



Case Report

Glioependymal cyst in the medulla oblongata – A case report

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ABSTRACT

Background: Glioependymal cysts (GECs) are rare benign lesions that can be found anywhere along the neuroaxis, with most of the reports denoting supratentorial location. Here, we introduce a rare case of successfully treated glioependymal cysts lying in an uncommon location, namely medulla oblongata.

Case Description: A 69-year-old lady presented with progressive unsteadiness and swallowing disturbances, and brain magnetic resonance imaging showed a dorsally located lesion within the medulla oblongata; based on the presentation and radiological features, surgical intervention was deemed mandatory. The suboccipital midline approach was used to perform marsupialization of the cyst with shunting through a syringosubarachnoid shunt to prevent future recurrence, and the patient outcome was improved.

Conclusion: Medulla Oblongata's location for glioependymal cysts proposed unique diagnostic and operative challenges that may require highlighting for practicing neurosurgeons.

Keywords: Benign intracranial cyst, Glioependymal cyst, Medulla oblongata, Neuroepithelial cyst

INTRODUCTION

Glioependymal cysts (GECs) are rare benign cystic lesions of congenital ectodermal origin.^[4] They develop early during the 3rd week of pregnancy, typically as single cysts; however, few were reported as multiloculated.^[3,13] Usually occurs alone as an isolated lesion. Also, an association with corpus callosum defect was reported.^[12]

Preoperative diagnosis is challenging, as GECs share many clinical and radiological characteristics with other anomalies, making histopathological studies the only way to differentiate between them. GECs can be found intra-axial or extra-axial, with the former being more common, predominantly in the frontal lobe.^[8] Only a few cases were reported within the posterior fossa, as in the cerebellum, near the fourth ventricle, making the medulla oblongata an extremely rare location.^[7,10]

Here, we report a rare case of surgically managed GEC located in the dorsal of the medulla oblongata.

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CASE SCENARIO

A 69-year-old lady had unsteadiness with swallowing difficulty that worsened gradually. Further complicated by severe aspiration pneumonia, which required hospital admission. The patient suffered from tetraparesis, with hoarseness of voice, curtain sign on the right side, and coarse nystagmus bilaterally without extraocular movement abnormality. Later on, she did a brain magnetic resonance imaging (MRI), which showed an intra-axial cystic lesion within the posterior part of the medulla oblongata. It became thin as paper, and the cyst extended superiorly to the lower end of the fourth ventricle and inferiorly to level the C1. It did not communicate with the fourth ventricle; both tonsils displaced laterally. The cystic content followed cerebrospinal fluid (CSF) sequences, not restricted to diffusion-weighted imaging (DWI), with a hypointense signal in T1 and a hyperintense signal in T2. In addition, the capsule was not enhanced [Figure 1]. We performed a partial resection for the cyst with a cysto-subarachnoid shunt due to the presenting symptoms.

Intraoperatively, the cyst wall was whitish, transparent, and laid on the dorsal surface of the medulla oblongata; superiorly, it embedded into the foramen Magendie; meanwhile, the cyst wall was incised, biopsied, and sent for histopathology; and the cyst content was CSF-like clear, colorless fluid gashed out, and the part of the cyst wall which adherent to the medulla was intentionally left. Then, a shunt tube was placed between the cyst cavity and cervical subarachnoid space and fixed on the medulla using surgical fibrin glue [Figure 2]. The histopathological finding came with glial fibrillary acidic protein (GFAP) positive and epithelial membrane antigen (EMA) negative, and hematoxylin and eosin stains showed stratified cuboidal epithelium with cilia and underlying neuroglial tissue in the cyst wall [Figure 3]. These findings were compatible with the gliependymal cyst.

Following one month, the patient's symptoms improved, and the follow-up MRI 3 months later showed a remarkable resolution of the cyst [Figure 4].

DISCUSSION

The gliependymal cyst (GEC) is a rare nonmalignant cyst that accounts for 0.01% of all central nervous system tumors. It is embryologically driven from ectopic rests of primitive neuroglial tissue, can occur in the midline or laterally, and has been reported both as a unilocular and multilocular anomaly.^[4] GEC tends to be more common in adults than children, with most cases being women, and the age average is 42.3 years, even though it can be found at any age.^[12] Our patient was a woman in her 60s.

GECs had been described by multiple terms, including neuroglial, neuroepithelial, and choroidal cysts, which were previously used to designate different types of intracranial benign cysts with varied histopathologies. That makes the final interpretation challenging, which was rationalized by Robles *et al.* Thus, the term “gliependymal cyst” should be used to describe a unique set of cysts with specific pathological and radiological features.^[9]

GECs, presentations were varied owing to their slow-growing nature, with diversity in location commonly causing seizure and compression of adjacent neuronal structure,^[13] with some reported cases found incidentally.^[4,9]

Owing to their neuroepithelial origin, they can be found along the neuroaxis, with the supratentorial location, like the frontal lobe, being the most common. Few articles report the posterior cranial fossa as in the cerebellum, pons, as a site for GECs. In our case, the GEC is located in the medulla oblongata, which was not reported before.^[2,8,11]

Radiologically, GECs are unilocular cysts with thin-walled morphology on computed tomography and MRI. In most reported papers, they were hypointense on T1-weighted and fluid-attenuated inversion recovery images and hyperintense on T2-weighted images, were neither restricted in diffusion-weighted sequence nor enhanced with contrast, and did not have edema. Robles *et al.* reviewed that most of the intracranial benign cysts showed the same radiological findings.^[9] Yet, the GEC can be distinguished radiologically from the arachnoid cysts by the absence of scalloping of

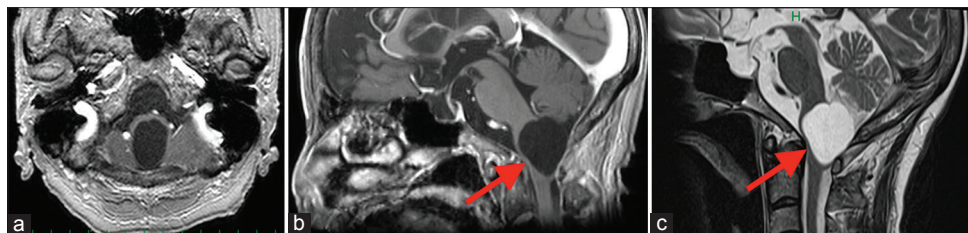


Figure 1: Preoperative magnetic resonance imaging. (a) The axial T1-weighted image (T1WI) with contrast. (b) sagittal T1WI with contrast. (c) T2-weighted image sagittal. demonstrates that the cystic lesion follows the cerebrospinal fluid signal and located posteriorly with thinning of medulla oblongata. The red arrow represent the gliependymal cyst (GEC) in both 1b and 1c.

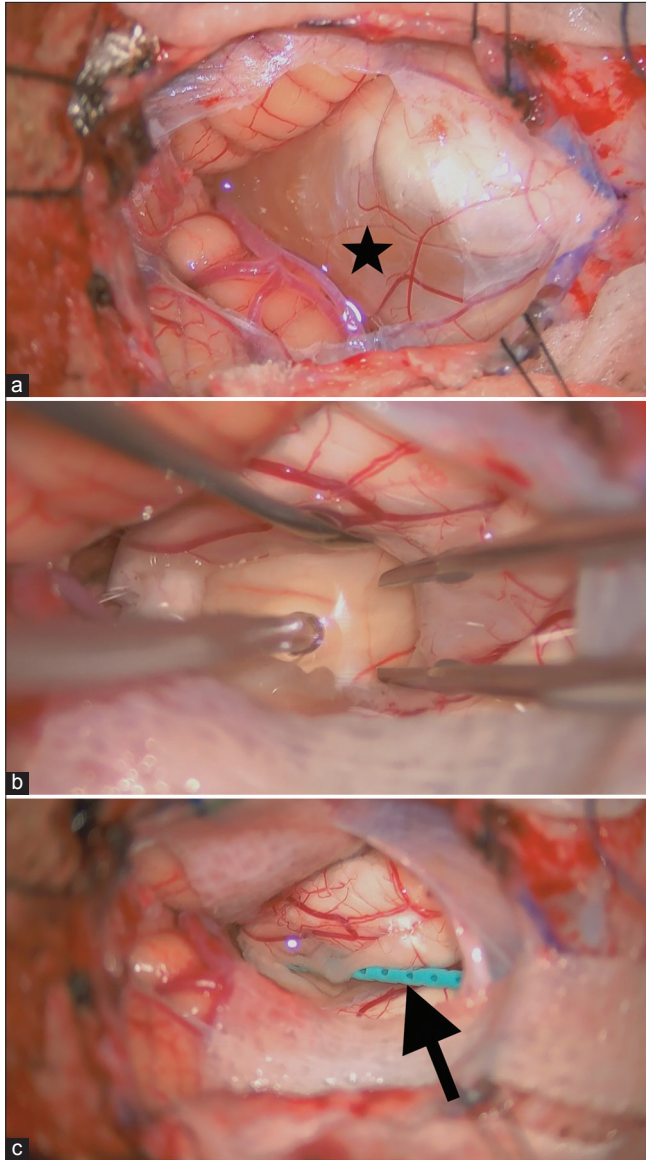


Figure 2: Intraoperative views through the posterior midline approach show the cyst at the dorsal part of the medulla oblongata. (a) Thin, translucent cyst wall (black star), (b) The colorless fluid content, and (c) cysto-subarachnoid shunt placement (syringosubarachnoid shunt, create Medic Co., Ltd., Kanagawa, Japan) (the black arrow).

the adjacent cranium. Despite this, the arachnoid cysts are still far more similar in appearance to GECs, especially if they occupy the cerebellopontine angle. GECs differ from epidermoid and dermoid cysts due to the restriction of their contents in DWI owing to their heterogeneity and differ from neuroentric cysts, which are iso to hyperintense on T1-weighted images and hypo to hyperintense on T2-weighted images. In addition, GECs can be distinguished from porencephalic cysts, which communicate with the lateral ventricle and show surrounding gliosis and spongiosis. Contrary to GECs, infectious cysts, such as neurocysticercosis, are smaller than 10 mm (about 0.39 in) and partly calcified and enhanced, usually isointense to hyperintense on T1-weighted MRI.^[1,6,9,12]

Treatment was directed for the symptomatic, progressive in nature cases, not for the incidentally discovered ones.^[13] As in our patient, who had worsening symptoms related to the mass effect of the cyst. Ideally, GECs are treated by a complete resection with the lowest possible morbidity. If this treatment is not feasible, further options include aspiration, fenestration into the cistern, open or endoscopic cystoventriculostomy, and cystoperitoneal, cystocisternal, or cystoventricular shunting, all of which have a high success rate and few recurrence cases.^[3,5] In the presented case, we perform marsupialization, leaving part of the cyst wall that was adherent to medullary parenchyma to avoid neuronal injury and worsening in symptoms with the installment of a cysto-subarachnoid shunt to avoid the recurrence.

Intraoperatively, GECs consisted of a thin translucent or grayish membrane with no communication with the subarachnoid or ventricular spaces and contain CSF-like clear fluid in most of the reported cases, with very slow growth due to the activity of the ependymal cells.^[5] Intraoperatively, we found a transparent cyst wall and CSF-like content.

Most reviewers described the wall of GECs as being formed by two or three different layers, and owing to their embryologic origin, they were also positive for GFAP and lacked reactivity for cytokeratin, carcinoembryonic antigen, and EMA.^[4] In our case, the histology confirmed the diagnosis with positive GFAP and negative EMA.

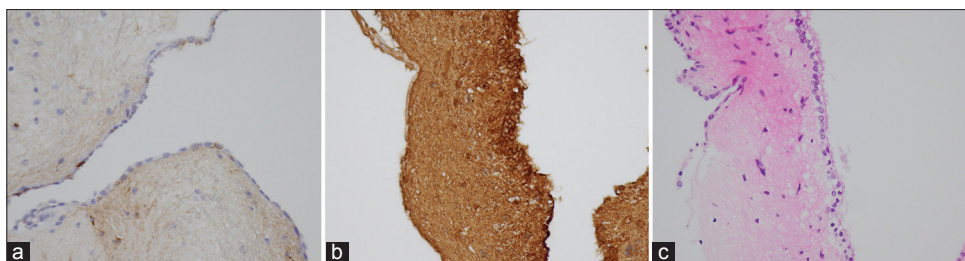


Figure 3: Histopathology of the resected lesion. (a) epithelial membrane antigen-negative, (b) glial fibrillary acidic protein-positive, and (c) Hematoxylin and Eosin stains showing stratified cuboidal epitheliums with cilia and underlying neuroglial tissue in the cyst wall.

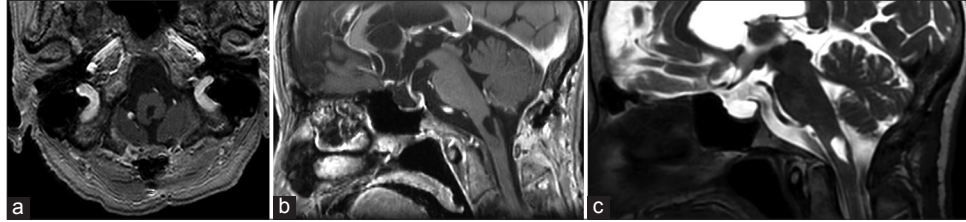


Figure 4: Postoperative magnetic resonance imaging. (a) The axial T1-weighted image (T1WI) with contrast, (b) sagittal T1WI with contrast, and (c) T2-weighted image demonstrates the resolution of the lesion.

In summary, GECs can be easily confused with another intra-axial benign cystic lesion. Owing to the similarity in radiology and presentation with other benign cystic lesions, we suggest that the differential diagnosis of posterior fossa cystic lesions be inclusive to GECs as a rare and unique lesion. We reported a case of a rare GEC location within the dorsal medulla oblongata. The cyst was successfully treated through partial cyst wall removal followed by cysto-subarachnoid shunt using a midline suboccipital approach. This approach will enable the avoidance of parenchymal injury of the medulla and prevent recurrence.

CONCLUSION

There are distinct diagnostic and operative challenges associated with the location of gliopendymal cyst in the medulla oblongata that may be required to be emphasized by daily practicing neurosurgeons.

Ethical approval

Not applicable.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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