




# Atypical Presentation of Idiopathic Intracranial Hypertension: A Case Series and Literature Review

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## Abstract

Idiopathic intracranial hypertension (IIH) is a condition in which intracranial pressure (ICP) increases without an apparent cause. Typically, patients present with headaches, dizziness, pulsatile tinnitus, visual disturbances, blurred vision, diplopia, photophobia, visual field defects, and papilledema on fundoscopy. The association between IIH, spontaneous cerebrospinal fluid (CSF) rhinorrhea, and arachnoid cysts has been discussed in the literature; however, there is no clear explanation for this association. We aimed to present a series of four patients with a confirmed diagnosis of IIH with atypical presentations, discuss the management of each case, and provide an explanation for this association to alert clinicians to the atypical presentation of IIH and facilitate early diagnosis and proper treatment of this condition by CSF diversion. This was a retrospective case series of all patients who were diagnosed with IIH and showed improvement after ventriculoperitoneal shunt insertion after failure of at least one operative intervention resulting from primary radiological and clinical findings in 2001 to 2022. Data on demographics, clinical presentation, radiological findings, surgical management, and diagnostic criteria for IIH were recorded. We identified four patients with a confirmed diagnosis of IIH who presented with atypical presentations as follows: intracranial arachnoid cyst, cervical spine arachnoid cyst, giant Virchow perivascular space, and spontaneous CSF (CSF) rhinorrhea. All patients responded to CSF diversion after failure of surgical treatment targeting the primary pathology. IIH should be suspected after the failure of primary surgical treatment in cases of spontaneous CSF rhinorrhea, spinal and cranial arachnoid cysts, and symptomatic ventriculoperitoneal shunt. Treatment in such situations should be directed toward IIH with CSF diversion.

## Keywords

- ▶ idiopathic intracranial hypertension
- ▶ arachnoid cyst
- ▶ spontaneous CSF rhinorrhea
- ▶ Virchow perivascular space
- ▶ lumboperitoneal shunt

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## Introduction

Idiopathic intracranial hypertension (IIH), previously known as pseudotumor cerebri, is a condition in which intracranial pressure (ICP) increases without an apparent cause. In Western countries, the incidence of IIH in the general population is approximately 1 to 2/100,000 per year and increases dramatically to reach 19/100,000 per year among females who are overweight by more than 20% of their optimal weight and who are of childbearing age (20–44 years). In Asian countries, the incidence is much lower, accounting for 0.03/100,000 per year, and is rare in men and children.<sup>1,2</sup> Obesity is considered a major risk factor for developing IIH and increases the morbidity of the disease.<sup>3,4</sup> The etiology is linked to the overuse of certain drugs, in particular growth hormone, tetracyclines, and excessive vitamin-A intake. Patients diagnosed with obstructive sleep apnea are also reported to have a higher risk of developing IIH.<sup>2–4</sup> Typically, patients present with headache, dizziness, pulsatile tinnitus, visual disturbances, blurred vision, diplopia, photophobia, visual field defects, and papilledema on fundoscopy.<sup>3,5–7</sup>

The association between IIH, spontaneous cerebrospinal fluid (CSF) rhinorrhea, and arachnoid cysts (ACs) has been reported; however, there is no clear explanation for this association. We aimed to present a series of four patients with a confirmed diagnosis of IIH with atypical presentations, discuss the management of each case, and provide an explanation for this association to alert clinicians to the atypical presentation of IIH and facilitate early diagnosis and proper treatment of this condition by CSF diversion.

## Materials and Methods

A retrospective search of the operative database was used to review the surgical records, clinical presentations, and radiological studies of patients who were diagnosed with IIH and improved after ventriculoperitoneal (VP) insertion after failure of at least one operative intervention related to the primary radiological and clinical findings. All the included patients underwent surgery at our institute between 2001 and 2022.

We excluded patients who were previously diagnosed with hydrocephalus or brain tumors, or who underwent CSF diversion procedures. IIH was suspected as the primary diagnosis if there was papilledema, normal brain parenchyma without evidence of hydrocephalus, a tumor mass on neuroimaging, or no abnormal meningeal enhancement on magnetic resonance imaging (MRI), with or without gadolinium. Moreover, elevated opening pressure ( $\geq 250$  mm CSF) in a properly performed lumbar puncture or more than or equal to 280 mm CSF in children, if not adequately sedated.

We included four patients who met our criteria. Data on demographics, clinical presentation, radiological findings, surgical management, and diagnostic criteria for IIH were recorded. All patients were followed up after surgery for clinical assessment and radiological investigation, if necessary.

## Results

### Demographics, Presentation, and Surgical Management

Based on the primary radiological findings, we identified four patients who underwent VP shunt insertion after failure of the surgical intervention. All patients were followed up post-VP shunt surgery and reported significant improvement in preoperative symptoms. There were no reported mortalities in the four patients with unusual IIH presentations. **Table 1** shows the demographics, clinical presentation, radiological findings, surgical management, and diagnostic criteria for IIH.

### Case Presentation

#### Case 1: Cranial Arachnoid Cyst

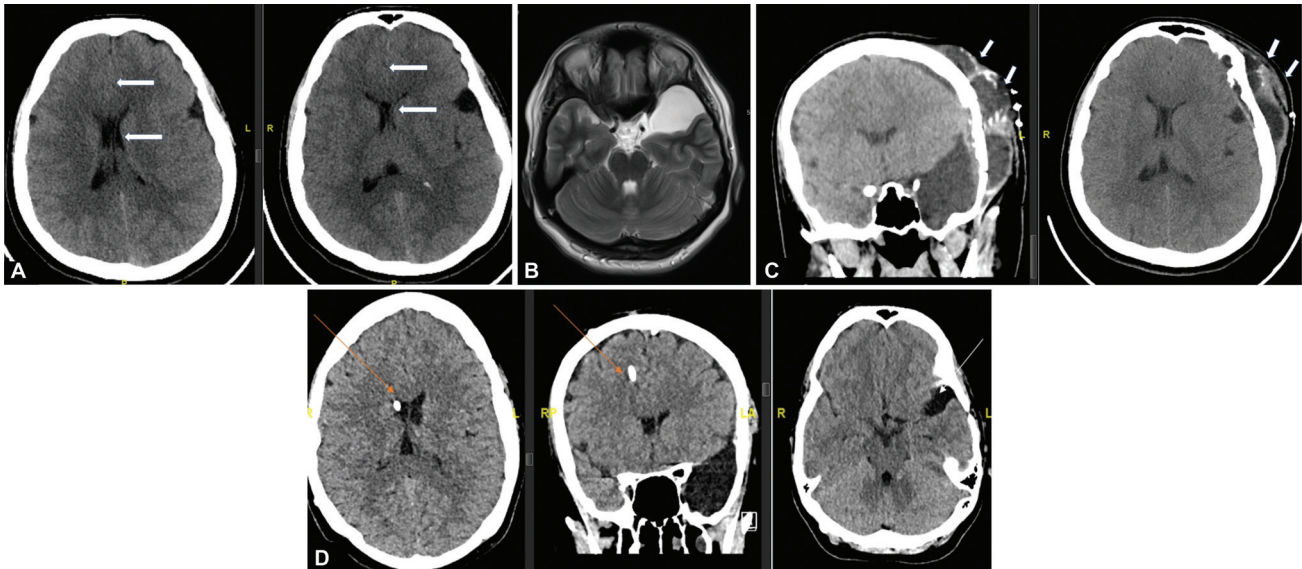
A 22-year-old female presented to the emergency department complaining of a mild occipital headache that had radiated to the cervical spine for 1 week and blurry vision. The headache responded to over-the-counter analgesia, and the patient reported nausea and a few episodes of vomiting for 3 days but denied any history of trauma, fever, photophobia, phonophobia, or tinnitus. Neurological examination revealed right gaze nystagmus, decreased visual acuity in the right eye, a right nasal visual field defect, and grade 4 bilateral papilledema with intraretinal hemorrhage in the right eye. Computed tomography (CT) showed a large left fronto-temporo-parietal AC causing a mass effect and a rightward midline shift (**Fig. 1A**). MRI revealed a large, well-defined, extra-axial, left-sided cystic lesion in the middle cranial fossa that followed the CSF signal on all sequences (**Fig. 1B**). The patient underwent a left-sided minicraniotomy with cyst fenestration.

Postoperative CT showed expected changes. The patient reported improvement in her headache 3 days postoperatively but continued to have blurry vision. One week later, the patient presented to the emergency department with headache, bilateral blurry vision, and left frontal swelling. CT (**Fig. 1C**) showed subgaleal CSF collection but no acute insults, and brain MRI was unremarkable. Lumbar puncture revealed a high opening pressure (300 mm of water), and CSF analysis was unremarkable. The diagnosis of IIH was raised and the patient underwent right frontal VP shunt insertion (**Fig. 1D**). The patient's symptoms improved dramatically after VP shunt insertion and resolved completely within a month.

#### Case 2: Cervical Arachnoid Cyst

A 69-year-old male diagnosed with hypertension and dyslipidemia presented in 2003 with a left cervical (C4–5) AC causing left brachialgia. He underwent corpectomy of C5 and cyst excision, and developed a CSF leak after surgery, which resulted in the insertion of a lumboperitoneal (LP) shunt.

The patient's symptoms improved, and his condition was stable until he presented in 2021 with severe disabling bilateral shoulder and arm pain along with headache; however, the pain was not associated with movements and was



**Fig. 1** (A) Preoperative axial computed tomography (CT) images showing the mass effect and midline shift (white arrows). (B) Axial T2-weighted magnetic resonance imaging showing the left temporal arachnoid cyst. (C) CT brain scan after craniotomy and microscopic fenestration of the cyst. Coronal and axial images showing the subgaleal cerebrospinal fluid collection (white arrows). (D) CT scan after insertion of a right frontal (red arrow) ventriculoperitoneal shunt.

difficult to control with strong analgesics. Radiologic MRI showed a left C4–C6 AC with long-standing bony changes (► **Fig. 2A**). Although fundoscopy showed no papilledema, we decided to revise the LP shunt, and the opening pressure was high (350 mm H<sub>2</sub>O). A new LP shunt was inserted, his symptoms improved dramatically after the revision, and he reported improvement of the headache postoperatively.

The patient underwent a second revision of the LP shunt 12 months later. In February 2023, he had a third attack of LP malfunction with recurrence of severe left-sided brachialgia, which was changed from an LP shunt to a left frontal VP shunt (medium-pressure valve); his condition improved, and in the last follow-up clinic visit, his condition was good and stable (► **Fig. 2B**).

### Case 3: Virchow Giant Perivascular Space

A 35-year-old female patient, with no previous medical problems, presented to the hospital with progressive headache and numbness in her right foot for 6 months. Neurological examination showed no motor weakness, and she had impaired pin prick, light touch, and temperature sensation in the right leg and foot. Fundus examination showed bilateral papilledema. CT and MRI revealed a multilocular non-enhancing cystic lesion involving the midbrain, extending to the pons and thalamus, along with mild ventricular dilation and a giant Virchow perivascular space (PVS; ► **Fig. 3A, B**). The patient underwent right frontal VP shunt insertion and, upon tapping the ventricle, it was noticed that the opening pressure was quite elevated (unfortunately, we did not measure the CSF pressure). Postoperatively, the patient's symptoms improved significantly, and the postoperative CT scan (► **Fig. 3C**) showed regression of hydrocephalic changes, which decreased in size after surgery but remained unchanged during follow-up; the patient was clinically free of symptoms.

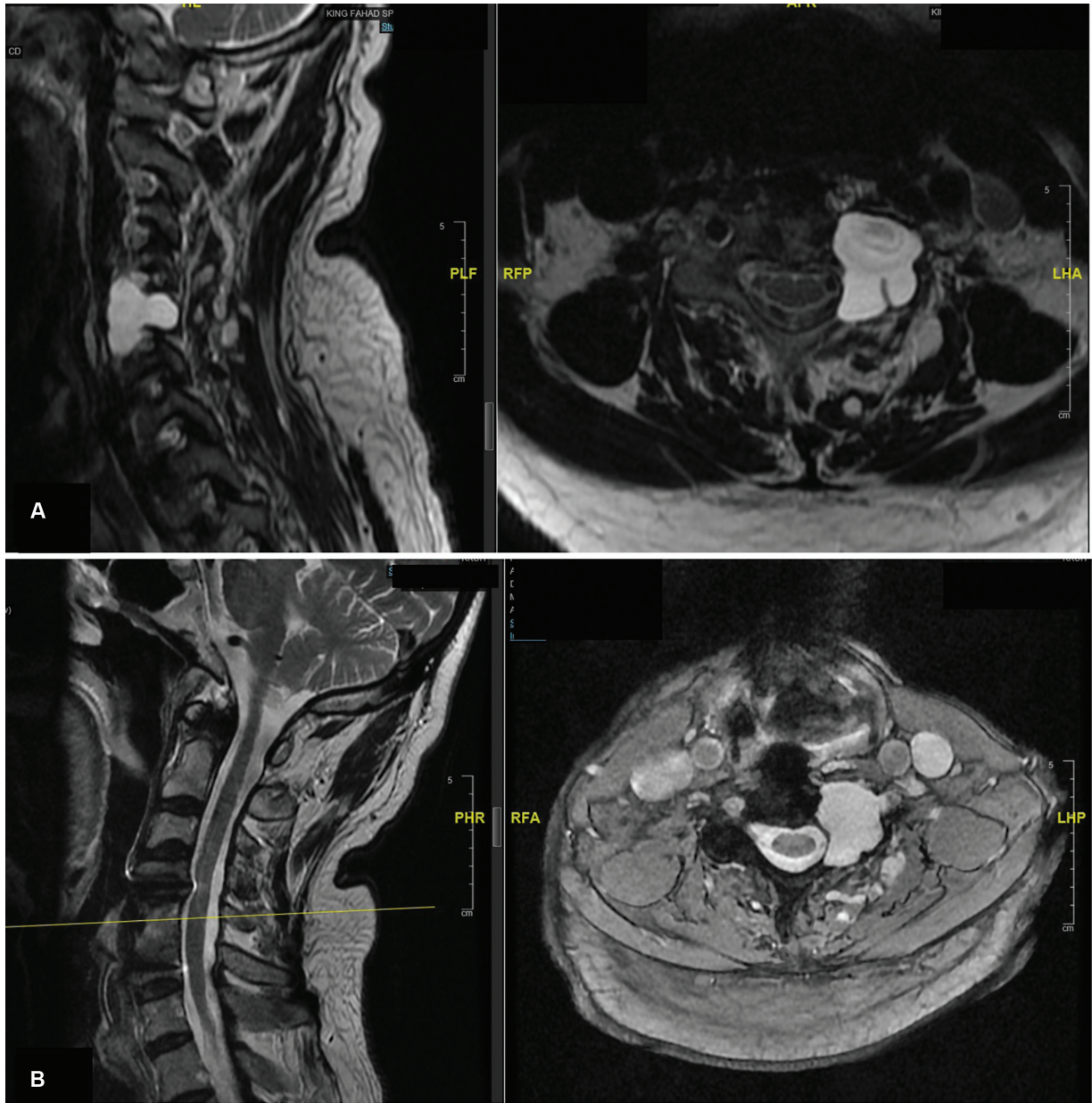
### Case 4: Spontaneous CSF Rhinorrhea

A 50-year-old gentleman presented with spontaneous CSF rhinorrhea in 1995 with no history of head trauma or surgery. Radiological studies (high-resolution CT of the head with thin coronal cuts of the anterior cranial fossa and radionuclide cisternography) confirmed the presence of a CSF fistula. Contrast CT cisternography via lumbar puncture revealed a fistula on the floor of the anterior cranial fossa and contrast extending from the intracranial subarachnoid space into the sphenoid and ethmoid air cells. Two attempts at surgical repair of the CSF fistula (first trans-sphenoid and second transcranial surgery) failed to treat the CSF rhinorrhea. At this point, a diagnosis of IIH was made, and the decision for CSF diversion was made. A VP shunt was inserted, the CSF leak stopped, and the patient remained well for 5 years before being lost to follow-up. ► **Table 2** summarizes the criteria used for the diagnosis of IIH in the four patients, which were primarily raised when there was a documented rise in ICP, the presence of papilledema, and a significant improvement after VP shunting.

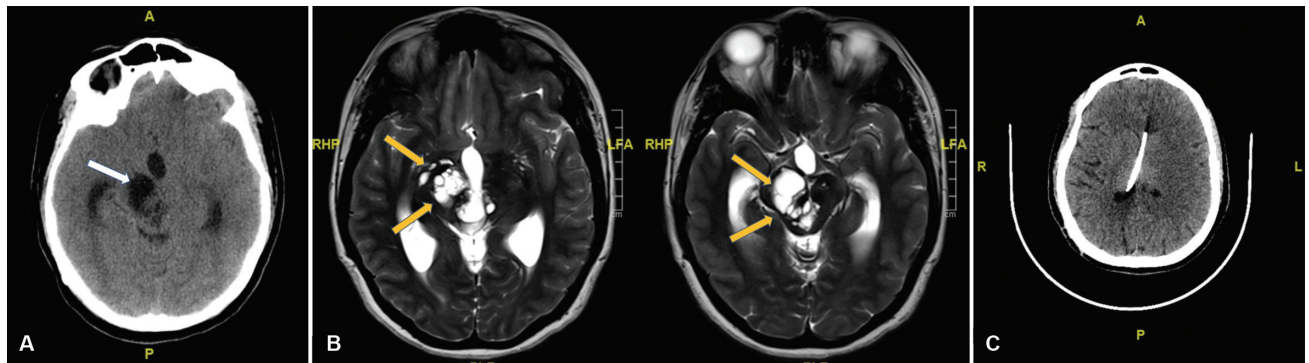
## Discussion

We identified four patients with a confirmed diagnosis of IIH who presented with atypical presentations as follows: intracranial AC, cervical spine AC, giant Virchow PVS, and spontaneous CSF rhinorrhea. All patients responded to CSF diversion after failure of surgical treatment targeting the primary pathology.

IIH is a disorder characterized by increased ICP without hydrocephalus or mass lesions and normal CSF composition. The diagnostic criteria for this disorder were described by Dandy in 1937 and were applied in clinical practice until they were modified in 1985.<sup>6,8</sup> The diagnostic criteria for IIH were updated in 2002 by the American Academy of Neurology,<sup>7,9,10</sup>



**Fig. 2** (A) T2-weighted magnetic resonance imaging (MRIs) showing the left side cervical arachnoid cyst at the intervertebral foramen. (B) T2-weighted MRIs showing regression of the cyst after ventriculoperitoneal shunt insertion.



**Fig. 3** (A) Axial computed tomography (CT) scan showing giant tumefactive perivascular space in the right-sided mesencephalon and thalamus (white arrow). (B) Axial T2-weighted magnetic resonance imaging showing the lesion the right aspect of the midbrain extending to the pons and thalamus on the ipsilateral side (white arrows). (C) Axial CT scan of the brain after insertion of the ventriculoperitoneal shunt.

**Table 1** Demographics, clinical presentation, radiological findings, and surgical management

Patient	Age (years)	Gender	Primary clinical presentation	Radiological finding	Primary surgery	Surgeries before VP shunt insertion	Papilledema
1	22	Female	Raised ICP	Left frontotemporal AC	Craniotomy and fenestration	1: Craniotomy and fenestration plus lumbar drain	Grade 4 with retinal hemorrhage
2	69	Male	Root irritation & severe brachialgia	Left C4–5 AC	Cyst excision	4: LP shunt insertion & 3 revisions of LP shunt	No
3	35	Female	Raised ICP, Rt. Side sensory deficit	Rt. Thalamic & midbrain giant PVS	Rt frontal VP shunt	None	Present
4	50	Male	Spontaneous CSF rhinorrhea	CSF fistula	1. Trans-sphenoid repair	2: Trans-sphenoid & Transcranial repair	Not examined

Abbreviations: AC, arachnoid cyst; ICP, intracranial pressure; LP, lumboperitoneal; PVS, perivascular spaces; VP, ventriculoperitoneal.

**Table 2** Diagnostic criteria for IIH

Patient	Diagnostic criteria
1	Symptoms of raised ICP, bilateral papilledema and high opening pressure during LP > 300 mm water
2	CSF opening pressure was high > 350 mmH <sub>2</sub> O
3	VP shunt; upon tapping the ventricle, CSF pressure was considerably high (unfortunately the CSF pressure was not measured). Bilateral papilledema
4	CSF leak reluctant to primary repair and improvement after VP shunt

Abbreviations: CSF, cerebrospinal fluid; LP, lumboperitoneal; ICP, Intracranial pressure; VP, ventriculoperitoneal.

and included papilledema; normal neurologic examination except for cranial nerve abnormalities, normal brain parenchyma without evidence of hydrocephalus, mass, or structural lesion on neuroimaging; and no abnormal meningeal enhancement on MRI, with and without gadolinium. Moreover, elevated opening pressure ( $\geq 250$  mm CSF) in a properly performed lumbar puncture or more than or equal to 280 mm CSF in children, if not adequately sedated.

### Arachnoid Cyst

ACs are congenital collections of CSF-like fluid within the arachnoid membrane, lined by arachnoid cells, and situated in the subarachnoid space of the cisterns and major cerebral fissures. It has been suggested that ACs arise during the early stage of subarachnoid space formation from the maldevelopment of the leptomeninges during the embryological period, which leads to disturbances in CSF hydrodynamics.<sup>11</sup> ACs account for nearly 1 to 1.4% of intracranial space-occupying lesions; most cases (approximately 75%) occur in the pediatric age group, and in most cases, they do not grow and remain clinically silent. Different theories explain their growth, including production of CSF from the cyst wall that contains some choroid plexus remnants, increased osmotically active protein composition in the cystic fluid, selective or active transport from the cyst-lining cells, or mechanical entrapment of the fluid inside the cyst.<sup>11,12</sup> Most ACs are clinically silent and have a benign course; however, some cysts can enlarge causing neurological symptoms and signs of raised ICP and local mass effect.<sup>12,13</sup> In 2020, Houlihan and Marks described four patients with intracranial ACs associated with IIH.<sup>14</sup>

In our study, two patients had symptomatic ACs and were found to have IIH and manifestations of raised ICP (high CSF opening pressure >250 mmH<sub>2</sub>O). To our knowledge, this is the first case in the English literature of IIH presenting with a cervical/spinal AC.

Both patients did not improve after primary surgery directed at the ACs, and both had persistent symptoms of high CSF pressure; they developed CSF leaks from their wounds, and both improved dramatically and rapidly after CSF diversion. Previous reports have not clearly explained the relationship between ACs and the development of IIH or

primary pathology. To date, there is no consensus on the optimal medical management to treating patients with both conditions simultaneously.<sup>13–16</sup>

We hypothesized that the development of symptoms in an otherwise silent AC is secondary to the development of IIH, and not vice versa. The presence of elevated CSF opening pressure may explain why symptoms recur quickly after the initial surgical treatment directed towards the cyst, as well as the rapid improvement of the symptoms after CSF diversion.

### Giant Virchow Perivascular Spaces

Cerebral PVS, also known as Virchow–Robin spaces, are physiological interstitial fluid-filled channels, typically less than 2 mm in diameter, that extend from the subpial space and form around arterial perforators as they course from the cortex into the brain parenchyma.<sup>17</sup> The precise functions of these structures have yet to be delineated, but predominant theories suggest, including facilitation of fluid movement between the basal cisterns and the interstitial space; modulation of immune responses by providing a conduit for macrophages and lymphocytes to reach the CSF; and forming part of the glymphatic system for metabolic waste product elimination.<sup>17,18</sup> The PVS are considered dilated when they become larger than 2 mm, frequently observed with advancing age, various neuropsychiatric disorders, multiple sclerosis, microvascular disease, and traumatic brain injury.<sup>18,19</sup>

The cause of PVS dilatation is unclear. Hydrodynamic disturbances in the CSF and interstitial fluid flow caused by slow-growing benign tumors or preexisting hydrocephalus have been suggested. Alternatively, increased vessel permeability with fluid exudation due to microvascular disease or ex vacuoperiarteriolar ischemic parenchymal injury, resulting in interstitial fluid leakage, has also been postulated.<sup>17–19</sup>

There is no clear etiology for giant symptomatic VP shunt; however, we assume that the progression of VP shunt is secondary to IIH, as all criteria for the diagnosis of IIH are met (symptoms of raised ICP, bilateral papilledema, and high opening CSF pressure) and a dramatic response to CSF diversion (VP shunt). Accordingly, we believe that IIH might be considered the primary diagnosis underlying the progression of PVS in our patient.

### Spontaneous CSF Rhinorrhea

Striking overlaps exist between the demographic, clinical, and radiological characteristics of patients with IIH and those with spontaneous CSF leakage, suggesting that the two entities are strongly related.<sup>20,21</sup> The association between CSF leaks through the skull base and raised ICP has been reported since the 1960s, and it has been suggested that spontaneous CSF leaks might represent a variant of IIH.<sup>20–22</sup>

Diagnosing IIH in patients with spontaneous CSF leaks represents a challenge for the treatment team. Data regarding funduscopic examinations in patients with spontaneous CSF leaks are deficient, and it is not clear how the diagnosis was made. Furthermore, other reports relied solely on an ICP greater than 25 cm of water to justify the diagnosis.<sup>22–27</sup> The pathophysiology is unknown but might stem from progressive

erosion of the thin bone of the skull base by persistent pulsatile high CSF pressure. Currently, there is no consensus regarding ICP management after spontaneous CSF leak repair when IIH is suspected.

Many patients with spontaneous CSF leaks may harbor initial symptoms and signs suggestive of raised ICP, such as headaches (60%), pulsatile tinnitus (20%), and diplopia in only 5% of cases.<sup>24–26</sup> These rates are lower than those associated with classic IIH (90, 60, and 30%, respectively), which is not surprising given the likelihood that the leak decompensates the IH and, therefore, relieves symptoms. This explains why papilledema is rarely reported. Additionally, spontaneous CSF leaks have a higher rate of failure after repair than those of traumatic origin, and closure of spontaneous leaks has occasionally been reported after interventions aimed at decreasing ICP.<sup>22,24,26</sup>

In our patient (case number 4), the CSF leak did not stop after trans-sphenoid and transcranial repair and stopped only after VP shunt insertion, which supports the assumption that spontaneous CSF rhinorrhea is associated with IIH. Although no consensus exists in the literature, a growing number of authors advocate for more aggressive postoperative management of ICP, such as with ICP-lowering agents (e.g., acetazolamide) or CSF diversion procedures.<sup>24</sup>

## Conclusion

A diagnosis of IIH should be made after failure of primary surgical treatment in cases of spontaneous CSF rhinorrhea, spinal and cranial ACs, and symptomatic VP shunt. Hydrostatic CSF pressure in cases of IIH forces the CSF in the direction of least resistance, causing empty Sella syndrome, spontaneous CSF rhinorrhea, growth of ACs, and VP shunt. Treatment in such situations should be directed toward IIH in the form of CSF diversion.

### Ethical Approval

The manuscript does not contain patient data or prospective clinical studies.

### Authors' Contributions

F.S.A. helped in conceptualization, methodology, software, validation, visualization, writing, review and editing. A.I.A. was involved in conceptualization, data curation, formal analysis, investigation, methodology, project administration, resources, validation, visualization, writing original draft, review and editing. S.A. and S.M. helped in data curation, investigation, resources, software, validation, and visualization. S.M.E. was involved in conceptualization, data curation, methodology, software, supervision, validation, visualization, writing review and editing.

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**Conflict of Interest**

None declared.

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