Interdisciplinary Neurosurgery: Advanced Techniques and Case Management 34 (2023) 101815

Contents lists available at ScienceDirect

Interdisciplinary Neurosurgery: Advanced Techniques and Case Management

journal homepage: www.elsevier.com/locate/inat

Case Reports & Case Series

Treatment for spontaneous intracranial hypotension associated with cerebral venous thrombosis resulting in severe neurologic deterioration

Cheol Young Lee

Department of Neurosurgery, Chung-Ang University Gwangmyeong Hospital, Gwangmyeong City, South Korea

ARTICLE INFO

Keywords:
Spontaneous intracranial hypotension
Cerebral venous thrombosis

ABSTRACT

Spontaneous intracranial hypotension is an uncommon condition, caused by spontaneous leakage of cerebrospinal fluid (CSF), but well-known cause of orthostatic headache. Although subdural fluid collection is a usual complication of SIH, SIH as a risk factor for cerebral venous thrombosis is not well known and a causal relationship has not been clearly established. Generally, spontaneous intracranial hypotension alone has been known to show clinically benign course, but this complication like cerebral venous thrombosis can be seizure and/or life threatening course. Each treatment for spontaneous intracranial hypotension and cerebral venous thrombosis is well known, however, its indication and treatment methods, in the setting of concurrent spontaneous intracranial hypotension with cerebral venous thrombosis is unknown, especially in severe neurologic deterioration status. Herein, we report a case of a patient was treated with emergency epidural blood patch followed by intra-venous direct thrombectomy and anticoagulant therapy for spontaneous intracranial hypotension complicated by cerebral venous thrombosis and would like to discuss treatment.

1. Introduction

Spontaneous intracranial hypotension is not only an increasingly recognized cause of new-onset headaches, particularly among young and middle-aged adults, but also a well-known cause of orthostatic headache. It is a condition of negative intracranial pressure due to cerebrospinal fluid (CSF) leakage from the dural sac [1–3]. Although subdural fluid collection and/or hemorrhage are usual complications of spontaneous intracranial hypotension, but there have been little known about spontaneous intracranial hypotension as a risk factor that can lead to cerebral venous thrombosis. In general, spontaneous intracranial hypotension has been known to show a clinically benign course, however, spontaneous intracranial hypotension can result in life-threatening conditions such as cerebral venous thrombosis, which can cause subarachnoid hemorrhage and seizures [4]. These conditions can eventually lead to death [5,6]. Approximately one-third of patients with cerebral venous thrombosis develop focal or generalized seizures prior to diagnosis [7]. When a patient has both spontaneous intracranial hypotension and cerebral venous thrombosis, a decision regarding the priority and modality of treatment is required. As we known, the standard first-line treatment for spontaneous intracranial hypotension is epidural blood patch (EBP), whereas that for cerebral venous thrombosis is anticoagulant therapy. However, treatment guidelines and indications in the setting of concurrent spontaneous intracranial hypotension and cerebral venous thrombosis are unclear. Moreover, for patients with a neurologically severe status, anticoagulant therapy alone may not be tolerable or may not prevent neurological aggravation. Here, we report a complex case of a patient with spontaneous intracranial hypotension complicated by cerebral venous thrombosis and would like to discuss the treatment methods.

2. Case report

A 38-year-old man had headache and neck pain for several weeks. A few days before admission, the patient presented decreased consciousness with seizure. Thus, he was transferred to our department. On admission, neurologic examination showed stupor consciousness (Glasgow Coma Scale: GCS 8), no verbal response, and motor weakness grade III. Brain CT/MR in another hospital revealed brain herniation, both sylvian fissure and basal cistern obliteration and bilateral subdural hemorrhage, suggesting intracranial hypotension (Fig. 1A, B and C). Under a diagnosis of intracranial hypotension, emergency EBP at the lumbar level with autologous blood patch was performed. A few hours after EBP, the neurologic status of the patient was gradually improved from GCS 8 to 10. However, the patient still showed drowsy consciousness and limitation in verbal response. Even on the next day, there

E-mail address: leecy009@hanmail.com.

https://doi.org/10.1016/j.inat.2023.101815
Received 16 March 2023; Received in revised form 15 June 2023; Accepted 26 June 2023
Available online 28 June 2023

was no improvement in neurological symptoms, and fluctuation of consciousness was observed. Additionally, brain MRI/A was performed. On radiologic imaging, venous thrombosis was observed in both transverse sinuses and superior sagittal sinus. And the left transverse and sigmoid sinus was not visualized (Fig. 2A, B and C). Under a diagnosis of cerebral venous sinus thrombosis, intra-venous direct thrombectomy with aspiration catheter was performed immediately. Through the right jugular vein, aspiration in both transverse and inion area was performed. Partial recanalization was achieved. However, left transverse-sigmoid sinus occlusion remained. The guiding catheter was moved to the left jugular vein, and several direct aspiration thrombectomy was performed. Multiple blood clots were removed, and finally, both transverse-sigmoid sinus was recanalized successfully (Fig. 3A, B, C, D and E). After direct thrombectomy, systemic heparinization was carried out despite both subdural hemorrahge. On the next day, the patient's neurological condition was further improved (GCS 12). However, the patient was not fully recovered yet. In comparison with the time of admission, the patient showed greater neurological improvement, so we decided to observe while performing conservative treatment. Four days after the first EBP, spine MR myelography was performed, and it showed the remaining CSF leakage, especially at the thoracic level (T9 ~ 10). Epidural blood was observed at several locations below and around the leakage site (Fig. 4). Despite conservative treatment after the first emergency EBP and direct thrombectomy treatment, the patient was not

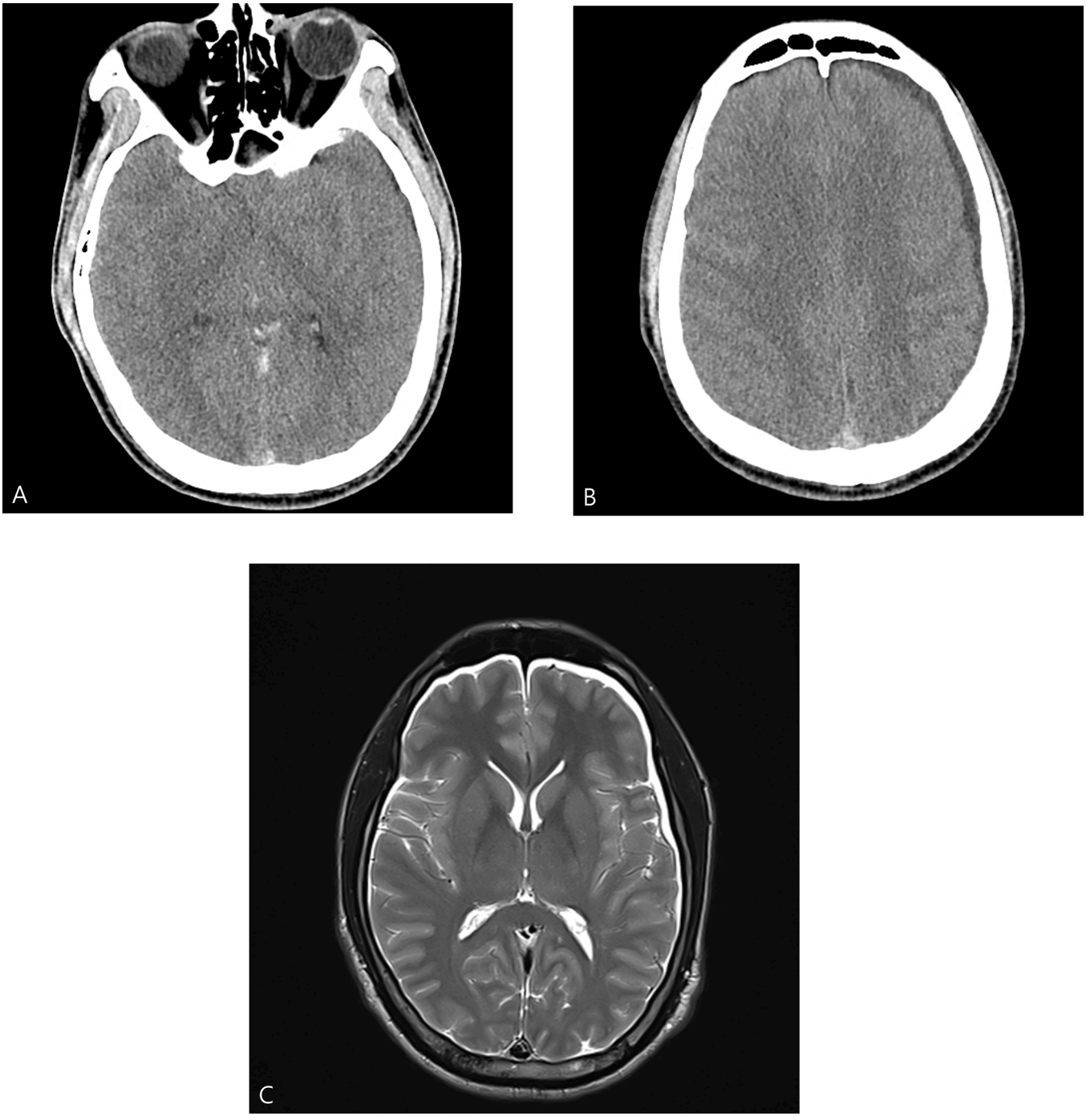

Fig. 1. (A and B) Brain CT/MR revealed brain herniation, both sylvian fissure and basal cistern obliteration, and (C) bilateral subdural hemorrhage, suggesting intracranial hypotension.

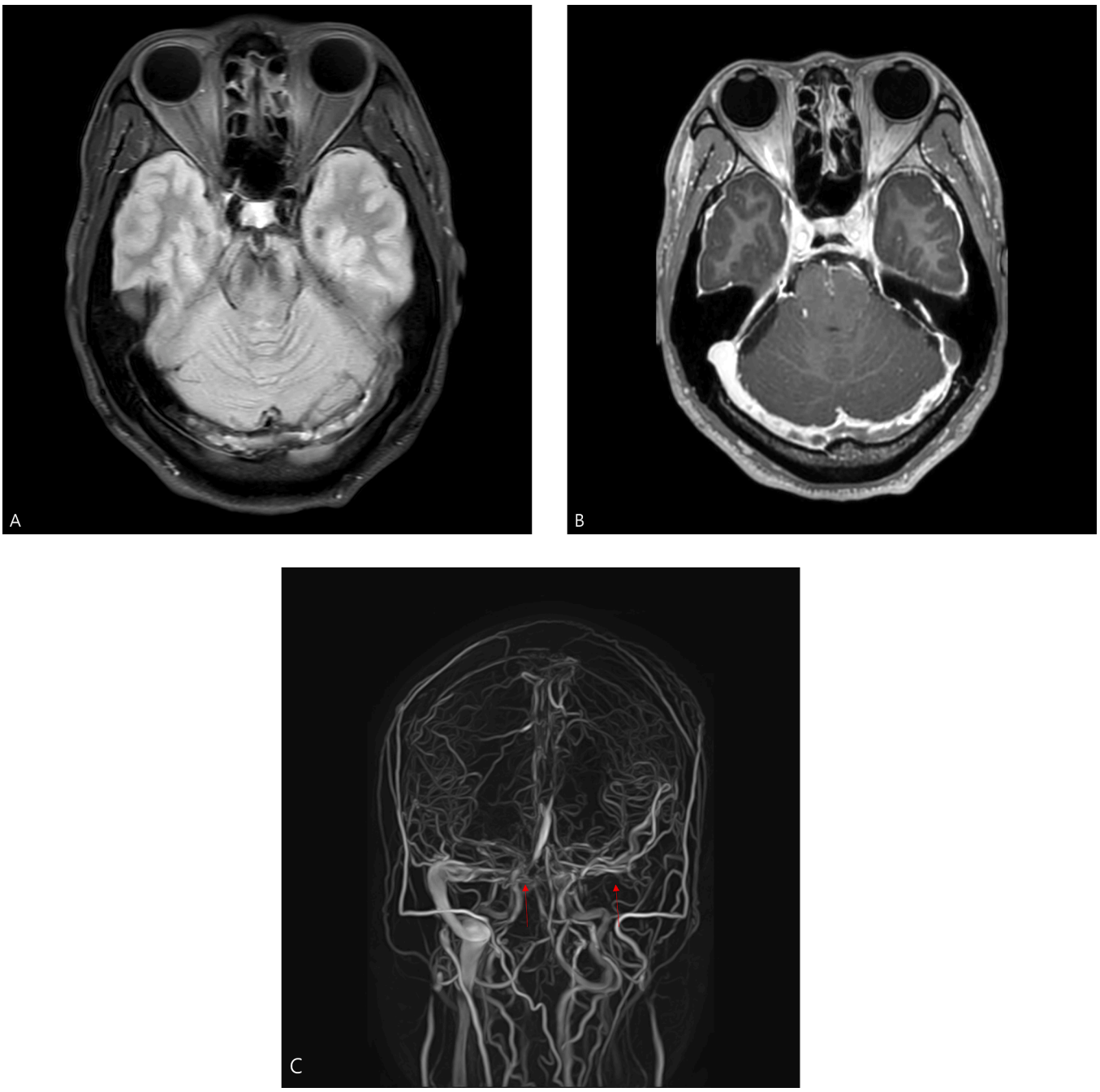

Fig. 2. (A and B) On MRI/A, cerebral venous sinus thrombosis was observed in both transverse sinuses and at the junction with the superior sagittal sinus, and (C) the left transverse and sigmoid sinus was not visualized (red arrow).

fully recovered. We thought that although a large amount of CSF leakage and cerebral venous thrombosis were resolved, a small amount of leakage could impede further neurological improvement. Therefore, additional EBP was performed at the thoracic level. On the next day, the patient was fully recovered (GCS 15) and discharged with no neurologic deficit. Five weeks later, follow-up CT angiography showed improved brain herniation, obliteration cisterns, and decreased subdural hemorrhage despite anticoagulant therapy. The basilar artery, which was bent and descended due to brain herniation on initial MRA, returned to its normal shape and position (Fig. 5A, B, C and D). In the treatment for this patient, the first EBP reduced CSF leakage, and as a result, this alleviated spontaneous intracranial hypotension and neurological symptoms. Second, brain perfusion was improved because cerebral venous thrombosis was resolved by mechanical thrombectomy and this result in further improvement. Third, because residual CSF leakage was stopped completely by second EBP, neurological symptoms were fully resolved. Based on the findings of this case, we believe that patients with severe neurological deterioration associated with spontaneous intracranial hypotension and cerebral venous thrombosis may require immediate treatment for CSF leakage and cerebral venous thrombosis through EBP and mechanical thrombectomy, if possible.

3. Discussion

Spontaneous intracranial hypotension is an uncommon condition caused by spontaneous leakage of CSF. The primary clinical feature is orthostatic headache, which is relieved when lying down, and it is a typical sign of spontaneous intracranial hypotension [1–3]. In addition

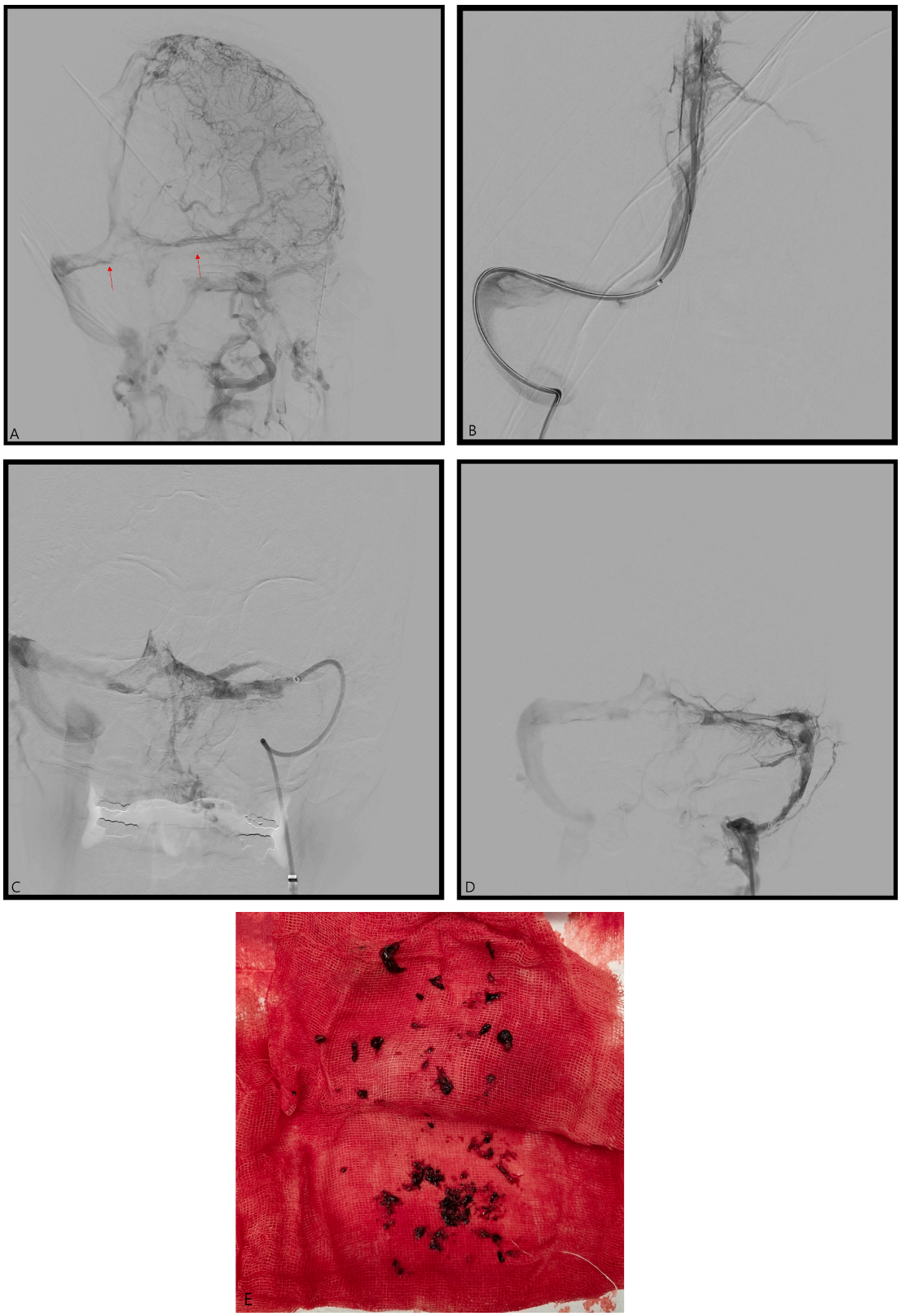

Fig. 3. (A) On conventional cerebral angiography, venous thrombosis was observed in both transverse sinuses and superior sagittal sinus (red arrow), and the left transverse and sigmoid sinus was not visualized. (B) Through the right jugular vein, intravenous direct thrombectomy with an aspiration catheter was performed. After aspiration of both the transverse and junctional areas, partial recanalization was achieved; however, left transverse-sigmoid sinus occlusion was remained. (C-E) The guiding catheter was moved to the left jugular vein, and several direct aspiration thrombectomy was performed. Multiple blood clots were removed, and finally, both transverse-sigmoid sinus was recanalized successfully. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

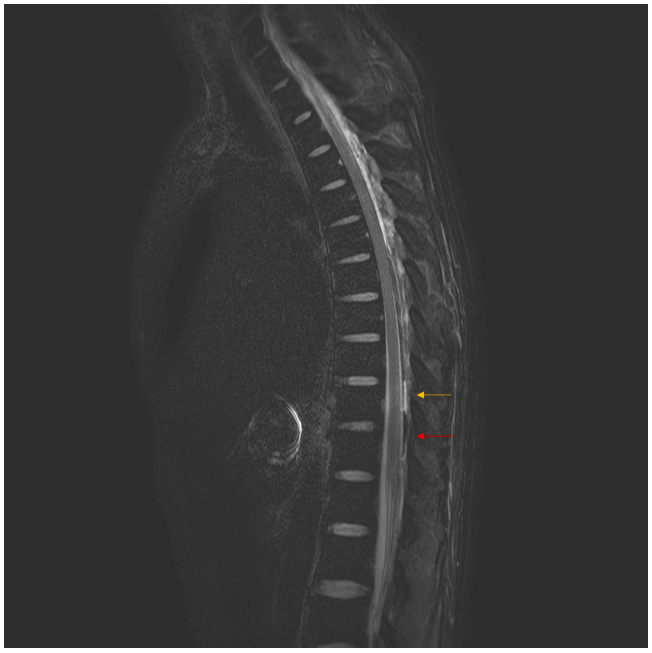

Fig. 4. Spine MR myelography showed the remaining CSF leakage at the thoracic level (yellow arrow, T9 ~ 10). Epidural blood (red arrow) was observed at several locations below and around the leakage site.

to various headache patterns, several associated symptoms have been reported [1–3,8,9]. Brain MRI is known to show findings such as pachymeningeal enhancement, subdural collections that may mimic a primary subdural hematoma, downward displacement of the cerebral tonsils, enlargement and congestion of cerebral venous structures, and pituitary hyperemia [1–3,10]. Despite the presence of some characteristic clinical symptoms and radiographic features, diagnosis is often delayed as these characteristics are not recognized or obscured. Recently, cases of cerebral venous thrombosis have been reported in patients with spontaneous intracranial hypotension. This condition is more difficult to diagnose and may lead to misdiagnosis [10–21]. However, the causal relationship between spontaneous intracranial hypotension and cerebral venous thrombosis has not been clearly established. Moreover, there are no definite guidelines for evaluation and management [22,23].

The association between spontaneous intracranial hypotension and cerebral venous thrombosis was first reported in 2004 [11]. Since then, a small number of cases have been reported as case reports [10–21,24]. Although cerebral venous thrombosis affects only approximately 5 persons per million (0.0005%) in the general population [25], the prevalence increases to 2% among patients with spontaneous intracranial hypotension [4]. In previous reports of spontaneous intracranial hypotension accompanied by cerebral venous thrombosis, it was generally difficult to clearly distinguish the sequence of events based on radiographic findings, because early MRI showed changes in both cerebral venous thrombosis and spontaneous intracranial hypotension [10–21,24]. In three recent reports [4,10,17], the initial MRI only showed changes in spontaneous intracranial hypotension, and cerebral venous thrombosis was diagnosed in subsequent MRI studies. Therefore, it suggested that spontaneous intracranial hypotension precedes cerebral venous thrombosis and may influence the occurrence of cerebral venous thrombosis. Although an association between spontaneous intracranial hypotension and cerebral venous thrombosis was not reported until 2004, the development of cerebral venous thrombosis following iatrogenic spinal CSF leak has been observed since the 1980 s [26–29].

Spontaneous intracranial hypotension may influence the occurrence of cerebral venous thrombosis through one or more mechanisms. First, according to the Monro-Kellie doctrine, the loss of one component must be compensated by the increase of another component in a closed compartment such as the intracranial and spinal subdural space. Therefore, any loss of CSF is replaced by an increase in the most readily expansible component, which is venous blood. Consequently, venous engorgement and congestion result in the slowing of venous blood flow [30]. Second, the downward sagging of intracranial structures due to the loss of CSF buoyancy causes traction on cerebral veins and sinuses, resulting in the mechanical distortion of the vessel wall, which leads to venous flow turbulence or stasis. Third, the loss of CSF decreases CSF absorption into the cerebral venous sinuses, leading to an increase in cerebral venous blood viscosity [1,10,31].

Initial and several subsequent reports of cerebral venous thrombosis associated with spontaneous intracranial hypotension have suggested that a change in headache patterns, e.g., from orthostatic to continuous, may be a characteristic symptom of the development of cerebral venous thrombosis in spontaneous intracranial hypotension [12,16,18,21]. In addition to headache, the clinical manifestations of cerebral venous thrombosis associated with spontaneous intracranial hypotension are generally similar to those of cerebral venous thrombosis, which include venous cerebral infarction, seizure, and the development of dural arteriovenous fistulas [10,13–15,17–20].

Uncertainty exists regarding the treatment of underlying spinal CSF leak in the setting of cerebral venous sinus thrombosis. Until now, the use of anticoagulants has been recommended as a first-line treatment for cerebral venous thrombosis. The current consensus regarding cerebral venous thrombosis treatment with anticoagulation is based on two small randomized studies demonstrating its safety and association with decreased risk of death and disability [32,33]. However, anticoagulation theoretically increases the risk of bleeding and subdural hematoma development, which is a concern among patients with spontaneous intracranial hypotension. Consequently, when anticoagulants are used in these circumstances, the benefit of anticoagulant therapy and the bleeding risk should always be considered, especially when subdural fluid collections are present [34]. The mainstay of spontaneous intracranial hypotension treatment is EBP placement. Dobroky and Farb et al [35,36] reported that method and importance. of searching leakage area. Searching for a CSF leak is not easy, but precisely locating the site of CSF leakage is fundamental to successful treatment, which includes a targeted epidural patch and surgical closure when conservative measures do not provide long-term relief. Increased awareness of spontaneous intracranial hypotension the need for disgnostic therapeutic guidelines. However, uncertainty exists regarding the treatment of the underlying spinal CSF leak in the setting of cerebral venous sinus thrombosis. Moreover, its effect on cerebral venous thrombosis development and recanalization is unknown. However, considering our case, particularly when symptoms of spontaneous intracranial hypotension persist, we suggest that aggressive treatment of the underlying CSF leak should be provided along with anticoagulation as a standard treatment for cerebral venous thrombosis. In addition, we think it is necessary to perform direct thrombectomy for cerebral venous thrombosis when the patient have poor neurologic or consciousness state. The primary treatment focus should be spontaneous intracranial hypotension, because cerebral venous thrombosis development is closely related to spontaneous intracranial hypotension-induced pathophysiological changes in the brain. Therefore, despite maintaining anticoagulant treatment after mechanical thrombectomy, the placement of a second EBP was attempted. There have been case reports of spontaneous intracranial hypotension and cerebral venous thrombosis; however, there are limited reports of patients with severe neurological symptoms such as deterioration of consciousness. In addition, in spontaneous intracranial hypotension patients with severe neurological symptoms, the effect of direct thrombectomy for cerebral venous thrombosis is unclear. Based on the findings of our case, we think that immediate treatment to prevent CSF leakage and improve cerebral circulation may be necessary for severe neurologically impaired patients with

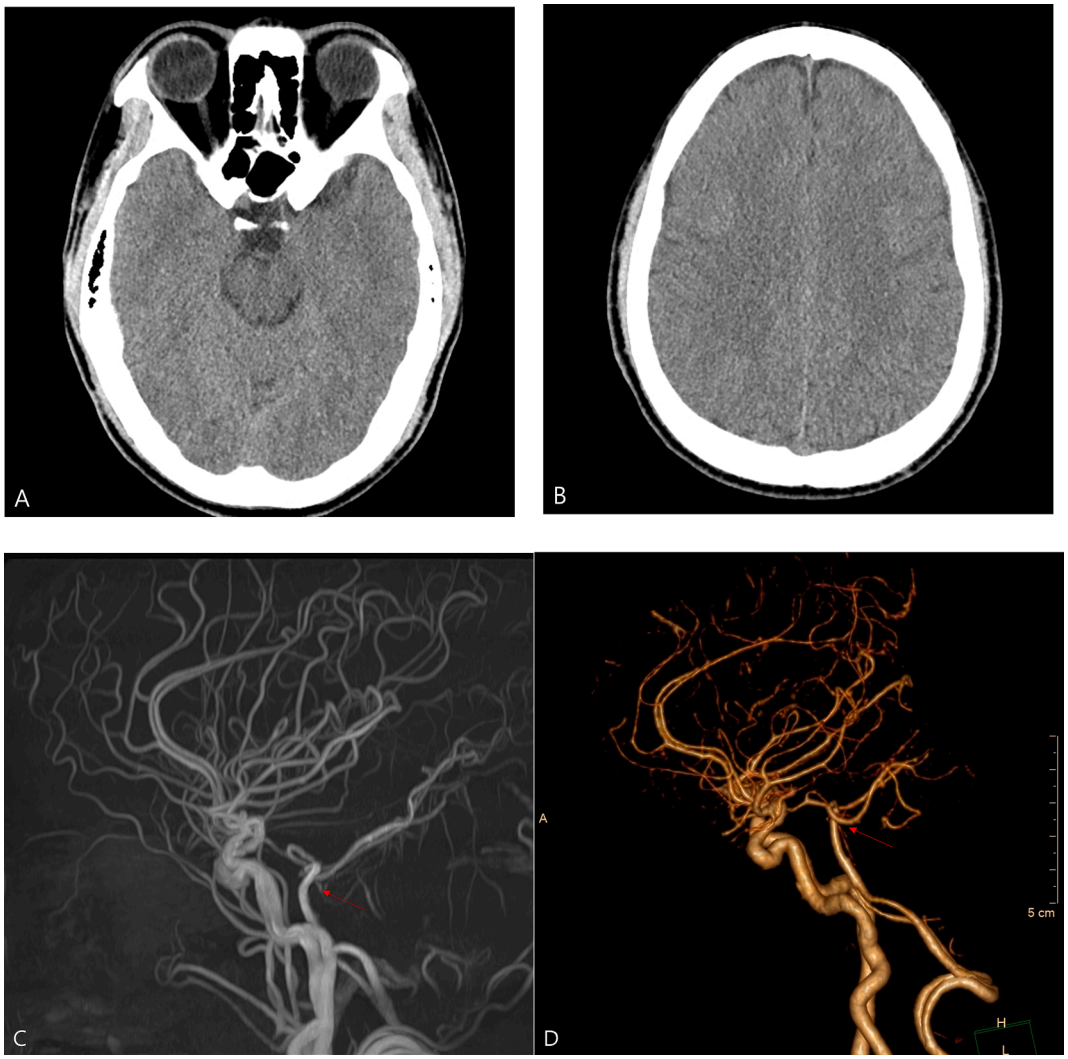

Fig. 5. (A and B) Follow-up CT angiography showed improved brain herniation, obliteration cistern, and decreased subdural hemorrhage despite anticoagulant therapy. (C and D) The basilar artery, which was bent and descended due to brain herniation on initial MRA, returned to its normal shape and position (red arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

spontaneous intracranial hypotension and cerebral venous thrombosis. We believe that only when brain sagging and perfusion are improved with EBP and direct recanalization, the patients' neurological status can be improved, and sequelae can be minimized. Both spontaneous intracranial hypotension and cerebral venous thrombosis are frequently misdiagnosed, resulting in treatment delay and increased risk of complications. In this case report, we highlighted the importance of prompt diagnosis and aggressive treatment.

4. Conclusion

In the treatment of spontaneous intracranial hypotension and cerebral venous thrombosis, EBP and anticoagulant therapy have been mainly performed. However, the priority and modality of treatment in cases of concurrent spontaneous intracranial hypotension with cerebral venous thrombosis have not been well known. In addition, anticoagulant therapy alone for cerebral venous thrombosis may not be tolerable to severe patients. For neurologically severe patients, it may be crucial to improve brain herniation and cerebral perfusion as soon as possible. If necessary, immediate EBP and direct thrombectomy should be performed to improve the patients' neurological status, and has to be followed by maintenance of anticoagulant therapy and accurate evaluation of the CSF leak site.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Acknowledgements

This research was supported by the Chung-Ang University Research Grants in 2023.

References

[1] W.I. Schievink, Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension, J. Am. Med. Assoc. 295 (19) (2006) 2286–2296.
[2] B. Mokri, Low cerebrospinal fluid pressure syndromes, Neurol. Clin. 22 (1) (2004) 55–74.
[3] T.J. Schwedt, D.W. Dodick, Spontaneous intracranial hypotension, Curr. Pain Headache Rep. 11 (1) (2007) 56–61.
[4] W.I. Schievink, M.M. Maya, Cerebral venous thrombosis in spontaneous intracranial hypotension, Headache 48 (10) (2008) 1511–1519.
[5] J.M. Ferro, M. Correia, M.J. Rosas, A.N. Pinto, G. Neves, Seizures in cerebral vein and dural sinus thrombosis, Cerebrovasc. Dis. 15 (1-2) (2003) 78–83.
[6] F. Masuhr, M. Busch, N. Amberger, H. Ortwein, M. Weih, K. Neumann, K. Einhaupl, S. Mehraein, Risk and predictors of early epileptic seizures in acute cerebral venous and sinus thrombosis, Eur. J. Neurol. 13 (8) (2006) 852–856.

[7] J.M. Ferro, Patrícia Canhão, M.-G. Bousser, J. Stam, F. Barinagarrementeria, Early seizures in cerebral vein and dural sinus thrombosis: risk factors and role of antiepileptics, Stroke 39 (4) (2008) 1152–1158.
[8] B. Mokri, Spontaneous low pressure, low CSF volume headaches: spontaneous CSF leaks, Headache 53 (7) (2013) 1034–1053.
[9] L. Bassani, et al., Noninvasive diagnosis and management of spontaneous intracranial hypotension in patients with marfan syndrome: Case Report and Review of the Literature, Surg. Neurol. Int. 5 (2014) 8.
[10] A. Haritanti, D. Karacostas, A. Drevelengas, V. Kanellopoulos, E. Paraskevopoulou, A. Lefkopoulos, I. Economou, A.S. Dimitriadis, Spontaneous intracranial hypotension: clinical and neuroimaging findings in six cases with literature review, Eur. J. Radiol. 69 (2) (2009) 253–259.
[11] D. Sopelana, et al., May intracranial hypotension be a cause of venous sinus thrombosis? Eur. Neurol. 51 (2) (2004) 113–115.
[12] K.D. Flemming, M.J. Link, Spontaneous CSF leak complicated by venous thrombosis and dural arteriovenous fistula, Cephalalgia 25 (9) (2005) 751–753.
[13] M. Savoiardo, S. Armenise, P. Spagnolo, T. De Simone, M.L. Mandelli, A. Marcone, G. Morciano, C. Andreula, E. Mea, M. Leone, L. Chiapparini, Dural sinus thrombosis in spontaneous intracranial hypotension: Hypotheses on possible mechanisms, J. Neurol. 253 (9) (2006) 1197–1202.
[14] P.-H. Lai, J.-Y. Li, Y.-K. Lo, M.-T. Wu, H.-L. Liang, C.-K. Chen, A case of spontaneous intracranial hypotension complicated by isolated cortical vein thrombosis and cerebral venous infarction, Cephalalgia 27 (1) (2007) 87–90.
[15] H. Kataoka, E. Tanizawa, S. Ueno, Spontaneous intracranial hypotension is associated with a risk of venous sinus thrombosis and subdural hematoma, Cerebrovasc. Dis. 23 (4) (2007) 315–317.
[16] S. Albayram, K.M. Tasmali, A. Gunduz, Can spontaneous intracranial hypotension cause venous sinus thrombosis? J. Headache Pain 8 (3) (2007) 200–201.
[17] M.Y. Lan, Y.Y. Chang, J.S. Liu, Delayed cerebral venous thrombosis in a patient with spontaneous intracranial hypotension, Cephalalgia 27 (10) (2007) 1176–1178.
[18] S. Richard, S. Kremer, J.-C. Lacour, H. Vespignani, P. Boyer, X. Ducrocq, Cerebral venous thrombosis caused by spontaneous intracranial hypotension: two cases, Eur. J. Neurol. 14 (11) (2007) 1296–1298.
[19] Y.-F. Wang, J.-L. Fuh, J.-F. Lirng, F.-C. Chang, S.-J. Wang, Spontaneous intracranial hypotension with isolated cortical vein thrombosis and subarachnoid haemorrhage, Cephalalgia 27 (12) (2007) 1413–1417.
[20] S. Takeuchi, et al., Spontaneous intracranial hypotension associated with dural sinus thrombosis, Neurol. Med. Chir. (Tokyo) 47 (12) (2007) 555–558.
[21] K. Tan, N. Venketasubramanian, C.Y. Hwang, C.C.T. Lim, My headache does not get better when I lie down: spontaneous intracranial hypotension complicated by venous thrombosis, Headache 48 (1) (2008) 149–152.
[22] W.I. Schievink, Misdiagnosis of spontaneous intracranial hypotension, Arch. Neurol. 60 (12) (2003) 1713–1718.
[23] W.I. Schievink, M.M. Maya, F. Moser, J. Tourje, S. Torbati, Frequency of spontaneous intracranial hypotension in the emergency department, J. Headache Pain 8 (6) (2007) 325–328.
[24] A. Yamamoto, Y. Hattammaru, S. Uezono, Spontaneous intracranial hypotension associated with cerebral venous thrombosis detected by a sudden seizure: a case report, JA Clin. Rep. 6 (1) (2020) 59.
[25] J. Stam, Thrombosis of the cerebral veins and sinuses, N. Engl. J. Med. 352 (17) (2005) 1791–1798.
[26] J. Schou, M. Scherb, Postoperative sagittal sinus thrombosis after spinal anesthesia, Anesth. Analg. 65 (5) (1986) 541–542.
[27] C.H. Hubbert, Dural puncture headache suspected, cortical vein thrombosis diagnosed, Anesth. Analg. 66 (3) (1987) 285.
[28] R.W. Hartley, A. Jackson, R.S. Cooke, Dural venous sinus thrombosis following myelography: demonstration by magnetic resonance imaging, Br. J. Radiol. 65 (780) (1992) 1134–1136.
[29] E. Wilder-Smith, I. Kothbauer-Margreiter, B. Lammle, M. Sturzenegger, C. Ozdoba, S.P. Hauser, Dural puncture and activated protein C resistance: risk factors for cerebral venous sinus thrombosis, J. Neurol. Neurosurg. Psychiatry 63 (3) (1997) 351–356.
[30] B. Mokri, The Monro-Kellie hypothesis: applications in CSF volume depletion, Neurology 56 (12) (2001) 1746–1748.
[31] J.H. Park, S.H. Yoon, New concept of cerebrospinal fluid dynamics in cerebral venous sinus thrombosis, Med. Hypotheses 70 (1) (2008) 143–147.
[32] K.M. Einhäupl, A. Villringer, S. Mehraein, C. Garner, M. Pellkofer, R.L. Haberl, H.-W. Pfister, P. Schmiedek, W. Meister, Heparin treatment in sinus venous thrombosis, Lancet 338 (8767) (1991) 597–600.
[33] S.F. de Bruijn, J. Stam, Randomized, placebo-controlled trial of anticoagulant treatment with low-molecular-weight heparin for cerebral sinus thrombosis, Stroke 30 (3) (1999) 484–488.
[34] Y.T. Mao, Q. Dong, J.H. Fu, Delayed subdural hematoma and cerebral venous thrombosis in a patient with spontaneous intracranial hypotension, Neurol. Sci. 32 (5) (2011) 981–983.
[35] T. Dobrocky, P. Nicholson, L. Häni, P. Mordasini, T. Krings, W. Brinjikji, J. K. Cutsforth-Gregory, R. Schär, C. Schankin, J. Gralla, V.M. Pereira, A. Raabe, R. Farb, J. Beck, E.I. Piechowiak, Spontaneous intracranial hypotension: searching for the CSF leak, Lancet Neurol. 21 (4) (2022) 369–380.
[36] R.I. Farb, et al., Spontaneous Intracranial Hypotension: A Systematic Imaging Approach for CSF Leak Localization and Management Based on MRI and Digital Subtraction Myelography, AJNR Am. J. Neuroradiol., 40 (4) (2019) 745–753.