Auditory and Vestibular Research

Spontaneous Intracranial Hypotension with Predominant Audiovestibular Symptoms: Two Cases and a Review of the Literature

Asst. Prof. M.D. Özden SAVAŞ
Department of Otorhinolaryngology, Dokuz Eylül University School of Medicine, Izmir, Turkey
Orchid Id: *0000-0002-3552-757X*

M.D. Kıvanç KORKMAZ
Department of Otorhinolaryngology, Dokuz Eylül University School of Medicine, Izmir, Turkey.
Orchid Id: 0000-0002-5963-1574

Prof. M.D. Süleyman MEN
Department of Radiology, Dokuz Eylül University School of Medicine, Izmir, Turkey.
Orchid Id: 0000-0002-8221-3194

Prof. M.D. Enis Alpin GÜNERİ
Department of Otorhinolaryngology, Dokuz Eylül University School of Medicine, Izmir, Turkey.
Orchid Id: 0000-0003-2592-0463

Corresponding Author:
M.D. Kıvanç KORKMAZ
Department of Otorhinolaryngology, Dokuz Eylül University School of Medicine, Izmir, Turkey.
e-mail adress: kivanc.korkmaz@deu.edu.tr

HIGHLIGHTS

- Audiovestibular symptoms can be the initial presentation of intracranial hypotension
- Consider SIH in the differential diagnosis of hydrops-like audiovestibular symptoms

ABSTRACT

Background: Spontaneous intracranial hypotension is a recognized clinical entity caused by spinal cerebrospinal fluid leakage and is classically characterized by orthostatic headache. However, some patients initially present with audiovestibular symptoms that may mimic primary inner ear disorders and lead to diagnostic delay. Recognition of this atypical presentation is essential to avoid misdiagnosis and to facilitate timely neuroimaging and appropriate management.

The Cases: Two patients with spontaneous intracranial hypotension are described, in whom tinnitus, sound sensitivity, aural fullness, and dizziness constituted the predominant initial manifestations. A cerebrospinal fluid–venous fistula at the level of the tenth thoracic vertebra was identified using computed tomography myelography, and symptoms resolved following transvenous embolization after unsuccessful epidural blood patch therapy. In another patient, no definite site of cerebrospinal fluid leakage could be localized, and conservative management was preferred. A review of the literature is also provided, focusing on the proposed pathophysiological relationship between reduced cerebrospinal fluid pressure and secondary endolymphatic hydrops.

Conclusion: Spontaneous intracranial hypotension should be considered in the differential diagnosis of patients presenting with hydrops-like audiovestibular symptoms, even in the absence of headache. Early recognition and appropriate neuroimaging are essential for accurate diagnosis and optimal management.

Keywords: Spontaneous Intracranial Hypotension, Cerebrospinal Fluid-Venous Fistula, Endolymphatic hydrops, Tinnitus

Introduction

Spontaneous intracranial hypotension (SIH) was first described by Schaltenbrand in 1938 (1). SIH is an important cause of secondary headaches resulting from spinal cerebrospinal fluid (CSF) leaks that have received increasing attention over the past few decades (2).

The incidence has been estimated at 5 per 100 000 per year, with a peak around 40 years of age. Women are affected more commonly than men (3). The most prominent feature of this syndrome is profound orthostatic headache provoked by sitting or standing that is relieved by lying flat, usually within 15–30 minutes (4). Other symptoms include neck pain, back pain, radicular upper-limb symptoms, nausea and vomiting, diplopia, visual blurring, and bitemporal hemianopsia (5,6). Furthermore, if left untreated, SIH may result in frontotemporal brain sagging syndrome, which is characterized by progressive cognitive impairment, reduced conscious state, ataxia or parkinsonism (7,8). Additionally, several patients with SIH (20–30%) present with audiovestibular symptoms such as tinnitus, dizziness, aural fullness, hyperacusis, and fluctuating hearing disturbances (9,10). These manifestations are thought to reflect secondary inner ear involvement and may clinically resemble endolymphatic hydrops rather than a primary vestibular disorder (11,12,13).

The presence of diffuse pachymeningeal gadolinium enhancement accompanied by subdural fluid collections (i.e., chronic subdural hematoma) on brain magnetic resonance imaging (MRI) and low CSF pressure in the lateral decubitus or sitting position has been identified as diagnostic findings in patients presenting with orthostatic headache (14). Imaging is essential both to confirm the diagnosis of intracranial hypotension and to localize the leak. Pachymeningeal thickening and enhancement is the most common brain MRI finding followed by dural venous engorgement, tonsillar herniation, and subdural collection (15). Further localization of the leak can be achieved using a variety of myelographic techniques including dynamic computed tomography (CT) myelography, digital subtraction myelography, fluoroscopic myelography, magnetic resonance (MR) myelography, and nuclear medicine myelography (5).

This report presents two cases of SIH in which audiovestibular symptoms constituted the initial clinical presentation, including tinnitus, hyperacusis, aural fullness, and dizziness. One case was attributed to a spinal CSF–venous fistula, while no definite CSF leak could be identified in the second case.

Case Presentatıon

CASE 1

A 61-year-old male patient presented with sound sensitivity, dizziness, aural fullness and bilateral non-pulsating tinnitus for one week in the absence of concomitant symptoms, such as hearing loss. The medical history was unremarkable except for hypertension and benign prostatic hyperplasia. There was no past or recent history of trauma or surgery. No abnormalities were noted on neurotological examination or positional tests. Pure tone audiometry with 15 dB air and bone conduction thresholds bilaterally was compatible with normal hearing. Tympanometry revealed type A tympanogram bilaterally. Within a few weeks after the onset of auditory symptoms, the patient developed severe orthostatic headaches. Comprehensive vestibular testing was not performed, as the rapid development of severe orthostatic headache prompted neuroimaging and neurosurgical evaluation prior to further audiovestibular workup. Contrast-enhanced brain and temporal MRI revealed subdural effusion in both hemispheres, diffuse dural contrast enhancement at the supratentorial and infratentorial levels, and enlargement of the subdural space with engorgement of venous structures on T2-weighted images, all consistent with intracranial hypotension (Figure 1). The orthostatic nature of the headache combined with characteristic neuroimaging findings effectively excluded Ménière's disease and vestibular migraine as primary diagnoses.

The contrast-enhanced spinal MRI revealed spinal cord indentation at the level of the 10th–11th thoracic vertebrae, without evidence of epidural fluid collection. Contrast leakage from the subarachnoid space into the paraspinal vein at the 10th thoracic vertebra level on CT myelography confirmed the diagnosis of CSF-venous fistula (Figure 2). Epidural blood patch was administered twice, with a four-day interval between applications, without any notable improvement in the symptoms. Thereupon, endovascular embolization with ethylene-vinyl alcohol copolymer was performed (Figure 3). The patient's symptoms improved dramatically after embolization and cranial MRI performed 4 months after embolization showed recovery of findings associated with intracranial hypotension.

CASE 2

A 67-year-old woman presented with a 2-month history of non-pulsating bilateral tinnitus and dizziness. The patient had a medical history of hypertension and coronary artery disease, and had undergone surgery for L4-L5 disc herniation 10 years prior. The patient did not report headache. Normal otoscopic findings were observed, and the patient's balance examination was unremarkable. Pure tone audiogram was compatible with normal hearing with type A tympanogram bilaterally. Videonystagmography revealed no spontaneous, gaze-evoked, or positional nystagmus. Caloric testing demonstrated bilateral symmetric, normal responses. Further audiovestibular investigations including vestibular evoked myogenic potentials and electrocochleography could not be performed, as the patient declined additional diagnostic procedures due to the mild nature of her symptoms. The absence of headache militated against vestibular migraine, while the lack of fluctuating sensorineural hearing loss and recurrent vertigo attacks was inconsistent with a diagnosis of Ménière's disease. The patient was diagnosed with spontaneous intracranial hypotension after the detection of subdural collection and pachymeningeal enhancement on contrast-enhanced MRI of the brain (Figure 4a and b). CT myelography for fistula localization showed no contrast leakage. Neurosurgical evaluation was performed, and given the absence of progressive neurological symptoms and the patient's preference to decline further invasive diagnostic procedures or treatment, conservative management with close clinical follow-up was chosen. At six-month follow-up, the patient reported complete resolution of dizziness, while tinnitus persisted at a similar level in the absence of headache.

Discussion

In recent years, spontaneous spinal CSF leaks have been visualized in SIH patients with various imaging techniques such as digital subtraction myelography (DSM) and decubitus CT myelography and have been shown to establish the etiology of SIH (3,5).

Spontaneous spinal CSF-venous fistulas were first reported in 2014 in a small group of patients with spontaneous intracranial hypotension by Schievink et al. (16). The exact pathogenesis of these lesions is unclear, with potential hypotheses being that these lesions represent aberrant connections between the nerve root sleeve and a paraspinal vein or that these lesions are secondary to increased CSF drainage via spinal arachnoid granulations into adjacent radicular veins (17).

Schievink et al. classified spinal CSF leaks into three types. Type 1 leaks are associated with dural tears, usually presenting with extradural fluid collection. Type 2 leaks (most common) are related to the presence of meningeal diverticula (less commonly presenting with extradural collection). Type 3 leaks result from direct CSF-venous fistulas and do not result in an extradural collection (18). In a notable proportion of cases (19%), the underlying cause remains undetermined (19).

Mild cases of SIH may resolve spontaneously with conservative measures including bed rest, increased oral fluid uptake, steroids, caffeine and analgesics. Non-targeted epidural blood patch (injection of autologous blood into the spinal epidural space) is the first line of treatment (3). Localization of the CSF leak is required for targeted treatments in cases where repeated non-targeted epidural blood patch treatment fails. Myelography with iodinated contrast CT, MR myelography and digital subtraction myelography are useful for guiding targeted treatment (5). Surgical interventions such as dural repair, laminectomy, and nerve root sleeve closure are indicated for patients in whom nonsurgical measures have been ineffective (3). In 2021, Brinjikji et al. defined transvenous paraspinal vein embolization to treat CSF-venous fistula patients by occlusion of the CSF draining vein with a liquid embolic agent leading to a reduction in shunting sufficient to result in symptomatic and radiographic improvement (17). CSF-venous fistula cases refractory to repeated epidural blood patch or percutaneous fibrin glue injections are considered suitable for embolization technique (20).

Audiovestibular symptoms have been reported in 20–30% of patients with SIH and may include tinnitus, aural fullness, dizziness, and fluctuating hearing loss (9,10). These symptoms may precede the onset of orthostatic headache and can constitute the initial clinical manifestation of the condition, reinforcing the need for awareness of SIH in audiovestibular clinical settings (11,21,22). The clinical presentation may closely resemble that of Ménière's disease or other primary inner ear disorders, leading to diagnostic delay (12,21).

Although exact pathophysiology remains unknown, possible relationship between endolymphatic hydrops and reduced CSF pressure was first suggested by Carlborg and Farmer in 1983 and supported by recent literature (12,21,23). It is hypothesized that decreased cerebrospinal fluid (CSF) pressure may be transmitted via a patent cochlear aqueduct, leading to a reduction in perilymphatic pressure and subsequent expansion of the endolymphatic compartment. This secondary mechanism may result in hydrops-like audiovestibular symptoms.

SIH may be an under-recognized cause of endolymphatic hydrops (9,11,12). The audiovestibular manifestations observed in SIH may therefore clinically resemble endolymphatic hydrops and lead to diagnostic confusion. However, the absence of characteristic fluctuating sensorineural hearing loss and the presence of intracranial hypotension support a distinct secondary pathophysiological process rather than primary Ménière's disease. High clinical suspicion and appropriate neuroimaging are essential for accurate diagnosis and timely management (21).

Conclusıon

Audiovestibular symptoms may constitute the initial manifestation of spontaneous intracranial hypotension, possibly mediated through a secondary endolymphatic hydrops mechanism, though the exact pathophysiology remains to be fully elucidated. Recognition of this atypical presentation is crucial for appropriate diagnostic evaluation and management. In the first case, the etiology of SIH was attributed to a CSF–venous fistula at the level of the tenth thoracic vertebra, a relatively recently recognized cause of intracranial hypotension. These findings underscore the importance of considering SIH in the differential diagnosis of patients presenting with hydrops-like audiovestibular symptoms.

Ethıcal Consıderatıons

Compliance with ethical guidelines

This study was approved by the Ethics Committee of Dokuz Eylül University (Code: 2026/06-04 in 09/02/2026). The authors confirm that informed consent for publication was obtained from all participants prior to submission, and that all non-essential identifying details were omitted to protect patient privacy. Participants were informed that the material may be published in a scientific journal and made publicly available in print and/or electronic form, and consent was provided voluntarily with full understanding of the publication process.

Funding

This research did not receive any grant from funding agencies in the public, commercial, or non-profit sectors.

Authors' contributions

ÖS: Acquisition of data, interpretation of the results, and drafting the manuscript. KK: Acquisition of data and drafting the manuscript. SM: Study design, supervision, and critical revision of the manuscript. EAG: Study design, supervision, and final approval of the manuscript. All authors read and approved the final version of the manuscript.

Conflict of Interest

There are no competing financial interests.

Acknowledgements

The authors would like to thank the patients for their cooperation and consent to publish. The authors also extend their gratitude to the Department of Otorhinolaryngology and the Department of Radiology, Dokuz Eylül University, for their institutional support and contributions to the management of these cases.

References

1. Schaltenbrand G. Neuere Anschauungen zur Pathophysiologie der Liquorzirkulation. Zentralbl Neurochir. 1938;3:290-300.
2. Ferrante E, Trimboli M, Rubino F. Spontaneous intracranial hypotension: review and expert opinion. Acta Neurol Belg. 2020;120(1):9-18. [DOI:10.1007/s13760-019-01166-8]
3. Dobrocky T, Nicholson P, Häni L, Mordasini P, Krings T, Brinjikji W, et al. Spontaneous intracranial hypotension: searching for the CSF leak. Lancet Neurol. 2022;21(4):369-80. [DOI:10.1016/S1474-4422(21)00423-3]
4. Wang SJ. Spontaneous Intracranial Hypotension. Continuum (Minneap Minn). 2021;27(3):746-66. [DOI:10.1212/CON.0000000000000979]
5. Cheema S, Anderson J, Angus-Leppan H, Armstrong P, Butteriss D, Carlton Jones L, et al. Multidisciplinary consensus guideline for the diagnosis and management of spontaneous intracranial hypotension. J Neurol Neurosurg Psychiatry. 2023;94(10):835-43. [DOI:10.1136/jnnp-2023-331166]
6. Shukla D, Sadashiva N, Saini J, Kamath S. Spontaneous Intracranial Hypotension - A Dilemma. Neurol India. 2021;69(Supplement):S456-62. [DOI:10.4103/0028-3886.332255]
7. Datta A, Fasano A, Lenka A. Movement Disorders in Brain Sagging Syndrome Due To Spontaneous Intracranial Hypotension: A Review. Tremor Other Hyperkinet Mov (N Y). 2024;14:44. [DOI: 10.5334/tohm.914]
8. Sugiyama A, Tamiya A, Yokota H, Mukai H, Otani R, Kuwabara S. Frontotemporal Brain Sagging Syndrome as a Treatable Cause Mimicking Frontotemporal Dementia: A Case Report. Case Rep Neurol. 2022;14(1):82-7. [DOI:10.1159/000521968]

9. Cahal M, Roth J, Ungar OJ, Brinjikji W. Fluctuating hearing loss secondary to spontaneous intracranial hypotension: A case report and review of the literature. Interv Neuroradiol. 2023:15910199231221863. [DOI:10.1177/15910199231221863]
10. D'Antona L, Jaime Merchan MA, Vassiliou A, Watkins LD, Davagnanam I, Toma AK, et al. Clinical Presentation, Investigation Findings, and Treatment Outcomes of Spontaneous Intracranial Hypotension Syndrome: A Systematic Review and Meta-analysis. JAMA Neurol. 2021;78(3):329-37. [DOI:10.1001/jamaneurol.2020.4799]
11. Kim JK, Lee HY, Kim HY, Kwak MY. A hidden pathophysiology of endolymphatic hydrops: case report of a patient with spontaneous intracranial hypotension presenting with sudden sensorineural hearing loss with vertigo. Front Neurol. 2024;15:1394534. [DOI:10.3389/fneur.2024.1394534]
12. Sakano H, Jafari A, Allehaiby W, Harris JP. Spontaneous Intracranial Hypotension May Be an Under-recognized Cause of Endolymphatic Hydrops. Otol Neurotol. 2020;41(7):e860-3. [DOI:10.1097/MAO.0000000000002665]
13. Venkatasamy A, Péporté ARJ. Secondary endolymphatic hydrops: a clinical and literature overview. Front Neurol. 2025;15:1525954. [DOI:10.3389/fneur.2024.1525954]
14. Headache Classification Committee of the International Headache Society (IHS) The International Classification of Headache Disorders, 3rd edition. Cephalalgia. 2018;38(1):1-211. [DOI:10.1177/0333102417738202]
15. Farnsworth PJ, Madhavan AA, Verdoorn JT, Shlapak DP, Johnson DR, Cutsforth-Gregory JK, et al. Spontaneous intracranial hypotension: updates from diagnosis to treatment. Neuroradiology. 2023;65(2):233-43. [DOI:10.1007/s00234-022-03079-5]
16. Schievink WI, Moser FG, Maya MM. CSF-venous fistula in spontaneous intracranial hypotension. Neurology. 2014;83(5):472-3. [DOI:10.1212/WNL.0000000000000639]
17. Brinjikji W, Savastano LE, Atkinson JLD, Garza I, Farb R, Cutsforth-Gregory JK. A Novel Endovascular Therapy for CSF Hypotension Secondary to CSF-Venous Fistulas. AJNR Am J Neuroradiol. 2021;42(5):882-7. [DOI:10.3174/ajnr.A7014]
18. Schievink WI, Maya MM, Jean-Pierre S, Nuño M, Prasad RS, Moser FG. A classification system of spontaneous spinal CSF leaks. Neurology. 2016;87(7):673-9. [DOI:10.1212/WNL.0000000000002986]
19. Carlton Jones L, Butteriss D, Scoffings D. Spontaneous intracranial hypotension: the role of radiology in diagnosis and management. Clin Radiol. 2022;77(3):e181-e94. [DOI:10.1016/j.crad.2021.11.007]
20. Callen AL, Timpone VM, Schwertner A, Zander D, Grassia F, Lennarson P, et al. Algorithmic Multimodality Approach to Diagnosis and Treatment of Spinal CSF Leak and Venous Fistula in Patients With Spontaneous Intracranial Hypotension. AJR Am J Roentgenol. 2022;219(2):292-301. [DOI:10.2214/AJR.22.27485]
21. Fontaine N, Charpiot A, Debry C, Gentine A. A case of spontaneous intracranial hypotension: from Ménière-like syndrome to cerebral involvement. Eur Ann Otorhinolaryngol Head Neck Dis. 2012;129(3):153-6. [DOI:10.1016/j.anorl.2011.08.005]
22. Sharp S, Silbert PL, Davies S, Friedland P. Spontaneous Intracranial Hypotension: An Uncommon Cause of Postural Vestibulocochlear Symptoms. Ear Nose Throat J. 2026;105(3):191-7. [DOI:10.1177/01455613231194431]
23. Carlborg BI, Farmer JC Jr. Transmission of cerebrospinal fluid pressure via the cochlear aqueduct and endolymphatic sac. Am J Otolaryngol. 1983;4(4):273-82. [DOI:10.1016/s0196-0709(83)80071-4]

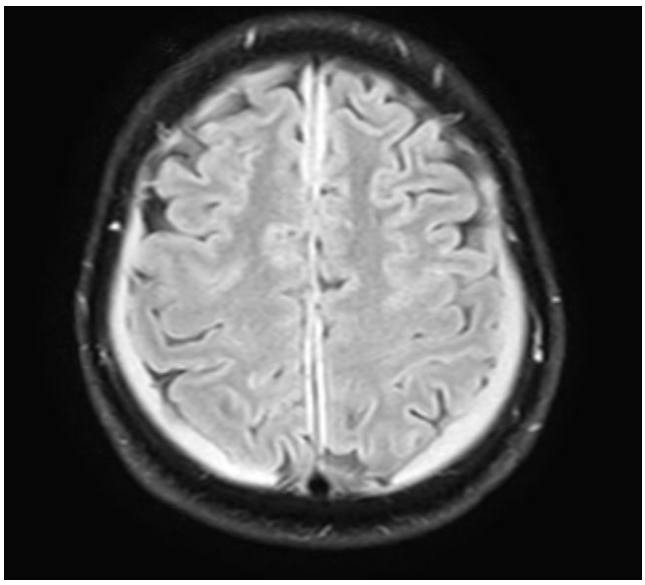

Figure 1 - Subdural effusion in both hemispheres, prominence of dural sinus structures, and diffuse dural contrast enhancement at the supratentorial and infratentorial levels.

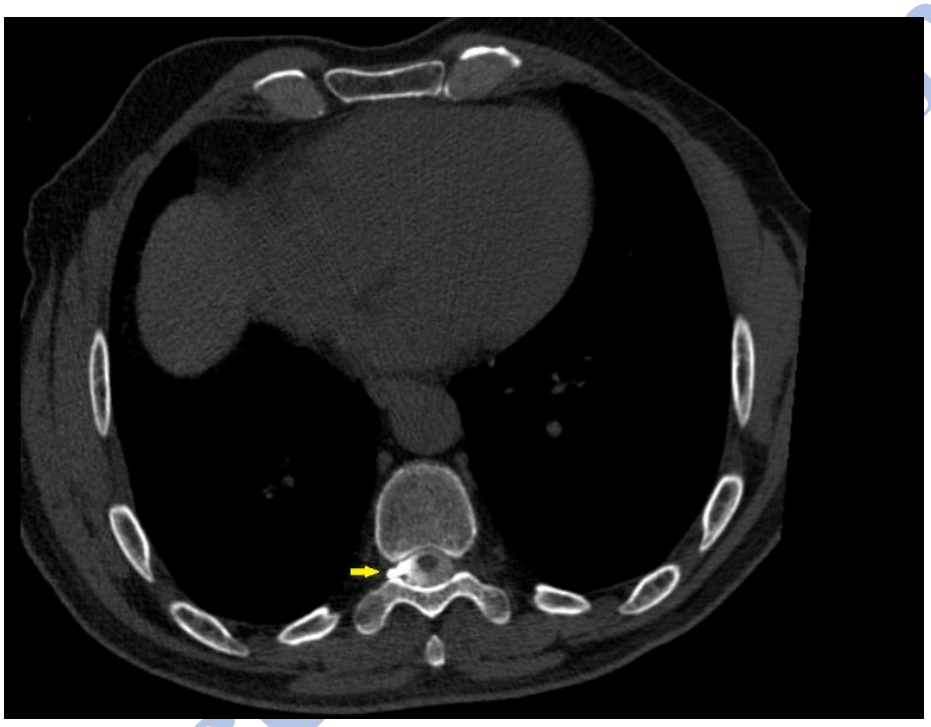

Figure 2 – Arrow shows contrast leakage from the subarachnoid space into the paraspinal vein at 10th thoracic vertebrae level.

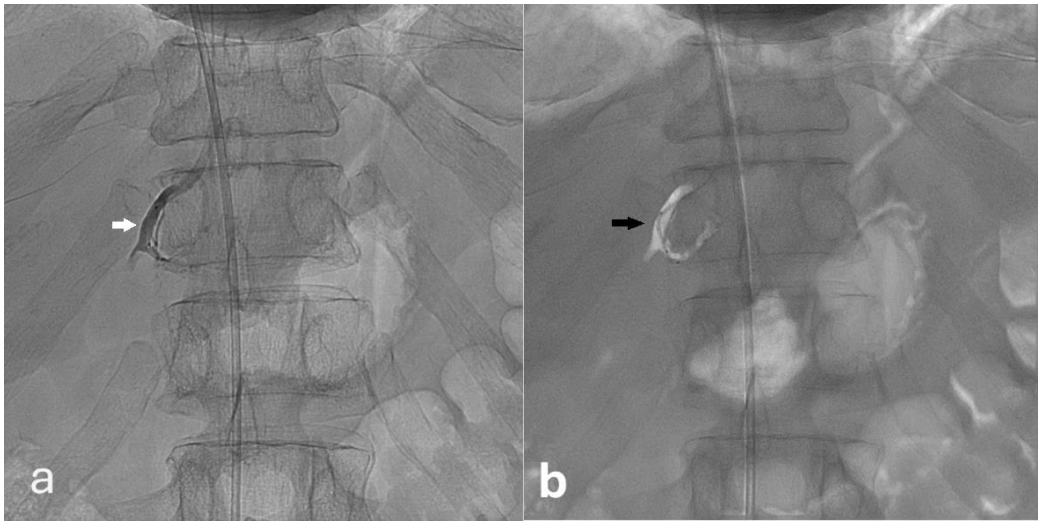

Figure 3 – Images of subtraction myelography, white arrow shows venous fistula before embolization (a) and black arrow shows same vein obstructed with ethylene-vinyl alcohol copolymer after embolization (b).

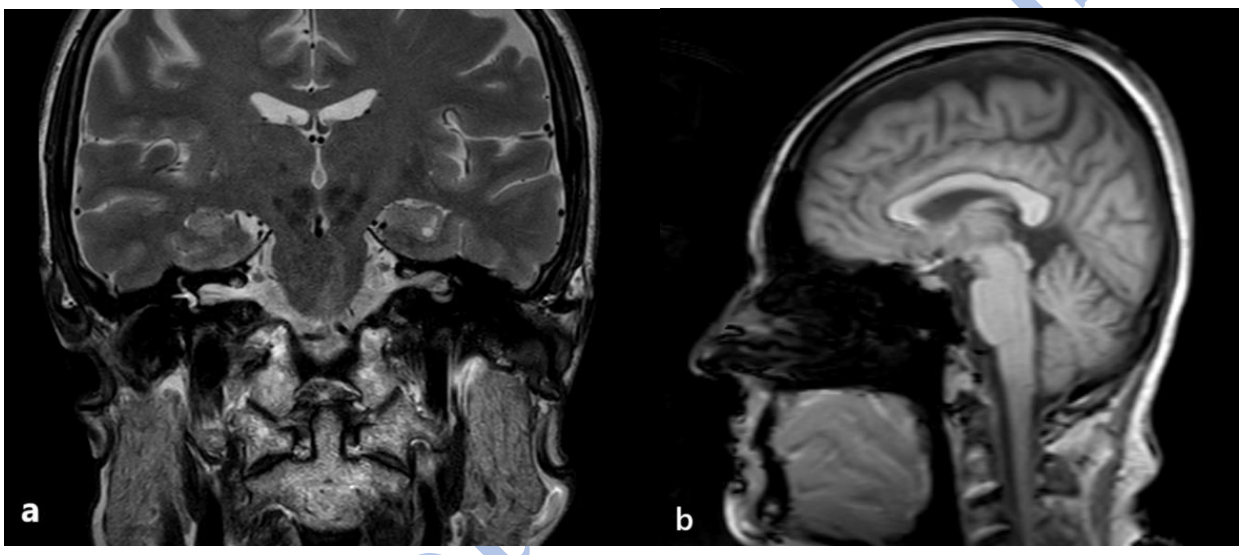

Figure 4- Cranial MR findings showing pachymeningeal enhancement and engorgement of venous sinuses on coronal T2 image (a) and sagittal T1 image demonstrating subdural fluid collection (b).