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Case Report

Giant tumefactive perivascular spaces ☆,☆☆

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ABSTRACT

Perivascular spaces are parts of the glymphatic pathway in the brain, which are microscopic but visible on magnetic resonance imaging when enlarged. Here, we described a case of a 16-year-old boy who presented with chronic headaches. Magnetic resonance imaging revealed giant perivascular spaces in the right centrum semiovale. Furtherly, we summarized the literature on classical and rare presentations of massive perivascular spaces and raised awareness that more clinical significance of the giant tumefactive perivascular spaces remains to be elucidated.

Introduction

Perivascular spaces (PVS), also referred to as Virchow-Robin spaces, are parts of the glymphatic pathway in the brain. PVS are spaces filled with interstitial fluid surrounding the perforating brain arteries, capillaries and venules, suggesting an important role in clearing harmful metabolites into meningeal and cervical lymphatic drainage vessels [1]. Normal PVSs are typically smaller than 3 mm in diameter. Symptomatic, massive enlargement of PVSs are referred to as giant or tumefactive PVSs, which are typically greater than 1.5 cm, which most appear in basal ganglia, subcortical white matter, and midbrain at the pontomesencephalic junction [2]. However, little is known about the clinical manifestations, management and pathologies of tumefactive PVS. Many aspects of perivascular spaces remain controversial and the lack of systematic data on tumefactive PVS raise awareness on these phenomena.

Case presentation

A 16-year-old boy with no known medical history presented with a 2-year history of bilateral headaches with visual aura (scintillating lightspot). The headache lasted 1-2 minutes each time and was of mild to moderate intensity. No accompanying neurologic symptoms were found. No family history was obtained. Follow-up examinations demonstrated vitamin B12 deficiency. The results of other routine laboratory tests were normal, as well as the metabolic screening and long-term video-electroencephalography (EEG) monitoring. Magnetic resonance imaging revealed giant perivascular spaces

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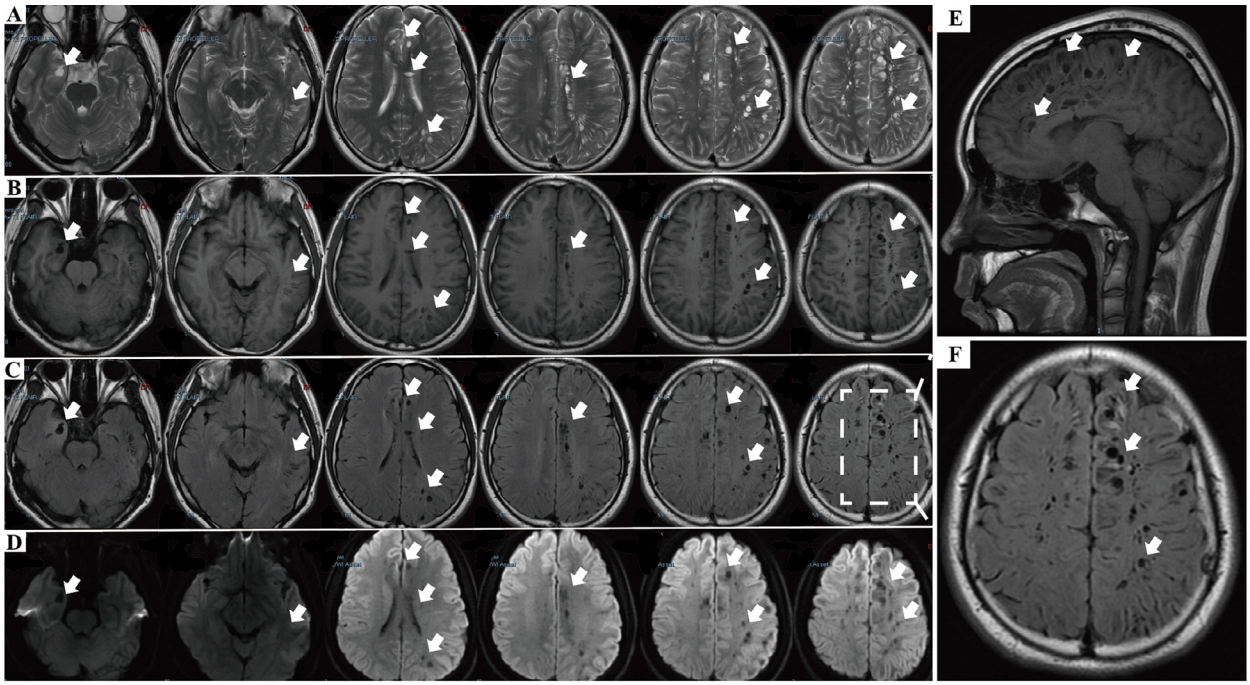

Fig. 1 – Perivascular spaces (white arrows) visualized with MRI on T2-weighted (A), T1-weighted (B), fluid attenuation inversion recovery (C), diffusion-weighted (D), and sagittal T1-weighted images (E).

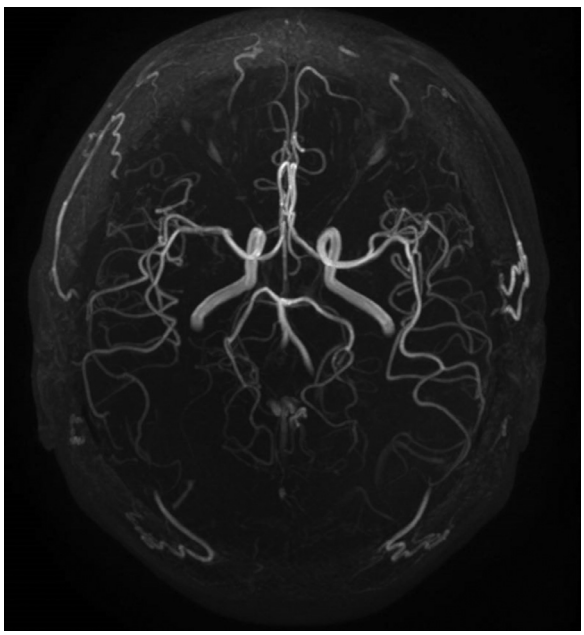

Fig. 2 – No abnormality was observed in the magnetic resonance angiography.

in both centrum semiovale (Fig. 1). while no abnormality was observed in magnetic resonance angiography (Fig. 2). Unfortunately, the patient refused a lumbar puncture. The change of CSF remains to be explored.

Discussion

As asymptomatic perivascular spaces are less likely to undergo brain MRI, the exact epidemiology of giant tumefactive perivascular spaces has yet to be delineated. Giant tumefactive perivascular spaces are associated with variable neurologic symptoms depending on their different size and locations [2–4]. Giant tumefactive perivascular spaces manifested as headache, chorea, dementia, stroke, and seizure depending on variable size and location. Patients usually present with headache and other nonspecific symptoms, while some having an initial insidious onset with sudden significant deterioration.

The signal intensity of perivascular space dilation is identical to that of cerebrospinal fluid on all MRI sequences, which revealed low signal intensity on T1 weighted images (WI), high signal intensity on T2 WI, and no appreciable contrast enhancement. Hyperintensity of the adjacent brain parenchyma on fluid-attenuated inversion recovery (FLAIR) was reported to be present in few patients. Of note, the enhanced PVS needs to be differentiated from vasculitis.

Perivascular spaces are a channel for interstitial fluid drainage. Normal perivascular space function is crucial for the maintenance of brain health, but the significance of giant perivascular space still remains unclear. Perivascular spaces dilation rarely appears in healthy people but more frequently in patients with stroke, dementia, or neuroinflammatory diseases. Previously, enlarged perivascular spaces have been thought to result from brain tissue loss during aging [1]. Increasing studies indicate that perivascular spaces have important roles in the movement and drainage of fluid in the

brain and enlarged perivascular spaces are not just an epiphenomenon associated with aging risk factors. The perivascular spaces dilation was associated with pericyte loss and blood-brain barrier breakdown occur in neurological disorders, including Alzheimer disease [5] and CADASIL [6]. Either arteriolosclerosis or venous collagenosis could contribute to perivascular space dilation [1]. In multiple sclerosis, perivascular space dilation has been observed during active inflammation indicating the development of arteriopathies and associated venular pathology [7]. Also, enlarged perivascular spaces are related to small vessel disease, even in early disease stages [6]. As yet, the available data are insufficient to illustrate the etiology of giant tumefactive perivascular spaces.

Misdiagnosis may result in unnecessary neurosurgical intervention. Reports of such cases have offered an extensive differential diagnosis that includes cystic neoplasms, brain abscess, cystic infarction and mucopolysaccharidosis. Burr hole and aspiration may be considered to resolve significant massive effect and improve symptoms as clinical deterioration. More research is needed to clarify the exact etiology of these giant tumefactive perivascular spaces.

Patient consent

Written informed consent was obtained from the individual for the publication of any potentially identifiable data included in this article.

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