

GIANT VIRCHOW ROBIN SPACES WITH TRIVENTRICULAR HYDROCEPHALUS- A CASE REPORT

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ABSTRACT

VRS are peri-vascular spaces which are fluid-filled spaces, pial-lined, accompanying perforating arteries and venules. Dilated VRS occur in three characteristic locations. Type I VRS occur along the lenticulo-striate arteries entering the basal ganglia through the anterior perforating substance. Type II VRS are seen over the high convexities along the course of perforating medullary arteries as they enter the cortical gray matter and extend into the white matter. Type III VRS are seen in midbrain. Expanding lacunae, giant or tumefactive Virchow Robin spaces in brainstem are rare and may cause of benign aqueductal obstruction with resultant proximal triventricular hydrocephalus. We report a case of 48-year old hypertensive patient who presented with giddiness, frontal headache, swaying towards right side with diplopia in right eye. MRI brain showed enlarged Virchow Robin spaces in pons and mid-brain on left side extending to left thalamus and left brachium pontis causing compression over fourth ventricle and aqueduct with proximal obstructive dilatation of both lateral ventricles and third ventricle.

Keywords: Virchow Robin spaces, Virchoma, Giant, Tumefactive, Lacunae.

Introduction

VRS are peri-vascular spaces which are fluid-filled spaces, pial-lined, accompanying perforating arteries and venules¹ (Fig. 1). They are commonly seen above the anterior perforating substance and anterior commissure along lenticulo-striate arteries and less commonly seen in subcortical and sub-insular regions.² Usually VR spaces are 'leave me alone' lesions. Dilated VRS occur in three characteristic locations. Type I VRS seen along the lenticulo-striate arteries entering the basal ganglia through the anterior perforating substance. Type II VRS seen along the course of perforating medullary arteries over the high convexities as they enter the cortical gray matter and extend into the white matter. Type III VRS are seen in mid-brain.

Expanding lacunae in brainstem is a rare cause of

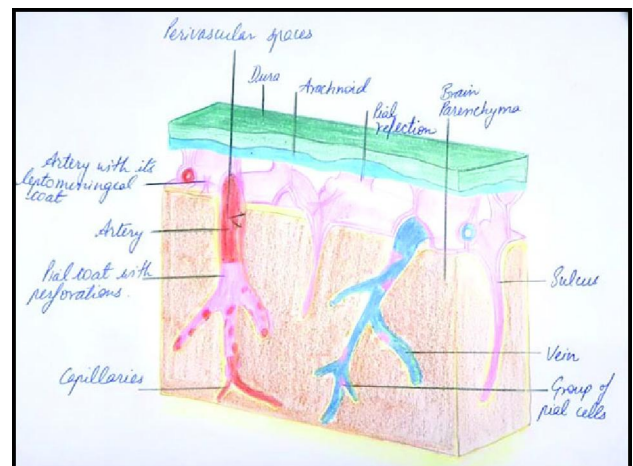


Figure1: Diagrammatic representation of Virchow Robin spaces.

benign aqueductal obstruction and non-communicating normal pressure hydrocephalus. They are also called expanding cerebral lacunae.³

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Expanding VRS causing mass effect and hydrocephalus are named as tumefactive VRS and are usually seen in mesencephalothalamic region. They are also called Virchoma or cavernous dilatation of VRS or giant VRS.²

Case Report

A 48-year old hypertensive male presented with chief complaints of diplopia in right eye, frontal headache, and giddiness since 3 weeks. He gave a history of swaying towards right side while walking, slowed and slurred speech since last 1 year. He also had history of seizures in his childhood.

CT scan of brain with contrast revealed large multilocular cystic lesion measuring approximately 37 (AP) x 25 (T) x 30 (CC) mm in size, in brainstem, involving left brachium pontis, pons and midbrain on left side, extending to left cerebral peduncle and superiorly into left thalamus (Fig. 2). Mass effect was noted on left antero-lateral aspect of fourth ventricle, aqueduct and posterior third ventricle with resultant proximal obstructive dilatation of both lateral ventricles. No

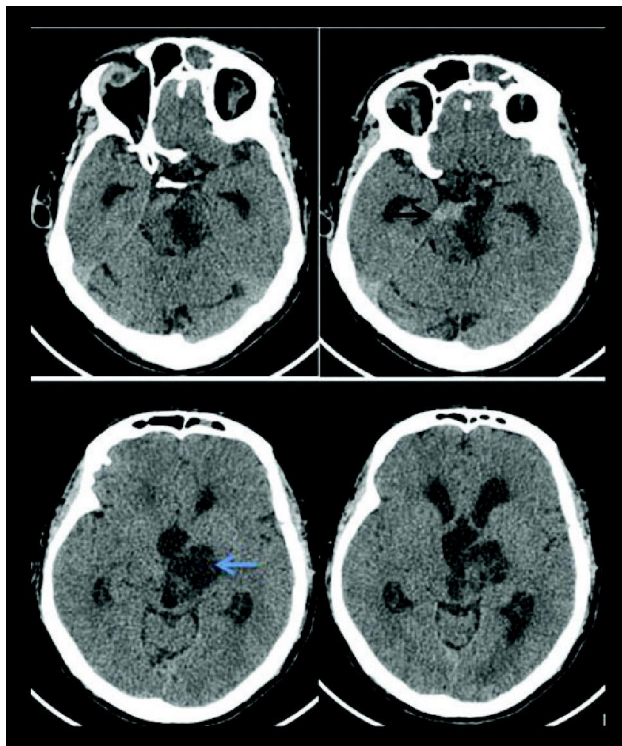


Figure 2: Non-contrast axial CT images showing hyper-density in region of basilar aneurysm (black arrow) and CSF-density giant VR spaces (blue arrow) in pons, midbrain and left thalamus.

enhancement was noted in post-contrast study (Fig. 3). No solid mural nodule was noted within the cystic lesion.

Incidentally, a saccular aneurysm measuring approximately 11 (CC) x 9 (AP) x 8 (T) mm was noted arising from terminal portion of basilar artery, extending along its right lateral aspect in contrast study. A peri-aneurysmal hematoma of size 17 x 15 mm was noted along its right postero-lateral aspect appearing hyperdense with respect to brain parenchyma on plain study. This was confirmed on CT cerebral angiography (Fig. 3).

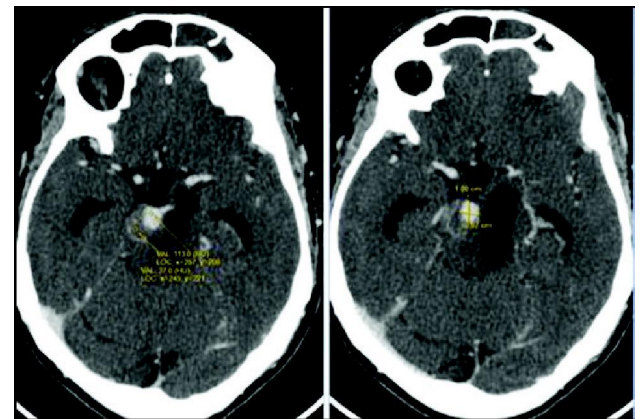


Figure 3: Post contrast axial CT images showing basilar artery saccular aneurysm with peri-aneurysmal hematoma adjacent to the non-enhancing giant VR spaces.

MRI brain showed CSF intensity multilocular cystic lesion and multiple small cysts adjacent to each other at above mentioned locations with ventricular mass effect and obstructive dilatation of both lateral ventricles (Fig. 4). Subtle areas of gliosis were noted in adjoining brain parenchyma in pons, mid-brain and left thalamus appearing hypointense on T1WI (Fig. 4,5) and hyperintense on T2WI (Fig. 6) and hypointense on FLAIR (Fig. 7), showing no diffusion restriction (Fig. 8). No solid component was noted. No abnormal enhancement or enhancing solid nodule was noted in contrast study. Saccular aneurysm was noted near the bifurcation of basilar artery which was seen as a prominent flow void on T2WI. This was confirmed on MR brain angiography (Fig. 9). Peri-aneurysmal hematoma was noted on right postero-lateral aspect of the aneurysm which appeared iso-intense on T1WI and hypointense on T2WI. On MR spectroscopy, no abnormal brain metabolites with normal Cho/Cr ratio and presence of lactate was detected without increase in choline.

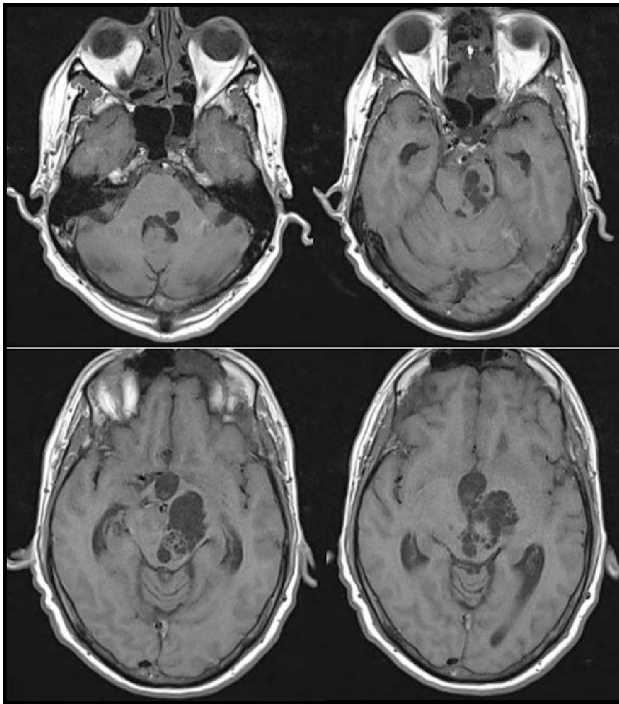


Figure 4: T1W axial images at level of pons, midbrain and thalamus depicting giant VR spaces with signal intensity similar to CSF.

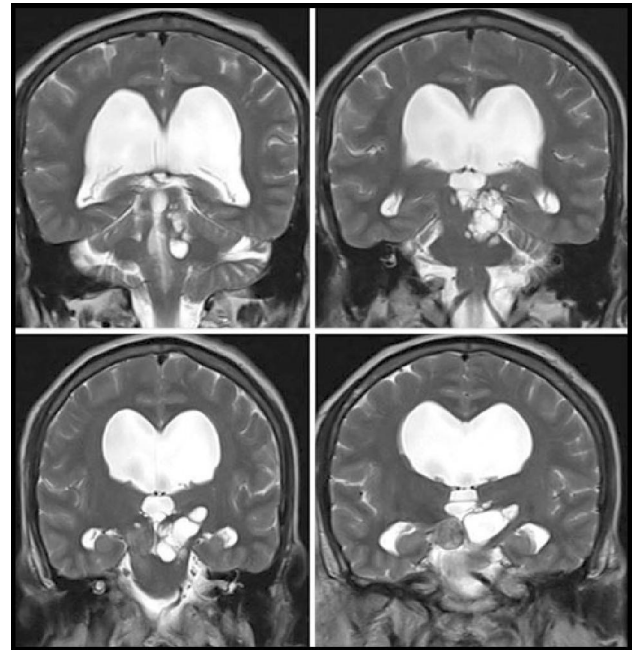


Figure 6: Coronal T2W images showing the cranio-caudal and transverse extent of giant VR spaces from brachium pontis to left thalamus. Also seen is a well-defined basilar artery aneurysm with peri-aneurysmal hematoma on the contra-lateral side.

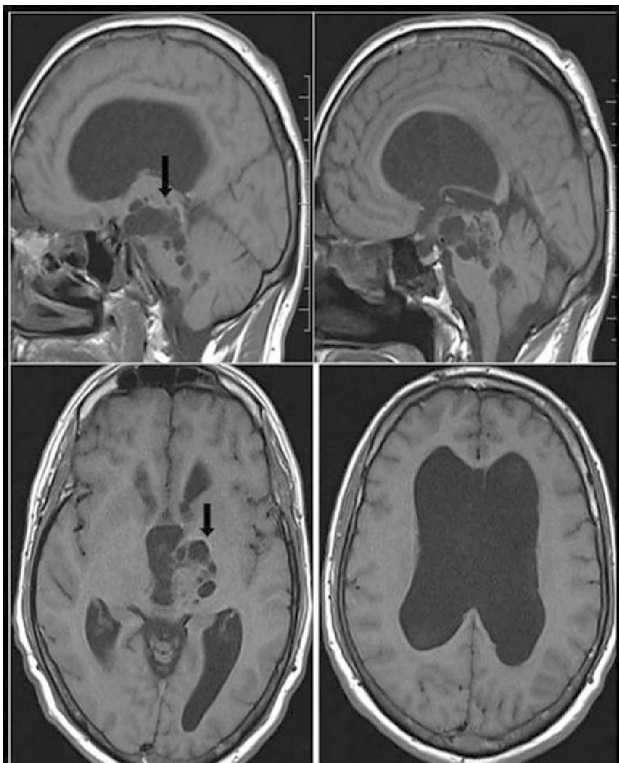


Figure 5: Sagittal and axial T1W images depicting giant VR spaces (black arrow) causing compression over the aqueduct with resultant tri-ventricular hydrocephalus.

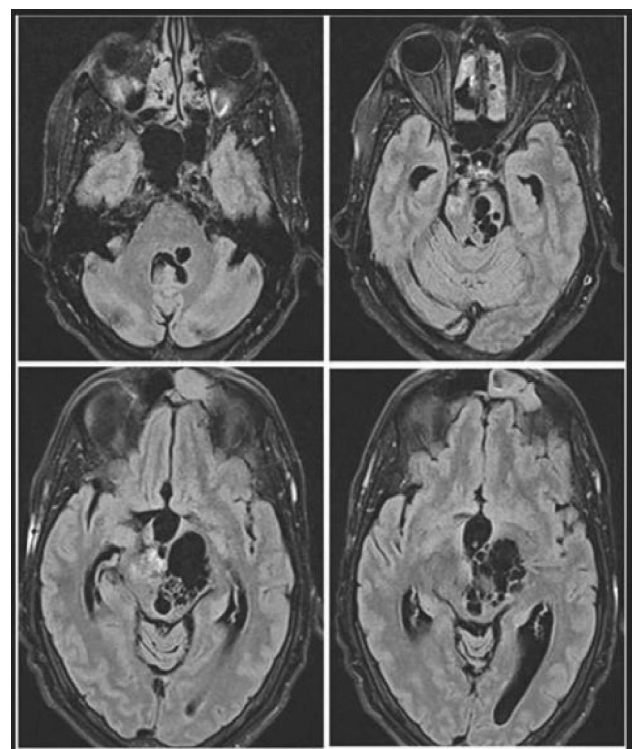


Figure 7: FLAIR sequence axial images at level of pons, midbrain and thalamus depicting giant VR spaces with signal intensity similar to CSF.

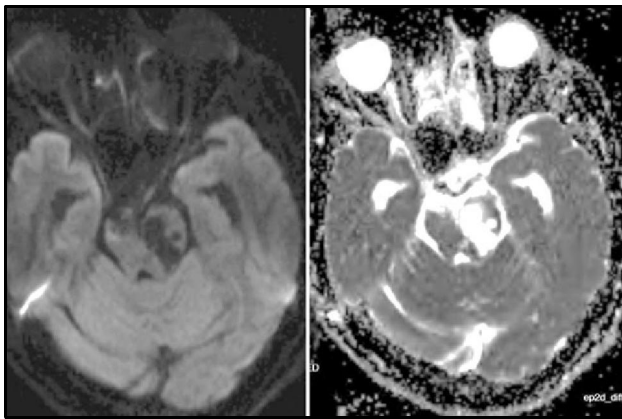


Figure 8: Diffusion weighted images showing no restriction in the region of giant VR spaces.

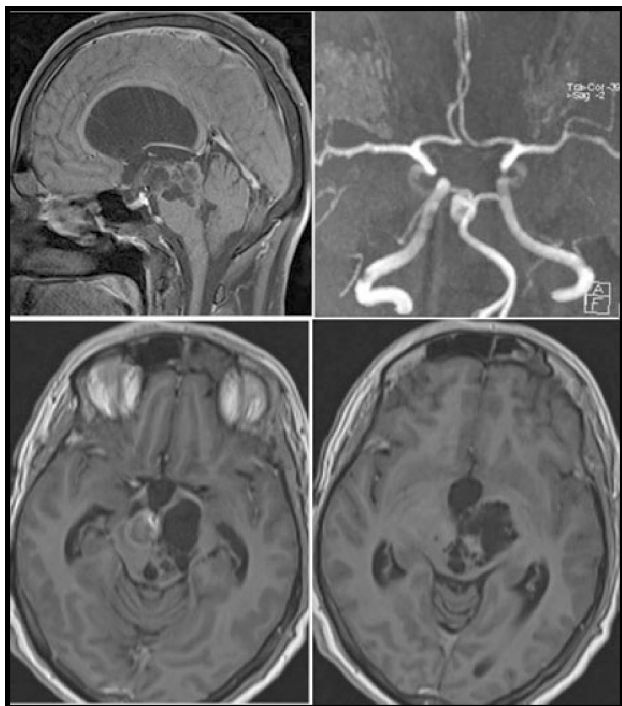


Figure 9: MR post contrast images and MR angiographic image demonstrating the basilar artery aneurysm along-with the non-enhancing giant VR spaces.

A diagnosis of giant type III Virchow Robin spaces in brainstem was made with tri-ventricular hydrocephalus.

Discussion

Pestalozzi 1849 was the first to describe the perivascular space. It came to be known as Virchow Robin spaces as it was described by Virchow in 1851, a German pathologist as a sub-adventitial space conti-

nuous along the capillaries.⁴

Brain lacunae based on pathological features is divided into 3 types by Pourier and Derouesne. Type I lacunae corresponds to lacunar infarction. Type II lacunae corresponds to small intra-parenchymal hematomas. Type III (expanding) lacunae corresponds to dilated perivascular VR spaces. Type III c lacunae correspond to perivascular dilatation at the entrance of a perforating artery into the lentiform nucleus.³ Typical VRS on MRI are seen as tiny to large CSF intensity spaces with well-defined smooth margins showing no enhancement after contrast administration and are surrounded by normal brain parenchyma.³

They follow the path of penetrating arteries and have no mass effect. Mild grade I VRS are <2 mm in diameter. Moderate grade II VRS are 2-3 mm in diameter. Marked grade III VRs are >3 mm in diameter.⁴

They can be found in brainstem along penetrating branches of collicular and accessory collicular arteries.⁵

Hypertension, dementia, advanced age and incidental white matter lesions are significantly associated with grade II and III VRS.⁴

Type III VRS are lined by a single stratum of epithelium like flat cells and contain a small artery/ arteriole. Adjoining brain parenchyma adjacent to expanding lacunae show limited edema, gliosis, spongiosis or demyelination.³

Abnormal dilatation of VR spaces/ expanding lacunae are rarely seen, they can be asymmetric as the lesions predominate at one side of the brain. They can cause hydrocephalus due to compression on adjoining cerebral aqueduct. Demyelination in adjoining brain is due to mass effect by enlarged VR spaces on the adjacent brain parenchyma.

Various mechanisms have been proposed.

1. Derouesne et al proposed that VR spaces arise from difficult drainage of interstitial fluid into ventricular system due to increased intra-ventricular pressure. This hypothesis can be accepted only if there is primary hypertension within the ventricular system.⁶
2. Mascalchiother mechanism is compression of aqueduct by enlarged VR spaces rather than its cause. This theory is supported by observation that treatment of hydrocephalus does not prevent the enlargement of VR spaces.⁷

3. Homeyer et al proposed that there is an initial disorder of the lymphatic drainage leading to an impaired interstitial fluid drainage with enlargement of VRS.⁸
4. Poirier and Derouesne 1984-described these lacunae as type III d. they thought that these enlarged VRS are due to changes in vascular permeability caused by vasculitis.⁹

Differential Diagnosis are parasitic cysts, infection, cystic tumours, epidermoid cysts. DWI can differentiate Virchow Robin spaces from epidermoid cysts which shows restricted diffusion related to CSF. MR Spectroscopy differentiates it from parasitic cysts, abscesses and tumours.

Cystic tumours will show decreased NAA, increased Cho with variable amount of lactate and lipids. Abscesses and parasitic cysts show varying amounts of acetate, lactate, succinate and amino acids with absence of normal brain metabolites. Mild increase in choline can be seen due to demyelinated areas surrounding these lesions.³

Location of cystic lesions corresponding to the path of penetrating arteries, multiplicity, absence of calcification and solid components, signal identical to CSF on all MR sequences, no contrast enhancement, negative laboratory findings for parasites, no abnormal metabolites on MR spectroscopy with normal NAA/ Cr and Cho/ Cr ratios with presence of lactate are useful in diagnosis of expanding VR spaces.³

Though asymptomatic, the expanding lacunae can cause aqueductal compression with resultant triventricular hydrocephalus and become symptomatic. Symptoms can occur due to compressive effects on adjacent brain parenchyma. Placement of a CSF shunt or ventriculo-cisternotomy can treat hydrocephalus but does not cause arrest of growth of lesion and do not modify the focal signs or symptoms.³

Conclusion

Expanding VRS in brainstem can become symptomatic by causing compression on aqueduct with resultant proximal obstructive hydrocephalus. Our case is unique as type III VRS were extending into brachium pontis as well as thalamus.

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