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Case Report

Neuroepithelial cyst causing homonymous hemianopia treated through surgical marsupialization under visual-evoked potentials: A case report

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ABSTRACT

Background: Neuroepithelial cysts (NECs) are rare entities, occasionally causing neurological symptoms that can be overlooked.

Case Description: A case of an occipital neuroepithelial cyst is discussed. The initial presentation consisted of mild homonymous hemianopia and gait impairment. Conservative management was suggested to start with, but at 6 months follow-up, the patient's symptoms were worsening. Surgery was performed under general anesthetic and using visual-evoked potentials. The cyst was marsupialized and connected with subdural space, and a few samples were sent for histological analysis. The patient experienced immediate improvement in her symptoms, and the visual tests at follow-up confirmed the resolution of the previously documented hemianopia.

Conclusion: NECs should be carefully assessed to rule out symptoms associated with mass effects. This case and others reported in the international literature show that occipital neuroepithelial cysts can benefit from surgical treatment with meticulous preoperative planning. The aid of neuromonitoring is crucial to identify anatomical variations and cortical functionality that are potentially distorted in the presence of these lesions.

Keywords: Glioependymal cyst, Neuroependymal cyst, Neuroepithelial cyst, Neuroglial cyst

INTRODUCTION

Neuroepithelial cysts (NECs), also known as neuroglial cysts (NGC), glioependymal cysts (GEC), or neuroependymal cysts, are benign developmental anomalies of the central nervous system (CNS) derived from ectodermal remnants. Their distinction from choroid plexus cysts (CPCs) is unclear, but the latest classifications tend to group these lesions.[23] While reported as silent in most cases,[6,7,9] they can cause localized pressure with subsequent neurology or even show symptoms of raised intracranial pressure.[1,4,21] They can arise from any region of the brain parenchyma, but the occipital location seems to be more uncommon, and symptoms can be subtle or underestimated by clinicians. We report a case of NEC presenting with mild left-sided hemianopia. The patient was initially treated conservatively but subsequently developed worsening symptoms with parietal syndrome and, therefore, underwent surgical fenestration

with full clinical recovery. The case highlights the importance of adequate clinical assessment, management, and follow-up in such a scenario, where a benign, congenital lesion might lead clinicians to underestimate the role of aggressive surgical management.

CASE REPORT

A 60-year-old right-handed lady was referred to our neurosurgery service by her general practitioner due to 3 months clinical history of headaches and suspected apraxia. The headache was reported as non-positional, intermittent, with a frequency of a few days, primarily retro-orbital, and not exacerbated by cough or sneezing. The patient also reported an increasingly noticeable worsening of her gait, with a tendency to trip over and deviate toward the right-side during walking.

Three different neurosurgical consultants and one neurologist saw the patient. One physician reported some possible signs related to the pathology, although there was a disagreement regarding the findings. A possible extremely subtle left-sided homonymous hemianopia located at the very extreme end of the visual field and an equivocal, minor degree of gait deviation toward the left was reported. Apart from these two possible findings, neurology was unremarkable. The patient was otherwise fit and well, working as a psychotherapist, exercising regularly, and having no significant comorbidities.

Magnetic resonance imaging (MRI) was performed to rule out an intracranial mass. The scan highlighted a right parieto-occipital multiloculated cyst. A first, more minor component was isointense to the cerebrospinal fluid (CSF) on fluid-attenuated inversion recovery sequences located in the inferoposterior portion of the cyst. A second, more prominent component showed suspected proteinaceous content and was located superior-anteriorly, in close contact with the atrium of the lateral ventricle on the same side. The cyst wall did not take contrast, and there was no restriction on diffusion sequences.

The case was discussed in the department multi-disciplinary team meeting (MDT), taking into account the high performance status of the patient and the concerns related to a potentially invasive procedure close to the visual cortex. Both surgical and conservative options were discussed with the patient. All factors considered, conservative management through a wait-and-watch strategy was initially recommended and agreed on by the patient. A formal ophthalmology assessment and an MRI scan were booked to be performed after 6 months after the initial visit. The patient was provided with active contacts during working hours and instructed to attend the hospital in an emergency setting in case of acute deterioration. At that point, on visual field assessment, the suspected homonymous hemianopia was confirmed to be present, worse on the left eye compared to the right and more marked than expected [Figure 1]. Fundoscopy showed no obvious papilledema, so there was no concern about a possible rise in intracranial pressure. However, the patient's symptoms were worsening at this point: her headaches were constant, and she started to notice problems in accurately reaching objects on the left side of her visual field. The repeated MRI scan is shown in Figure 2. On repeated counseling, the patient expressed the will to consider surgery as an option. She was extensively informed about the risks, specifically regarding the possibility of cyst re-filling, possible further visual deterioration, and lack of neurological improvement, which she fully understood.

Surgical management and postoperative course

The surgical strategy was discussed in the MDT, and several options were considered. The aim of the surgery would have been to decompress the cyst and get a biopsy to exclude any malignancy or potentially growing lesion. Endoscopic fenestration was considered an option, but the chance of an inconclusive biopsy was considered too high. Moreover, given the expected variation in functional anatomy due to a suspected long-standing lesion, endoscopic manipulation near the visual cortex without adequately exposing the area was deemed to put the patient at risk of further visual loss. Open surgery was therefore considered, although the question of whether to put the cyst in communication with the ventricle remained. The available literature evidence was reviewed (see Discussion section): The concern of spreading proteinaceous content inside the ventricle and the literature trend that most of these procedures report successful open fenestration with no rate of recurrence,[2,3,7,11,24] open fenestration under neuromonitoring was chosen as the best course of action.

Surgery has been performed under total intravenous anesthesia under neurophysiological monitoring. Specifically, somatosensory-evoked potentials and visual-evoked potentials (VEPs) were running during the procedure. The patient has been positioned prone on a Montreal mattress, with the head fixed on a Mayfield clamp, and neuronavigation has been set up to identify the entry area. After disinfection and draping, a parasagittal approach and a right-sided parieto-occipital craniotomy have been performed to access the lesion.

As soon as the bone flap was removed, the VEPs changed in amplitude and shape. The dura has been, therefore, opened in a curvilinear fashion with the pedicle flap towards the midline, and the cyst position has been checked using intra-operative neuronavigation. At this point, cortical stimulation has been performed to identify the calcarine cortex and the corresponding primary visual area. Interestingly, the most superficial area of the cyst was located under a highly responsive

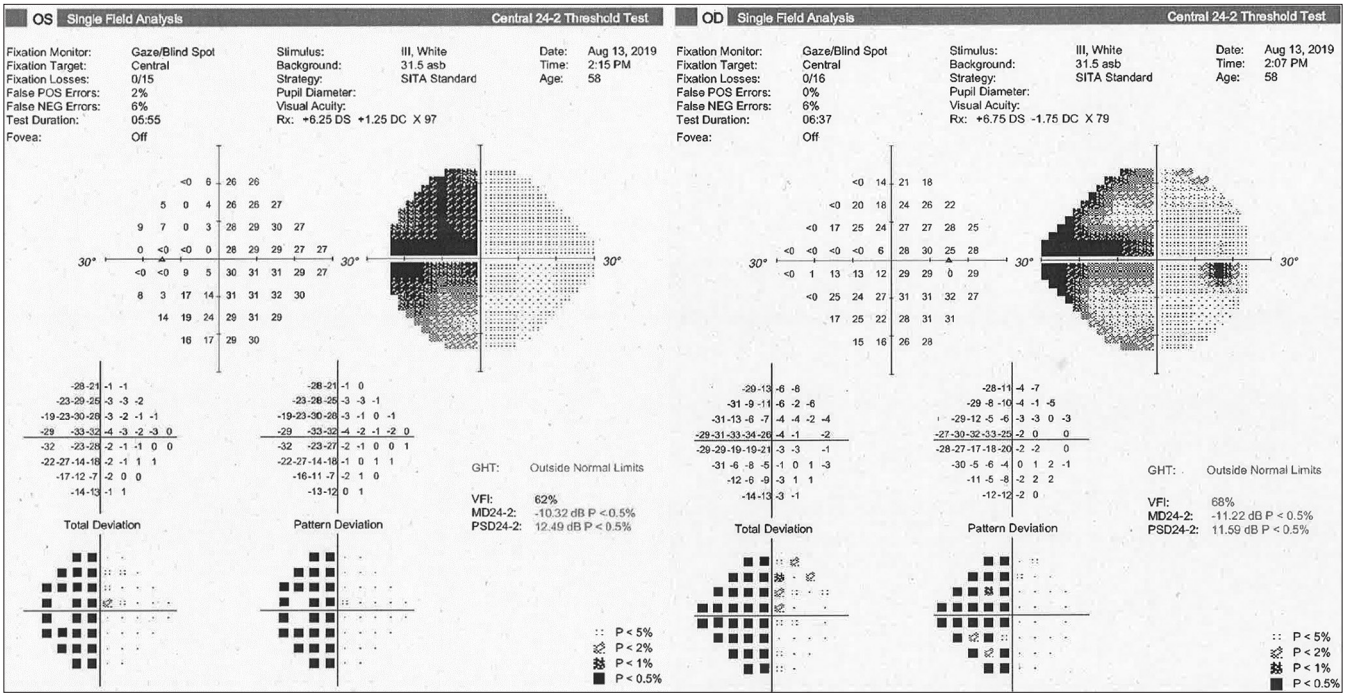

Figure 1: Preoperative Humphrey visual test of the patient showing marked left homonymous hemianopia.

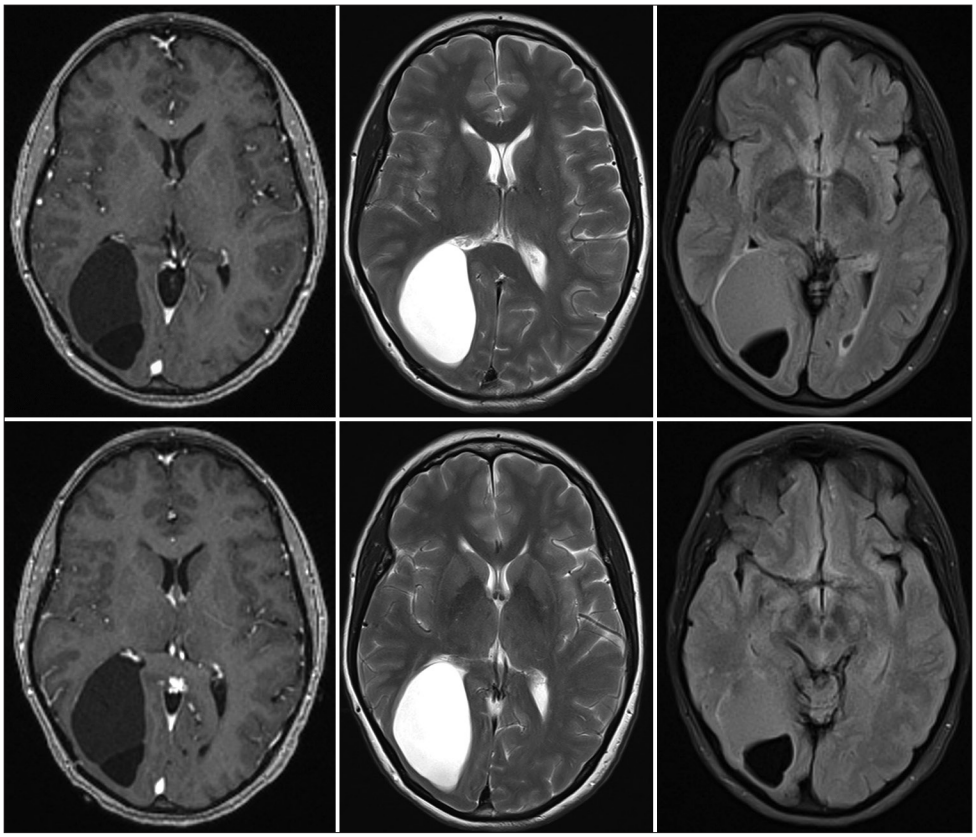

Figure 2: Preoperative magnetic resonance imaging scan, axial sequences: T1 with contrast (left), T2 (center), fluid-attenuated inversion recovery (right) showing a suspected neuropithelial cyst arising from the trigone and occupying the parieto-occipital parenchyma.

area, where clear interruption of the VEPs was achieved with a minimal amount of cortical stimulation (<3 mA). After completing the cortical mapping, a safe entry point has been established at the level of the parieto-occipital sulcus, just beneath the pre-cuneus, above a bridging vein [Figure 3] medially. A shallow subcortical dissection of approximately 1 cm depth was performed under microscope magnification, and the cyst wall at this point was identified and opened. Xanthochromic fluid came out from the cyst at moderate pressure and has been collected for cytology. A septation in the context of the inferior-medial portion of the cyst was also identified and fenestrated. Finally, samplings of the cyst wall were sent off for histology, and the cyst was left fenestrated with the subdural space. The dura was closed with 3-0 vicryl stitches, the bone flap was repositioned using titanium microplates and screws, and the remaining pericranium, sub-cutaneous, and cutaneous layers were closed using vicryl stitches and clips.

On awakening, the patient was neurologically intact, with no new neurological deficits on four limbs, cranial nerves, or changes in higher cognitive performances. She reported immediate resolution of the preoperative headache. On mobilization the next day, she did not show any obvious gait impairment, her postoperative course was uneventful, and she was discharged on day 2 postoperative. The postoperative MRI scan revealed a significant reduction in the size of the cyst on both its components [Figure 4]. At a 1-month clinic follow-up, the patient reported a complete resolution to her previously noticed gait problems and pain symptoms. A new Humphrey visual field test showed complete resolution of the hemianopia [Figure 5].

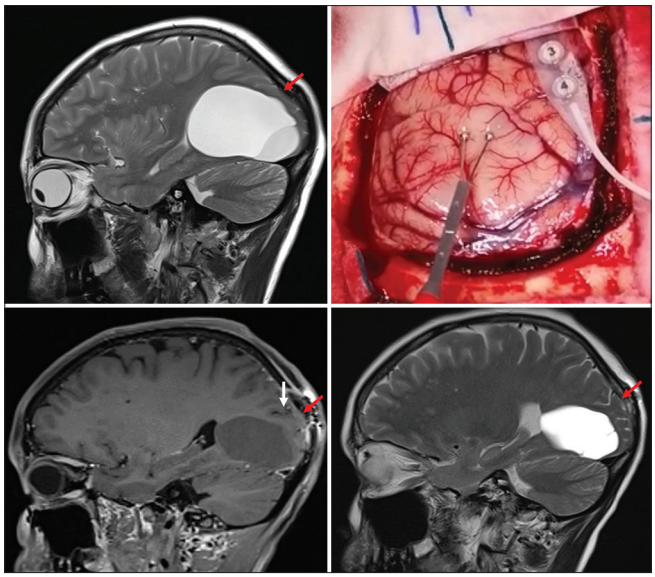

Figure 3: (Top left) Preoperative magnetic resonance imaging scan (sagittal) showing the marked compression of the occipital cortex. This was initially interpreted as non-functional/atrophic. Red arrow points towards the re-expansion of the brain tissue before and after fenestration. However, on intraoperative stimulation (top right), the most superficial point of the cortex overlying the cyst appeared to be functionally active; therefore, a more superior entry point was chosen above the draining vein seen in the surgical field. (Bottom left and bottom right) Red arrows show the re-expansion of the thin occipital cortical mantle, and white arrows point out the sulcus chosen as an entry point.

The histology revealed a neuroepithelial cyst with cuboidal epithelium, positive for cytokeratin, epithelial membrane antigen (EMA), glial fibrillary acid protein (GFAP), and negative for S-100.

DISCUSSION

NEC is rare but often misinterpreted as congenital lesions of the CNS. According to the latest classifications, the larger NEC group includes ependymal cysts and CPCs.[23] On examination under optical microscopy, all these lesions share standard features: columnar and/or cuboidal, mostly ciliated epithelium, with a variable degree of stroma and basal membrane.[7] Immunohistochemistry is variable, with various degrees of expression of cytokeratin, S-100, GFAP, and EMA.[6,11,14] NEC can either be clinically silent or symptomatic, depending on their location and size.[6] Several authors argue that NECs are clinically silent in most cases,[6,7,9] although the exact epidemiology is unclear. The prevalence seems to overlap between males and females, and the age varies from the peri-natal diagnosis[13] to late adulthood.[23] While typically isolated findings in adults, NEC and CPC show a stronger association with a variety of structural and genetic anomalies in the pediatric population, including corpus callosum dysgenesis[13,16,17] and several chromosome anomalies.[22] When symptomatic, they can show a variety of clinical presentations, including focal neurological symptoms,[1] seizures,[21] and even life-threatening increased intracranial hypertension.[4] Locations are also variable. Most authors tend to refer to the cysts inside the ventricles as CPC,[4,8,20,22] while cysts found embedded into the brain parenchyma are mostly referred to as NEC/NGC/GEC.[1,6,12,21]

To the best of our knowledge, four reports for six cases of symptomatic occipital NEC in adults are reported in the recent medical literature.[2,5,18,21] In three of such cases,[2,18] the initial presentation consisted in hemianopia. In all these cases, the radiological intensity of the cyst on the MRI signal was identical to that of the CSF, with one case showing a denser fluid level[18] suspicious of a proteinaceous component, comparable to the one we have found in our case. All these cases were treated with surgery, and all showed complete recovery.[2,5,18,21] Interestingly, one case was treated through complete cyst resection,[18] while the others were treated with fenestration,[2,5,21] but the outcome was equivalent, with no case of recurrence reported in this region. Recent mini-series and case reports regarding surgical treatment of NEC or CPC

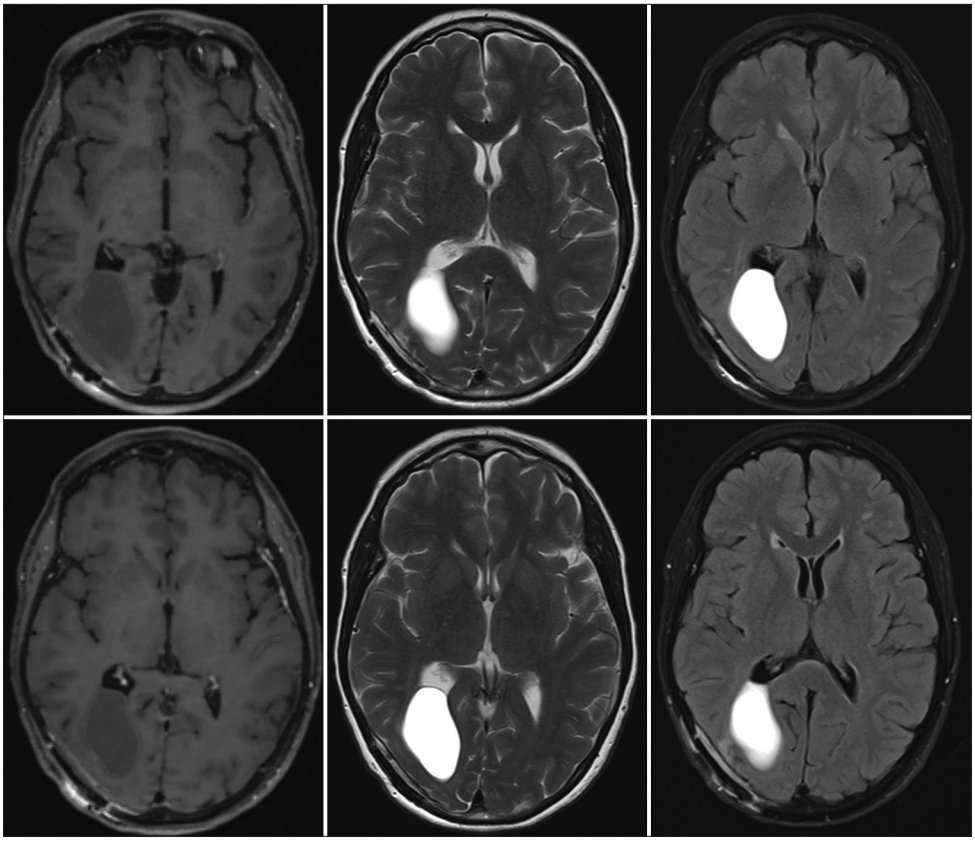

Figure 4: Postoperative magnetic resonance imaging scan, axial sequences: T1 with contrast (left), T2 (center), fluid-attenuated inversion recovery (right) showing the significant reduction in the size of both components of the cyst.

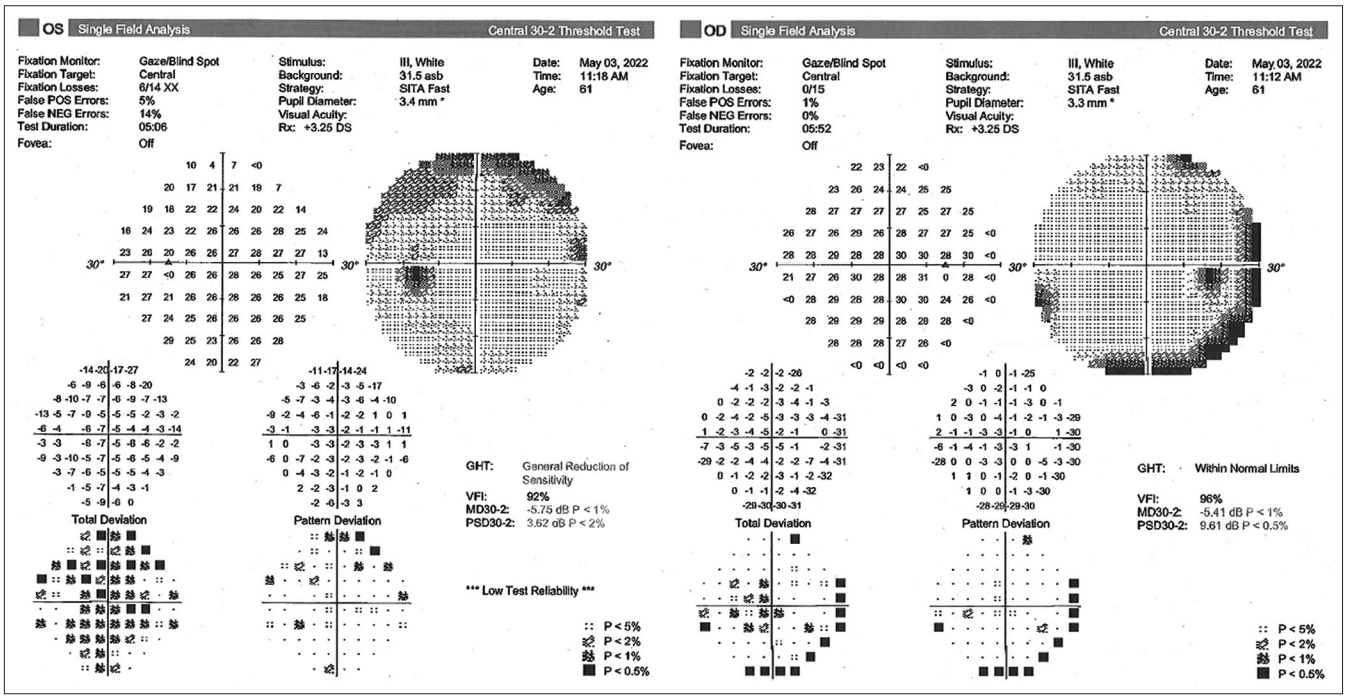

Figure 5: Postoperative Humphrey visual test showing near-complete resolution of the hemianopia.

in different locations describe various surgical strategies, showing good outcomes in most cases.[2,3,7,11,24] However, transient neurological deterioration and stroke complications were described after endoscopic fenestration,[4,19] which is one of the reasons why we opted for open surgery. The other consideration concerned the chance of expected variability in functional anatomy due to a long-standing lesion near the visual cortex.[15] In our case, a carefully planned trans-cortical approach was performed strongly considering the availability of VEPs. The cortical mapping highlighted an unexpectedly positive response in an area that was erroneously suspected to be cortical atrophy.

Interestingly, VEPs appeared to change as soon as the dura was opened. While this finding might be interpreted as a reliable adjustment of the cortex to the relieved local pressure from the cyst, VEPs are notoriously volatile and show a relatively high percentage of false positives. Therefore, caution is advised by several authors regarding their interpretation.[10] However, we have assumed that the repeated cortical mapping was correct in confirming VEP impairment on direct stimulation of the area overlying the cyst; therefore, a more superior corticectomy was performed. The postoperative radiological and clinical course confirmed an almost complete resolution of the previously diagnosed hemianopia. The remarkable regression of symptoms after surgical treatment stresses the importance of not underestimating the clinical presentation of these cysts and their natural history. Even if clinically silent for a prolonged period, they can undergo changes that can eventually cause increased intramural pressure and neurological symptoms. This eventuality should always be considered. Based on this case and the others reported in the literature, surgery appears to be a safe and resolutory option to treat symptomatic NEC.

CONCLUSION

Occipital neuroepithelial cysts are rare lesions that can occasionally present with symptoms and signs indicative of localized neurological compromise. Clinical manifestations may be subtle, necessitating comprehensive evaluation prior to discard any aggressive approach. Surgical intervention, when indicated, must be meticulously planned due to the cyst's proximity to critical structures, particularly the visual cortex and optic radiations. Despite limited data, current literature evidence suggests a favorable prognosis with a high recovery rate following surgical fenestration.

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Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

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

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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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