



Image Report

Giant tumefactive perivascular spaces in a pediatric patient: A rare radiological entity

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ABSTRACT

Background: Giant tumefactive perivascular spaces (TPVS) are radiological rarities and may mimic other neurological structural lesions. Fewer than 80 cases have been reported in the literature with even fewer in the pediatric population.

Case Description: The authors present an image report showcasing a 3-year-old boy presenting with uncontrolled seizures despite multiple anti-epileptic medications. His magnetic resonance imaging showed multiple, non-contrast enhancing cyst clusters within the left parieto-occipital region that was hyperintense on T2-weighted imaging, and isointense to cerebrospinal fluid. Due to a characteristic absence of perilesional edema seen on fluid-attenuated inversion recovery imaging or diffusion restriction on diffusion-weighted imaging (DWI) sequences, this was diagnosed as a giant TPVS.

Conclusion: Accurate diagnosis of these rare radiological entities is based on pathognomonic findings that can help prevent unnecessary surgery and guide management for patients, particularly in the pediatric population as seen in our case.

Keywords: Giant tumefactive perivascular space, Neuro-radiology, Pediatric, Virchow-robin space

INTRODUCTION

Perivascular (Virchow-Robin) spaces (PVSs) were initially described by Virchow in 1851 and Robin in 1859 as pial-lined fluid filled structures accompanying penetrating arteries as they enter the cerebrum.^[7] Cases have shown that the enlargement of PVSs can cause mass effect; therefore, the term “tumefactive” is applied. Often they are mistaken for different pathological processes on brain imaging, particularly cystic neoplasms.

The rarity of these lesions is such that fewer than 80 giant (larger than 15 mm) tumefactive perivascular spaces (TPVSs) have been reported in the literature, and even fewer in the pediatric population.^[13] We believe it is important to present a case encountered at our institution due to such a low incidence.

CLINICAL IMAGE

A 3-year-old boy presented to our clinic with partial clonic seizures for the past 1½ years. These lasted for 30 min and were terminated with an anticonvulsant. Every episode occurred

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in the same fashion and on the same side of the body. He had no prior history of loss of consciousness, intractable vomiting, fall, or trauma. Despite being on two anti-epileptic medications from his previous medical team, he had another seizure a few months before presentation. On examination, he was awake, alert, and his speech was intact. He was able to follow commands with no motor deficits. He did not exhibit any cerebellar signs or gait abnormalities. Systemic examination was also unremarkable.

On magnetic resonance imaging (MRI) brain with contrast imaging, there was an irregular and abnormal signal intensity lesion in the left parieto-occipital region [Figure 1]. It appears primarily hypointense, with multiple cystic regions on T1-weighted imaging, and hyperintense with a “bubbly” appearance on T2-weighted images. These cystic spaces are seen to form clusters. The lesion appears to be causing a mass effect over the occipital horn of the left lateral ventricle. Perilesional edema was not seen on fluid-attenuated inversion recovery (FLAIR) sequences. Diffusion-weighted imaging (DWI) and apparent diffusion coefficient sequences did not reveal any signal restriction within the lesion. There is no enhancement seen within the cysts after contrast administration.

The lesion remained stable on follow-up imaging and was labeled as giant TPVS.

DISCUSSION

In one of the largest retrospective studies of 37 patients with giant TPVSs, lesions were predominant in male patients (1.8:1) and ranged from 6 to 48 years in age.^[8] Most were unilateral and presented as multiple and clustered cysts. This is similar to our case where the lesion was localized to the left parieto-occipital region with numerous cysts included in the study. However, only one patient in that cohort had seizures as the presenting complaint, similar to our case. Giant TPVS is relatively uncommon in the pediatric age group and Salzman *et al.* were only able to find four such cases <18 years of age. Few case reports have been published detailing pediatric population cases, often incidentally discovered.^[1,9]

Commonly, perivascular spaces present near the lenticulostriate arteries.^[4] They have also been identified along the subinsular region, dentate nuclei, and cerebellum.^[11] When PVs develop into larger dilatations, they present a diagnostic challenge and are often mistaken for other pathological processes. Typical appearances include round and oval shaped clusters that do not show contrast-enhancement and are isointense to cerebrospinal fluid (CSF). DWI sequences also often show no diffusion restriction.^[3] The most common site for giant TPVS is the mesencephalothalamic region and causes mass effect. Interestingly, FLAIR imaging shows signal

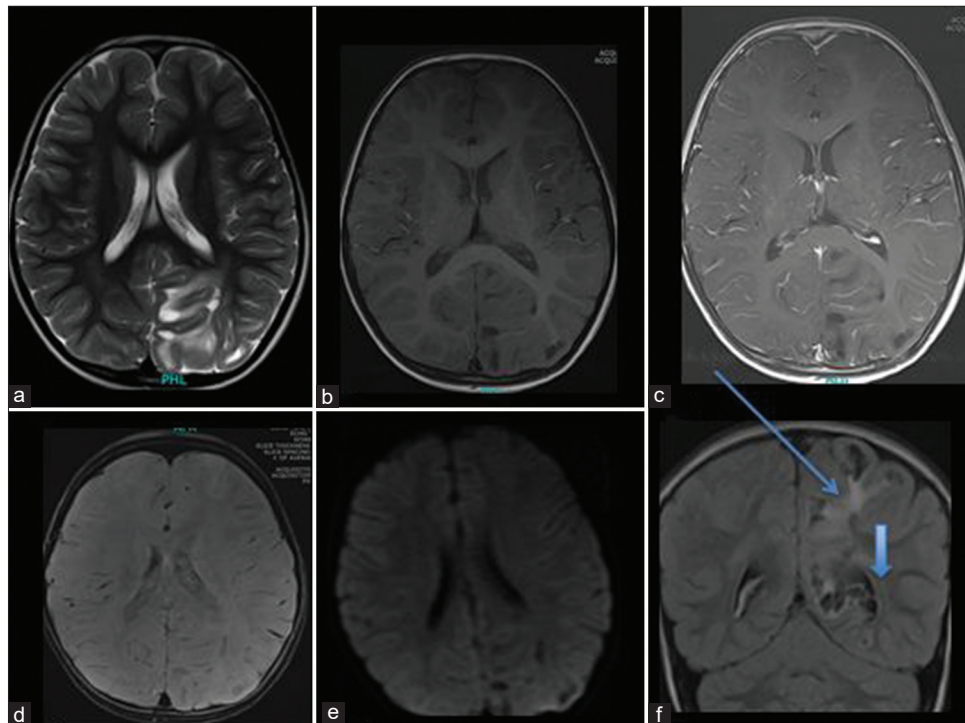


Figure 1: Axial T2 (a), Axial T1 plain (b), and post-contrast (c). In addition, axial susceptibility weighted imaging (SWI) (d), diffusion (e), and coronal fluid-attenuated inversion recovery (f) show diffuse and confluent white matter hyperintensity (marked by arrow) surrounding the giant perivascular spaces (arrow head). There is associated gyral expansion over the perivascular spaces with no enhancement. The SWI shows no hemorrhage or calcifications. There is no diffusion restriction (e) or post-contrast enhancement (c).

intensity suppression without adjacent edema noted in the brain parenchyma.^[8]

Due to their compressive nature, these lesions may be mistaken for cystic neoplastic conditions.^[10] Therefore, it is essential that physicians, surgeons, and radiologists that encounter such lesions be aware of their pathognomonic features: locations near to penetrating arteries, the lesion following CSF signals, does not enhance on contrast, and with normal adjacent brain matter. This can save patients from unnecessary surgical biopsies or resection when not indicated. Mass effect is almost always in proportion to the size and location of the giant TPVS.^[3]

For clinical management, dilated TPVS are subdivided into categories according to their anatomical relationship with penetrating arteries.^[5] For asymptomatic cases, no intervention is needed with surveillance MRI imaging recommended.^[12] Symptomatic cases require surgery to address hydrocephalus and mass effect on adjacent structures. CSF diversion through shunt procedure can be useful as a temporizing procedure, while some authors have reported success with cyst drainage and fenestration to relieve focal mass effect.^[6,13] Since CSF diversion would not cause most TPVSs to self-resolve, they should be strictly followed up for symptomatic recurrence as has been seen before.^[2]

CONCLUSION

Giant TPVS may be mistaken for other pathological processes, particularly due to their rarity in pediatric patients. However, pathognomonic radiological characteristics can help identify cases that may not require surgical intervention.

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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