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TECHNICAL NOTE - NEUROSURGICAL TECHNIQUES

Cyst-cisternal shunting for cystic multirecurrent brainstem epidermoid: case report and literature review

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Abstract
Background Surgical treatment of recurrent, posterior cranial fossa epidermoids in multioperated patients carries significant morbidity, mainly due to tumor adhesion to cranial nerves and vessels, and brainstem involvement. Radical resection is often not feasible; therefore surgery should aim to restore cerebrospinal fluid circulation, release engulfed neurovascular structures, and relieve brainstem compression. Intra-axial epidermoids are extremely rare. We present an innovative surgical technique of a cyst-cisternal shunting to treat cystic recurrent, unresectable brainstem epidermoids.
Methods The surgical technique is stepwise described and a case illustration is reported. The pertinent literature has been reviewed.
Results Few cases of brain stem epidermoid tumors have been described to date. The surgical steps of this technique and related intraoperative images are provided. One case illustration regarding the resection of a large recurrent cystic intra-axial brainstem epidermoid is reported to demonstrate the application of the technique in a clinical setting. The patient was followed up for 14 years and did not experience any recurrence, showing a stable disease at the last follow-up control. A systematic review of the competent literature has been provided.
Conclusions Cyst-cisternal shunting in case of recurrent, brainstem epidermoid is a safe and long-term effective technique to relieve mass effect into the brainstem.

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Keywords Epidermoid · Brainstem · Shunt · Cyst · Brain tumor

Introduction

Epidermoids are rare, benign, slow-growing lesions that arise from retained ectodermal remnants. They account for 0.2–1.8 % of all intracranial tumors and up to 7 % of cerebellopontine angle (CPA) tumors [23, 24, 29]. Lesions characteristically spread along anatomical cleavage planes, progressively filling the subarachnoidal spaces.

Despite being histologically benign, epidermal cysts pose a neurosurgical challenge, mainly in case of recurrent tumors. Surgical removal is the treatment of choice, which should aim for an extensive resection with preservation of the patient's neurological function [29].

Radical surgery is not always achievable and carries significant risk of perioperative complications including cranial nerve deficits, hydrocephalus, and aseptic meningitis, due to the release of epidermoid breakdown products [2]. Because of the high surgical morbidity, balance between extent of resection and a patient's functional preservation might be taken into consideration, especially in multioperated patients, harboring recurrent lesions with intra-axial extension.

The authors describe an innovative technique for cisternal shunting of recurrent, cystic, unresectable brainstem epidermoids.

Study design

The authors describe the case of a recurrent cystic brainstem (BS) epidermoid tumor. A literature review has been performed on PubMed and Google Scholar, using the search

terms "brainstem, epidermoid, cerebellopontine angle, cyst, and recurrent".

A total of 24 papers reporting the management of BS epidermoid tumors resulted from the electronic search. Papers report both pediatric and adult cases. Table 1 summarizes data obtained from the literature review.

Case report

A 5-year-old female was referred to our institution in January 2001 with cognitive impairment, dysphagia, and gait disturbances.

The neurological examination at hospital admission revealed left hemiparesis and a complete right VI cranial nerve palsy. A contrast-enhanced MRI showed a voluminous cystic lesion of the prepontine cistern extending into the pons.

A right retrosigmoid approach was performed; because of brainstem invasion, tumor capsule could not be completely peeled off. Histological examination was diagnostic for epidermoid tumor. Postoperative course was uneventful, with a complete recovery of the hemiparesis and of the VI cranial nerve palsy.

In September of 2002, a contrast-enhanced MRI pointed out a small intraparenchymal cystic tumor recurrence at the medullary-pontine junction (Fig. 1a, b); since the patient was asymptomatic, a wait-and-see approach was undertaken.

In January of 2003, the patient started complaining of worsening headaches, with a MRI showing a volumetric increase of the intraparenchymal cystic component, which split brainstem white matter fibers, causing severe mass effect.

The patient underwent right retrosigmoid approach with magnum foramen decompression. Cisternal arachnoid adherences were resected through microdissection technique, freeing neurovascular structures from scaring adhesions in order to restore cerebrospinal fluid circulation.

The intraparenchymal lesion component was opened through smooth dissection and the lining tissue gross-totally removed, taking care to avoid spillage of cyst contents into the subarachnoid space. Perioperative administration of corticosteroid and intraoperative irrigation with hydrocortisone was adopted to minimize the risk of chemical meningitis. Once the cyst was completely emptied, useless attempts to peel tumor capsule, tightly attached to the brainstem, were avoided.

Residual cyst was shunted into the cisternal space using a small catheter, which was left in place at the opening site of the brainstem lesion. The catheter was modeled in a J-shaped fashion using a 3-0 silk stitch, which helped to maintain the shunt system in place and to direct drainage flow according to spinal fluid circulation (Fig. 2).

The postoperative course was uneventful (Fig. 1c, d). The patient was followed up for 142 months after the shunting operation. At the last clinical evaluation, the neurological exam was within the norm without radiological evidence of recurrence (Fig. 1e, f).

Discussion

Internal shunting of intracranial cysts has already been described in rare instances, for example in case of cystic craniopharyngiomas or arachnoidal cysts, to release tumor-related compression to the surrounding tissue, as an alternative therapeutic option in patients not eligible for surgical resection [3, 7].

To the best of our knowledge, only 24 cases of brainstem epidermoid tumors have been previously reported, both in the pediatric and adult population, with disappointing results in terms extent of surgical resection, recurrence rate, and postoperative morbidity (Table I) [1, 4–6, 8, 10–22, 25–28, 30, 31]. The present case represents the one with the longest follow-up ever published in the literature.

As widely known, since epidermoids are not sensitive for radiation/chemotherapy, making surgery the treatment of choice for these lesions. Over the last decades, the spread of endoscopy and the advancements made in neurosurgical techniques have dramatically improved the results of CPA epidermoid surgery. Nevertheless, resection of these tumors still requires high technical skills, mainly because of their invasive growth pattern, which put tumor bulk in tight relationships to critical neurovascular structures.

A crucial point in a surgeon's decision-making is weather to stop or proceed with the peeling of the tumor capsule from neurovascular structures. Being non-neoplastic, slow-growing lesions, loss of function in order to achieve complete tumor removal is not justifiable. This means that the surgeon should take care to prevent any morbidity and should not attempt to remove the portion of tumor capsule tightly attached to neuro-vascular structures [24, 29]. For this reason, compared to overall data referred to extent of resection of intracranial epidermoids, CPA lesions carry a relatively lower gross-total resection rate, which account for less than 75 % of cases [23].

Subtotal resection can lead to progressive production of keratin, resulting in cyst recurrence and risk of chemical meningitis [24, 29]. Notably, it occurs in less than 40 % of patients undergoing subtotal resection and it is directly related to the amount of residual cyst [9, 23]. Nevertheless, perioperative administration of corticosteroid and intraoperative irrigation of the surgical site with hydrocortisone has been proven to be effective in minimizing that risk. Moreover, the use of steroids is reported to reduce the risk of postoperative communicating hydrocephalus, which

Table 1 Review of the literature

Author, year	Age/Sex	Location	Clinical presentation	Signs	Management	Outcome
Bhatia et al. [4]	3.5 years/M	PM	Ataxia Delayed milestones	Hemiparesis, papilledema V, VI, VII CN deficit	Shunting, aspiration	Meningitis Death 2 weeks after surgery
Leal and Miles [15]	3.5 years/F	Medulla	Meningitis Hemiparesis	Hemiparesis, papilledema, VI, VII CN deficit	Aspiration STR	Minimal improvement Posterior fossa abscess, Death 2 months later
Schwartz and Balentine [25]	14 years/M	Pons	Meningitis Hemiparesis	Hemiparesis, VII CN deficit	Shunting	Progressive BS function deterioration; Death
Weaver and Coulon [28]	1 year/M	Pons	Diplopia Facial weakness	VI, VII CN deficit	Aspiration STR	Meningitis, Symptoms recurrence after 10 weeks
Ogawa et al. [19]	38 years/F	PM	Diplopia, ataxia	Hemiparesis, VI, VII CN deficit	Aspiration STR	Meningitis, pneumonia, death after 3 months
Guy et al. [11]	25 years/F	Pons	Meningitis, ataxia	Hemiparesis	Evacuation	Tracheostomy, improvement
Iihara et al. [12]	32 years/M	Pons	Diplopia, ataxia	Gaze palsy, VII CN deficit	STR	Meningitis, improvement
Obana and Wilson [18]	27 years/M	Pons	Diplopia, ataxia	Gaze palsy	STR	Improvement
	27 years/F	Medulla	Hemiparesis	Gaze palsy	GTR	Improvement
	37 years/M	PM	Ataxia, hearing impairment	Hemiparesis	STR	Improvement
Fournier et al. [8]	14 months/M	PM	Gait disturbances	Quadriparesis Ataxia, VII CN deficit	STR	Multiple recurrences, Death 19 months after surgery
Radha Krishnan et al. [21]	13 years/F	PM	Headache Ataxia Dysphagia Diplopia	Papilledema Gaze palsy Gag reflex deficit VII CN deficit	Shunting RT	Death
Kuzeyli et al. [14]	2 years/M	Pons	Headache	Hemiparesis VII CN deficit	STR	Asymptomatic at 5 months FU
Yoshizato et al. [30]	69 years/F	Pons	Hemiparesis	Hemiparesis	GTR	Improvement
Malcolm et al. [16]	25 years/M	PM	Ataxia	VI CN deficit	GTR	Improvement
Kachhara et al. [13]	55 years/M	Medulla	Ataxia	Hemiparesis	GTR	Improvement
Sinha et al. [26]	38 years/F	Pons	Hemiparesis, ataxia	VI, VII CN deficit	STR	Improvement
Caldarelli et al. [5]	18 months/F	PM	Irritability Behavioral alterations Ataxia	Mild neck stiffness	Aspiration GTR	Recurrence at 18 months FU, Surgery (STR)
Caldarelli et al. [6]	2 years/F	PM	Behavioral disturbance, cerebellar symptoms	NA	STR	Recurrence, surgery (STR)
Ziyal et al. [31]	5 years/F	Medulla	Dysphagia Diminished gag reflex, hoarseness	IX, X, XI, XII CN deficit	GTR	CN deficits gradually improved
Recinos et al. [22]	17 months/F	PM	Hemiparesis, facial weakness Ataxia	VII CN deficit Gaze paresis	GTR	Asymptomatic at 2-year FU
Takahashi et al. [27]	10 years/F	PM	Dysphagia Post-prandial vomiting	Bilateral VI CN deficit	ETV Aspiration STR	Surgery for multiple recurrence Persistent VI and VII CN deficit
Ahmed et al. [1]	13 months/F	PM	Recurrent meningitis	NA	STR	Meningitis, Death after 33 months
Gopalakrishnan et al. [10]	6 years/F	Pons	Intermittent headache	None	Aspiration GTR	No postoperative deficits
	2 years/M	PM	Hemiparesis	Hemiparesis	Aspiration	Hemiparesis improvement

Table 1 (continued)

Author, year	Age/Sex	Location	Clinical presentation	Signs	Management	Outcome
					GTR	
Patibandla et al. [20]	5 years/F	PM	Headache	Swallowing impairment	STR	Survived with small recurrence
Mishra et al. [17]	15 years/F	PM	Quadriparesis Diplopia Dysphagia Dysarthria	Quadriparesis V, VI, IX, X, XII CN deficit	GTR	Mild transient VI CN deficit

M male, *F* female, *CN* cranial nerve, *FU* follow-up, *BS* brainstem, *RT* radiotherapy, *ETV* endoscopic third ventriculostomy, *NA* not available, *PM* ponto-medullary, *STR* subtotal tumor resection, *GTR* gross total tumor resection

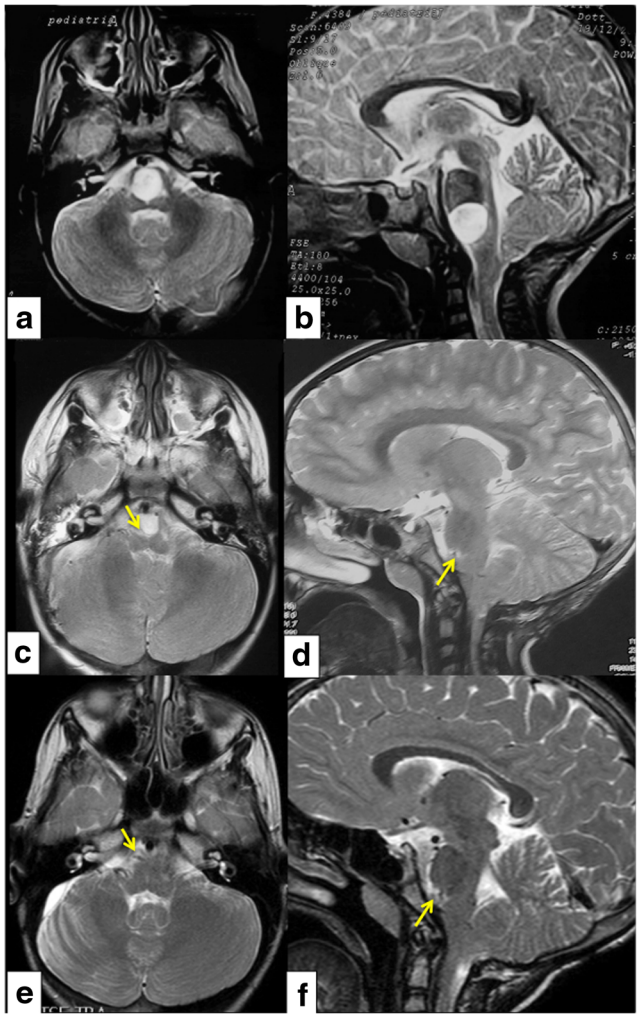

Fig. 1 MRI study showing tumor recurrence on axial (**a**) and sagittal (**b**) T2-weighted images; postoperative MRI study showing the presence of the cystic-cisternal shunt (*yellow arrow*), with complete relief of tumor bulk-related mass effect on axial (**c**) and sagittal (**d**) T2-weighted images; MRI study, performed at last FU, showing the persistence of the cyst-cisternal shunt (*yellow arrow*) without disease progression on axial (**e**) and sagittal (**f**) T2-weighted images

may develop after an intense period of meningitis or following leakage of the cyst contents [23, 29].

Besides the complication following subtotal removal, the main factors, which have to be considered, in terms of extent

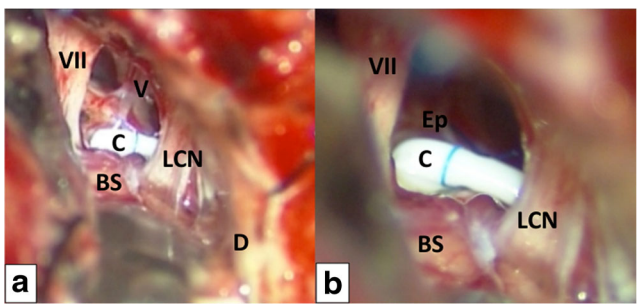

Fig. 2 The intraoperative placing of the cyst-cisternal shunt: overview of the CPA after the placing of the shunting system (**a**); particular of the catheter at higher magnification (**b**). *BS* brainstem, *C* catheter, *D* dura, *Ep* epidermoid, *LCN* lower cranial nerve, *V* fifth cranial nerve, *VIII* eighth cranial nerve

of surgical resection, are the patient's age, adhesion of the capsule to surrounding neurovascular structures, number of previous surgeries, and the patient's preoperative neurological status. Surgery of recurrent intra-axial lesions should aim to relieve brainstem compression due to tumor mass effect. In these patients, balance between extent of resection and functional preservation might lead to subtotal removal, carrying a low risk of long-term recurrence. Even in case of subtotal resection, indeed, the overall estimated recurrence rate of these tumors is 24 % of cases [2].

The authors described an innovative surgical technique for the shunting of recurrent, cystic, unresectable epidermoids into the subarachnoid space through a J-shaped ventricular catheter. The described procedure avoids dangerous attempts to peel the tumor capsule, often not distinguishable from the brainstem parenchyma. The catheter is modeled in order to stay in place indefinitely, acting as a shunt system for the cyst contents clearance.

Conclusions

Cyst-cisternal shunting in case of recurrent, intra-axial brainstem epidermoids appears to be a safe and long-term effective technique to relieve mass effect and brainstem compression.

Compliance with ethical standards

Conflict of interest The authors report no conflicts of interest.

Funding No funding was received for this research.

Ethical standards All procedures were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed consent Informed patient consent was obtained from next of kin for inclusion in this technical note.

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