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Case Report

Cerebrospinal fluid hydrodynamics in arachnoid cyst patients with persistent idiopathic intracranial hypertension: A case series and review

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ABSTRACT

Background: A clear connection has been established between arachnoid cysts (ACs) and the evolution of idiopathic intracranial hypertension (IIH), a connection, which is presently not well understood. Cerebrospinal fluid (CSF) is an integral element of this condition. Little is known about either the influence of AC on CSF hydrodynamics or the specific nature of CSF, which contributes to the complex pathology of IIH.

Case Description: This study aimed to chronicle in detail four patients with previously treated intracranial ACs, who developed persistent IIH. This series and review aims to identify and qualitatively analyze the multiple constituents, which could possibly elucidate the intrinsic relationship between arachnoid cyst-induced IIH and CSF hydrodynamics. A retrospective analysis of the medical records of four patients admitted to the institution's neurosurgery department during the period of 1994–2013 was completed. This study investigated discernible aspects linking CSF pathophysiology with the development of IIH in AC patients. Four male patients, ranging from 3 to 44 years of age at presentation, had a left-sided arachnoid cyst treated surgically. All four patients subsequently developed IIH. Three patients remain persistently symptomatic.

Conclusion: IIH associated with AC is a hydrodynamic disorder. The full discovery of its fluctuant pathophysiology is the only way to identify an effective standard for the management and treatment of this condition.

Keywords: Arachnoid cyst, Cerebrospinal fluid, Hydrodynamic, Idiopathic intracranial hypertension, Intracranial pressure

BACKGROUND

Idiopathic intracranial hypertension (IIH) is defined as both the clinical and neurological manifestation of increased intracranial pressure (ICP) in the absence of any secondary cause.[17] These include trauma, space occupying lesion, enlarged ventricles, or meningeal inflammation or penetration. IIH remains a diagnosis of exclusion, the title given to a condition of poorly understood etiology.[38] It has a heterogenic phenotype; however, typical patient presentation includes headache, transient visual obscuration, pulsatile tinnitus, blurred vision, diplopia, photophobia,[42] and papilledema on fundoscopy.[5,20,43]

The annual incidence of IIH is 1–2/100,000.[15,33] Classically, obese women of childbearing age[11,26,42] are identified as the high-risk population[6,19,21,22,25,33,42] in cases of IIH. This disorder rarely occurs

in men and children.[11,42] Various medications and exogenous substances have been associated with IIH such as growth hormone,[41] tetracyclines,[17] and hypervitaminosis A.[14,18]

Arachnoid cysts (ACs) are congenital collections of benign fluid contained within the arachnoid membrane, lined by arachnoid cells, and situated in the subarachnoid space of the cisterns and major cerebral fissures.[24,32] It has been suggested that ACs arise from disturbance of cerebrospinal fluid (CSF) hydrodynamics in the early phase of subarachnoid space formation due to leptomeningeal maldevelopment during the embryologic period.[1,34,35] These cysts constitute approximately 1% of all intracranial mass lesions.[1] The incidence of asymptomatic AC is increasing as more patients undergo neuroimaging procedures for unrelated symptoms.[27,30] A prevalence of 1.4% has previously been identified in a retrospective review of over 48,000 consecutive adult brain magnetic resonance imaging (MRI); of these, only 5% were symptomatic. About 75% of the symptomatic AC occurred in children.[2]

CSF physiology is based on the concept of maintaining hydrodynamic order. Hydrodynamic order, in turn, entails preserving a balance between CSF production, circulation, and reabsorption as well as compensating for deviations by appropriately readjusting the unaffected parameters so as to restore equilibrium between formation and absorption.[23] Contributions by Dandy, Cushing,[8,10] and Weed[44,45] have hugely increased the neurosurgical community understanding of the multitudinous aspects of CSF flow both from an anatomical, physiological, functional, and clinical perspective. The accepted principles of CSF state that the fluid is formed in the choroid plexus situated in the frontal and occipital horns of the lateral ventricles, circulates through the ventricular system, and thereafter enters the arachnoid space where it is reabsorbed by the arachnoid villi (AV). Despite these crucial developments, there are still deficits in our knowledge and uncertainties in relation to the definitive physiology and pathology of CSF reabsorption.

Although IIH by definition is idiopathic, research has postulated a link between IIH and multiple disorders. In the literature, there are various cases documenting this ambiguous connection.[27,30] This association between the development of IIH after surgical treatment of AC is extremely rare, and presently, there is no national or international incidence rate, which accurately measures this obscure relationship. Furthermore, the pathophysiological mechanism, which causes this condition, is unknown. Since the etiology is uncertain, there is no efficacious therapy or global consensus on optimal medical management for treating patients suffering with this disorder.

CSF disturbance has been identified as an important constituent in the development of IIH. It has also been proposed as the sole cause of the disorder and multiple hypotheses have postulated the etymology.[13,32] At present, the accepted principle is that there is hydrodynamic disequilibrium between the production and absorption of fluid.[23] A clear link has yet to be determined between CSF hydrodynamics and the onset of IIH in patients post-AC,[23] but the investigation of such a hypothesis is relevant and noteworthy.

This retrospective case review series describes four patients admitted during the period of 1994–2013 suffering with IIH in the setting of AC. This study investigated and analyzed the clinical aspects linking CSF pathophysiology with the development of IIH in patients who had AC treated surgically.

CASE REPORT

Case report 1

A 34-year-old man presented with headache of gradual onset, which continued intermittently over the subsequent fortnight. The pain was most severe over the left frontal region radiating down the left side of his face, associated with dizziness. Computed tomography (CT) brain identified a superficial AC over the surface of the left cerebellar hemisphere with no mass effect or evidence of hydrocephalus. The patient underwent a burr hole craniotomy, cyst drainage, and excision of a portion of the cyst wall. CSF pressure in the cyst upon measurement was 25 cm H_2O.

The patient's symptoms had not resolved 3 months postoperatively and a follow-up CT revealed reoccurrence of the AC. A posterior fossa craniotomy cyst drainage and more extensive excision of the cyst wall were undertaken. CSF pressure postoperatively was persistently 25 cm H_2O.

The patient did not recover well from the second surgery and presented 6 months later with low-grade meningitis, worsening headache, fever, nausea, and vomiting. Lumbar puncture (LP) was performed and revealed an elevated white blood cell count with no organisms, for which he was treated with a course of antibiotics. The patient's severe headaches persisted.

LP was performed 3 months later, which found a CSF pressure of 29 cm H_2O. MRI and magnetic resonance venography (MRV) demonstrated a collapsed cyst with no evidence of venous sinus thrombosis (VST) or any other changes suggestive of increased ICP. Papilledema was not present. A diagnosis of IIH was established and a lumboperitoneal shunt was inserted. The patient's symptoms improved, and he was subsequently prescribed amitriptyline 25 mg at night, to alleviate any periodic headache.

The patient presented 5 years later with severe occipital headache, dizziness, and vertigo. LP showed a CSF pressure of 32 cm H_2O. MRI and MRV demonstrated no reoccurrence of the superficial AC with suspected narrowing of the posterior third of the sagittal sinus. The lumboperitoneal shunt was replaced with a medium pressure valve cystoperitoneal shunt.

Three years later, the patient presented with dizziness following two episodes of severe vertigo accompanied by nausea and vomiting. A further MRI revealed no abnormality apart from the previously identified collapsed AC. On shunt flushing, a CSF pressure of 2 cm H_2O was noted. Low-pressure symptomatology was identified. A high-pressure valve was inserted. This further aggravated the patient's symptoms. CSF pressure was 15 cm H_2O on measurement and further valve pressure downgrading was achieved by insertion of a variable pressure shunt set at 7 cm H_2O replacing the high-pressure shunt. This patient remains persistently symptomatic.

Case report 2

A left-sided temporal AC [Figure 1] was an incidental finding on CT scan in an infant male undergoing investigation for the cause of his cleft lip and palate. There was no mass effect or evidence of hydrocephalus.

This cyst remained benign until the age of 3, on which the patient presented with a 3-week history of progressively worsening headache, vomiting, lethargy, and irritability. No papilledema was noted. A cystoperitoneal shunt was inserted. A postoperative CT revealed a collapsed cyst and the patient experienced symptomatic relief.

At 7 years of age, the patient presented with episodic severe headache and vomiting and no other signs or symptoms. CT scan revealed no evidence of cystic fluid collection and the ventricles were normal in size, shape, and position. The patient was treated symptomatically, and his complaints resolved.

The patient presented at 11 years of age to the emergency department with diplopia, photophobia, headache, and vomiting. Bilateral abducens nerve palsy was identified with bilateral papilledema noted on fundoscopy. CT scan revealed slit ventricles and no residual temporal AC. A cystoperitoneal
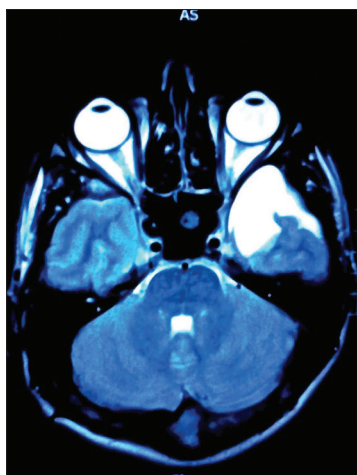

Figure 1: Computed tomography scan of Patient 2 showing a left temporal arachnoid cysts.

shunt was *in situ* and no malfunction was identified. MRI scan showed a Chiari malformation. There were no associated syrinx or tussive headaches in the occipital region on coughing or straining.

Intracranial hypertension without dilated ventricles was diagnosed. The cystoperitoneal shunt was revised, but when the patient showed no improvement, a right frontal ventriculoperitoneal shunt was inserted which appeared to relieve the patient's symptoms. The patient presented with two further symptomatic episodes due to shunt malfunction. A new lumboperitoneal shunt was inserted, and the symptoms resolved, although multiple shunt revisions were necessary.

A year later, the patient presented with a short history of recurrent headache, for which the lumboperitoneal shunt was revised. CT venogram showed patent venous sinuses with normal deep venous system and no evident stenosis. The diagnosis of IIH was reaffirmed.

At 17 years of age, the patient underwent foramen magnum decompression for a Chiari malformation and presented 3 weeks later with severe headaches. LP revealed a CSF pressure of 48 cm H_2O and slit-ventricle syndrome. A right-sided ventriculoperitoneal catheter was inserted, and his symptoms improved. To date, this patient has undergone eight shunt revision procedures.

Case report 3

An 8-month-old male presented with a short history of absence seizures. A large left frontotemporal AC was identified on CT scan with no mass effect or evidence of hydrocephalus. A cystoperitoneal shunt was inserted. This resulted in complete resolution of the cyst.

Over the following 10 years, the patient presented with migraine-like attacks with no contributing evidence on CT scan and no residual cyst formation. He was treated with intranasal sumatriptan 20 mg prn to be administered on the onset of headaches.

At 13 years of age, the patient was referred after a persistent headache of 1 week's duration and three episodes of vomiting. No papilledema was noted on fundoscopy. LP revealed CSF pressure of 17 cm H_2O. The patient continued to suffer with episodic severe headache and seizures. The patient went on to develop blurred vision with normal visual acuity and bilateral papilledema. CT scan revealed slit ventricles and no evidence of cyst reoccurrence. ICP monitor recorded consistently high pressures ranging from 20 cm to 60 cm H_2O. The patient was diagnosed with IIH and a ventriculoperitoneal shunt was inserted.

The patient experienced reoccurrence of headaches 8 months later. LP showed a CSF pressure of 25 cm H_2O. A cephalic

block was identified, and a new ventriculoperitoneal catheter was inserted along the track of its predecessor. The patient's slit-ventricle syndrome persisted.

The patient became symptomatic 5 months later. LP showed a pressure of 25 cm H_2O. A lumboperitoneal shunt was inserted. The patient subsequently developed low-pressure symptoms – headaches worse during the day instantly relieved by lying down. A high-pressure valve replaced the low-medium pressure Hakim valve. The patient was well for a week, until sudden onset of worsening severe headaches. LP showed CSF pressure of 14 cm H_2O, a pressure apparently too high for the patient to tolerate. A medium-high pressure Hakim valve replaced the high-pressure valve.

The patient continued to experience persistent, if not milder episodic headaches. MRI identified a Chiari malformation with cerebellar tonsillar protrusion approximately 1 cm below the level of the foramen magnum [Figure 2]. There was no associated syrinx.

At 16 years of age, the patient had three complex partial seizures. An electroencephalogram showed excess intermittent rhythmical slow-wave activity over the left hemisphere. This implied the potential for partial and generalized seizures, findings consistent with an underlying structural abnormality in the left temporal region, the area of the collapsed AC. The patient was treated with Keppra, 750 mg qAM and 1000 mg qPM and Lamictal, 50 mg twice daily.

The patient remained reasonably well for 2 years. He once more experienced similar episodes of headache, vomiting, and tonic seizures. ICP monitor identified markedly raised pressure of a minimum of 30 cm H_2O. The lumboperitoneal shunt was removed and a new cystoperitoneal shunt was inserted.
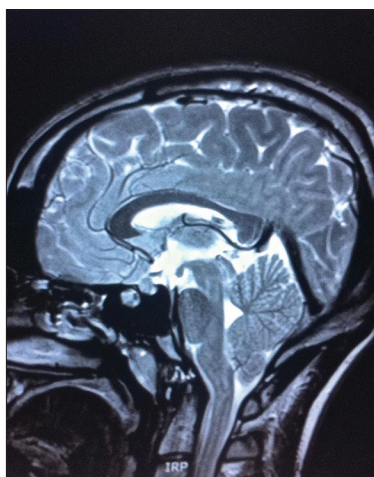

Figure 2: Magnetic resonance imaging of Patient 3 depicting cerebellar tonsillar herniation.

Case report 4

A 4-year-old male presented with intermittent severe headache and irritability. On physical examination, a substantial left-sided temporal bulge was found. CT scan identified a large left temporal AC with no mass effect or evidence of hydrocephalus.

The patient's headaches continued to worsen, and a craniotomy was undertaken with excision of the cyst wall. There was moderate symptomatic improvement for 1 month. A repeat CT scan showed that the cyst had decreased in size by 50%. A trial of acetazolamide was prescribed, which produced little symptomatic improvement. LP revealed a CSF pressure of 28 cm H_2O. A ventriculoperitoneal shunt was inserted. The patient is currently well.

DISCUSSION

There are conflicting views as to the composition of AC fluid. It was thought that arachnoid cells lining the cyst secrete CSF, but recent chemical analysis has revealed that AC fluid is not identical to CSF.[4] Although the osmolarity is the same, there is a difference in the concentrations of certain compounds and ions.

This compositional difference between AC fluid and CSF is relevant as it presents an alternative mechanism of cystic fluid accumulation. This evidence supports the concept of active transport as a mechanism underlying AC filling.[4]

It must be questioned whether this mechanism modifies the composition and substrate concentration of CSF. In turn, such changes in CSF could modify the rate or amount of fluid absorbed by the AV, thus causing an elevation in ICP, a possible causal agent or compounding factor leading to, or augmenting IIH. Although there is no demonstration of this concept in the above cases, it is an idea worth discussing in this review.

Factors that influence the production of CSF can be classified as exogenous or endogenous. Neural influences affecting fluid production include sympathetic, cholinergic, and peptidergic fibers.[31] From a treatment perspective, a prime example of utilization of this information is pharmacological therapeutic agents such as acetazolamide to decrease CSF production[36] so as to decrease ICP and ameliorate the symptoms of IIH. The issue with such a treatment is the lack of therapeutic effect,[30] as illustrated in Patient 4.

Approximately 15–20 ml of CSF is produced every hour. An increase in CSF volume can be due to increased production but can also be associated with hydrostatic, hydrodynamic, and obstructive mechanisms as a possible reason for elevated CSF pressure.[30] Consequently, quantifying actual circulating CSF volumes is essential in differentiating the origin of an equilibrium disorder.

Assessing volume is complex and difficult.[16,37] General initial evaluation presently depends on radiological and LP measurements. In addition, a normal CSF value does not necessarily rule out an underlying disorder.[16,37] This is true in IIH where the condition arises in the presence of both normal and elevated CSF pressure.[39] This was particularly evident in Patients 1 and 3 who presented symptomatically, but both had ICP readings within normal limits.

Although an established entity in human adults, AV are not found in the human infant.[12,28] This information is extremely relevant. ACs develop during the embryological period of fetal maturation and can arguably present before and during the development of the AV. It is logical to question the influence of these cysts in the formation of the AV on the arachnoid membrane, as well as the physiological change which occurs in the subarachnoid space when surgical intervention induces cyst collapse or removal as seen in Patients 1, 2, 3, and 4.

If the AV exist in an environment, which include cyst-like masses and adjust accordingly during development, what effect does obliteration of the cyst have on the villi? The presence of an AC can be considered a structural abnormality and a potential obstruction, modifying fluid circulation, and absorption. As discussed in relation to AV and absorption, the embryonic origin of such cysts undoubtedly alters orthodox CSF anatomy and possibly its physiology.

Patients 1, 2, 3, and 4 all underwent surgical intervention to treat AC intending to alleviate symptoms [Table 1]. In addition, all four patients were subsequently managed with CSF diversion at the reoccurrence of symptoms. Removal of the established structure may incur the onset or exacerbation of CSF disequilibrium. This idea can also apply to the alteration of circulation during shunting. Such modifications can affect the flow of CSF as well as compartmental distribution and CSF perfusion pressure.

VST is also recognized as a possible cause, consequence, or compounding factor in the development of IIH. Patient 1 exhibited narrowing of the posterior third of the sagittal sinus on MRV, which was detected 6 years after initial symptomatic presentation. Including and identifying this agent are noteworthy, but it does not contribute to the present discussion, as there is no known association between AC and venous sinus dysfunction.

CSF pressure values and assessment formulate our present concept of CSF equilibrium, the balance between the production and absorption of CSF. The most recent revision of the Dandy criteria diagnoses IIH in the presence of signs and symptoms of elevated ICP with a CSF opening pressure of >25 cm H_2O, normal CSF constituents, and no alternative explanation of increased ICP.[17] Radiological imaging should be unremarkable. Recent studies have attempted to outline the range of normal CSF pressure (10–25 cm H_2O), in the aim of stratifying the diagnosis of IIH to individuals above this range. However, a normal opening pressure of up to 28 cm H_2O was identified in certain patients.[46]

Table 1: Patient clinical characteristics after surgical removal of AC.

Pt No.	Presenting age	AC location	ICP (method)	Neuroimaging identifying other intracranial pathology	Shunt Hx	Additional treatment	Current health status
1	34 years (1994)	L cerebellar hemisphere	25, 29, 32, 2, 15 cm H_2O (LP opening pressure)	A collapsed cyst with suspected narrowing of the posterior third of the sagittal sinus.	Shunt revisions = 8 Lumboperitoneal, Cystoperitoneal medium pressure valve	Amitriptyline 25 mg nocte	Symptomatic
2	3 years (1998)	L temporal	48 cm H_2O (LP opening pressure)	Chiari 1 malformation	Shunt revisions = 8 Cystoperitoneal, Frontal ventriculoperitoneal, Lumboperitoneal	-	Symptomatic
3	8 months (1996)	L frontotemporal	17, 25, 14, cm H_2O (LP opening pressure) 30, 20–60 cm H_2O (intracranial pressure monitor)	Chiari 1 malformation	Shunt revisions=8 Cystoperitoneal, Ventriculoperitoneal, Lumboperitoneal	Sumatriptan 20 mg prn, Keppra 750 mg qAM 1000 mg qPM, Lamictal 150 mg twice daily	Symptomatic
4	4 years (2013)	L temporal	28 cm H_2O (LP opening pressure)	-	Shunt revisions = 0 Ventriculoperitoneal	Acetazolamide	Well

Pt: Patient, No.: Number, AC: Arachnoid cyst, ICP: Intracranial pressure, CSF: Cerebrospinal fluid, cm H_2O: Centimeters of water, Hx: History, L: Left, mg: Milligrams, prn: As required, qAM: Taken every morning, qPM: Taken every night

Our guiding CSF pressure range of normal can be misleading on the backdrop of IIH, which can manifest symptomatic disequilibrium at various fluid pressures as is evident in Patients 1, 2, 3, and 4. As depicted in the reported cases, IIH may present in the presence of what is classified as both increased and normal CSF pressure. Volume usually correlates directly with pressure, but this relationship does not necessarily correlate with the manifestation of IIH.

Rate of CSF production (15–20 ml/h) has been acknowledged as acting independently to CSF pressure levels except in cases of severely elevated cerebral pressures.[29] If the rate of CSF production and its pressure levels acts independently of one another, it is rational to assume that attempting to decrease CSF formation will not have a drastic effect on ICP and its clinical manifestation. This idea corresponds with the variability of CSF pressures in IIH and how a measurement may present within normal limits, but still depict an intracranial hypertensive picture clinically.

Shunt success

Conceptually, shunting is the most direct and logical method of rapidly decreasing elevated ICP. CSF shunting can be achieved through various different diversion pathways. Despite the multiple potential complications associated with shunting, it is by far the most commonly used treatment in IIH and its incidence is rising.[7] In IIH patients, shunting appears to have a temporary period of success.

CSF diversion as a treatment option in AC patients creates shunt dependency.[29] Therefore, not only must we worry about complications arising from the use of shunts but also the consequences of shunt malfunction. It is the author's opinion that shunt complications or malfunction is not a credible cause of IIH, as there is an already existent disorder of ICP present. This is substantiated by the above cases of CSF pressure dysfunction and symptomatic patients in the presence of a fully functioning shunt system, seen in Patients 1, 2, and 3.

Although shunt complications and malfunction are a very prevalent possibility, repeated CSF divertive action was undertaken by the neurosurgery department aimed to relieve elevated ICP. As seen in Patients 1, 2, and 3, symptomatic episodes did occur in the presence of no shunt dysfunction, yet alleviation of ICP was necessary. At present, it is rational to suggest that the most therapeutic method of improving a symptomatic patient is through CSF diversion techniques.

Slit-ventricle syndrome

Slit-ventricle syndrome is the decrease in size or collapse of the ventricles on radiological imaging.[29] This is exhibited in Patients 2 and 3 who had such a presentation in partial or complete shunt obstruction, over drainage with low ICP, or idiopathic as a possible consequence of the dynamic nature of IIH. Slit-ventricle syndrome in a patient with previously normal sized ventricles may reduce CSF volume, circulation, distribution, and perfusion but can also predispose the patient to increased pressure.

Shunt revision

Shunt systems are unreliable, difficult to evaluate from a functional level,[3,40] and as is clearly evident in the illustrated cases, in continuous need of revision. Although shunting undoubtedly decreases ICP, full resolution of hydrodynamic order is not always restored. Shunt revision is a very probable outcome[16,37] and a necessity.

Out of the four patients listed, above three required surplus CSF diversion. This idea is further perpetuated by the negative consequences of indirect shunting, including ventricle collapse and its sequelae slit-ventricle syndrome, low-pressure syndromes, and structural displacement, most specifically, the acquired Chiari malformation, as seen in Patients 2 and 3.

Indirect shunting aims to ameliorate increased ICP and restore equilibrium. In actuality, the procedure may inadvertently exacerbate a previously established hydrodynamic disorder.

Patients 1, 2, and 3 all remain symptomatic despite multiple and varied therapeutic efforts. Although the respective treatments appear successful and provide symptom relief, the respite period is finite, and the patients once more become symptomatic. The persistence of the condition may be attributed, to on-going issues with indirect shunting, limited treated options, or simply a failure to ascertain a fully efficacious treatment.

This continuous transformation strengthens the link between persistent progressive IIH and the hydrodynamic status of CSF. The advancing nature of the disease also authenticates the inability to decipher a solid evidence-based strategy toward the management of IIH, as our present view identifies IIH as a static condition. The balancing act and continuous remodeling demonstrated with therapeutic CSF diversion as seen in Patients 1, 2, and 3, reinforced the idea of IIH as a dynamic evolving disorder.

This study is a retrospective case series. The study is limited as it allows for documentation and commentary on the patients and their condition. It is only useful for descriptive, exploratory analysis of the patients who have this disorder. It is also unknown whether these results are applicable to a wider population. Furthermore, replication is difficult. Researcher bias is also present as the researchers can only assess the data subjectively.

The patient ages ranged from 3 to 44 years. This is the largest series to date of patients suffering with IIH in the presence

of AC, although the sample size is small. This is due to the extremely rare nature of the condition, as well as the collected sample being limited to patients attending the neurosurgery department from 1994 to 2013. This leads to issues with interpretation of the qualitative analysis. Regardless, this case series provides rich, qualitative data on a condition, and obscure association where there is, as of yet, very little knowledge and understanding. This study is not designed to result in strong conclusions; rather, these data should be applied to the format of larger confirmatory studies and testable hypotheses.

This study attempts to link anatomical, physiological, and pathophysiological principles with the patients' clinical characteristics [Table 2]. Our extensive literature review did not identify any other studies, which ventured into such intricate etiology. This is the only study, which not only documents the patients' condition and its progression but also strives to distinguish connections, which could lead to larger authentic investigations and, moreover, the discovery of this condition's etymology.

Table 2: Identification of clinical characteristics associated with the pathophysiology of IIH after AC removal.

Clinical characteristic	Pathophysiologic constituents of IIH post-AC removal
Pharmacological management of IIH (CR 4)	CSF production
ICP within normal limits (CR 1, 3)	CSF volume
Surgical removal of AC (CR 1, 2, 3, 4)	AV and CSF absorption CSF circulation CSF diversion
IIH symptomatology at normal and elevated ICP (CR 1, 2, 3, 4)	CSF pressure ranges
IIH symptomatology in CSF diversion (CR 1, 3)	CSF dysfunction in functioning shunt system
IIH symptomatology in CSF diversion (CR 2, 3)	Slit ventricle syndrome Chiari 1 malformation
IIH symptomatology in CSF diversion (CR 1, 2, 3)	Shunt revision
Symptomatic patient despite treatment (CR 1, 2, 3)	Persistent IIH

IIH: Idiopathic intracranial hypertension, AC: Arachnoid cyst, CR: Case report, CSF: Cerebrospinal fluid, ICP: Intracranial pressure

CONCLUSION

If we can infer anything from the above evidence, it is the irrefutable fact that IIH, in the setting of AC, is a hydrodynamic condition of fluctuant nature. Identification of this disequilibrium's pathophysiology in AC patients will result in an efficacious treatment and subsequently a resolution of IIH.

This review increases our depth of knowledge into this obscure condition and links clinical characteristics with

various anatomical and pathophysiological elements to enhance our understanding. It also highlights how much there is yet to be discovered.

A clear guide to the management of this condition can only be definitively established when the pathophysiologic etiology is fully understood. It is for this reason that further physiological studies of AC and CSF hydrodynamics are necessary in this field.

IIH is a diagnosis of exclusion. It is so, because much has still to be clarified about the condition. Conceivably, the cause of IIH may never be discovered. This condition encompasses all disorders of elevated ICP and until the origin and pathological mechanisms are gradually illuminated, these illnesses will remain classified as idiopathic.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

Conflicts of interest

There are no conflicts of interest.

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