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Giant Cystic Virchow-Robin Spaces with Adjacent White Matter Signal Alteration

Komşu Beyaz Madde Değişiklikleri ile Birlikte Dev Kistik Virchow-Robin Boşlukları

Nisar A WANI[1], Farooq MIR[1], Irshad M BHAT[1], Tariq GOJWARI[1], Salma BHAT[2]

[1]*Sher-I-Kashmir Institute of Medical Sciences, Deparatment of Radiodiagnosis & Imaging, Srinagar, India*
[2]*Sher-I-Kashmir Institute of Medical Sciences, Department of Pathology, Srinagar, India*

Correspondence address: Nisar A WANI / E-mail: nisar.wani@yahoo.com

ABSTRACT

Perivascular spaces surround the small arteries and veins as they enter into the brain parenchyma from the subarachnoid spaces. Also called as Virchow-Robin spaces, these are prominent in the basal ganglia and high convexity white matter of the elderly. Occasionally VR spaces may get massively enlarged and may mimic a cystic mass lesion. The typical CSF-like signal intensity of the cysts and location on MRI, in the absence of a neurological abnormality help in the diagnosis of the giant VR spaces and thus biopsy is avoided. Typically there is no significant adjacent brain abnormality; however FLAIR images may sometimes reveal perilesional white matter hyperintensity, which may be an indication of gliosis due to the mass effect of the lesion. Such a signal alteration should not deter one from making a diagnosis of giant Virchow-Robin spaces when the rest of the imaging findings are typical. We describe a case of a 50-year-old female with incidental giant Virchow-Robin spaces in the right hemispheric subcortical white matter with adjacent white matter hyperintense signal intensity on T2-weighted and FLAIR images.

KEYWORDS: Fluid attenuation inversion recovery (FLAIR) sequence, MRI, Virchow-Robbin spaces

ÖZ

Subaraknoid boşluklardan beyin parenkimine girdikleri zaman küçük arter ve venleri perivasküler boşluklar sarar. Virchow-Robin boşlukları olarak da isimlendirilen bu boşluklar yaşlılarda bazal ganglialarda ve yüksek konveksite beyaz maddede belirgindir. Zaman zaman bu VR boşlukları çok büyüyebilirler ve kistik kitle lezyonlarını taklit edebilirler. Bu kistlerin tipik BOS benzeri sinyal özellikleri ve MRG deki yerleşimleri, nörolojik anomali yokluğunda dev VR boşluklarının teşhisinde yardımcı olur ve böylece biyopsiden kaçınılabilir. Tipik olarak komşu beyinde belirgin bir anomali yoktur fakat FLAİR görüntüler bazen beyaz madde perilezyonel hiperintensitesi gösterebilir ki bu da lezyonun kitle etkisine bağlı gliozisin bir göstergesidir. Böyle bir sinyal değişikliği diğer görüntüleme bulguları tipik olduğunda Virchow-Robin boşlukları teşhisinin konulmasını engellememelidir. Biz T2 ağırlıklı ve FLAİR görüntülerde komşu beyaz madde hiperintens sinyal intensitesi gösteren sağ hemisferik subkortikal beyaz madde dev Virchow-Robin boşlukları insidental olarak saptanan 50 yaşında bir bayan hasta sunuyoruz.

ANAHTAR SÖZCÜKLER: Fluid attenuation inversion recovery (FLAIR) sekans, MRG, Virchow-Robbin boşlukları

INTRODUCTION

Enlarged perivascular spaces, also known as Virchow-Robin spaces, are pial-lined interstitial fluid-filled structures that accompany penetrating vessels as they enter into the brain parenchyma. They frequently appear in the inferior basal ganglia, clustering around the anterior commissure and surrounding the lenticulostriate arteries as they course superiorly. Other common locations include the midbrain, deep white matter, and subinsular cortex. They can also be found in the region of the thalami, dentate nuclei, corpus callosum, and cingulate gyrus (5). They are common, incidental, "leave me alone" lesions that should not be mistaken for more ominous disease when they are significantly and massively enlarged (giant VR spaces), which is an occasional occurrence. Knowledge of their signal intensity characteristics and localization helps in this differentiation, which is important for correct patient management (3).

CASE REPORT

A 50-year-old, normotensive, non-diabetic female patient with known manic-depressive illness, presented with a history of headache for the last month. She was well controlled by antidepressant medication including lithium and her lithium levels were within acceptable limits. She did not have any history of fever, vomiting or any neurodeficit accompanying the headache. Her relatives reported that she had banged her head against the wall sometime in the past. On examination, she was afebrile and her blood pressure was 120/70 mmHg. Her higher functions were intact with normal cranial nerves on examination of the central nervous system and no sensory or motor deficit. There were no signs of meningeal

irritation. MRI examination of the brain was performed with a 1.5T superconducting magnet, using T1W spin-echo, T2W turbo spin-echo, FLAIR and post gadolinium T1W sequences in various orthogonal planes, to rule out a possibility of an intracranial space occupying lesion/ subdural collection. The non-contrast study showed a cluster of multiple, well-defined, thin-walled cysts in the subcortical white matter of the right parietal and occipital lobes of brain (Figure 1). The individual cysts measured 1-2 cm in diameter and showed hyperintense signal intensity on T2-weighted and hypointense signal intensity on T1-weighted sequences, in both cases isointense to the ventricular CSF signal intensity. There was no significant mass effect on the ventricle and no midline shift was seen. The axial fluid attenuated inversion recovery (FLAIR) sequence showed suppression of the T2W hyperintense signal intensity of the cystic fluid similar to the ventricular CSF, while the adjacent white matter around the cysts showed hyperintense signal intensity (Figure 2). Close examination showed tiny, millimeter size, cystic foci in the left high parietal white matter on T2W images. There was no evidence of restricted diffusion on the diffusion-weighted images. Post-gadolinium T1W images did not show any contrast enhancement of the cyst wall or the adjacent brain (Figure 3). These imaging findings in the absence of any neurological abnormality were highly suggestive of giant Virchow-Robin spaces. Adjacent white matter signal alteration was supposed to be related to the gliosis resulting from the mass effect by the cysts. No active surgical intervention was performed and follow-up with serial MRI was decided instead for the patient.

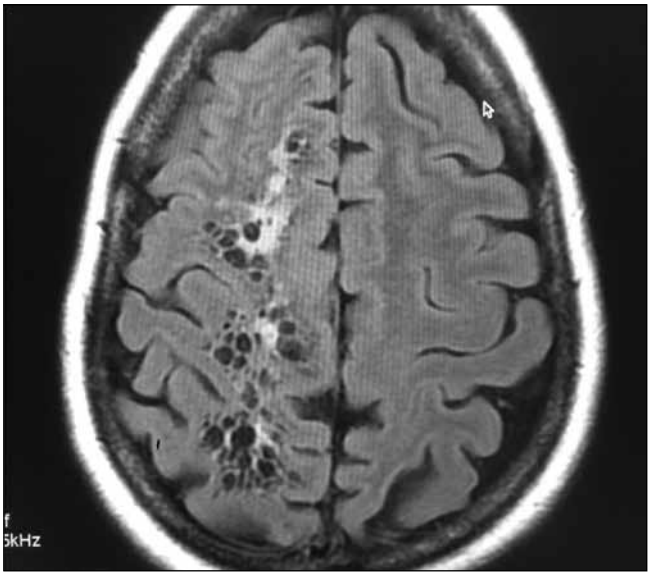

Figure 2: Axial FLAIR image of the brain showing suppression of the signal in the right hemispheric subcortical white matter lesions. Discrete areas of hyperintense signal intensity are seen in the adjacent white matter.

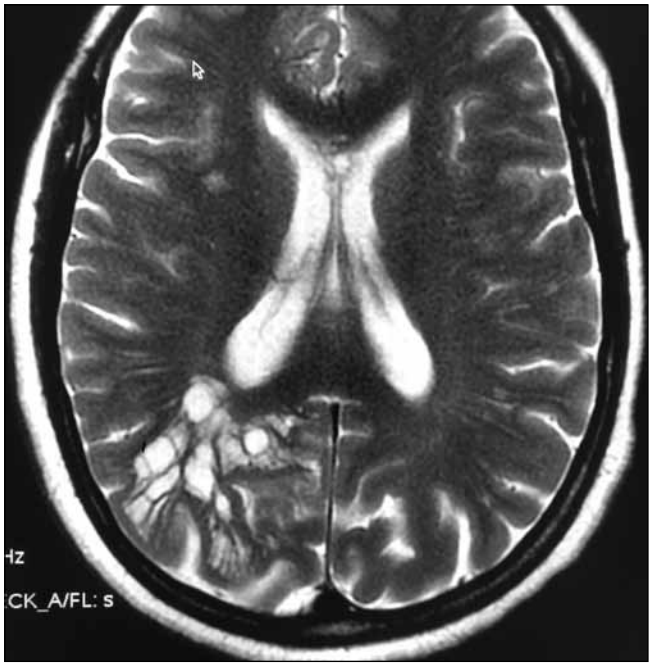

Figure 1: T2-weighted axial MR image showing a cluster of thin walled, well defined, rounded and oval, 1-2 cm hyperintense signal intensity lesions in the right (occipito-parietal) white matter. The signal of the lesions is same as that of the CSF and there is no significant mass effect. There are areas of hyperintense signal intensity in the white matter outside the hypointense walls of the well-defined lesions.

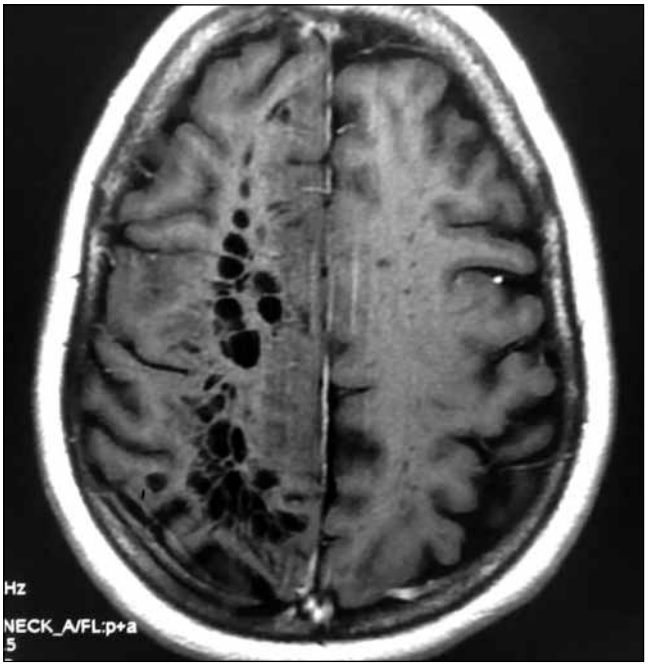

Figure 3: Axial post gadolinium T1W image showing no evidence of contrast enhancement in the right hemispheric subcortical white matter cystic lesion.

DISCUSSION

VR spaces surround the walls of arteries, arterioles, veins, and venules as they course from the subarachnoid space through the brain parenchyma. The subarachnoid space does not communicate directly with the VR spaces, which are, in effect, the lymphatic drainage pathways of the brain. Small VR spaces appear in all age groups. VR spaces are found with increasing frequency and are larger with advancing age (3). The postulated mechanisms underlying expanding VR spaces include increasing permeability of the arterial wall, disturbance of the drainage route of interstitial fluid due to cerebrospinal fluid (CSF) circulation in the cistern, spiral elongation of blood vessels, brain atrophy, gradual leaking of the interstitial fluid from the intracellular compartment, and fibrosis and obstruction of VR spaces along the length of arteries and consequent impedance of fluid flow (3,8). Dilated VR spaces typically occur in three characteristic locations: Type I VR spaces appear along the lenticulostriate arteries entering the basal ganglia through the anterior perforated substance. Type II VR spaces are found along the paths of the perforating medullary arteries as they enter the cortical gray matter over the high convexities and extend into the white matter. Type III VR spaces appear in the midbrain (3,8).

VR spaces have typical MR imaging features. These are round or oval with a well-defined, smooth margin, occur along the path of penetrating arteries, are isointense relative to CSF, and demonstrate no enhancement following contrast medium administration. When VR spaces become enlarged, they are known as giant VR spaces. They differ from typical VR spaces in that they are larger in size and may have an associated focal mass effect (8). Dilations of perivascular spaces, even if giant, are, in most cases, fortuitously discovered because they do not induce any clinical abnormality (4) However, some atypical presentations have been reported including an acute obstructive hydrocephalus due to the compression of the aqueduct of Sylvius (6).

Unusually dilated Virchow-Robin spaces typically appear on MR imaging as round, oval, or curvilinear lesions with a well-defined smooth margin, with signal intensities identical to the CSF on all pulse sequences. Lesions are located along the path of the perforating arteries, have some mass effect and do not show any contrast enhancement (2). Most patients have unilateral lesions in the convexity white matter, predominantly located in the occipital and parietal lobes (2,7). FLAIR imaging typically shows complete signal intensity suppression without abnormalities in the adjacent parenchyma. Discrete foci of increased signal intensity on FLAIR images have been reported adjacent to giant VR spaces have been reported in a few cases (7). In elderly patients, a possible theory is that this associated signal intensity alteration may represent advanced chronic ischemic change related to the mass effect of the VR spaces (2). In younger patients, the changes in the white matter adjacent to the cystic VR spaces may be secondary to gliosis or spongiosis (8). Alternatively, the signal intensity changes may be related to multiple tiny tightly clustered VR spaces that are too small to be discriminated on the basis of current MR imaging findings (8).

As most of the markedly enlarged, giant VR spaces with mass effect border a ventricle or subarachnoid space, they may be misinterpreted as other pathologic processes, most often a cystic neoplasm (8). Other pathologies in the differential diagnosis include parasitic cysts, cystic infarctions, nonneoplastic neuroepithelial cyst, deposition disorders such as mucopolysaccharidosis and periventricular leukomalacia. Cystic neoplasms rarely exhibit signal intensity exactly like the CSF. Neurocysticercosis cysts may have a scolex (parasite head), and the cyst walls often enhance. Neurocysticercosis cysts may be multiple but do not typically occur in clusters within the brain parenchyma. Lacunar infarcts are associated with clinical features of stroke or neurodeficit, and on imaging can usually be distinguished from VR spaces since they exhibit adjacent parenchymal hyperintensity more commonly. Patients of mucopolysaccharidosis are usually children with typical facies, clinical features and other imaging findings, which are all absent in a case of giant VR spaces. Periventricular leukomalacia, usually seen in premature infants, shows marked loss of periventricular white matter, predominantly in the periatrial regions, and compensatory focal ventricular enlargement adjacent to regions of abnormal white matter signal intensity. The involvement tends to be symmetrical. Corpus callosal thinning can be seen as a secondary manifestation (3,5,8,9).

When the lesions in question occur in a characteristic location described for VR spaces, follow CSF signal intensity on all MRI sequences, do not enhance with contrast material, and have normal adjacent brain parenchyma or small areas of increased signal intensity on FLAIR, their appearance is virtually pathognomonic of giant VR spaces (4,7,9).

Invasive diagnostic examinations for unusually dilated Virchow-Robin spaces are unnecessary. However, careful follow-up examination using FLAIR MR imaging can detect chronological changes in such lesions. Most of the patients with giant VR spaces do not need any surgical treatment, thus qualifying these spaces as leave alone lesions (3,9). However, rare cases with significant mass effect and or hydrocephalus may need surgical decompression or CSF shunting such as cystocisternostomy or ventriculocystostomy (1,7).

We conclude that, well-defined round or oval clustered cystic lesions with typical CSF signal intensity on all MR Imaging sequences, in high parietal white matter and without any contrast enhancement are diagnostic of giant VR spaces. Adjacent white matter signal alteration on FLAIR images may be related to the mass effect of these cystic lesions and do not necessarily contradict the imaging diagnosis. Biopsy is unnecessary for this benign leave alone lesion.

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