

Giant Perivascular Spaces Presenting As Obstructive Hydrocephalus

Rahul Dev*, Venkata Subbaih A, Navojit Chatterjee, Smily Sharma, Navneet Kumar, Sumit Kumar, Mohit, Roshni A, Udit Chauhan, Khanak Nandolia, Mohit Tayal, Ashish Kaushik, Divya Pandey, Sandeep Kumar, Sudhir Saxena

Department of Radiodiagnosis and Imaging, All India Institute of Medical Sciences, Rishikesh, Uttarakhand India

*Corresponding author

Rahul Dev

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Abstract: Virchow-Robin (VR) spaces are an entity seen across all age groups which occasionally enlarge enough to cause mass or pressure effects. A middle age patient presented to outpatient clinic with long standing progressive headache. MRI findings revealed multiple CSF intensity non enhancing lesions obstructing ventricular outflow tract and extending through it with resultant obstructing hydrocephalus.

Keywords: Giant Perivascular space, VR spaces, CSF signal intensity, Basal cisterns, Hydrocephalus.

INTRODUCTION

Perivascular spaces (PVS) also known as Virchow robin (VR) spaces are pial lined structures accompanying the penetrating vessels as they pass from subarachnoid space into the brain parenchyma typically located in the basal ganglia, along the cerebral convexities and in brainstem [1]. Routinely, these spaces can be identified on MR imaging obtained in patients of all ages measuring upto 2mm in diameter. Rarely these spaces may show gross enlargement and assume bizarre configuration attributed for definite neurological abnormality [2]. Giant or Tumefactive PVS are extremely rare and invariably require neurosurgical intervention. These are also seen to have a positive correlation with small vessel disease and cognitive decline [3].

In the present report we present a rare case of Giant PVS in an adult patient presenting with complaints of progressive headache for last 2 years with subsequently imaging showing the presence of large CSF intensity cyst within brain parenchyma compressing ventricular system and obstructing ventricular outflow tracts leading to obstructive hydrocephalus.

CASE REPORT

A 54-year-old patient presented to outpatient clinic with the history of mild to moderate headache for the duration of past two years which was progressively increasing in severity. There was no history of any fever, convulsions, blurring of vision, tinnitus or any other neurological symptoms. Family history and past history were unremarkable except placement of ventriculo-peritoneal shunt in view to correct the hydrocephalus at a different centre. The patient underwent contrast enhanced MRI of the brain after excluding contraindications to the procedure and taking written informed consent and screening for any metallic prosthesis. MRI examination revealed multiple variable size round to oval lesions following CSF signal intensity on all sequences, compressing upon lateral and third ventricles with few lesions occupying outflow tract of third as well as fourth ventricle with resultant obstructive hydrocephalus as seen on axial and sagittal

T2 weighted sequences (Figure-1). Axial FLAIR, Diffusion weighted images and corresponding ADC map show no hyperintensity, no diffusion restriction, no area of blooming on susceptibility weighted sequence (SWI) (Figure-2). Post contrast axial T1 weighted axial and sagittal images showed no wall enhancement with no intralesional enhancing solid component or eccentric nodule (Figure 3). The lesions appear stable without any change in size and appearance as compared to previous imaging done two years back at another centre. Patient clinical condition remains stable and is in close follow up.

DISCUSSION

Giant PVS are seen as round to oval well defined smooth walled cystic lesions following CSF signal intensity on all sequences, showing no post contrast enhancement with absence of enhancing solid component, blooming or diffusion restriction and unremarkable surrounding parenchyma on FLAIR sequence [1, 4]. Small VR spaces less than 2mm in size appear in all age groups. With advancing age, VR spaces are found with increasing frequency and apparently larger in size. Occasionally VR spaces may be markedly dilated, assume bizarre shapes, may show mass effect and are usually encountered bordering a ventricle or a subarachnoid space. This is most commonly seen when disease distribution in the

mesencephalothalamic region leading to obstructive hydrocephalus as in our case [4, 5]. The lesions are also found with a relatively lesser frequency in cerebral parenchyma notably frontal lobes. The etiology of this entity is uncertain leading to formulation of various theories including altered arterial permeability, altered interstitial fluid drainage, elevated CSF pressure, lymphatic blockade and inflammation. In vast majority of cases these lesions are asymptomatic with no neurological abnormality [6]. This entity need to be differentiated from various pathological entities as discussed further. Periventricular leukomalacia shows similar appearance however they are seen in pre perinatal hypoxic injury or show stigmata of previous haemorrhage. Chronic lacunar infarcts are differentiated from Giant PVS as they are relatively larger in size, irregular in shape and asymmetrical distributed [7]. Neoplastic lesions may show restricted diffusion or contain solid enhancing component. Infectious etiology like neurocysticercosis may show a spectrum of appearances encompassing peripheral rim enhancement, intralesional hypointense scolex, or perilesional white matter oedema, whereas neurosarcoidosis may show

leptomeningeal or dural enhancement as well as parenchymal mass lesions [6]. There are documented reports of these benign spaces attributing to neurological symptoms. Salzman *et al.* in their study mentioned headache to be the most common presenting symptom similar to seen in our case [4]. Other relatively less common symptoms included visual disturbances, unstable gait and changes of dementia. However, the clinical symptoms rarely correlate with the size of the lesions. These lesions have favourable outcome irrespective of surgical treatment and almost no tendency for spontaneous regression. Rarely these cystic lesions may further increase in size and number despite surgical intervention or otherwise. This phenomenon was documented by Fujimoto *et al.*, in a case showing reappearance of dilated VR spaces after a gap of more than a decade presenting clinically as cranial nerve palsies [8]. A genetic correlation between dilated perivascular spaces and small vessel ischaemic changes was postulated in particular reference to white matter changes [9].

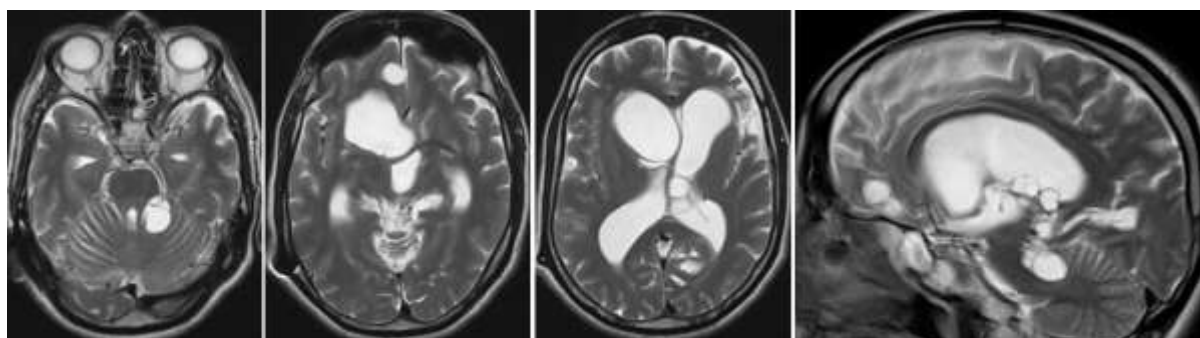


Fig-1: Axial T2 weighted images show multiple variable sizes well defined hyperintense cysts at the level of basal cisterns, with a few cysts extending superiorly compressing the third and right lateral ventricle. Sagittal T2 weighted image shows T2 hyperintense cysts along the base of anterior cranial fossa as well as occupying and obstructing third ventricle outflow tract with one of the cysts seen to extend via the foramen of Luschka.

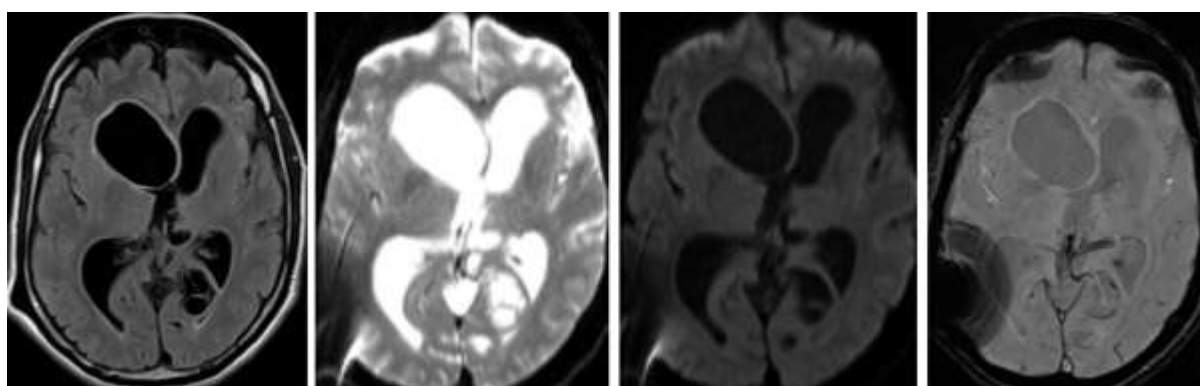


Fig-2: Axial FLAIR, diffusion weighted, ADC and gradient images (in sequence) show that the lesions are following CSF signal intensity with no perilesional edema or adjacent brain parenchymal gliosis, no diffusion restriction or blooming.

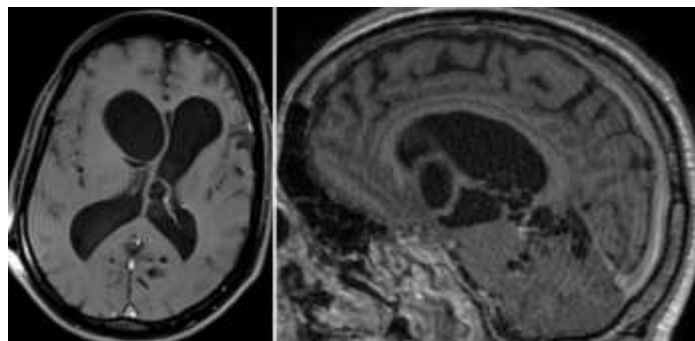


Fig-3: Axial and Sagittal T1 postcontrast images depict that the cystic lesions show no wall enhancement, absence of adjacent meningeal enhancement, no enhancing solid component or eccentric enhancing nodule

CONCLUSION

Giant PVS are fluid filled structures of varied morphology seen in close relation to penetrating vessels of the brain most commonly in mesencephalothalamic region. They follow CSF signal intensity on all sequences, show no enhancement, diffusion restriction or blooming with unremarkable adjacent brain parenchyma. These are asymptomatic in vast majority and rarely enlarge to significant proportions causing mass effect on adjacent brain parenchyma or ventricular system causing obstructive hydrocephalus which requires surgical intervention. They need to be differentiated from infectious and non-infectious inflammatory as well as neoplastic etiologies of varied nature. Being aware of their characteristic appearance and location is prerequisite to definite diagnosis and obviating inadvertent medical or surgical treatment as well as repeated imaging.

REFERENCES

1. Ahmad M, Narayanasamy S, Siddiqui MA, Ahmad I. Giant perivascular spaces: utility of MR in differentiation from other cystic lesions of the brain. *Journal of the Belgian Society of Radiology*. 2014 Nov 1;97(6).
2. Sankararaman S, Velayuthan S, Ambekar S, Gonzalez-Toledo E. Giant tumefactive perivascular spaces: A further case. *Journal of pediatric neurosciences*. 2013 May;8(2):108.
3. Ding J, Sigurðsson S, Jónsson PV, Eiriksdóttir G, Charidimou A, Lopez OL, van Buchem MA, Guðnason V, Launer LJ. Large Perivascular Spaces Visible on Magnetic Resonance Imaging, Cerebral Small Vessel Disease Progression, and Risk of Dementia: The Age, Gene/environment Susceptibility–reykjavik Study. *JAMA neurology*. 2017 Sep 1;74(9):1105-12.
4. Salzman KL, Osborn AG, House P, Jinkins JR, Ditchfield A, Cooper JA, Weller RO. Giant tumefactive perivascular spaces. *American journal of neuroradiology*. 2005 Feb 1;26(2):298-305.
5. Kwee RM, Kwee TC. Virchow-Robin spaces at MR imaging. *Radiographics*. 2007 Jul;27(4):1071-86.
6. Mathias J, Koessler L, Brissart H, Foscolo S, Schmitt E, Bracard S, Braun M, Kremer S. Giant cystic widening of Virchow-Robin spaces: an anatomofunctional study. *American Journal of Neuroradiology*. 2007 Sep 1;28(8):1523-5.
7. Rudie JD, Rauschecker AM, Nabavizadeh SA, Mohan S. Neuroimaging of Dilated Perivascular Spaces: From Benign and Pathologic Causes to Mimics. *Journal of Neuroimaging*. 2018 Mar;28(2):139-49.
8. Fujimoto K, Kuroda JI, Hide T, Hasegawa Y, Yano S, Kuratsu JI. Giant tumefactive perivascular spaces that expanded and became symptomatic 14 years after initial surgery. *Surgical neurology international*. 2012;3.
9. Duperron MG, Tzourio C, Sargurupremraj M, Mazoyer B, Soumaré A, Schilling S, Amouyel P, Chauhan G, Zhu YC, Debette S. Burden of Dilated Perivascular Spaces, an Emerging Marker of Cerebral Small Vessel Disease, Is Highly Heritable. *Stroke*. 2018 Feb 1;49(2):282-7.