



Case Report

Giant tumefactive perivascular spaces: A case report

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ABSTRACT

Background: Perivascular spaces are interstitial fluid-filled regions located deep to the pia mater. They play roles in lymphatic drainage and the central nervous system immunological function. When they enlarge, they are referred to as giant tumefactive perivascular spaces. Often misdiagnosed as cystic neoplasms, they require a high degree of clinical suspicion and key radiological features to be accurately diagnosed. We describe an interesting case in which a man presented with worsening headache, subsequently found on neuroimaging to have this phenomenon.

Case Description: A 32-year-old man with low testosterone presented to the ER for worsening headache, blurred vision, and photophobia. Computed tomography of the brain showed hydrocephalus with follow-up magnetic resonance imaging revealing several enlarged cystic spaces within the brain, concerning for neoplasm. He ultimately left against medical advice before the further evaluation was done. He followed up with a neurosurgeon as an outpatient, where further review showed characteristic features indicative of giant tumefactive perivascular spaces, thus avoiding the need for unnecessary biopsy and potential surgery.

Conclusion: Often misdiagnosed as cystic neoplasms, giant tumefactive perivascular spaces are benign processes that can have a broad presentation with the most common finding being a headache. Key radiologic features, including smooth margins, isointensity to cerebrospinal fluid, and lack of postcontrast enhancement, are crucial to diagnosis, preventing unnecessary surgery with increased morbidity.

Keywords: Cystic, Giant tumefactive perivascular spaces, Pia mater

BACKGROUND

Perivascular spaces, also referred to as Virchow–Robin spaces, are interstitial fluid-filled regions that lie deep to the pia mater.^[1-3] They follow cerebral arteries and veins as they penetrate the cortex and are considered a normal anatomical structure, hypothesized to function in both a lymphatic and immunological capacity.^[1] However should these spaces enlarge they can exert mass effect, resulting in complications such as obstructive hydrocephalus.^[2] It is at this point that they are referred to as giant tumefactive perivascular spaces. Understanding their presentation and imaging features is key so as to avoid unnecessary invasive procedures. We describe for you a case involving a 32-year-old male who initially presented to the ED for headache, later found to have this very phenomenon. This case reveals how a high index of clinical suspicion, in addition to characteristic imaging criteria, is necessary for diagnosis, as it prevented said patient from undergoing unnecessary brain biopsy, associated with its own degree of morbidity.

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CASE DESCRIPTION

A 32-year-old male presented to Delray Medical Center Emergency Department for acute worsening headache, along with blurred vision and photophobia. Before presenting to the ED, he had been following with his PCP for persistent headache and vision changes. His medical history was significant only for low testosterone, for which he was on clomiphene and Arimidex. He denied tobacco, alcohol, and illicit drug use, and his family history was significant for a paternal grandfather with brain cancer. During his 5-day hospital course, he underwent a CT scan of his head, significant for hydrocephalus. Follow-up magnetic resonance imaging (MRI) confirmed hydrocephalus but also revealed numerous, enlarged cystic spaces within the brain parenchyma, and concerned for a cystic neoplasm. His laboratories were negative for infectious serologies, with the only abnormality being a mildly elevated erythrocyte sedimentation rate. He was advised during his hospitalization to undergo neurosurgical evaluation and potential biopsy; however, he refused and left against medical advice. On discharge, he followed up with his PCP, who referred him to a neurosurgeon for further evaluation. Based on imaging features, his symptoms were attributed to obstructive hydrocephalus in the setting of this cystic mass, thought to be chronic in nature. Given his continued symptoms, the original plan was to perform an endoscopic third ventriculostomy with external ventricular drain placement and CSF analysis. If this failed to resolve the patient's symptoms, the next step would be to perform a ventriculoperitoneal shunt. However, at the time of this article, the patient had been lost to follow-up.

CONCLUSION

Perivascular spaces are interstitial fluid-filled potential spaces that traverse along cerebral arteries and veins. They are located in the subpial space and are surrounded by a basement membrane.^[1,4] While not completely understood, several hypotheses have been proposed regarding their function. They are thought to drain interstitial fluid into the lymphatic vessels of the head and neck. By doing so, they expose indwelling macrophages to a variety of different antigens, which are ultimately phagocytosed and presented to B-cells and Helper T-cells, resulting in their activation.^[1] These spaces are most commonly found along the lenticulostriate arteries, but can be located anywhere in the brain, including the midbrain, white matter, and thalamus.^[1-3]

When these perivascular spaces enlarge, typically greater than 1.5 cm in size, they are referred to as giant or tumefactive perivascular spaces.^[3] The condition is relatively uncommon, with one retrospective review by Salzman *et al.* showing only 37 cases from 1988 to 2004.^[2] They occur in both genders and

at any age; however, the average age of discovery is around 46 years old.^[2] While a definitive pathophysiologic mechanism has not been established, several theories have been proposed. This includes spiral elongation of penetrating blood vessels secondary to hypertension, increased pulsations within cerebrospinal fluid, impaired interstitial fluid drainage, and increased permeability of the arterial wall in the setting of vasculitis.^[1,2]

Patients are usually asymptomatic, with identification occurring as an incidental finding. However, if symptomatic and clinical presentation is broad, with the most common finding being a headache. Alternatively, patients may present with dizziness, vision changes, syncope, seizure, or problems with memory, balance, and concentration.^[1,2,4] The differential diagnosis for these lesions is broad, with the most often being mistaken for cystic neoplasms. As a result, patients often undergo extensive workup and procedures, including biopsy and surgery unnecessarily for a relatively benign process. Besides malignancy, these giant perivascular spaces can be misdiagnosed as infectious (HIV, parasitic cysts, and fungal *Cryptococcus*) or vascular (cystic infarction and vascular dementia) in origin.^[1,2] Once discovered, giant perivascular spaces are left untreated unless complications from their presence arise.^[3] The biggest complication observed is obstructive hydrocephalus secondary to enlargement with associated mass effect.^[2,4] The degree of hydrocephalus may become so severe that surgical intervention and intraventricular shunt placement may be indicated.

The key to correctly diagnosing giant tumefactive perivascular spaces is recognizing the hallmark radiological findings. The most common imaging studies utilized are CT and MRI. They are delineated by their round to ovoid shape with smooth margins. They can present as a solitary lesion or in clusters. They are isointense relative to the CSF and do not show postcontrast enhancement or distort adjacent brain parenchyma.^[1-4]

In conclusion, this case depicts the discovery of a rarely observed, anatomical variant found incidentally on imaging. This case depicts the discovery of a rarely observed, the anatomical variation found incidentally on imaging. While initial radiographic findings may be concerning for a more ominous disease process such as neurological malignancy, a high degree of clinical suspicion along with characteristic imaging features is needed to assist in the diagnosis. By doing so, this prevents these patients from having to undergo invasive and high-risk neurointervention to achieve a diagnosis, for which treatment is often not indicated.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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