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**Supratentorial hydrocephalus secondary to vertebrobasilar dolichoectasia: A rare complication –
Case report and literature review**

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ABSTRACT

Introduction: Intracranial Arterial Dolichoectasia (IADE) is an uncommon cerebrovascular illness marked by elongation, dilatation, and tortuosity of cerebral arteries, most typically of the basilar and vertebral arteries. Its most common subtype is vertebrobasilar dolichoectasia (VBDE), for which diagnostic criteria have been established by imaging modalities, most commonly magnetic resonance angiography (MRA), which identifies features, such as a basilar artery (BA) diameter greater than 4.5mm. VBDE is often asymptomatic; however, can result in significant complications, including ischemic events, cranial nerve compression and obstructive hydrocephalus because of CSF pathway obstruction.

Case Report: We present the case of a 53-year-old South Indian female who arrived at the emergency department with altered sensorium and vomiting. VBDE was demonstrated on radiological investigation with a dilated basilar artery compressing third ventricle and aqueduct of Sylvius. According to Smoker's criteria, findings were scored 5, indicative of severe VBDE. CSF flow study confirmed supra-aqueductal stenosis and subsequent monoventriculoperitoneal shunt surgery led to complete resolution of symptoms.

Conclusion: This case is a major highlight of timely diagnosis and management of VBDE with obstructive hydrocephalus. Earlier management can be achieved by better imaging and clinical evaluation that will prevent worsening of neurological dysfunction and enhance patients' outcomes. This case underscores the further study of the etiology and treatment of VBDE.

Keywords: intracranial arterial dolichoectasia, vertebrobasilar dolichoectasia, hydrocephalus, basilar artery, smoker's criteria

Abbreviations:

CSF: Cerebrospinal Fluid

IADE: Intracranial Arterial Dolichoectasia

MRA: Magnetic Resonance Angiography

VBDE: Vertebrobasilar Dolichoectasia

INTRODUCTION

An important rare cerebrovascular disease characterized by elongation (*dolikhós*) and dilatation (*ektasis*) of intracranial arteries, usually of the basilar and vertebral arteries, is termed as Intracranial Arterial Dolichoectasia (IADE). This leads to dilation and tortuosity of the affected vessels causing significant structural changes [1]. It is an important risk factor for advanced age, hypertension, and higher prevalence in males; prevalence estimates in adult populations have ranged from 0.06% to 5.8%. Variations are produced by differences in diagnostic criteria, imaging techniques, or the demographic characteristics of studied populations [2,3]. The most common subtype of VBD is Vertebrobasilar Dolichoectasia (VBD), and structural changes in the basilar artery are visualized using magnetic resonance angiography (MRA). In order to diagnose VBD, the diameter must be greater than 4.5 mm, or the lateral deviation from the midline must be greater than 10 mm [4,5].

Clinically IADE can be asymptomatic, however, it can result in serious complications, which can be grouped as ischemic, hemorrhagic and mass effects. One of the main causes of death for people with IADE is ischemic stroke, which is the most prevalent and severe form of condition that affects brainstem as well as posterior cerebral artery region. In mass-effect cases, IADE may produce symptoms of brainstem and cranial nerve compression, such as trigeminal neuralgia, hemifacial spasm and motor deficits [1]. IADE related hydrocephalus is a rare but notable complication mainly due to vertebrae basilar arteries, which causes direct compression of cerebrospinal fluid pathways and shows the diversity and the potentially severe outcome of this condition [6,7].

Vertebrobasilar dolichoectasia (VBD) causing obstructive supratentorial hydrocephalus is a rare condition, and given the potential for severe clinical outcomes, authors present the case of a 53y/o female patient with obstructive hydrocephalus secondary to VBD, in order to contribute to the literature. It describes clinical and radiologic findings of compressive effects of VBD on cerebrospinal fluid pathways leading to hydrocephalus. The present case stresses the need for further research on the pathophysiology of VBD and a comprehensive diagnostic and management approach. Early intervention is necessary for the prevention of progressive neurological decline and the improvement of patient outcomes [8].

CASE PRESENTATION

A 53-year-old South Indian woman arrived at Saveetha Medical College's emergency room with a six-hour history of three vomiting episodes and impaired sensorium. In the preceding days, she had experienced intermittent confusion and disorientation, which had now progressed to persistent drowsiness. She also

reported a two-week history of chronic headaches, occasionally associated with blurred vision, light sensitivity, and urinary incontinence.

On examination, the patient was afebrile, normotensive, and maintained adequate oxygen saturation on room air. In addition to being confused about time, place, and people, she appeared sleepy, which affected her cooperation and attentiveness. Her estimated Glasgow Coma Scale (GCS) scores were M4 (withdrawal to pain), V2 (incomprehensible sounds), and E1* (eye response unassessable due to drowsiness). Due to limited cooperation, assessment of eye movements and bedside visual acuity was challenging. There was no relative afferent pupillary deficit (RAPD) and both pupils were receptive to light, indicating normal pupillary responses. Upon fundoscopic examination, there were no indications of papilledema and the disc margins were normal with a dull macula.

A history of gait instability before this episode of altered sensorium, brisk reflexes in the lower limbs, and mild weakness were noted on neurological examination. Examinations of cardiovascular and respiratory systems were unremarkable. Based on these findings a provisional diagnosis of encephalopathy was made, and then neuroimaging, a metabolic panel, and a large number of comprehensive blood investigations were performed to look for underlying causes of her altered mental status. Routine blood investigations were normal, and therefore, all would be consistent with a metabolic or systemic cause for the patient's symptoms. A CT scan of the brain then showed diffuse cerebral edema, which appeared as effacement of adjacent sulcal spaces on neuroimaging. In addition, ventriculomegaly, with marked ventricular enlargement of both lateral ventricle, foramen of Monro, and third ventricle, was seen on the scan. In addition, periventricular hypodensities suggestive of CSF seepage were observed (Figure 1). Vertebrobasilar dolichoectasia (VBDE) was suggested by the dilated and tortuous BA compressing left hemipons as well as floor of third ventricle (Figure 2).

Subsequent to these initial CT findings, a CEMR of the brain was performed to better evaluate the patient's condition. The MRI was consistent with dolichoectasia of the vertebrobasilar system, with the basilar artery approximately 5.7 mm in diameter. This enlarged artery, which is situated lateral to the clivus and dorsum sellae and extends to bifurcate at the level of third ventricle's floor, was seen to compress the anterolateral aspect of left hemipons (Figures 3 and 4). These imaging findings demonstrate the extent of vascular anomalies responsible for the patient's clinical symptoms and provide support for a diagnosis of VBDE with obstructive effect on surrounding brain structures.

The severity of vertebrobasilar dolichoectasia was evaluated by smoker's criteria. On this scale, one point was given to a BA diameter >4.5mm. Also, the artery was displaced laterally from the clivus by an average of 2 points beyond the lateral border. Additionally, this bifurcation height raised the third ventricle's floor by two points. Five of the smoker's criteria point to a high risk of vertebrobasilar dolichoectasia. The extent of this suggests that the vasculature is abnormally encroaching on the brainstem and other tissues and that this is likely to be causing the patient's clinical signs. This evidence corroborates the need for a specific strategy to address the structural and clinical features of this disorder.

Further evaluation of cerebrospinal fluid (CSF) flow study was unremarkable for CSF flow beyond the aqueduct of Sylvius, a characteristic of supra aqueductal stenosis. This finding is consistent with supra aqueductal stenosis (FIGURE 5). This means that the dilated basilar artery is compressing on the floor of

third ventricle, preventing CSF flow (Figure 5) for supra aqueductal stenosis. The development of supratentorial hydrocephalus and patient's neurological symptoms are due to this obstruction. The findings further demonstrate that her observed hydrocephalus and the development of her clinical presentation are mostly caused by the structural impact of dolichoectatic BA on the third ventricle's floor.

Additional MRI findings included vertical tortuosity of the bilateral optic nerves and partially empty sella, all of which are suggestive of intracranial hypertension (Figure 6). In addition, these anatomical changes support the diagnosis of elevated intracranial pressure probably resulting from the obstructive effects of the dilated basilar artery on CSF flow at the aqueduct of Sylvius. These findings also include supratentorial hydrocephalus and features of intracranial hypertension, which further emphasize the effects of vertebrobasilar dolichoectasia on a patient's neurological status and emphasize a comprehensive management strategy.

Obstruction was localized at floor of third ventricle (Figure 7), and the decision was made to proceed with mono-ventriculoperitoneal shunt surgery. The procedure was uneventful, and the postoperative course was uneventful. After one month, the patient fully recovered and had significant improvement. One month after surgery, a follow up CT scan showed relieved hydrocephalus (Figure 8). These results confirm the efficacy of shunting targeted to the site of obstructive hydrocephalus caused by vertebrobasilar dolichoectasia.

DISCUSSION

Vertebrobasilar dolichoectasia, or dolichoectasia of vertebrobasilar artery (DVA), is a rare vascular anomaly characterized by basilar artery elongation, dilation, and tortuosity. VBDE occurs principally in older individuals, with a mean age of presentation of about 60.5 years, and its prevalence has ranged from 0.06% to 5.8% [6,7]. Often asymptomatic, VBDE can result in severe complications like ischemic strokes, brainstem compression, cranial nerve palsies, as well as hydrocephalus, less commonly hydrocephalus [8, 9]. The literature regarding VBDE related hydrocephalus is limited, and this case further illustrates the diagnostic and management complexity of this condition.

Diagnostic criteria and imaging

The diagnosis of VBDE is primarily based on imaging studies; Smoker's criteria are key to assessing VBDE severity. This criterion is used to evaluate basilar artery diameters larger than 4.5mm and structural deviations (lateral displacement, or high bifurcation level) [10]. Prolonged hypertension and possibly congenital factors are often blamed for the structural degeneration often seen in dolichoectatic arteries, including thinning of the arterial wall, loss of elastic fiber, and smooth muscle atrophy [11]. Also similar to these, Siddiqui et al. [6] and Kansal et al. [7] documented the use of MRI and MRA to describe the VBDE's effect on midbrain and cerebral aqueduct, leading to obstructive hydrocephalus. These two cases highlight how imaging plays an essential role in understanding the effect of VBDE on neighboring structures.

Complications and mechanisms of hydrocephalus

The hydrocephalus observed in VBDE is typically obstructive, resulting from direct compression of the cerebral aqueduct or Monro foramina, which stops the CSF from flowing. Two types of VBDE-induced

hydrocephalus are identified: obstruction: visible and obstruction: invisible. The latter is associated with a secondary "water hammer" effect due to pulsatile forces of the ectatic artery on CSF dynamics, comparable to normal pressure hydrocephalus (NPH) [12]. Zisimopoulou et al. [9] and Ebrahimzadeh et al. [10] report VBDE induced aqueductal compression producing obstructive hydrocephalus, and emphasize the necessity of prompt intervention to relieve intracranial pressure and control symptoms.

Management and outcomes

Treatment of VBDE related hydrocephalus is by ventriculoperitoneal (VP) shunting, with good outcome in most cases. Surgical planning, especially shunt placement, requires preoperative imaging, such as Smoker's criteria and CSF flow studies [13]. In some patients with foramina of Monro compression, biventricular shunting is indicated because the obstruction is bilateral, as in the case of Weber et al. [14]. However, patients with "obstruction invisible" hydrocephalus caused by the 'water hammer' effect have variable responses to shunting, and therefore thorough CSF flow dynamics assessment is needed to inform treatment strategies [7].

In this case, placement of a monoventriculoperitoneal shunt resulted in rapid symptom resolution and normalization of CSF flow, as has been observed with favorable outcomes by Kansal et al. [7] and Celik et al. [8]. VBDE anatomically presents challenges due to third ventricular compression, especially in need of individualized surgical strategies. According to the studies of Thiex and Mull [15] and Lee et al. [16], cases with acute and progressive symptoms should be immediately intervened to avoid further neurological decline.

Prognosis and clinical implications

The prognosis of VBDE-induced hydrocephalus is highly variable based on a review of available case reports (Table 1), and appears to depend upon the degree of CSF obstruction, coexisting vascular conditions and the timing of intervention. Symptomatic relief after shunting is the rule, but outcomes are less favorable when complications such as ischemic events or NPH are present. Early diagnosis and treatment are important because advancements in imaging and surgical techniques have decreased the risk of long-term neurological damage [17].

CONCLUSION

Vertebrobasilar dolichoectasia (VBDE) is a rare but important cause of obstructive hydrocephalus and this case serves to highlight the marked neurological symptoms caused by it. MRI and CSF flow studies proved critical in identifying the cause and differentiating VBDE from similar conditions, including normal pressure hydrocephalus (NPH). Smoker's criteria were applied to the application of severity of VBDE; cerebrospinal fluid studies confirmed the existence of supra aqueductal stenosis due to basilar artery compression. Targeted management alleviated symptoms and allowed for full neurological recovery by prompt surgical intervention, namely monoventriculoperitoneal shunt placement. The variable presentation and complex pathophysiology of VBDE emphasize the need for a complete diagnostic and therapeutic approach. The importance of early recognition, extensive imaging, and individualized treatment strategies

to prevent progressive neurological deterioration and improve clinical outcomes in patients with VBDE associated obstructive hydrocephalus is highlighted in this case.

Patient consent

Written informed consent was obtained from the patient (or their legal guardian) for the publication of this case report and any accompanying images. All identifying information has been removed or anonymized to protect patient privacy

Conflict of interest

There are no conflicts of interest

Author's contributions

- **Aadithiyan Sekar:** Conceptualized the case report, conducted clinical assessment, and drafted the initial manuscript.
- **Rashi Seetha:** Interpreted diagnostic images, contributed to the radiology section, and reviewed for accuracy.
- **Abdul Majith:** Assisted in patient management, contributed to the discussion, and reviewed the manuscript for clinical relevance.

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FIGURES AND TABLES

TABLE

TABLE 1. Summarizes VBDE-induced hydrocephalus cases, detailing clinical presentations, compression levels, treatments, and patient outcomes across studies

Authors	Age/Sex	Journal	Clinical Presentation	Level of Compression	Treatment	Outcome
¹ Siddiqui et al ^[6]	71, F	British Journal of Radiology	Gait disturbances, Headache, Urinary incontinence, Vomiting, Left sixth nerve palsy	Midbrain – Cerebral aqueduct – third ventricular outflow	Monoventriculoperitoneal shunting	Good recovery
Kansal et al ^[7]	60, M	Journal of Neurosciences in Rural Practice	Urinary incontinence, Gait disturbances, Headache, Papilloedema	Midbrain – Cerebral aqueduct – third ventricular outflow	Monoventriculoperitoneal shunting	Good recovery
Celik et al ^[8]	47, M	Journal of Neurological Surgery, Part A	Headache, Confusion, Vomiting, Right-sided face as well as hand numbness	Midbrain – Cerebral aqueduct – third ventricular outflow	Monoventriculoperitoneal shunting	Good recovery
Zisimopoulou et al ^[9]	48, M	Clinics and Practice	Urinary incontinence, Disinhibition	Midbrain – Cerebral aqueduct – third ventricular outflow	Monoventriculoperitoneal shunting	Good recovery
Ebrahimzadeh et al ^[10]	68, M	Surgical Neurology International	Headache, Gait Disturbance, Urinary Memory Loss, Incontinence, Visual Loss, Papilloedema	Midbrain – Cerebral aqueduct – third ventricular outflow	Monoventriculoperitoneal shunting	Good recovery
Zdravkovic et al ^[11]	85, M	American Journal of Forensic Medicine	Sudden onset coma	Concurrent midbrain and pons compression	Did not survive	Did not survive



¹ Mohammed et al ^[12]	51, F	Surgical Neurology International	Vomiting, Left facial palsy, Acute confusion	Midbrain – Cerebral aqueduct – third ventricular outflow	Monoventriculoperitoneal shunting	Good recovery
Oishi et al ^[13]	57, M	Journal of Stroke and Cerebrovascular Diseases	Urinary Incontinence, Headache, Gait Disturbance	Midbrain – Cerebral aqueduct – third ventricular outflow	Monoventriculoperitoneal shunting	Good recovery
Weber et al ^[14]	Not mentioned	Der Radiologe	Acute consciousness disturbance	Foramina of Monro compression	Biventriculoperitoneal shunting	Good recovery
Thiex and Mull ^[15]	Not mentioned	Surgical Neurology	Basilar megadolicho trunk causing obstructive hydrocephalus	Foramina of Monro	Ventriculoatrial shunting	Good recovery
Lee et al ^[16]	52, M	Medicine (Baltimore)	Gait disturbance, Left-side homonymous hemianopsia, Memory impairment, Headache, Visual field impairment,	¹ Midbrain – Cerebral aqueduct – third ventricular outflow	Monoventriculoperitoneal shunting	Good recovery
Umana et al ^[17]	Not mentioned	Journal of Neurological Surgery Part A	Hypoplastic third ventricle, Biventricular hydrocephalus, Vertebrobasilar dolichoectasia	Third ventricular outflow	Biventriculoperitoneal shunting	Not specified

FIGURES



FIGURE 1. (A and B): Non-enhanced computed tomography (CT) axial images of the brain demonstrate dilatation of the bilateral lateral ventricles, foramen of Munro, and third ventricle, accompanied by hypodense areas in the periventricular region (white arrow), suggestive of periventricular seepage. Diffuse cerebral edema is also noted, indicated by the effacement of adjacent sulcal spaces.

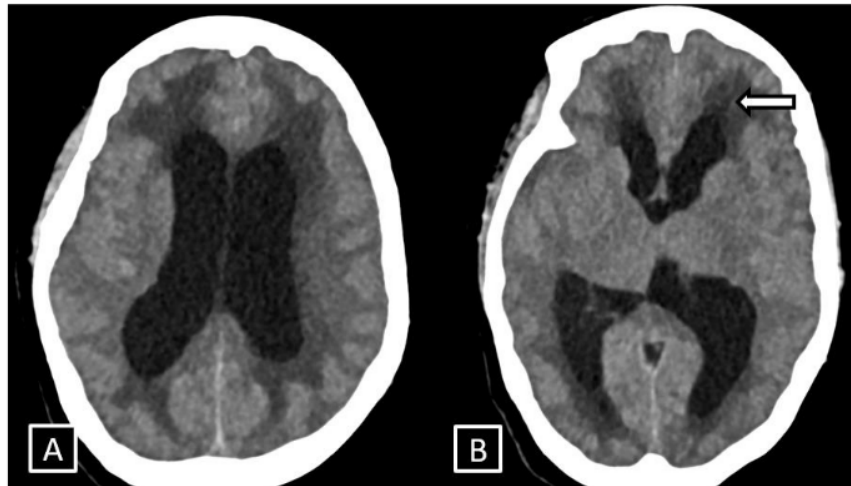


FIGURE 2. Axial (A and B) and sagittal (C) non-enhanced computed tomography (NECT) images of brain. These images reveal a dilated, tortuous basilar artery compressing the left hemipons (white arrow head) and the floor of third ventricle (white arrow)

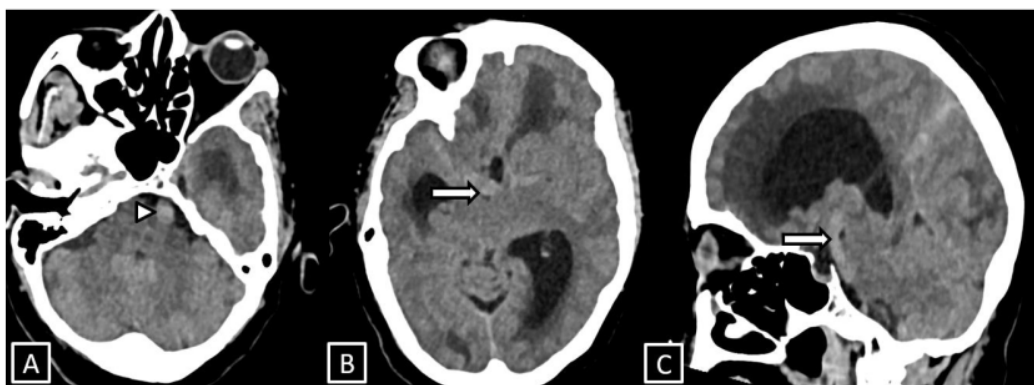




FIGURE 3. (A and B): Magnetic Resonance Imaging (MRI) T2 weighted and Magnetic Resonance Angiography (MRA) axial images of the brain shows dilated tortuous basilar artery measuring ~ 5.7mm

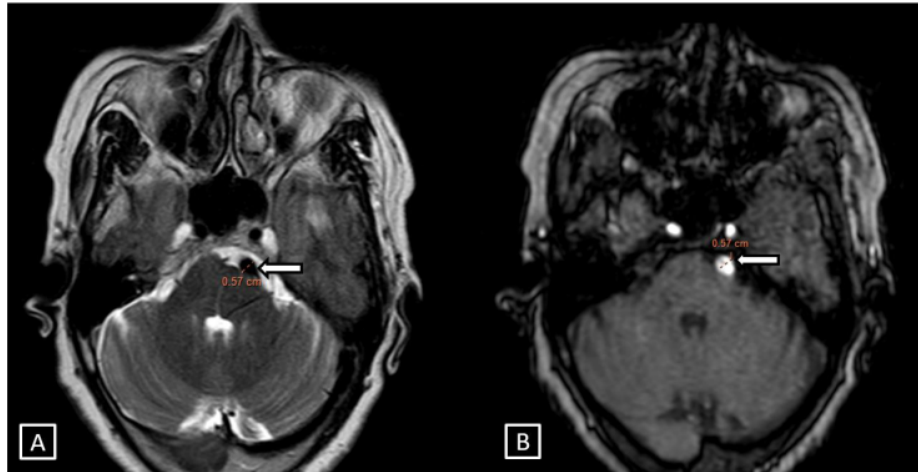


FIGURE 4. (A, B, and C): Contrast Enhanced Magnetic Resonance (CEMR) images of brain. (A and B) Sagittal and axial views show a dilated, tortuous basilar artery compressing the floor of third ventricle (white arrows). (C) Coronal CEMR image reveals the basilar artery coursing laterally to the margin of the clivus (white arrowhead) and bifurcating at a high level at the floor of third ventricle (white arrow)

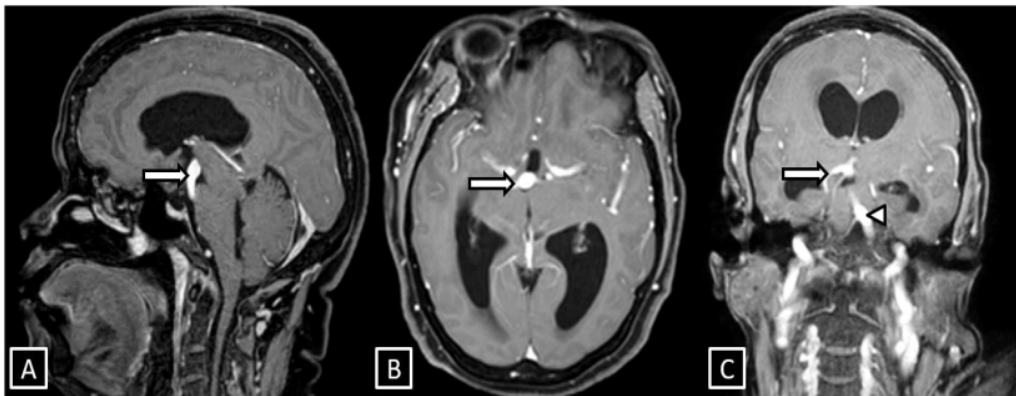




FIGURE 5. (A, B, and C): Magnetic Resonance CSF flow study images—rephased, magnitude, and phase views—demonstrate an absence of CSF flow beyond the site of stenosis to the posterior fossa (white arrows), indicating significant obstruction at this level

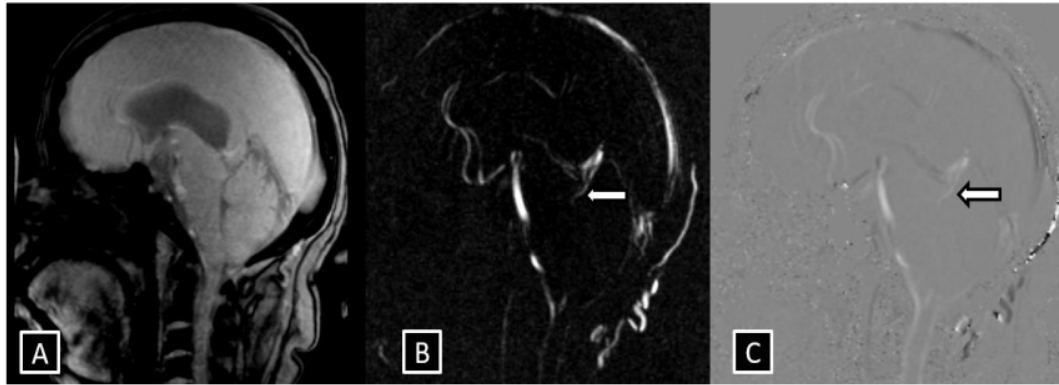


FIGURE 6. (A, B, and C): Magnetic Resonance T2-weighted axial images (A and B) show a prominent, tortuous vertical course of the bilateral optic nerves with an enlarged subarachnoid space (white arrows). (C) Magnetic Resonance T1-weighted sagittal image reveals a partially empty sella (white arrowhead), suggesting features indicative of intracranial hypertension.

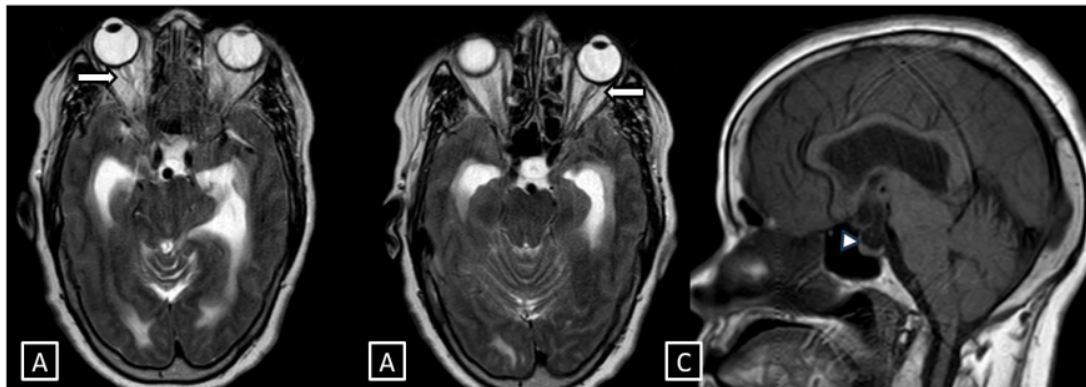




FIGURE 7. (A and B): Non-enhanced computed tomography (CT) axial images of the brain demonstrate the status post-monoventriculoperitoneal shunting. The intraventricular drain tube courses along the right parietal lobe, with its tip positioned within the frontal horn of the left lateral ventricle.

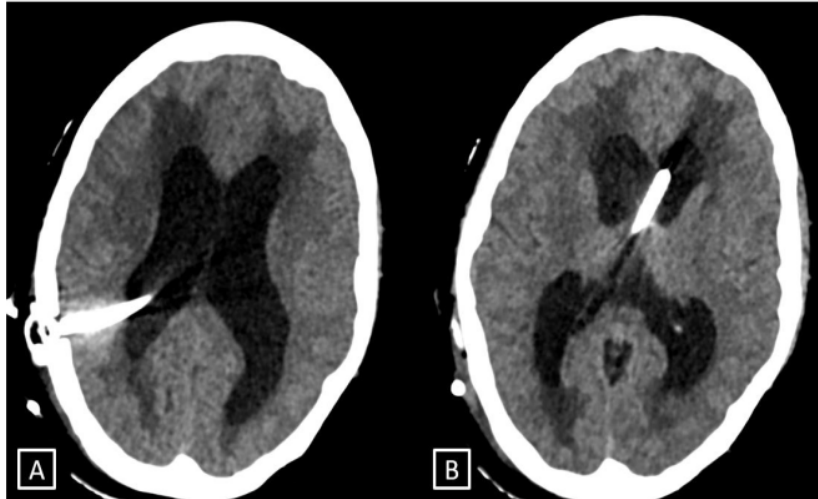


FIGURE 8. (A and B): Non-enhanced computed tomography (CT) axial images of the brain, taken one month postoperatively, show a normal-appearing ventricular system with the intraventricular drain tube in situ, suggestive of relieved hydrocephalus following monoventriculoperitoneal shunting.

