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Original Article

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Evaluation of Radiological Signs/ Findings of Normal Pressure Hydrocephalus in Patients Presenting with Hakim Triad

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ABSTRACT

Objectives:

- To evaluate different radiological signs / findings of Normal pressure hydrocephalus (NPH) in MRI, in patients with atleast 2 symptoms of hakim triad.
- The role of different radiological signs in differentiating patients with NPH and patients without NPH
- To rule out other closely related differential diagnosis for NPH.

Materials & Methods: This study was performed using 1.5 Tesla Philips D stream MRI machine. Imaging and clinical data of 50 patients with atleast 2 symptoms from Hakim triad from 2022-2018 were reviewed retrospectively and different radiological signs are evaluated for NPH.

Results & Conclusion: Out of 50 cases in our study, Small Callosal angle, Disproportionately enlarged subarachnoid space hydrocephalus, Wider temporal horns and idiopathic NPH RAD scale helps to differentiate, patients with NPH from patients without NPH. Out of all D/D, Alzheimer's being most closely related D/D.

Key Words: *Pressure Hydrocephalus, NPH*

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INTRODUCTION

Normal pressure hydrocephalus (NPH) is a potentially reversible syndrome characterized by enlarged cerebral ventricles (ventriculomegaly) and hakim triad (cognitive impairment, gait apraxia and urinary incontinence) [1, 2].

Classification of Normal pressure hydrocephalus [3]

PRIMARY	SECONDARY
Idiopathic NPH	SAH Meningitis Intracerebralhaemorrhage Brain tumors Traumatic brain injury Ischaemic stroke

International guidelines for the diagnosis of idiopathic NPH [4]

1) Ventricular enlargement with Evan's index > 0.3
2) Absence of macroscopic obstruction to the CSF flow
3) At least one of these supportive features –

- Enlarged temporal horns of lateral ventricles, not entirely attributable to hippocampal atrophy.
- Callosal angle of 40^0 or greater

- Periventricular signal changes on CT and MRI due to altered brain water content and not attributable to microvascular ischaemic changes ordemyelination.
- Flow void in sylvian aqueduct or 4thventricle on MRI

Japanese guidelines for diagnosis of NPH [5]

- Narrowing of sulci and subarachnoid spaces over high convexity and midline surface of brain
- Enlarged sylvian fissure and basal cisterns

Periventricular signal changes are not essential, according to Japanese guidelines.

Radiological signs of NPH*:*

- Evan's ratio >0.3
- Callosal angle (40^0-90^0)
- Wider temporal horns
- Disproportionately enlarged subarachnoid space hydrocephalus(DESH)
- Dilated 3rdventricle
- Idiopathic NPH RAD score > 8

PURPOSE OF THIS STUDY:

To evaluate different radiological signs / findings of normal pressure hydrocephalus in MRI, in patients with atleast 2 symptoms of hakim triad. We have taken even 2 symptoms as positive clinical outcome for the purpose of this study.

MATERIALS & METHODS:

- Imaging and clinical data of 50 patients with atleast 2 symptoms from hakim triad from 2022-2018 were reviewed retrospectively and different radiological signs are evaluated for NPH
- All MRI examinations were conducted through 1.5 Tesla Philips scanner d Stream MRI machine.

RESULTS:

Radiological signs of NPH	***No of cases***
Evans ratio > 0.3	37
Callosal angle 40^0-90^0	27
Wider temporal horn(>6mm)	31
NPH RAD score >8	23
DESH	25
Dilated 3rd ventricle	35

D/D of Hakim triad	***No of cases(total=50cases)***	***Percentage***
NPH	27	54%
Alzheimer's	13	26%
MSA	2 cases of MSA-P	4%
	3 cases of MSA-C	6%
PSP	5	10%

Out of 50 cases in our study, Callosal angle, Disproportionately enlarged subarachnoid space hydrocephalus, Wider temporal horns and idiopathic NPH RAD scale helps to differentiate, patients with NPH from patients without NPH.

DISCUSSION

RADIOLOGICAL SIGNS OF NPH

EVANS INDEX:

It is ratio of maximum width of the frontal horns of lateral ventricles to the maximum width of inner table of cranium at the level of frontal horns [6]. (or)

The ratio of maximum width of the frontal horns of lateral ventricle to the largest inter diameter of the skull.

This ratio was measured in axial plane, on the same axial level. Evans index > 0.3indicates ventriculomegaly. This index is not precise and informative enough to help with the differential diagnosis of NPH. This index can also be greater in atrophy (like Alzheimer's) This index cannot differentiate NPH and atrophy.

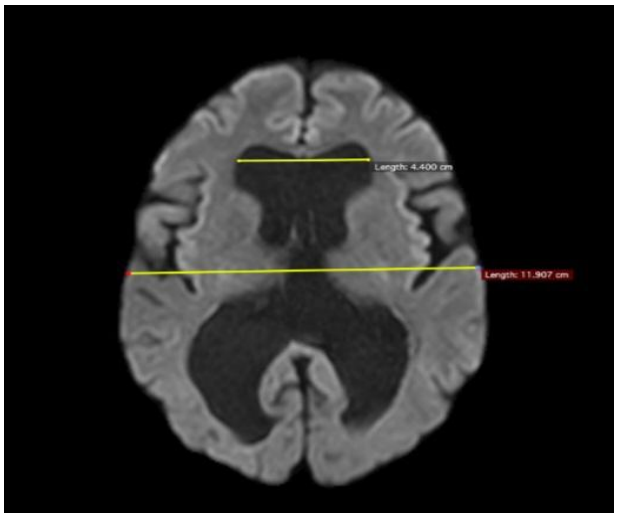

Figure 1: This is an axial FLAIR MRI image showing dilated lateral ventricles with evans ratio of > 0.3 indicating ventriculomegaly

CALLOSAL ANGLE:

Angle was made by drawing 2 lines tangentially aligned with superior medial walls of lateral ventricles [7]. It is the radiological sign that can distinguish NPH and atrophy Eliminates non hydrocephalus patients Small callosal angle < 90^0 – present in NPH Callosal angle 100^0-120^0- present in normal individuals and atrophy that occurs in alzheimer's and lewy body dementia Callosal angle is best evaluated on coronal plane

Figure 2: This coronal T2W MRI image showing dilated lateral ventricles with narrow callosal angle < 90^0

DILATED 3RD VENTICLE:

In idiopathic NPH cerebrospinal flid flows back and forth through aqueductdue to cardiac pulse, while net direction is towards 4th ventricle [8]. In addition to the dilated lateral ventricles, 3rd ventricle also dilates. Walls of 3rd ventricles bows outward in idiopathic NPH Concave walls of 3rd ventricle present in normal people. Dilated 3rd ventricle can be best evaluated in coronal and axial planes.

WIDER TEMPORAL HORNS:

Temporal horns or inferior horns of lateral ventricles project anteriorly and inferiorly in to temporal horn of brain [9] Width of temporal horns 6mm or more -seen in idiopathic NPH Wider temporal horns is typical of idiopathic NPH Wider temporal horns best evaluated in axial planes.

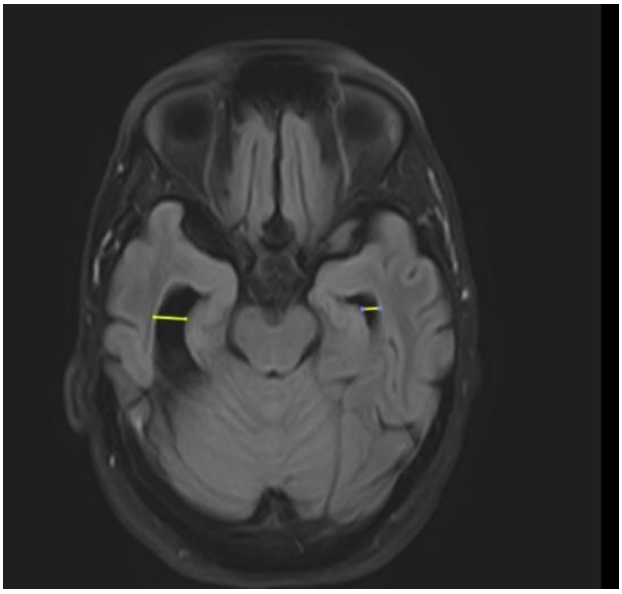

Figure 3: This is an axial FLAIR MRI image showing wider temporal horns or inferior horns of lateral ventricles having width of 8mm on right side and 6mm on left side.

DISPROPORTIONATELY ENLARGED SUBARACHNOID SPACE HYDROCEPHALUS (DESH) [10]:

It is characterized by –
More crowded sulci near superior cortex, while larger spaces seen inferiorly where CSF accumulates
+
Enlarged sylvian fissure and basal cisterns

DESH is the promising sign for idiopathic NPH

Narrowed sulci and focally dilated sulci were best evaluated on both coronal & axial planes Sylvian fissure was best evaluated on coronal planes

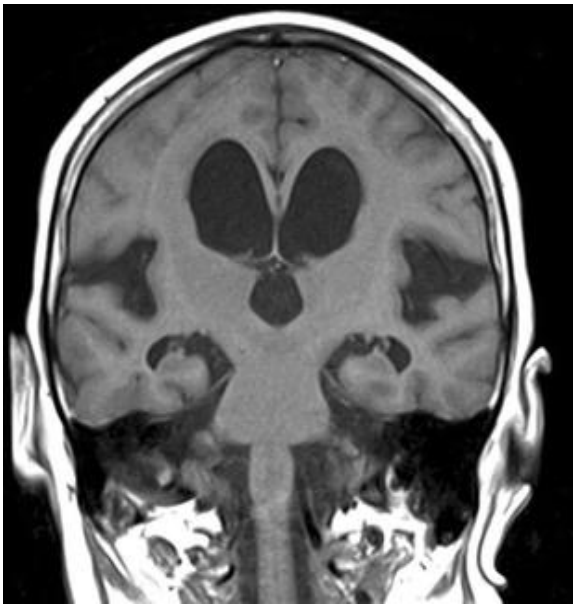

Figure 4: This is coronal FLAIR MRI image showing crowded sulci at superior cortex with enlarged sylvian fissure and basal cisterns.

IDIOPATHIC NPH RAD SCALE:

Idiopathic NPH RAD score ranges from 0 to 12 Idiopathic rad score could work as a diagnostic tool to detect idiopathic NPH. It contains total of 7 parameters that includes- Evans index, callosal angle, size of temporal horns, narrow high-convexity sulci, dilated Sylvian fissures, focally dilated sulci, peri-ventricular hypodensities.

Idiopathic NPH RAD score > 8 - suggests diagnosis of idiopathic NPH. Idiopathic NPH RAD score< or equal to 4 present in patients without NPH. Bulging of the lateral ventricular roof, DESH(the combination of high-convexity tightness, Sylvian fissure dilation, and ventriculomegaly) were not included in the score .

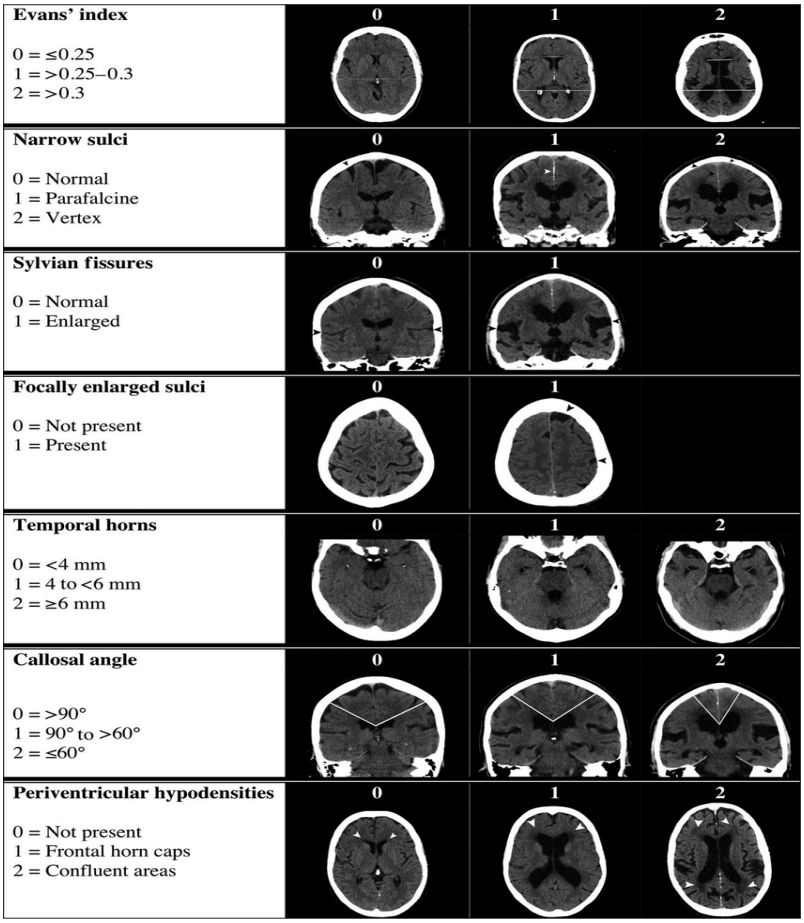

Figure 5: represents parameters of Idiopathic NPH RAD score

FAZEKAS SCALE:

The scale divides the white matter in periventricular and deep white matter, and each region is given a grade depending on the size and confluence of lesions [11]

- Periventricular white matter (PVWM)
- 0 = absent
- 1 = "caps" or pencil-thin lining
- 2 = smooth "halo"
- 3 = irregular periventricular signal extending into the deep white matter
- Deep white matter (DWM)
- 0 = absent
- 1 = punctate foci
- 2 = beginning confluence
- 3 = large confluent areas

PVWM &WM were best evaluated on FLAIR, T2WI axial planes

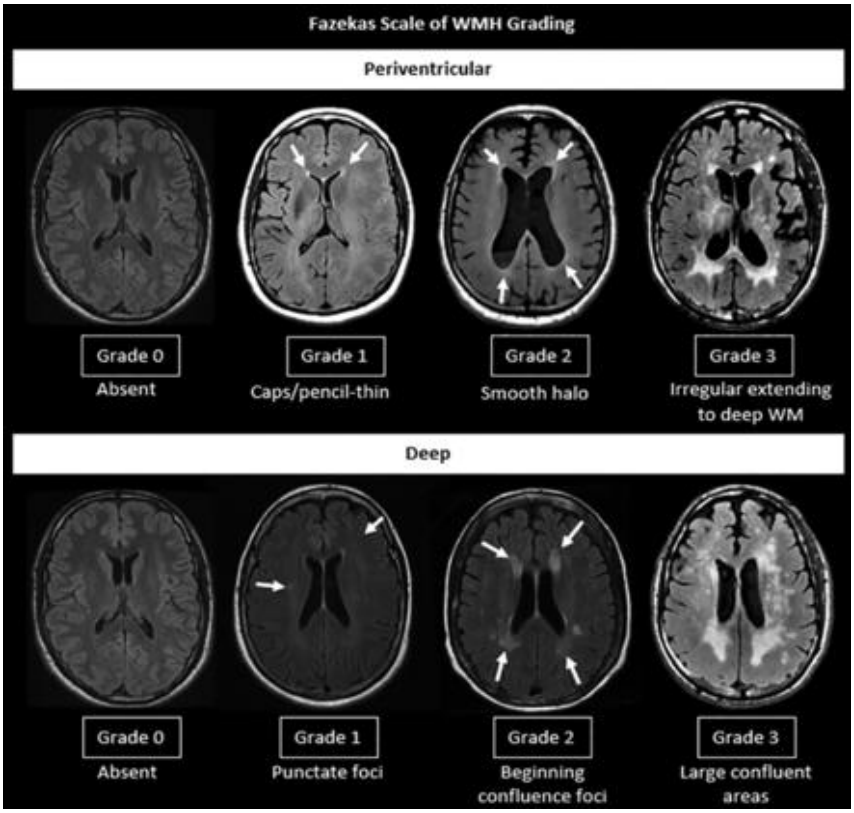

Figure 6: Represents Fazekas scale of grading PVWM &DWM changes

LIMITATIONS OF THIS STUDY:

As this is a retrospective study and due to lack of clinical data, we have data of only limited number of patients who have underwent lumbar puncture & ventriculo-peritoneal shunting. Follow up of these patients whether they had improvement of these symptoms or not is also limited.

As there were limited number of patients presenting with all the three clinical feature of this triad, we had taken patients with atleast two symptoms of this triad as the positive clinical outcome for the purpose of this study.

CONCLUSION

Smaller callosal angle, wider temporal horns, disproportionately enlarged subarachnoid space hydrocephalus (DESH), idiopathic NPH RAD scale helps to differentiate patients with NPH from patients without NPH. Out of all differentials for the NPH, Alzheimer's being most closely related differential for NPH.

LIST OF ABBREVIATIONS:

NPH- Normal pressure hydrocephalus
DESH-Disproportionately enlarged subarachnoid space hydrocephalus
MSA-Multisystem atrophy
MSA-P-Multisystem atrophy Parkinson’s type
MSA-C-Multisystem atrophy Cerebellar type
PSP-Progressive supranuclear palsy
PVWM-Periventricular white matter
DWM-Deep white matter

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