Case Report

A Giant Tumefactive Perivascular Space: A Rare Cause of Obstructive Hydrocephalus and Monoparesis

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Abstract

Cerebral perivascular spaces (PVSs), otherwise known as Virchow-Robin spaces, are interstitial fluid-filled channels, <2 mm in diameter that form around arterial perforators as they course from the cortex into the brain parenchyma. In contrast, a giant tumefactive PVS is a rare entity comprising of clusters of such channels larger than 15mm resembling a neoplastic process as the name suggests. We report a 55-year-old male who presented with unsteady gait, cognitive decline, and left lower limb weakness for 6 months. Magnetic resonance imaging revealed a noncontrast enhancing multicystic intraaxial lesion of the right mesencephalon-diencephalon junction extending into the anterior third ventricle causing obstructive hydrocephalus. A ventriculoperitoneal shunt was inserted with a complete reversal of his neurological symptoms. Such PVSs can easily be misidentified for a cystic tumor, and their unique radiological features are discussed to prevent unnecessary surgery. We also demonstrate that when they cause hydrocephalus and midbrain compression symptoms cerebrospinal fluid shunting alone can result in excellent outcomes.

Keywords: *Cerebrospinal fluid shunting, giant tumefactive perivascular space, hydrocephalus, Virchow–robin space*

Introduction

Intracranial giant tumefactive perivascular spaces (TPVS) are rare clusters of nonneoplastic cysts >15 mm in size.[1] They are pial-lined, interstitial fluid-filled structures that accompany penetrating arteries and are generally located at the mesencephalothalamic region.[1] Fewer than 80 cases have been reported in the literature, and surgical intervention may be necessary when they become symptomatic. We describe a patient that experienced neurocognitive decline and limb weakness that was subsequently diagnosed to have a giant TPVS of the mesencephalon-diencephalon junction with obstructive hydrocephalus.

Case Report

A 55-year-old male experienced frequent falls for 6 months associated with progressive memory loss. Physical examination revealed left lower limb weakness of Medical Research Council Grade 4/5. The Neurobehavioral Cognitive State Examination (NCSE) and the Montreal Cognitive Assessment (MOCA) revealed severe deficiencies in short-term memory with the latter score being 26/30. Magnetic resonance imaging (MRI) depicted an irregular noncontrast enhancing multicystic lesion of the right cerebral peduncle extending into the third ventricle that caused obstructive hydrocephalus at the level of the Foramen of Monro [Figure 1a-f]. Contrast T1-weighted sequences showed that the thalamoperforating arteries coursed through the lesion at the level of the mesencephalon-diencephalon junction [Figure 1d]. Perilesional edema could not be demonstrated on the fluid-attenuated inversion recovery sequence. Diffusion weighted-imaging and apparent diffusion coefficient sequences did not reveal signal restriction within the cysts [Figure 1g and h]. MR perfusion showed perilesional decreased cerebral blood volume. MR spectroscopy showed no increased choline content with normal choline/creatine and choline/N-acetylaspartate ratios. The radiological findings were highly suggestive of a giant TPVS. A ventriculoperitoneal shunt with a programmable valve was inserted uneventfully. During shunt placement, the cerebrospinal fluid (CSF) opening

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pressure was relatively high at 22 cmH_2O (16 mmHg). Collected CSF specimens showed no evidence of tumor cells and no microorganisms were cultured. Six weeks after the operation, the patient experienced full neurological recovery. His MOCA score was 30/30, and all NCSE domain scores were within the normal range. A year later, the patient remained asymptomatic, and a follow-up computed tomography scan showed resolution of the transependymal edema with no significant change in TPVS and ventricular size [Figure 1i and j].

Discussion

Cerebral PVSs, also known as Virchow–Robin spaces, are physiological interstitial fluid-filled channels typically <2 mm in diameter that extends from the subpial space and form around arterial perforators as they course from the cortex into the brain parenchyma.[1] The precise functions of these structures have yet to be delineated, but predominant theories suggest that they: (1) Facilitate fluid movement between the basal cisterns to the interstitial space, (2) Modulate immune responses by providing a conduit for macrophages and lymphocytes to reach CSF, and (3) Forms part of the glymphatic system for metabolic waste product elimination.[2] PVSs are considered dilated when they become larger than 2 mm and are frequently observed with advancing age, various neuropsychiatric disorders, multiple sclerosis, microvascular disease, and traumatic brain injury.[1] A retrospective review of 816 MRI scans performed for various indications found that 38% of adult patients had dilated PVSs.[3]

The cause for PVS dilatation is unclear, hydrodynamic disturbances in CSF and interstitial fluid flow caused by slow-growing benign tumors or preexisting hydrocephalus have been suggested.[4] Alternatively, increased vessel permeability with fluid exudation due to microvascular disease or ex vacuo periarteriolar ischemic parenchymal injury resulting in interstitial fluid leakage have also been postulated.[5,6]

Dilated PVSs can be classified into three types with respect to their anatomical arterial relationship.[7] Type I lesions are located along the lenticulostriate arteries as they cross through the anterior perforated substance into the basal ganglia. Type II PVSs surround the cortical medullary arteries as they descend into the gray-white matter junction. Type III PVSs are located in the mesencephalic region and may follow the collicular, thalamoperforating, paramedian

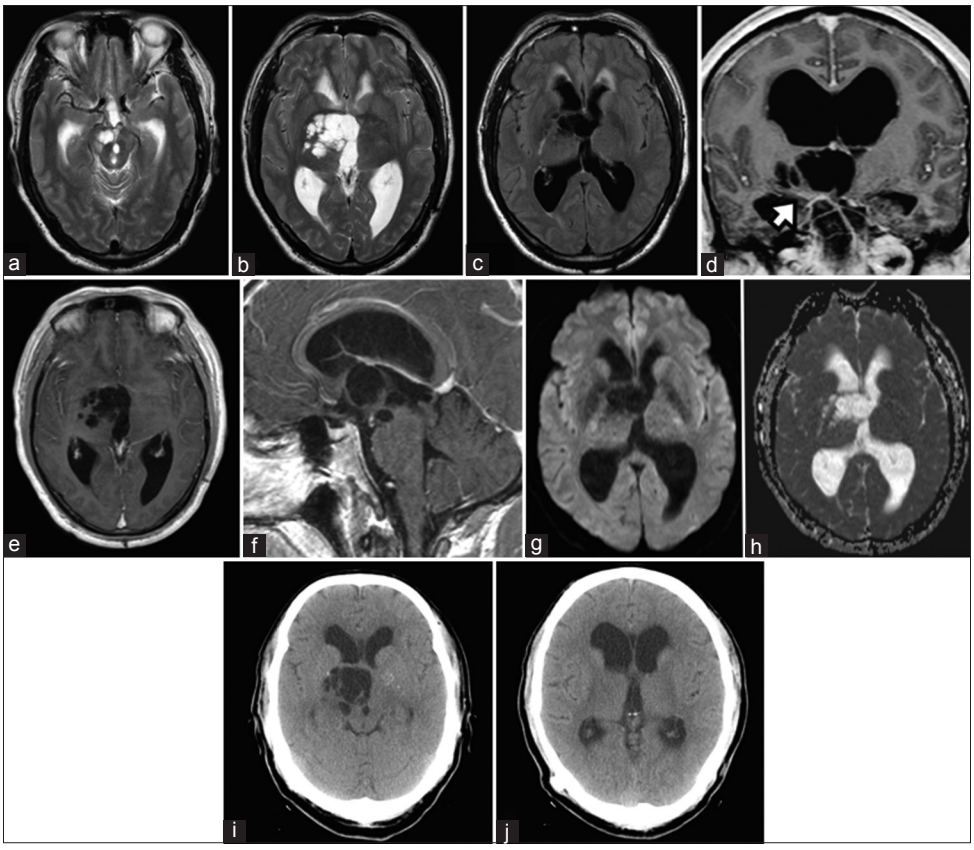

Figure 1: Multicystic perivascular spaces at the right cerebral peduncle of the midbrain that extended to the mesencephalon-diencephalon junction (a and b) Fluid-attenuation inversion recovery imaging revealed an absence of perilesional edema, but the presence of transependymal edema secondary to hydrocephalus (c). Thalamoperforating arteries as they coursed through the perivascular spaces (d, white arrow). The perivascular spaces did not display contrast enhancement (e and f). Diffusion weighted-imaging (g) and apparent diffusion coefficient (h) sequences showed that perivascular spaces fluid content had no restricted diffusion. One-year postoperative computed tomography scans showed no change in perivascular spaces (i) and ventricular size (j)

mesencephalothalamic, and circumferential penetrating arteries.

Although dilated PSVs are commonly encountered, fewer than 80 giant TPVSs (defined as being >15 mm) have been reported in the literature. Among this group of patients, 45 (61%, 45/74) had obstructive hydrocephalus and 37 (50%, 37/74) required CSF diversion. All 37 TPVSs were Type III lesions due to their proximity to the third ventricle and the Sylvian aqueduct [Table 1]. Cases reported by Salzman *et al*.[6] were excluded since no individual clinical or radiological features were described in their study to allow in-depth analysis.[1,4,8-28] Our pooled analysis showed that 49% (18/37) of TPVSs in this group had accompanying midbrain-localizing neurological signs. Apart from hemiparesis, patients were reported to have rubral tremors, oculomotor nerve palsy, Benedikt's syndrome, Parkinsonism, Parinaud's syndrome, and cerebellar ataxia.[4,8,10,12-20,29] The mean age of diagnosis was 41-year-old (range: 6–74) with a female-to-male ratio of 1: 1.9. Patients with giant TPVSs were considerably younger than those with dilated PVSs *per se*.[3]

Giant TPVSs can morphologically resemble neurocysticercosis, cystic low-grade gliomas, porencephalic cysts, ventricular diverticulae, and protein deposition disorders such as mucopolysaccharidosis on MRI.[6,7] However, PVSs are typically sharply demarcated, nonenhancing, purely cystic (displaying signal intensities similar to CSF on all sequences) and are often located along characteristic perforator vessel locations as described by Kwee[6,7] In contrast, the presence of perilesional edema, cyst content exhibiting restricted diffusion, intracystic solid, or enhancing components are more indicative of a neoplastic or infectious process.[6]

The management of Type III TPVS should address

Table 1: Reported cases of giant tumefactive perivascular spaces with obstructive hydrocephalus treated by cerebrospinal fluid diversion

Author/year	Age/sex	Presenting symptoms	Symptom duration	Location	Kwee type	Surgery type	Postoperative regression of cyst (yes/no)
Poirier *et al*./1983[16]	54/female	Cognitive decline, apathia, apragmatism, and ataxia	≥1 year	Mesencephalon-diencephalon junction	III	CSF shunt	NA
Derouesné *et al*./1987[4]	66/female	Parinaud's syndrome with unsteady gait and urinary continence	NA	Mesencephalon-diencephalon junction	III	VA shunt	No
Ono *et al*./1994[29]	26/male	Cognitive decline, headache, unstable gait, urinary incontinence, and Benedikt's syndrome	6 years	Mesencephalon-diencephalon junction	III	VP shunt and cystectomy	Yes
Schroeder *et al*./1996[17]	32/female	Headache and hemiparesis	16 years	Mesencephalo-diencephalon junction	III	ETV and cyst fenestration	Yes
Homeyer *et al*./1996[10]	42/male	Parinuad syndrome and blurring of vision	7 years	Mesencephalo-diencephalon junction	III	VP shunt	NA
Mascalchi *et al*./1999[8]	58/female	Parinaud's syndrome, tremors, unstable gait and urinary incontinence	3 months	Mesencephalon	III	VP shunt	NA
	55/male	Cognitive decline and unsteady gait	1 year	Cerebral peduncle and mesencephalic tegmentum	III	ETV	NA
Kanamalla *et al*./2000[11]	35/female	Headache and cognitive decline	NA	Mesencephalon	III	VP shunt	No
Papayannis *et al*./2003[12]	57/female	Tremors, unsteady gait, and bradykinesia	6 months	Mesencephalon	III	VP shunt	N
House *et al*./2004[30]	35/male	Headache and blurring of vision	NA	Mesencephalon	III	ETV and cyst fenestration	NA
	57/male	Cognitive decline and drowsiness	NA	Mesencephalon	III	ETV and cyst fenestration	NA
	44/female	Cognitive decline and unsteady gait	NA	Mesencephalon	III	ETV	NA
	56/female	Cognitive decline and poor memory	NA	Thalamus	III	VA shunt and cyst fenestration	No

Contd...

Table 1: Contd...

Author/year	Age/sex	Presenting symptoms	Symptom duration	Location	Kwee type	Surgery type	Postoperative regression of cyst (yes/no)
	40/male	Headache	NA	Mesencephalon	III	Cyst fenestration	NA
	47/female	Headache	NA	Mesencephalon-diencephalon junction	III	CSF shunt	NA
	35/male	Headache and confusion	NA	Thalamus	III	CSF shunt	NA
	49/female	Headache	NA	Thalamus	III	ETV	NA
Lee *et al*./2005[19]	8/male	Tremor	2 years	Mesencephalon-diencephalon junction	III	VP shunt	No
Rohlfs *et al*./2005[18]	50/male	Hemihypesthesia	NA	Mesencephalon-diencephalon junction	III	ETV and cyst fenestration	Yes
Fayeye *et al*./2010[13]	6/male	Parinaud's syndrome, ataxia, oculomotor, abducens, and facial nerve palsies	6 weeks	Mesencephalon	III	Cyst fenestration	Yes
Flors *et al*./2010[21]	10/female	Headache	4 months	Mesencephalon	III	VP shunt	No
Sturiale *et al*./2011[14]	38/male	Hemiparesis Benedikt's syndrome	Acute onset	Mesencephalon-diencephalon junction	III	VP shunt	Yes
Baldawa *et al*./2011[22]	46/female	Headache and unsteady gait	3 months	Mesencephalo-diencephalon junction	III	ETV	No
Fujimoto *et al*./2012[31]	17/male	Headache	NA	Mesencephalon-diencephalon junction	III	ETV and cyst fenestration	Increase in size
Rocha *et al*./2013[23]	52/female	Cognitive decline and unsteady gait	1 year	Mesencephalon-diencephalon junction	III	VP shunt	No
Fiorindi *et al*./2013[15]	43/female	Tremor and hemiparesis	NA	Mesencephalon-diencephalon junction	III	ETV and cyst fenestration	Y
	52/female	Tremor and visual disturbance	5 years	Mesencephalon-diencephalon junction	III	ETV and cyst fenestration	Yes
	29/male	Diplopia and anisocoria	NA	Mesencephalon	III	ETV and cyst fenestration	Yes
	19/male	Tremor, dizziness and oculomotor nerve palsy	NA	Mesencephalon-diencephalon junction	III	Cyst fenestration	Yes
Ottenhausen *et al*./2013[20]	43/female	Drowsiness, vomiting, and diplopia	3 days	Mesencephalon-diencephalon junction	III	ETV	NA
Choh *et al*./2014[24]	NA/male	Headache and unsteady gait	2 years	Mesencephalon-diencephalon junction	III	VP shunt	NA
Revel *et al*./2015[25]	74/male	Cognitive decline, unsteady gait and urinary incontinence	5 months	Mesencephalon-diencephalon junction	III	VA shunt	No
Kumar *et al*./2015[26]	30/male	Headache and cognitive decline	3 years	Mesencephalic tegmentum	III	VP shunt and ETV	No
Smith *et al*./2015[27]	50/male	Unsteady gait, headache, and blurring of vision	NA	Mesencephalon	III	ETV and cyst fenestration	Yes
Donaldson *et al*./2017[28]	31/male	Pulsatile tinnitus and headache	1 month	Mesencephalon-diencephalon junction	III	ETV	No
Al Abdulsalam *et al*./2018[1]	35/female	Headache and right foot numbness	6 months	Mesencephalon-diencephalon junction	III	VP shunt	Yes
Current study/2018	55/male	Cognitive impairment, unsteady gait, and lower limb weakness	6 months	Mesencephalon-diencephalon junction	III	VP shunt	No

CSF – Cerebrospinal fluid, VA – Ventriculoatrial, VP – Ventriculoperitoneal, ETV – Endoscopic third ventriculostomy, NA – Not available

both the hydrocephalus and its mass effect on the midbrain.[30] Twenty-three patients (62%) with hydrocephalus had their symptoms relieved by endoscopic third ventriculostomy (ETV) or shunt placement alone [Table 1]. In addition, among those with focal neurological symptoms, ten patients (59%) experienced sustained improvement with CSF diversion without the need for direct cyst manipulation. This finding lends support to the important role of the PVS in CSF-interstitial fluid hydrodynamics.[2,7] Modulating the fluid pressures in the ventricular and PVS compartments by CSF diversion could mean that additional cyst fenestration may be unnecessary. Fiorindi *et al*. further advised against cyst manipulation due to the risks of tearing its arterial perforators that perfuse the midbrain, thalamus, and basal ganglia.[15] It is evident that endoscopic cyst fenestration can lead to lesion regression on serial imaging, but whether this could lead to superior functional outcomes compared to CSF diversion alone is unclear. For this reason and due to concerns that the PVS wall would prohibit clear visualization of the third ventricular floor, we decided for shunt placement instead of ETV for our patient. Since most TPVSs do not involute with CSF diversion alone, as observed in our patient, one should vigilantly follow-up these patients as symptomatic reexpansion has been reported to occur more than 10 years after surgery.[31]

Conclusions

Giant TPVSs are rare entities and Type III lesions may present with hydrocephalus and focal neurological deficits due to their involvement of the midbrain. The clinician should be cognizant of the existence of such rare lesions by carefully evaluating the MRI so as to avoid unnecessary biopsies or excisions of these lesions. Our case illustrates that symptoms of midbrain compression can be completely reversed by CSF shunting without direct cyst decompression.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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