

Giant perivascular spaces: an uncommon cause of obstructive hydrocephalus

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A 55-year-old man with good past health presented to the emergency department with unsteady gait for 6 months with recent mild left-sided weakness. Urgent computed tomography (CT) scan of the brain showed a multiseptated cystic lesion in the right mesencephalothalamic region with pressure effect on the third ventricle causing obstructive hydrocephalus (Fig 1). Urgent magnetic resonance imaging (MRI) scan (Fig 2) of the lesion showed no post-gadolinium enhancement, no restricted diffusion, complete suppression of the cystic areas on T2 fluid attenuation inversion recovery (FLAIR) sequence and no abnormal parenchymal signal intensities compared with normal brain parenchyma. These imaging findings are consistent with tumefactive perivascular space. Invasive biopsy and surgical excision were avoided, and the patient underwent surgery for ventricular drain insertion.

Dilated perivascular spaces (PVSs) in the brain are interstitial fluid-filled structures lined by pia-mater that have accompanying patent penetrating arteries within, most commonly seen along the

lenticulostriate arteries.¹ They can be unilocular or multilocular and may have a radial pattern along the course of the penetrating arteries. The PVSs occur across all age-groups and are more frequent and larger with advancing age. The cause of dilated PVS remains unknown though numerous theories have been postulated including increased permeability of arterial wall and obstruction/disturbance of interstitial fluid drainage/flow. Dilated PVSs may be associated with microvascular diseases, trauma, non-vascular dementia, multiple sclerosis, and the mucopolysaccharidoses.

Rarely PVSs are markedly expanded and are termed tumefactive or giant PVSs. Some authors define tumefactive PVSs as those >1.5 cm. Tumefactive PVSs are most commonly located in mesencephalothalamic region.² They are also seen in cerebral white matter and in the cerebellar dentate nuclei. They can exhibit mass effect and cause obstructive hydrocephalus when occurring in the mesencephalothalamic region as in our case. The MRI signal intensities of typical PVSs should follow cerebrospinal fluid in all sequences including FLAIR imaging with no post-gadolinium enhancement. There is no restricted diffusion as the compartments are communicating. Tumefactive PVSs in cerebral white matter may have perilesional abnormal T2 and FLAIR hyperintensities in up to 50% of cases. The mass effect of the tumefactive PVS may cause chronic ischaemic change in adjacent white matter.³ Histopathological results typically show a pial-lined cyst with no evidence of neoplasm or infection.

Differential diagnoses include cystic infarction, tumour, and infection.⁴ Cystic infarctions assume a slit-like or ovoid shape whereas PVSs are more rounded or linear. The cystic content of tumours is usually not isointense to cerebrospinal fluid on MRI. Solid components are often present, which may enhance after contrast and are surrounded by oedema. Parasitic infections have a range of appearances on CT or MRI scans with contrast enhancement and oedema during the active phase and calcifications in the quiescent phase.

Asymptomatic tumefactive PVSs can be managed by follow-up imaging for stability in size. Spontaneous regression of tumefactive PVSs without surgical intervention is rare. Tumefactive PVSs with mass effect and obstructive hydrocephalus can be treated surgically with ventriculostomy, cyst fenestration, ventriculoperitoneal shunting,

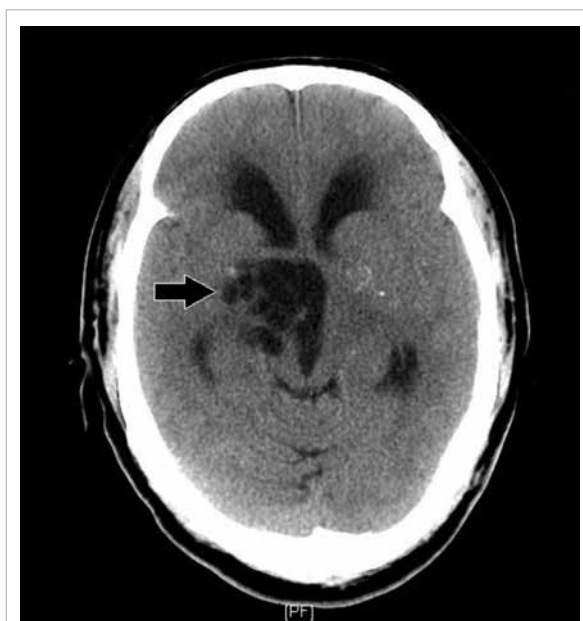


FIG 1. Plain computed tomography scan of the brain showing a multicystic lesion (arrow) in right mesencephalothalamic region with dilated frontal horns of the lateral ventricles and periventricular white matter hypodensities suggestive of transependymal oedema

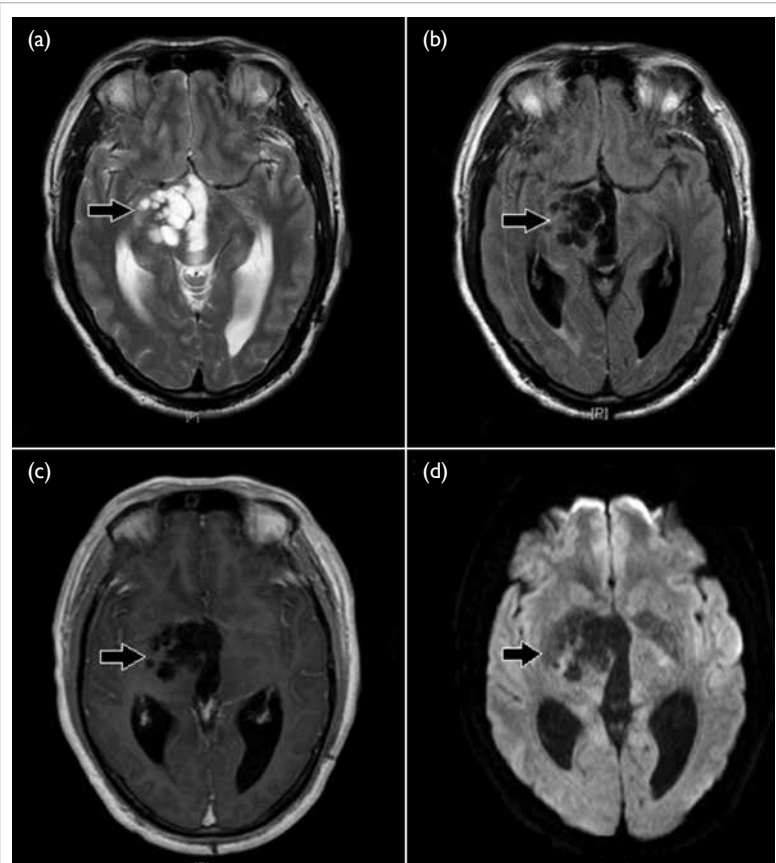


FIG 2. Magnetic resonance images showing tumefactive perivascular space (arrows): (a) T2-weighted sequence; (b) T2 fluid attenuation inversion sequence with gadolinium showing complete suppression of fluid signal in the multicystic lesion; (c) T1-weighted sequence with gadolinium showing no enhancement; and (d) diffusion-weighted imaging showing no restricted diffusion

or cystoperitoneal shunting. When the appearance is typical, surgical biopsy or excision should be avoided.

Author contributions

All authors contributed to the concept, acquisition and interpretation of data, drafting of the manuscript, and revision for important intellectual content. All authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

Conflicts of interest

All authors have disclosed no conflicts of interest.

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Ethics approval

The patient was treated in accordance with the Declaration of Helsinki. The patient provided informed consent for all procedures.

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