ORIGINAL RESEARCH PAPER

Radiodiagnosis

GIANT MESENCEPHLOTHALAMIC VIRCHOW ROBIN SPACES CAUSING OBSTRUCTIVE HYDROCEPHALUS

KEY WORDS: Virchow robin space Magnetic resonance imaging Hydrocephalus Mesencephalothalamic region

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ABSTRACT

Virchow-Robin (VR) spaces in the brain are pial-lined perivascular spaces which surround the perforating arteries and arterioles. These interstitial fluid filled spaces are well appreciated on magnetic resonance imaging (MRI). VR spaces are small in size and do not cause any clinical symptoms however gross dilations can present with varied clinico-radiological manifestations and complications. We report such a case of a 43 year old female patient presenting with complains of headache, and progressive cognitive decline accompanied by memory loss for the past 1 year. Magnetic resonance imaging revealed extreme dilatation of the VR spaces in the mesencephalothalamic regions with mass effect in the form of obstructive hydrocephalus and a diagnosis of Giant VR spaces was made. Occurrence of such large cystic spaces can mimic various pathologic conditions making it imperative to differentiate it from them, owing to the differences in their management.

Abbreviations:
VR- Virchow Robin
CSF- cerebrospinal fluid
OPD- outpatient department
MRI- Magnetic resonance imaging
FLAIR- fluid attenuation inversion recovery
ADC- apparent coefficient gradient

Introduction-

The Virchow-Robin space was first described in 1851 by Rudolf Virchow (German pathologist, 1821–1902) and later confirmed in 1859, by Charles Philippe Robin (French anatomist, 1821–1885) as perivascular spaces that surround the walls of vessels as they course from the subarachnoid space through the brain parenchyma.[1] However they do not communicate directly with the subarachnoid space. These spaces form a major pathway for lymphatic and interstitial fluid drainage from the brain, hence playing a significant role in the immune response to infections and in the clearance of cellular debris by phagocytosis.[2] VR spaces are commonly seen at magnetic resonance imaging (MRI) as small interstitial fluid filled spaces measuring <2mm[3] with signal intensities near to those of CSF with all pulse sequences. Dilatations of these perivascular spaces can occur occasionally causing specific clinical manifestations [2,4] depending on the location[4] and degree of tissue compression. We report a rare case of the giant dilations of the VR spaces in mesencephalothalamic region causing obstructive hydrocephalus. Magnetic resonance imaging findings of such giant dilatations can be similar to various pathological conditions.

Case report:

A 43 year female presented to the OPD with complains of headache since 2 years. The patient in addition also complained of imbalance and progressive cognitive decline accompanied by memory loss for the past one year. On neurological examination no significant sensory or motor deficit was noted with mild decline in the cognitive function.

Magnetic resonance imaging of the brain was performed which revealed multiple variable sized thin walled cystic lesions involving bilateral thalami and the midbrain. They appeared isointense to CSF on all pulse sequences [Figure 1(a-e)] with no restriction on diffusion [Figure 1(g-h)]. On post contrast study no enhancement was noted (Figure 2).The adjacent brain parenchyma showed normal signal intensity although there was significant compression of the third ventricle causing obstructive hydrocephalus and mild periventricular ooze [Figure 3(a-c)].

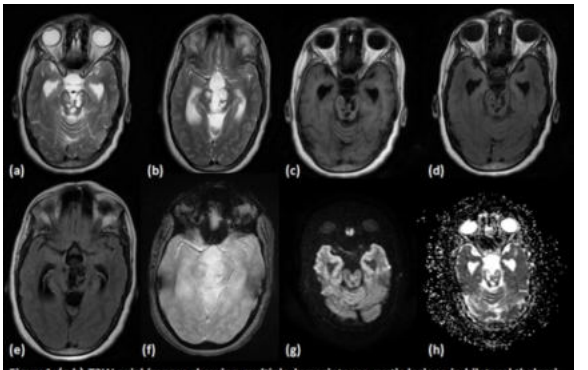

Figure1: (a,b) T2W axial images showing multiple hyperintense cystic lesions in bilateral thalami and the midbrain, with dilatation of the temporal horn of bilateral lateral ventricles.
(c)T1W axial image shows multiple hypointense lesions in the midbrain with dilated temporal horns of bilateral ventricles.
(d,e) FLAIR axial images shows multiple large well defined hypointense lesions in the midbrain with dilatation of bilateral lateral ventricles.
(f) Gradient echo image shows no blooming in the midbrain.
(g,h) Diffusion and ADC images show no evidence of restriction of the cystic lesions in the midbrain.

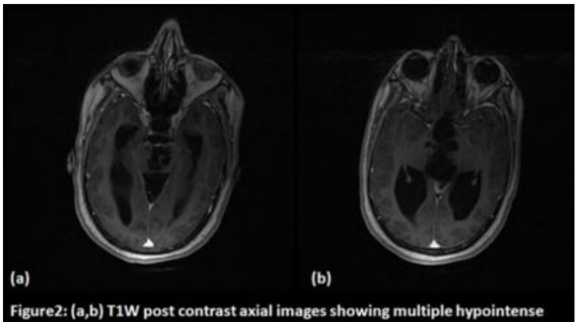

Figure2: (a,b) T1W post contrast axial images showing multiple hypointense cystic lesions in bilateral thalami and the midbrain without any evidence of post contrast enhancement. The bilateral lateral and third ventricle appear dilated.

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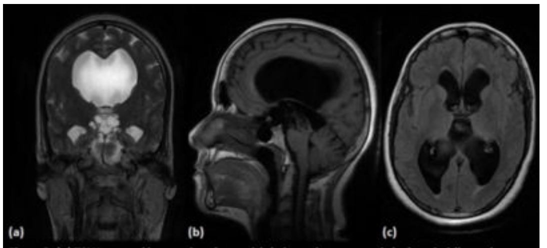

Figure3: (a) T2W coronal image showing multiple hyperintense cystic lesions in bilateral thalami and the midbrain largest measuring 10mm, with dilatation of the temporal horn of bilateral lateral ventricles.
(b)T1W sagittal image shows multiple hypointense lesions in the midbrain with dilated lateral and third ventricles and a normal 4th ventricle suggestive of obstructive hydrocephalus.
(c) FLAIR axial image shows dilated frontal and occipital horns of bilateral lateral ventricles with mild periventricular hyperintensities.

Discussion:

VR or perivascular spaces are normal anatomic structures which surround the walls of arteries, arterioles, veins, and venules as they course from the subarachnoid space through the brain parenchyma.[1] They are extensions of the subpial space however they do not have a direct connection with the subarachnoid space.[5,6] Hence the fluid within them is of slightly different composition as compared to the CSF. Small VR spaces (<2 mm) can occur in all age groups.[2,3] But with advancing age VR spaces are found with increased frequency and larger size (>2 mm).[3] Dilatation of VR spaces was first described by Durant-Fardel in 1843. These dilatations are cavities which always contain a patent artery. The causative factors for expanding VR spaces are still unknown. Several hypotheses have been put forward, normally high arterial wall permeability due to vasculitis[7], impaired interstitial fluid drainage due to lymphatic obstruction, impaired interstitial fluid drainage into the ventricles due to increased intraventricular CSF pressure, spiral elongation of penetrating blood vessels.[2,4,5] VRS dilations can also be a cause of hydrocephalus, due to local compressions altering CSF circulation and absorption.

Depending on their location, dilated VR spaces can be categorised into three types[2]:

Type I: located along lenticulostriate arteries entering towards the basal ganglia;

Type II: located in the cortex following the path of the medullary arteries;

Type III: situated in the midbrain. This type corresponds to the diagnosis of our patient.

Diagnosis of dilated VR spaces can be made with certainty on MRI.[8] VR space dilations appear as interstitial fluid-filled cavities, isointense to CSF (ie, T1-hyposignal and T2-hypersignal) on all pulse sequences. They show no enhancement on post contrast studies. The common locations are in the inferior third of the basal ganglia, in the mesencephalon, or in the hemispheric white matter in the subcortical regions of the convexity.[2,4]

VR dilations are usually small and asymptomatic and are viewed as non-pathological anatomic variations. However dilations >2 mm are more common among the elderly and are associated with microcirculation alterations and subcortical atrophy. They can also be observed in patient with cognitive decline[9] or strokes. These giant dilatations (>10mm) observed in our case in the midbrain have rarely been reported.

Clinically, giant VR spaces are associated with headache, dizziness, visual changes, dementia, seizure, syncope, stroke and cognitive impairment.[8,9] When the dilatations are large enough, symptoms related to mass effect may be observed. Giant VR spaces in the mesencephalothalamic region can cause complication of obstructive hydrocephalus [10] by compressing the third ventricle as seen in our case.

Asymptomatic VR dilations require no specific treatment and should be monitored only if they are numerous. Treatment of symptomatic VR dilatations depends on the type as well as the localisation of the lesions. Significant dilations often require surgical treatment [4,10,11], especially in cases of cranial hypertension where the purpose of the surgery is to aid in CSF diversion, while cyst drainage and fenestration is required to relieve any focal mass effect. Third ventriculocisternostomy and ventriculoperitoneal shunt are commonly employed surgical procedures with third venriculocisternostomy [12] being the preferred one.

Differential diagnosis:

Dilated VR spaces can be easily misinterpreted for a cystic neoplasm.[2] Other common differentials include lacunar infarction, cystic periventricular leukomalacia, parasitic infections, multiple sclerosis, neuroepithelial cysts, arachnoid cysts, or mucopolysaccharidosis[2,10]. Absence of perilesional gliosis, non enhancement, absence of soft tissue or solid component and typical location of the cysts helped us narrow down the differential to tumefactive VR spaces.

Conclusion:

Giant tumefactive VR spaces are interstitial-fluid filled spaces that follow the arteries and arterioles as they penetrate the brain. They are round or oval, single or multilocular cystic lesions isointense to CSF with no enhancement on post contrast studies. Mesencephalothalamic region is the most commonly involved and giant dilatations in this region as in our case (>10mm in size) can be associated with obstructive hydrocephalus. Knowledge of their imaging characteristics and localization helps in differentiating them from different pathologic conditions.

Learning points:

1. Virchow-Robin spaces are normal pial lined spaces which surround the vessels as they enter the brain parenchyma.
2. They are asymptomatic when small, large dilatations are associated with various clinical symptoms depending on the location of occurrence.
3. Hydrocephalus is the most frequently encountered complication of giant VR dilations, but stroke or dementia can be observed.
4. MRI is the modality of choice for its diagnosis as it follows CSF signal intensities on all pulse sequences.
5. It is imperative to differentiate it from various pathological conditions as the treatment strategy varies.

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