

Frequency of Central Nervous System Anomalies Associated with Hydrocephalus Diagnosed by Magnetic Resonance Imaging in all Ages

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Abstract

Hydrocephalus is a medical condition which is physiologically recognized by elevated intracranial pressure. It occurs mostly as a consequence of certain central nervous system pathologies such as Chiari malformation type I and II, myelomeningocele, space occupying lesions, TBM, Bacterial meningitis, glioblastoma, traumatic brain injuries along with hemorrhage. It is clinically recognized by many stellar signs such as seizures, dementia, gait disturbances and bradycardia along with sleep apnea. The main purpose of our research paper was to carefully evaluate the frequency of different CNS pathologies which represent as the etiology of raised intracranial pressure in all ages. The duration of study was from 01 June 2018 to 01 June 2019 at Lahore General Hospital, Pakistan. A retrospective cross-sectional study included 120 patients which were admitted to Neuro-Radiology department with provisional diagnosis of hydrocephalus along with the associated clinical manifestations. The patients were chosen through convenient sampling technique. All the examined patients included males and females of all ages. Overall 120 patients were included in this study. Out of 120 patients; males were 70 (41.6%) and females were 50 (33.6%) in this study population.

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The central nervous system pathology which was found to be most frequently associated with hydrocephalus was space occupying lesion 25 % (30cases) with mean age of 38.8, seconded by TBM 11.6 % (14cases) with the mean age of 37.2; glioma 9.1% (11cases) with mean age of 42 and SAH 8.3% (10cases) with the mean age of 48.8. In conclusion, MRI was the gold standard modality for the evaluation of hydrocephalus due to central nervous system pathologies.

Keywords: Magnetic Resonance Imaging; Cerebrospinal fluid; Central nervous system pathologies; Hydrocephalus.

1. Introduction

Hydrocephalus preponderance varies from 0.36- 0.75 per 1000 live birth[1]. The most prevalent association of hydrocephalus is found to be linked with the most complicated type of open neural tube defect known as myelomeningocele or spina bifida which is visually recognized as a protrubence of meninges along with the spinal cord through an open defect from the arches of vertebral column(cervical or lumbar) its preponderance mostly ranges from 0.2 -2 per 1000 live birth and the most clinical manifestation associated with it is ventriculomegaly which later embarks itself by disturbing hydrodynamics of CSF circulation and become the pathological leading cause of hydrocephalus which according to statistics range from 57-86% in all cases of myelomeningocele[2]. The other leading cause is Chiari type 2 malformation which is the most commonly aberrant anomaly which is manifested by the downward displacement of 4th ventricle and other structures with closed association with brainstem such as cerebellar tonsils and vermian via foramen magnum, more than 90% of victims associated with Chiari malformation type 2 have hydrocephalus[3]. Aqueductal stenosis is one of the other most leading cause of hydrocephalus , it is most recently reported that there are approximately 30-40% of cases of aqueductal stenosis that varies prevalently with 10-40% with the adult type of hydrocephalus and 15-60% with the congenital type of hydrocephalus[4]. Tuberculosis meningitis accounts 65% of association with hydrocephalus[5]. Choroid plexus papilloma of neuroectodermal origin assigned grade 1 by WHO has close association with obstructive hydrocephalus[6]. Hydrocephalus also exhibit strong staggering association with space occupying lesions such as ependyoma , choroid plexus papilloma , pilocystic astrocytoma and the most persistent of them all is tectal glioma which along with other disrupted physiologic function of brain causes raised intracranial pressure[7]. Hydrocephalus physiologically occurs due to the disruption of normal dynamic of CSF fluid circulation and anatomically in most cases it is characterized by enlarged lateral ventricles , enlarged sylvan fissures and subarachnoid spaces and tapering of sulci over protruded midline space of brain[8]. Hydrocephalus associated signs and symptoms varies according to its type which is based upon the obstruction of cerebrospinal fluid pathway, it might be obstructive /ex vacuo hydrocephalus or non – obstructive /in vacuo hydrocephalus or last most crucial type which occurs most commonly due to traumatic injury or any infectious pathology is non –pressure hydrocephalus but some most common sign and symptoms are vomiting ,nausea ,dementia ,seizures ,gait dyspraxia ,papilledema , decreased upward elevation of eyeball , the classic sun-down sign” of eyeballs and have difficulty elevating their head ,and according to some researches some victims can also have early parkinsonism signs[9]. The clinical manifestation relating to hydrocephalus is physically and mentally very paramount and physical but most customary its is usually diagnosed by constellation of these 3 physical sign known as gait disturbance , urinary continence and dementia

.the other most prominent physical feature that is most commonly examined by physical conduct known as McEwen 's sign in which a striking voice is heard from mostly an infantile enlarged, out spread fontanelle which physically appears like pot head, and it appears more paramount due to more prominent and non pulsatile scalp vein, the striking split pot sound is heard after a gentle percussion to an infantile head. Optic atrophy can eventuate due to confining of the chiasm and optic nerves by amplified 3rd ventricle[10,11]. MRI not only provides with mere diagnosis but helps the physician to accurately plans the treatment of hydrocephalus by detecting its cause which is mostly due to a central nervous system anomalies that precedes to with increased cerebrospinal fluid accumulation, MRI is now advancing with passing time and becoming the gold standard diagnostic tool for hydrocephalus by providing advanced sequences such as heavily T2 weighted sequence and contrast material enhanced MR cisternography (CE-MRC) and 3D MRI T2 weighted sequence which is also the most commonly used sequences in clinical setting, which gives the most integral anatomy of brain because of high resolution reformation due to 3D spacing without increasing specific absorption rate which can sometimes leads to hazardous effect. And the other most widely used sequence is PC-MRI which not only gives the quantitative but also qualitative analysis of cerebrospinal fluid circulation which help a physician to diagnose the associated cause and also to plan the treatment such as shunt placement surgery to reduce the accumulated cerebrospinal fluid, and there are certain radiological or MRI signs which is most commonly seen in the diagnosis of hydrocephalus due to various central nervous system anomalies such as enlarged frontal and temporal of lateral ventricles, and decreased angle of frontal horn, agenesis and elevation of corpus callosum, disrupted integrity of cortical sulci and hyper-intensities seen in the white matter due to interstitial edema caused by hydrocephalus and last but not the least the most superseding sign is flow void signal from the stenosed cerebral aqueduct, the leading cause of hydrocephalus[12] important MRI features which makes the diagnostic accuracy of hydrocephalus very high includes enlarged lateral ventricles; known as ventriculomegaly which is confirmed by Evan's Index. Evan's Index of more than 0.3 confirms ventriculomegaly, which is one of the characteristic sign of hydrocephalus on MRI and can be carefully visualized in all the sequences that is; T1W, T2W, FLAIR and DWI sequences. Dilatation of the recesses of the 3rd ventricle, narrowing and lifting of the corpus callosam and the hyperintensities in the periventricular region can also be easily appreciated on T2W, FLAIR sequences and DWI sequence along with ADC maps. The other principal sign is the aqueductal flow void phenomena on the T2W sequence which is the differentiating sign between communicating and non communicating hydrocephalus. Other techniques used also include phase-contrast MRI (PC-MRI), three-dimensional (3D) heavily T2W sequences and contrast-enhanced MR Cisternography (CE-MRC) for more extensive studies[13]. For this research purpose MRI was considered as the imaging modality which not only introduce novelty through its sequences but also elaborate the best visual acuity anatomy of the structures through which the CSF passes and also explain its circulatory physiology which not only make it most reliable modality to diagnose the disrupted dynamics of CSF known as hydrocephalus but also pin point the associated anomalies which serves as the main reason for the hydrocephalus it also enhance the paradigm for a neuroradiologist and surgeons to select the candidate for surgical intervention such as whether the patient is suitable for lumbar puncture, VP shunting and ETV. MRI sequence also helps to the check the patency of VP shunts; it can be easily evaluated by the 3D-SPACE technique with small isotropic voxels.

2. Methods and Materials

A total of 120 sample size of the patients was considered after calculating the prevalence of hydrocephalus associated with central nervous system anomalies. All of the patients which were hospitalized with the provisional diagnosis of hydrocephalus was undertaken for the purpose of research without any boundaries towards gender and age. GE 3T MRI machine at Lahore General Hospital was used to investigate the research studies. All the sequences which were present in the GE brain protocol were used along with certain special sequences such as ADC and MR spectroscopy. The descriptive data was elaborated in the form of standard deviation, mean, frequency and age statistics. MR images of hydrocephalic brain were obtained by using heavily T1W and T2W FSE sequences along with FLAIR and DWI sequences. MR criteria for hydrocephalus include Evan's Index to determine dilated ventricles.

3. Results

Overall 120 patients were included in this study. Out of 120 patients, 70 were males (58.3%) and 50 were females (41.6%).

Table 1: Gender wise distribution of the frequency and percentage of the cases

	Frequency	Percentage	Mean Age
Male	70	58.3	33.6
Female	50	41.6	36.62

According to table 2 data, out of 120 patients that were enrolled in our study with the provisional diagnosis of hydrocephalus had these clinical manifestations which are explained in the following table.

Table 2: Frequency and percentage of clinical features associated with patients of hydrocephalus

Clinical Features	Frequency	Percentage
Seizures	40	33.6
Dementia	20	16.6
Visual Impairment	12	10
Gait Disturbances	10	8.3
Urinary Incontinence	10	8.3
Bulged Fontanelles	10	8.3
Headache	10	8.3
Vomiting	8	6.6

Out of 120 patients that were included in our study and were admitted with provisional diagnosis of hydrocephalus due to following central nervous system pathologies w

Table 3: Frequency of CNS Pathologies

CNS Pathologies	Frequency	Percentage
Space occupying lesions	30	25
CVA Ischemia	7	5.8
Cerebral Atrophy	6	5
Tuberculous Meningitis	14	11.6
Glioblastoma Multiforme	11	9.1
Bacterial Meningitis	4	3.3
Cerebral Aqueductal Stenosis	6	5
Chiari Malformation II	8	6.6
Subarachnoid Hemorrhage	10	8.3
Meningioma	3	2.5
AVM	1	0.83
Other rare pathologies	10	8.3

4. Discussion

Hydrocephalus is a progressive extension of ventricular structure of the brain that is mostly occurs idiopathically or due causative agent which triggers the excessive production of CSF from the choroid plexus that reside within it and causes CSF retention within the intrinsic circulation. Oktay Algin conducted a research study which descriptively elaborates the clinical manifestation of patient with hydrocephalus, in his study he concluded that protuberant fontanelles, detached sutures and enlarged scalp veins were the prevalent clinical symptoms associated to stimulated brain distention seen in patients of hydrocephalus. Reduced papillary light retort, nystagmus, abducens nerve paresis and “sunset sign” were also seen in his study. Signs of raised intracranial pressure were also found to be frequently associated with focal neurological inadequacy and fits[14]. Our study is mostly based on collecting the frequency of the CNS pathology that mostly affect the intrinsic circulation of CSF and become the pathological factor of elevated intracranial pressure.. In our study we observed the similar variety of clinical manifestations associated with hydrocephalus. The most common attributably sign seen in patients of hydrocephalus was seizures (33.3%), dementia (16.6%), reduced vision (10%), gait disturbance (8.3%), urinary incontinence (8.3%), bulged fontanelles (8.3%), vomiting (8.3%) and lethargy in children (6.6%). Andreas M. Stark conducted a research study on tectal. In the study, the main clinical feature that was seen in the majority of patients at the time of diagnosis was raised intracranial pressure; seen in 10/12 patients (83%)[15]. The similar pattern was seen in our study which concluded that the frequency of hydrocephalic patients associated with gliomas were 11/120 (9.1%). In a study conducted by Juan F. Martinez –Lage he updated detailed article regarding an association of arachnoid cyst with hydrocephalus. According to his study the prevalence of hydrocephalus with arachnoid cyst ranged from 2% - 80% based on data collected from several reports[16][17]. In the study conducted by us the total number of patient diagnosed with hydrocephalus associated with arachnoid cyst was 1/120 (0.83%). In a study conducted by James M. Johnston a total of 695 patients of both genders were included. The conclusion drawn from this study was that hydrocephalus was the most common complication associated with SAH[18][19][20]. During our study we observed that the total patients of hydrocephalus associated with SAH were 10/120 (8.3%). Mei-Ling Sharon Tai conducted a research study in 2017 which included a total of 54 patients. She concluded in her research that frequency of the patients of hydrocephalus associated with meningitis were 38/54 (70.4%)[21].In the study

conducted by us we noted that the patients of hydrocephalus associated with meningitis were 4/120 (3.3%). Brian j Dlouhy conducted study which mainly emphasized on the correlation of neural tube defects such as myelomeningocele and chiari malformation type I and II with elevated intracranial pressure, in his study he elaborated a fact which states that CM type II is the most frequent central nervous system pathology that simply and severely associates with CSF flow abnormalities and plays a major role in the disrupted CSF flow mechanism, in his study he reported that 90% of cases who had CM type II had elongated ventricles along with displaced vermis and narrowed cerebral aqueduct had hydrocephalus. and CM TYPE I also showed connection with hydrocephalus, according to his study 10% of cases who had CM type I had the rare type of triventricular hydrocephalus which is easily appreciated on the MRI with a stenosed cerebral aqueduct mostly due to tectal mass known as tectal glioma which then leads to tonsillar herniation as well[22,23]. In a study conducted by Wende N Gibbs diagnostic spectra of hydrocephalus was emphasized. The author explained the stellar role of MRI in the diagnosis and clinical management of hydrocephalus. He also explained certain guidelines equipped by MRI to not only pinpoint hydrocephalus but also elaborate the associated pathologies which lead to the pathogenesis of hydrocephalus. The most common diagnostic signs were raised callosal angle along with ballooning of frontal and occipital horns of lateral ventricles[24,25].

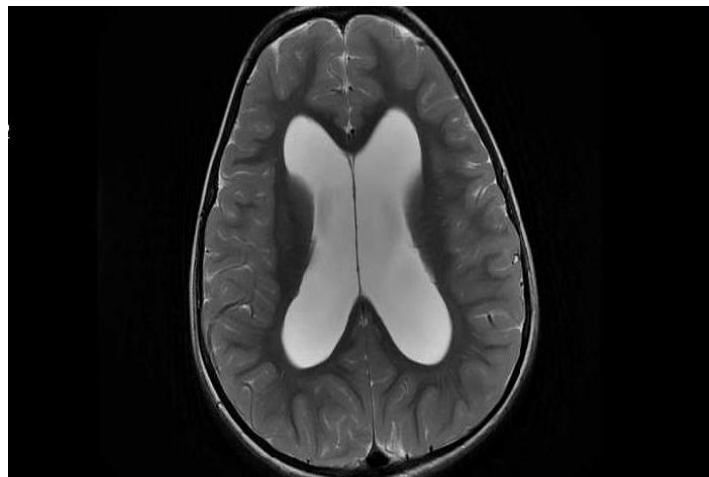


Figure 1: Hydrocephalus due to Bacterial Meningitis

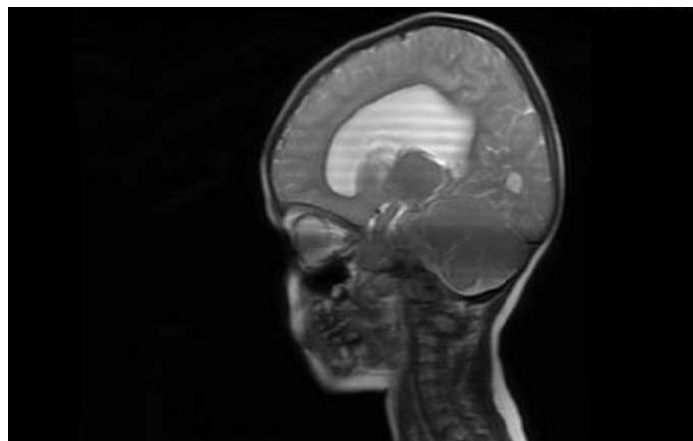


Figure 2: Hydrocephalus due to Chiari Malformation II

5. Conclusion

It was concluded that hydrocephalus was associated with almost 20 central nervous system pathologies which was predominantly found in males. The male to female ratio was 1.4:0.7 with mean age of 33.6 in males and the mean age of 36.62 in females. The central nervous system pathology which was found to be most frequently associated with hydrocephalus was space occupying lesion 25 % (30cases) with mean age of 38.8, seconded by TBM 11.6 % (14cases) with the mean age of 37.2; glioma 9.1% (11cases) with mean age of 42 and SAH 8.3% (10cases) with the mean age of 48.8. The statistics and figures provided by our research would help the neuroradiologists and neurosurgeons to devise different strategies to tackle with hydrocephalus due to different possible etiologies.

6. Recommendation

It is very important to first take careful physical examination of the patient and carefully correlate it with the clinical manifestations of the patient. And it is very important to have descriptive knowledge of the pathophysiology of any CNS pathology that results in hydrocephalus and you have to carefully observe the MRI characteristics of increased intracranial pressure and always calibrate the dilated ventricles through Evan index and callosal angle.

7. Author's contribution

Amna Babar and IqraAslam collected and analyzed the data .Sidra Azam, and Sana Waris designed the research plan. Aliza Gulzar Bukhari, Afra Mariam and Amna Babar write the research work and performed experimental works. All authors jointly prepared the manuscript, critically revised and finalized the manuscript.

8. Conflict of interest

The authors declared that there is no conflict of interest with present publication.

9. Financial Disclosure

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10. Ethics Committee Approval

This study was approved by scientific Researchers Ethical Committee of The University of Lahore

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