## Case Studies

# Report of Dramatic Improvement after a Lumboperitoneal Shunt Procedure in a Case of Anticoagulation Therapy-Resistant Cerebral Venous Thrombosis

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Cerebral venous thrombosis (CVT), which typically progresses from either acute or subacute onset, presents with symptoms related to intracranial hypertension (e.g., headache and papilledema) and brain parenchymal lesions (e.g., aphasia and hemiplegia). Anticoagulation therapy is generally accepted as a treatment for CVT and often leads to good clinical outcomes. However, we experienced a case of CVT with an uncommon clinical course. The patient was a 63-year-old man who presented with headache, papilledema, visual loss, and diplopia; his condition gradually deteriorated, and he was diagnosed with CVT via cerebral angiography. The sinus thrombus was extensive and resistant to anticoagulation therapy, and lumbar puncture revealed a progressive increase in cerebrospinal fluid (CSF) pressure. We performed a lumboperitoneal (LP) shunt procedure, which yielded marked improvement in the symptoms. The main mechanism of neurological dysfunction in CVT is venous outflow obstruction caused by venous thrombus, which results in brain edema, and/or venous infarction, which induces focal neurological signs. Another mechanism is impaired CSF absorption in the thrombosed sinuses, resulting in intracranial hypertension. We speculated that the latter mechanism strongly influenced our case, thus explaining the uncommon clinical course and effectiveness of the LP shunt procedure. Although LP shunting is not a common treatment for CVT, this case report could indicate the usefulness of this procedure for CVT with chronic progression and resistance to anticoagulation therapy. Key Words: Cerebral venous thrombosis-extensive thrombus-impaired cerebrospinal fluid absorption-resistance to anticoagulation-lumboperitoneal shunt.

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### Introduction

Cerebral venous thrombosis (CVT) is a rare type of stroke (.5%-1.0% of all strokes).<sup>1</sup> CVT often exhibits an acute or subacute onset with various symptoms. Ferro et al<sup>2</sup> re-

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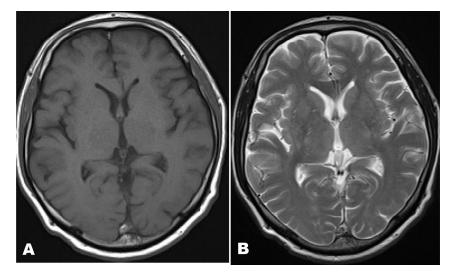
ported that more than 90% of CVT cases exhibited acute or subacute onset, with less than 10% of cases featuring chronic onset. The clinical manifestations of CVT may include symptoms related to intracranial hypertension, such as headache and papilledema, or focal neurological symptoms, such as aphasia, hemiplegia, and seizure, caused by brain parenchymal lesions (e.g., venous infarction and/or hemorrhagic infarction).<sup>3</sup> To date, various treatments for CVT have been reported; of these, anticoagulation is a therapeutic mainstay, and its usefulness has been supported by 2 previous randomized controlled trials.<sup>4,5</sup> Ferro et al<sup>2</sup> indicated the use of anticoagulation therapy in most cases of CVT and reported a relatively good prognosis with approximately

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**Figure 1.** T1-weighted magnetic resonance image (A) and T2-weighted image (B). These images indicated signal intensity alternation in the posterior aspect of the superior sagittal sinus (A and B).

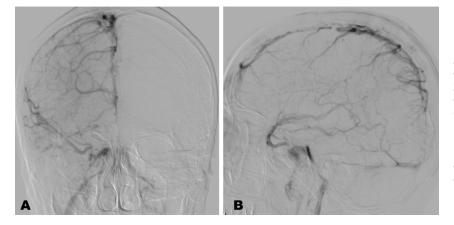
80% of patients achieving a complete recovery. More recently, endovascular treatments for CVT have been frequently reported. Some case series have indicated direct catheter thrombolysis and mechanical thrombectomy as effective therapies for CVT. Siddiqui et al<sup>6-9</sup> reported that more than 70% of CVT cases achieved near to complete recanalization, and more than 80% achieved a good outcome (modified Rankin Scale score 0-2). Notably, no relevant large prospective studies have been conducted. However, the American Heart Association guideline recommends considering endovascular intervention if deterioration occurs despite intensive anticoagulation treatment.<sup>10</sup>

Herein, we describe our experience with a case of chronically progressing CVT that exhibited resistance to anticoagulation therapy. As endovascular treatment was not available, we selected a shunt procedure, an uncommon treatment for CVT that was effective in our case and led to a good clinical outcome.

#### Case Report

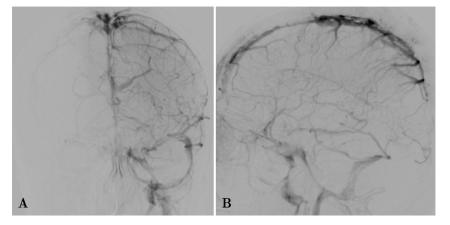
A 63-year-old man with a history of diabetes mellitus, hypertension, and chronic otitis media was admitted to the department of internal medicine in our hospital with complaints of headache, nausea, and mild dizziness of 5 days' duration. He did not present with paralysis or disturbed consciousness. Plain computed tomography (CT) did not indicate any abnormal signs, but magnetic resonance imaging (MRI) demonstrated altered signal intensity in the superior sagittal sinus (Fig 1). This finding suggested a CVT, and the patient was subsequently referred to our department 7 days after symptom onset.

Cerebral angiography revealed a long segment thrombus from the superior sagittal sinus to both transverse sinuses. Collateral venous drainage, however, was relatively abundant, with principal drainage via the right sylvian vein into the jugular vein and the left vein of Labbe into the sigmoid sinus (Figs 2, 3). Although



**Figure 2.** Venous-phase right carotid angiography (anteroposterior view: A, lateral view: B) shows a filling defect in the posterior aspect of the superior sagittal sinus, the right transverse sinus, and the right sigmoid sinus, which caused a slight delay in the visualization of cerebral venous structures (not shown). The flow from the right sylvian veins to the cavernous sinuses and into the jugular veins is the main collateral venous drainage route and is relatively enlarged because of the large amount of venous drainage.

**Figure 3.** Venous-phase left carotid angiography (anteroposterior view: A, lateral view: B) shows the failure to visualize the sinus from the posterior part of the superior sagittal sinus to the left transverse sinus. The main collateral venous drainages are the flow from the left sylvian vein and the vein of Labbe into the left jugular vein.



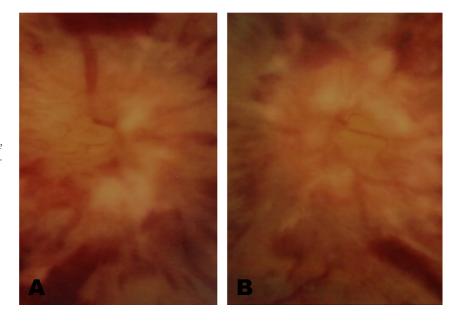
endovascular therapy was considered as a treatment option, it was deemed inappropriate because of the extensive thrombus and was therefore not performed.

Although these findings indicated a definitive diagnosis of CVT, the patient's symptoms were not severe and not specific for CVT with an extensive and long thrombus. Given the uncertain association of these symptoms with CVT, prophylactic anticoagulation therapy with oral warfarin, rather than intravenous heparin, was initiated 8 days after symptom onset to avoid an increase in thrombus size. The patient was discharged from our hospital 15 days after symptom onset, once his condition had stabilized with the intent to conduct outpatient follow-ups.

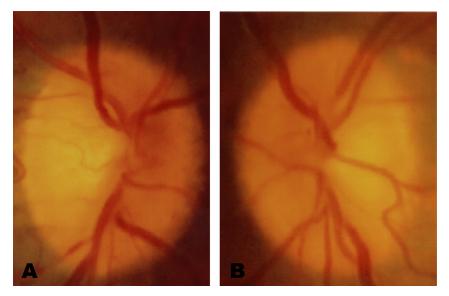
Thirty days after discharge, the patient was readmitted to our hospital with complaints of vision loss (right vision = .1, left vision = .2) and diplopia. A physical examination revealed bilateral papilledema (Fig 4) and bilateral abducens paresis. At that time, the lumbar puncture opening pressure had increased to  $28 \text{ cmH}_2\text{O}$ , which corresponded with symptom deterioration. Although the prothrombin time–international normalized ratio was within the optimal range at 2.9, cerebral angiography revealed no change in the venous thrombus despite the continued administration of warfarin at a daily 3-mg dose. Intravenous glycerin and oral acetazolamide (1000 mg/ day) administration were initiated. Despite these treatments, the symptoms remained and gradually deteriorated, and 55 days after symptom onset, the lumbar puncture opening pressure increased further to 37 cmH<sub>2</sub>O.

Sixty days after symptom onset, a lumboperitoneal (LP) shunt procedure was performed to decrease the intracranial pressure. Oral warfarin therapy was suspended 3 days before this procedure. The shunt pressure was gradually adjusted to a final value of 14 cmH<sub>2</sub>O.

The patient's headache and nausea disappeared immediately after LP shunting, and considerable improvement in his papilledema was observed approximately 1 month after LP shunting (Fig 5). The patient's visual acuity also improved gradually as his papilledema improved (right vision = .3, left vision = .4); his oculomotor nerve palsy



**Figure 4.** Right eye (A) and left eye (B). Severe papilledema is visible before LP shunting. Abbreviation: LP, lumboperitoneal.



**Figure 5.** Right eye (A) and left eye (B). Papilledema is dramatically ameliorated after LP shunting. Abbreviation: LP, lumboperitoneal.

also improved, and diplopia resolved completely. Anticoagulation therapy was not resumed. One year after the LP shunt procedure, follow-up angiography indicated no change in the venous thrombosis and continued occlusion in the previously affected sinuses. However, the patient did not experience recurrence of symptoms.

#### Discussion

CVT, unlike arterial stroke and intracranial hemorrhage, occurs mainly in young women. The risk factors for CVT in young women include oral contraceptive use, pregnancy, and puerperal status. However, the patient in our case was an elderly man with a history of chronic otitis media and related hearing loss. The cause of CVT was therefore presumed to be a parameningeal infection related to otitis media, which has been reported as a common risk factor for CVT.<sup>2,11</sup>

The case described in our present report was suggestive of chronic onset and exhibited slow progression despite anticoagulation therapy. The initial symptoms of headache and nausea gradually deteriorated into papilledema, vision loss, and diplopia, all of which suggested elevated intracranial pressure. No focal neurological signs related to brain parenchymal lesions were evident.

The main mechanism of neurological dysfunction in CVT is thought to be impaired venous drainage resulting from a venous thrombus that leads to tissue congestion, which may cause brain parenchymal lesions.<sup>1,12</sup> These lesions are recognized as brain edema, venous infarction, or hemorrhagic infarction via neuroimaging, and can induce various focal neurological signs. Ferro et al<sup>2</sup> reported that more than 60% of patients with CVT exhibited parenchymal lesions on CT/MRI.

However, in our case, altered signal intensity in the sinuses on MRI provided the only evidence of a parenchymal lesion, and the patient's symptoms were principally induced by intracranial hypertension. Therefore, another pathological mechanism must be considered. The present case was ultimately presumed to be a failure of cerebrospinal fluid (CSF) absorption, an arachnoid granulationrelated malfunction wherein the thrombus may block the outflow of CSF. A deterioration in CSF absorption would provoke intracranial hypertension.<sup>1,12</sup> Additionally, an extensive thrombus may induce such a malfunction over a wide area of arachnoid granulations and could thus easily elevate the intracranial pressure.

The patient in our case featured relatively welldeveloped collateral venous drainage, as indicated by cerebral angiography, enabling relief of the venous outflow obstruction. This feature might have reduced the effect of the primary pathogenic mechanism on our case. On the other hand, the thrombus, which was too extensive to treat via endovascular methods, might have caused the deterioration in CSF absorption; consequently, the other mechanism described above might have strongly influenced our case. Accordingly, we speculated that the principal pathogenesis of the neurological dysfunction observed in our case was a failure of CSF absorption, which could explain both the unusual progression and the effectiveness of the LP shunt procedure.

Although CT/MRI did not indicate ventriculomegaly in our case, previous reports that suggested the safety and effectiveness of LP shunting for idiopathic intracranial hypertension led us to believe that this procedure would successfully control the CSF outflow.<sup>13,14</sup> Shunt procedures are rarely used to treat CVT.<sup>15</sup> Although Ferro and Canhao<sup>11</sup> suggested repeated lumbar punctures or an LP shunt as potential treatment for intracranial hypertension that remains unresolved by other therapies, a review of the literature revealed few previous case reports involving LP shunting for CVT, except for a report by Hokari et al.<sup>16</sup> The present case report might therefore support the validity of shunting procedures for the treatment of CVT, particularly in cases involving chronic progression and resistance to anticoagulation therapy.

#### References

- Stam J. Thrombosis of the cerebral veins and sinuses. N Engl J Med 2005;352:1791-1798.
- 2. Ferro JM, Canhao P, Stam J, et al. Prognosis of cerebral vein and dural sinus thrombosis: results of the International Study on Cerebral Vein and Dural Sinus Thrombosis (ISCVT). Stroke 2004;35:664-670.
- Bousser MG, Chiras J, Bories J, et al. Cerebral venous thrombosis—a review of 38 cases. Stroke 1985;16:199-213.
- 4. de Bruijn SF, Stam J. Randomized, placebo-controlled trial of anticoagulant treatment with low-molecular-weight heparin for cerebral sinus thrombosis. Stroke 1999;30:484-488.
- 5. Einhaupl KM, Villringer A, Meister W, et al. Heparin treatment in sinus venous thrombosis. Lancet 1991;338:597-600.
- Mokin M, Lopes DK, Binning MJ, et al. Endovascular treatment of cerebral venous thrombosis: contemporary multicenter experience. Interv Neuroradiol 2015;21:520-526.
- Canhao P, Falcao F, Ferro JM. Thrombolytics for cerebral sinus thrombosis: a systematic review. Cerebrovasc Dis 2003;15:159-166.
- 8. Stam J, Majoie CB, van Delden OM, et al. Endovascular thrombectomy and thrombolysis for severe cerebral

sinus thrombosis: a prospective study. Stroke 2008;39:1487-1490.

- 9. Siddiqui FM, Dandapat S, Banerjee C, et al. Mechanical thrombectomy in cerebral venous thrombosis: systematic review of 185 cases. Stroke 2015;46:1263-1268.
- 10. Saposnik G, Barinagarrementeria F, Brown Jr RD, et al. Diagnosis and management of cerebral venous thrombosis: a statement for healthcare professionals from the American Heart Association/American Stroke Association. Stroke 2011;42:1158-1192.
- 11. Ferro JM, Canhao P. Cerebral venous sinus thrombosis: update on diagnosis and management. Curr Cardiol Rep 2014;16:523.
- Wasay M, Bakshi R, Bobustuc G, et al. Cerebral venous thrombosis: analysis of a multicenter cohort from the United States. J Stroke Cerebrovasc Dis 2008;17: 49-54.
- Friedman DI. Pseudotumor cerebri. Neurol Clin 2004;22:99-131, vi.
- Yadav YR, Parihar V, Agarwal M, et al. Lumbar peritoneal shunt in idiopathic intracranial hypertension. Turk Neurosurg 2012;22:21-26.
- Einhaupl K, Stam J, Bousser MG, et al. EFNS guideline on the treatment of cerebral venous and sinus thrombosis in adult patients. Eur J Neurol 2010;17:1229-1235.
- 16. Hokari M, Kuroda S, Ishikawa T, et al. Improvement of cerebral hemodynamic and metabolic parameters in a patient who presented intracranial hypertension due to superior sinus thrombosis after lumbo-peritoneal shunt: case report. No Shinkei Geka 2005;33:281-286.