Giant Tumefactive Perivascular Spaces: A Case Report and Literature Review

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Key words
- Cerebrospinal fluid
- Giant tumefactive perivascular spaces
- Hydrocephalus
- Perivascular spaces
- Virchow—Robin spaces

Abbreviations and Acronyms
CSF: Cerebrospinal fluid
GTPVS: Giant tumefactive perivascular spaces
MRI: Magnetic resonance imaging
PVS: Perivascular spaces
VP: Ventriculoperitoneal

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Citation: World Neurosurg. (2018) 112:201-204. https://doi.org/10.1016/j.wneu.2018.01.144

Journal homepage: www.WORLDNEUROSURGERY.org

Available online: www.sciencedirect.com

BACKGROUND: **Perivascular spaces (PVS), also known as Virchow—Robin spaces, are pial-lined, interstitial fluid-filled structures in the brain that accompany cerebral vessels as they penetrate the cerebral substance. In healthy individuals, a PVS diameter of <2 mm is considered normal and can typically be seen within the white matter on magnetic resonance imaging (MRI). When PVS dilate, they are considered benign lesions and are associated with aging and other risk factors. These dilated PVS can cause neurologic symptoms, depending on their size and location. Symptomatic, massive enlargement of PVS are referred to as "giant" or "tumefactive" PVS; these are extremely rare and require neurosurgical intervention.**

CASE DESCRIPTION: **We present a rare case of giant tumefactive PVS (GTPVS) associated with hydrocephalus in a female patient who presented with progressive headache for 6 months. The patient was found to have giant tumefactive dilatation of PVS involving the right midbrain, with extension to the pons and thalamus, and with supratentorial moderate hydrocephalus. She was treated with cerebrospinal fluid diversion alone.**

CONCLUSIONS: **PVS are found on MRI in healthy people; rarely, they may dilate and cause neurologic symptoms. GTPVS are rare and can be misdiagnosed as central nervous system tumors; however, their imaging characteristics facilitate diagnosis. It has been postulated that these expanding PVS are due to defects in the drainage of interstitial fluid, where it enters into the ventricular system, and they are not the result of increased intraventricular pressure. We hypothesize that this may have been the case for the patient in our study, as the GTPVS collapsed following the insertion of a ventriculoperitoneal shunt. However, more recent literature provides evidence to support the idea that hydrocephalus is the consequence, and not the cause, of aqueduct compression by the lesion.**

INTRODUCTION

Perivascular spaces (PVS), also known as Virchow—Robin spaces, are pial-lined, interstitial fluid-filled structures in the brain that accompany cerebral vessels as they penetrate the cerebral substance.[1] These spaces are believed to form a major lymphatic drainage pathway; hence, they play a significant role in the immunology of the brain.[2] In healthy individuals, a PVS diameter of <2 mm is considered normal and can typically be seen in the basal ganglia and supratentorial white matter on magnetic resonance imaging (MRI).[3] When PVS dilate, they are considered benign lesions and are found to be associated with aging, hypertension, and dementia.[4,5] Although frequently asymptomatic, these dilated PVS can cause neurologic symptoms depending on their size and location, or they may result in hydrocephalus due to a mass effect.[4-6] Conversely, the symptomatic, massive enlargement of PVS can occur and is referred to as "giant" or "tumefactive" PVS. This is extremely rare and requires neurosurgical intervention.[7]

In this report, we present a rare case of giant tumefactive PVS (GTPVS) with associated hydrocephalus in a female patient who presented with progressive headache for 6 months. The patient was found to have a giant, tumefactive dilatation of PVS associated with hydrocephalus, and was subsequently treated with cerebrospinal fluid (CSF) diversion.

CASE PRESENTATION

A 35-year-old female patient presented to our hospital with progressive headache and a history of numbness in her right foot of 6 months duration. She had no other significant past medical or surgical history. MRI sequences revealed a multilocular cystic lesion involving the right midbrain with extension to the pons and thalamus, accompanied by supratentorial moderate hydrocephalus (Figure 1A—F). The lesion displayed no contrast enhancement or calcification.

The patient underwent ventriculoperitoneal (VP) shunt insertion. Following VP shunt insertion, the patient's symptoms improved. A postoperative computed tomography (CT) scan revealed regression of hydrocephalic changes, reduced lesion size, and a space-occupying effect (Figure 1G). The lesion size remained

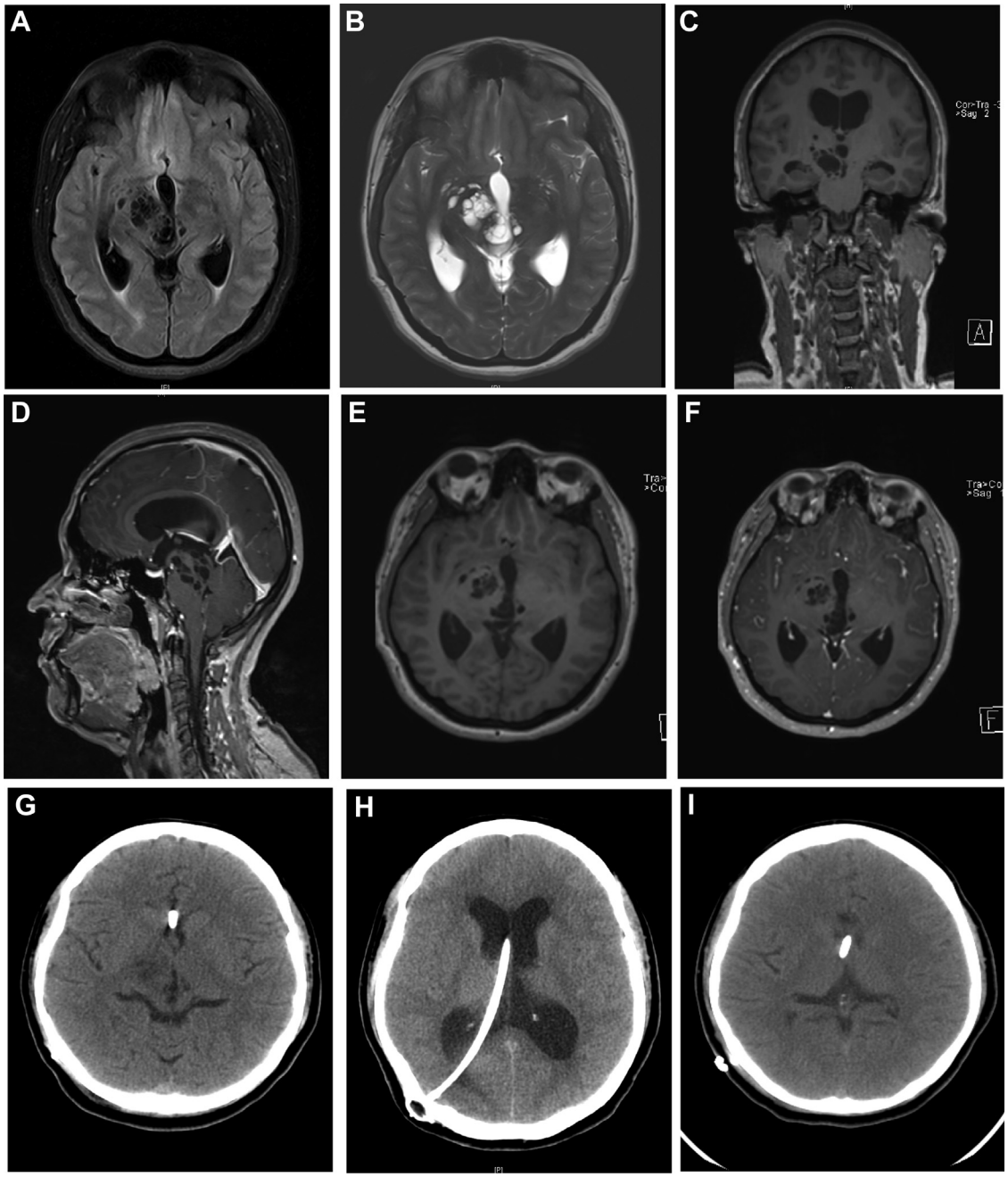

Figure 1. Axial fluid-attenuated inversion recovery-weighted (**A**), axial T2-weighted (**B**), coronal T1-weighted (**C**), sagittal T1-weighted (**D**), axial T1-weighted (**E**), and contrast-enhanced axial T1-weighted (**F**) magnetic resonance imaging showing a giant tumefactive perivascular space (GTPVS) in the right-sided mesencephalon and thalamus. The GTPVS is isointense relative to cerebrospinal fluid (CSF), involving and expanding into the right aspect of the midbrain with extension to the pons and thalamus on the ipsilateral side. The GTPVS is associated with supratentorial hydrocephalus. The lesion is characterized by its CSF signal intensity; it does not enhance following the administration of a contrast agent. (**G**) Postoperative computed tomography (CT) scan showing a regression in hydrocephalic changes and a significant reduction in the size of the lesion. (**H**) CT scan performed 6 months postsurgery showing the presence of acute hydrocephalus due to shunt malfunction. (**I**) CT scan performed following ventriculoperitoneal shunt revision demonstrating regression of hydrocephalus, as well as another remarkable reduction in the size of the lesion.