Surgical Management of Intracranial Giant Epidermoid Cysts in Adult: A Case-Based Update

Abstract

Epidermoid cysts (ECs) are benign and slow-growing lesions that account for about 0.2%–2% of all intracranial tumors. Symptoms appear slowly and tumors may have already grown to giant proportions when patients receive their first diagnosis. The optimal treatment for ECs is surgical removal, which includes the total resection of the entire capsule of the lesion in order to minimize the risk of malignant transformation associated with partial removal. However, considering the giant size that the ECs can reach at the time of the diagnosis, and their adherence to the surrounding structures, the risks and benefits of total versus subtotal resections in the short- and long-term patients' outcome are still under debate. Here, we report a case of an extensive giant EC and offer a discussion of its characteristics, surgical management, and postoperative outcome, taking a cue to argue about the recent literature based in the latest case studies.

Keywords: *Adult, brain cyst, giant epidermoid, surgical treatment, total resection*

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Introduction

Epidermoid cysts (ECs) are benign and slow-growing masses that account for about 0.2%–2% of all intracranial tumors.[1] ECs are congenital and formed out of aberrant ectodermal cells, which become trapped during the embryogenesis between the 3rd and 5th gestational weeks.[2] They histopathologically appear as pearly lesions lined with stratified squamous keratinizing epithelium.[3]

Intracranial ECs are most often intradural, but they can also occur extradurally in the intradiploic space in up to 10% of cases.[1] They tend to insinuate into several intracranial compartments by filling the subarachnoid space. The most common intradural locations are cerebellopontine angle (60%), fourth ventricle (5%–18%), parasellar area, and middle cranial fossa (15%); less frequently, they are located within ventricles and brain parenchyma.[4,5] ECs of the posterior fossa usually arise in the lateral subarachnoid cisterns, and those involving the brainstem and spine are rare.[6-11]

Surgical resection is the only affective treatment,[3,12] but the debate is still open regarding the advantages of a total resection versus a decompressive procedure with subtotal removal. Relatively few case series have been yet reported on giant ECs, and accounts vary in consideration of recurrence risk and patient outcomes in the short and long term [Table 1].[3,12-21]

Here, we report a case of a giant EC and provide a discussion of its characteristics, surgical management, and postoperative outcome, taking a cue to argue about the recent pertinent literature.

Case Report

A 60-year-old woman presented with a history of mild right hemiparesis, complex partial seizures, aphasia, and gait disturbances, without headache. Neurological examination confirmed the right hemiparesis, gait ataxia, and nystagmus on lateral gaze. Magnetic resonance imaging (MRI) showed a giant cystic lesion (90 mm × 80 mm × 60 mm) centered in the third ventricle, with bilateral extension toward cerebral hemispheres. The cyst was isointense on T1- and T2-weighted images and hyperintense on diffusion-weighted imaging (DWI), without contrast enhancement. It was composed of an anterior part compressing the septum pellucidum and extending in the intraventricular space; an anteroinferior

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portion insinuating toward the posterior fossa compressing the brainstem, aqueduct, basal cisterns, and left cerebral peduncle; and a superoposterior portion occupying the semioval centrum, with bilateral mesiotemporal extension to the hippocampus regions. Perilesional edema was not present [Figure 1].

A median occipital craniotomy was performed in prone position. After bilateral dural opening hinged on sagittal sinus, the cyst was evident on both sides beneath the pia mater, which was easily distinguished as a pearly and avascularized lesion. A small bilateral corticotomy was performed and the cyst was gradually suctioned and microsurgically separated from the surrounding structures of the internal venous system. The capsule appeared tenaciously attached to the neurovascular structures, impossible to be removed. The postoperative course was uneventful, with progressive improvement of the preoperative motor and visual field deficits. Histopathologic diagnosis consisted with epidermoid tumor.

Postoperative MRI revealed subtotal resection with residual tissue deeply located [Figure 2].

Neurological examination at discharge (1 week after surgery) revealed a complete resolution of the hemiparesis but with the persistence of gait ataxia and nystagmus.

At 1-year follow-up, gait ataxia and nystagmus were completely absent and the patient did not complain headache anymore.

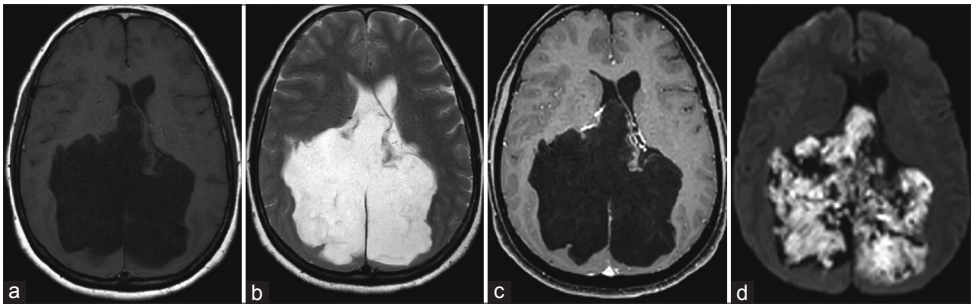

Figure 1: Preoperative MRI. Axial T1-weighted (a) and T2-weighted (b) MRI showing the low- and high-intensity signal of the lesion which seems to be pineal gland centered with extending from the posterior to the anterior fossa and insinuating into the brainstem, aqueduct, basal cisterns, and left cerebral peduncle. The lesion has no contrast enhancement (c). Diffusion-weighted imaging (d) shows the typical hyperintense and bright tones of the lesion. MRI – Magnetic resonance imaging

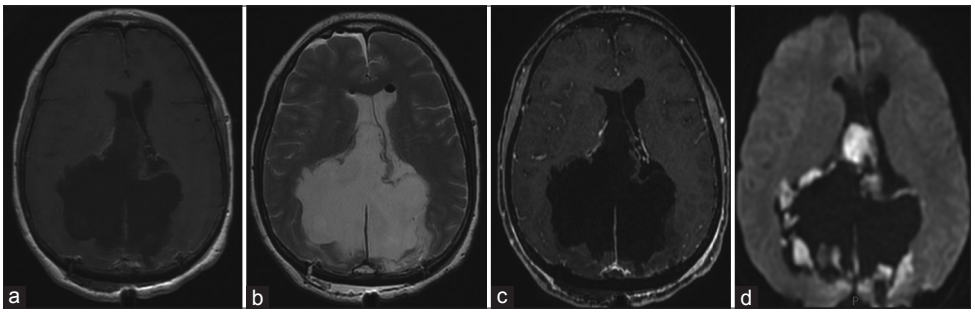

Figure 2: Postoperative MRI. Axial T1-weighted (a), T2-weighted (b), T1-gadolinium-enhanced (c), and DWI (d) images showing the subtotal resection of the lesion. DWI images show the residual pathological tissue around the surgical cavity, mainly in the mesial temporal lobe bilaterally and perimesencephalic cistern. MRI – Magnetic resonance imaging; DWI – Diffusion-weighted imaging

Table 1: Summary of studies (case series) about surgery of giant epidermoid cysts

Authors & Year	No. of cases	Tumor location	Total removal	Post-operative CN deficits	Recurrence	Mean follow-up (yrs)	Aseptic Meningitis	Long-term CN deficits
Yaşargil *et al*., 1989[21]	35	Supra/infratent	97%	20%	0	5.2	18.06%	2.85%
Darrouzet *et al*., 2004[17]	8	CPA	62.5%	50%	0	3.7	12.5%	37.5%
Safavi-Abbasi *et al*., 2008[22]	12	CPA	75%	16%	0	2.25	0	8%
Chowdhury *et al*., 2013[23]	23	Supra/infratent	73.9%	8.7%	0	3.25	0	8.7%
Mohanty *et al*., 1997[18]	25	Supra/infratent	48%	50%	0	3.6	0	8%
Goel *et al*., 2006[20]	96	Supra/infratent	48%	19%	2.1%	4.3	2.1%	16%
Lynch ey al., 2014[15]	33	Supra/infratent	72.7%	33%	9%	7.2	0	21%
Kato *et al*.,2010[16]	24	Supra/infratent	95%	45%	16.6%	5	4.1%	40.9%

CN: cranil nerve, CPA: cerebellopontine angle, yrs: years

MRI appeared superimposable to the postoperative one, showing neither cyst regrowth nor signs of chemical meningitis.

Discussion

Benign ECs are prevalently diagnosed in males during the 3rd–4th decade, but turn malignant predominantly in females.[22,23] ECs form from the accumulation of keratin and cholesterol, which desquamate into a pearly material within their walls, growing in the cisternal spaces and remaining asymptomatic for years due to the absence of initial mass effect.[11,22]

The usual pattern of EC growth is linear, and symptoms appear when ECs have grown to giant proportions, varying according to their location. They include hearing loss, dizziness, gait disturbance, trigeminal neuralgia, tinnitus, diplopia, visual impairment, apathy, headache, gait ataxia, epilepsy, raised intracranial pressure, and recurrent aseptic meningitis.[3] EC rupture is uncommon and may cause chemical meningitis and hydrocephalus.[22]

On computed tomography scans, benign ECs appear as hypodense or isodense areas, without contrast enhancement, although the presence of protein, lipid, calcium, and hemosiderin can occasionally make them appear hyperdense.[24]

On MRI, ECs show a multilobulated appearance and are hypointense on T1-weighted and hyperintense on T2-weighted images and DWI.[25] Benign ECs usually exhibit no contrast enhancement and peritumoral edema.[26,27] Fluid-attenuated inversion recovery and DWI sequences represent the main sequences to test for solidity and diagnose ECs. DWI offers a superior view of the EC borders and their characteristic bright tones, which help to distinguish them from arachnoid cysts, dermoids, lipomas, cholesterol granulomas, hamartomatous, cystic neoplasms, neurocysticercosis, and neuroenteric cysts.[8,22,28] The presence of edema, tissue invasion, rapid growth, and new contrast enhancement suggests malignant transformation.[29,30]

Hydrocephalus is not typical, as the ECs' pace of growth is gradual and its crevices occasionally awash with cerebrospinal fluid.[31]

Surgical debulking with capsule removal is a definitive treatment,[3,32] although it presents numerous challenges, especially in case of giant multicompartmentalized ECs because of the capsule adherence to many neurovascular structures.[19] The cyst content can be easily suctioned, but its spillage into the subarachnoid space may cause postoperative aseptic meningitis (2%–50% of cases), leading to dense adhesions between the residual capsule and cranial nerves and vessels.[31] A limited resection is sometimes recommended to minimize comorbidities,[19,22] but it needs to take into account the higher risks of recurrence (occurring on an average after a symptom-free interval of 7.74 years in 1%–54% of cases), and more rarely, of malignant transformation.[13,23]

In consideration of the higher morbidity of recurrent ECs, the latest studies take the view that total resection is associated with improved function and low mortality and should be recommended as the ideal goal of treatment.[3,22]

In our view, the case for total or subtotal resection should be evaluated on the basis of the following demographic, clinical, and neuroradiological parameters. First, older patients have a lower risk of recurrence compared to younger ones. Second, in patients with minor symptoms, the primary goal of treatment is not to cause iatrogenic morbidity. Third, resectability depends, in all cases, on preoperative neuroradiological considerations about size, number of compartments, and relationships with neurovascular structures.

Conclusion

In the management of giant intracranial ECs, a total debulking with capsule removal dramatically reduces the risk of recurrence and malignant transformation. In the surgical setting, however, it is crucial to consider all the features associated with a risk of iatrogenic morbidity, which is best avoided in case of benign lesions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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