].

The etiology of supratentorial neurenteric cysts is still unclear at the moment. Several theories have been proposed, the most relevant being the one described by Graziani et al. [23]. They suggested that supratentorial neurenteric cysts, Rathke’s cleft cysts, and colloid cysts originate from Seessel’s pocket migrating cells: Rathke’s cleft cysts occur from an intrasellar residue, colloid cysts are formed in the third ventricle, and neuroenteric cysts appear in the
A RARE CASE OF NEUROENTERIC CYST INVOLVING THE ANTERIOR FOSSA

presellar or retrosellar location. Another theory was proposed by Mittal et al. [19] suggesting that neuroenteric cysts may occur due to an abnormal endodermal cell migration dorsally through the neuroenteric canal into the ectoderm.

Most of the endodermal cysts, described in the literature, up to this moment, have been found in the spine, usually ventral to the spinal cord, intracranial lesions being far less common [2, 9, 10].

Due to their origin, endodermal cysts usually arise in the midline, most of the intracranial lesions described so far being typically identified in the posterior fossa [4, 11], usually anterior to the brainstem [11] or in the cerebellopontine angle [12]. Instead neuroepithelial cysts, tend to have a more variable development area. Supratentorial neuroenteric cysts are extremely rare, extra-axial lesions [13] and usually located in the supra and parasellar region [42, 43], near the septum pellucidum [44], third ventricle [45], anterior fossa [25, 35, 38, 39, 46] and along the optic nerve [37].

In the latest 20 years of neurosurgical experience, we encountered only one case of neuroenteric cyst. Initially we presumed a preoperative diagnosis of dermoid cyst, due to the lack of continuity between the cyst and surrounding dura-mater.

A review of the literature shows that neuroenteric cysts appears in all age patients with a slight male predominance (male-to-female ratio of 3:2) [18, 19]. We present a case of a 30 y/o male patient with neuroenteric cyst.

Histologically, neuroenteric cysts are benign lesions formed by simple or pseudostratified cuboidal or columnar epithelium cystic walls, located on a basement membrane with intestinal – like structure [20]. In our study the histological examination revealed a neuroenteric cyst with its wall formed by pseudostratified columnar epithelium and a basement membrane with intestinal – like structure.

Based on their imaging aspect and location, differential diagnosis of neuroenteric cysts includes Rathke’s cleft and colloid cysts epidermoid, dermoid, arachnoid, parasitic, neoplastic cysts [21, 22]. Histologically, neuroenteric cysts, Rathke’s cleft cysts and colloid cysts are similar.

A review of the literature reports that supratentorial neuroenteric cysts display variable imaging characteristics. On CT scans they might be identified as hyperattenuating, hypoattenuating, isoattenuating or an iso-hypoattenuating mass lesions [5, 12, 13, 24-39]. These findings may occur due to the protein levels of the cystic contents. On T1-weighted MR Imaging, neuroenteric cysts can display a hyperintense, isointense or iso-hypointense appearance meanwhile on T2-weighted MRI these lesions can appear hyperintense or mixed hyper/isointense. On FLAIR pulse sequences, they may occur hyperintense. There are no reports of neuroenteric cysts enhancing with gadolinium contrast, (MRI) or iodine agents (CT).

In our study gadolinium-enhanced T1-weighted MR images demonstrated a well-defined frontal mass with ring-like enhancement and extension into the cribriform plate of the ethmoid bone and into the left nostril. The lesion was measuring 10/10/20 mm. No significant vasogenic edema was identified. The MR images also revealed a cortical malformation – frontal polymicrogyria – previously undiagnosed.

Clinical presentation of neuroenteric cysts is influenced by its location, the most common symptom being represented by seizure [5, 12, 13, 24-39]. Some authors reported chemical meningitis in neuroenteric cysts and was presumed to be due to the irritant cyst contents [30–39]. In our study, the patient had no other symptoms except rhinoliquorrhea. Patient’s history revealed an acute episode of meningitis at 14 y/o, which was presumed to be due to the leakage of the irritating cystic fluid into the surrounding cerebral structure.

Treatment should aim for complete surgical excision of the cyst and contents, cyst aspiration, fenestration or partial resection being avoided, if possible. We performed a total surgical removal of the cyst, including the cystic walls. No cystic remnants were noted on the postoperative CT scan images.

Recurrent neuroenteric cysts have been reported, and this risk is mostly dependent by the extent of the resection that is achievable [4, 15].

Leventer et al. [20] reported early recurrences in cases where cysts were drained or cyst biopsy was attempted. They assumed that the cystic wall residue could be responsible for
the recurrence. Thus, intraoperatory, the remaining cystic walls should be distinguished from arachnoid membrane and coagulated.

Conclusion

Supratentorial neurenteric cysts involving the anterior fossa are rare. Radiological diagnosis is challenging due to the variability of imaging aspects. Therefore, intracranial neurenteric cysts should be differentiated by any well-demarcated cystic tumors, without enhancement on MRI. The gold standard treatment remains complete surgical resection with favorable outcome.

References

A RARE CASE OF NEUROENTERIC CYST INVOLVING THE ANTERIOR FOSSA