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Case Report

A Case of Idiopathic Intracranial Hypertension Treated by Transverse Sinus Stenting

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Objective: We report a patient with chronic headache due to idiopathic intracranial hypertension (IIH) associated with transverse sinus (TS) stenosis. The symptom improved after stent placement at the site of stenosis.

Case Presentation: The patient was a 37-year-old woman with progressive headache and diplopia as chief complaints. She had severe bilateral papilledema. Magnetic resonance imaging (MRI) and angiography revealed stenosis of the bilateral TS. Lumbar puncture demonstrated raised intracranial pressure and IIH was tentatively diagnosed. Visual impairment progressed despite oral acetazolamide therapy. A venous pressure gradient was monitored during stent placement. The pressure gradient improved after stenting. Dual antiplatelet therapy was initiated 1 week before the procedure. Papilledema and headache resolved immediately after the procedure. No in-stent stenosis or occlusion occurred during the follow-up period.

Conclusion: Stent placement for TS stenosis can improve the cerebral venous return in IIH patients. Although restenosis is possible, venous sinus stenting is considered an effective treatment.

Keywords ▶ intracranial hypertension, transverse sinus, stenting, papilledema

Introduction

Idiopathic intracranial hypertension (IIH) was first reported as "meningitis serosa" by Quincke in 1893. Thereafter, this disease was termed "pseudotumor cerebri."[1)] It is characterized by signs and symptoms associated with intracranial hypertension, such as headache diplopia, papilledema, and elevated cerebrospinal fluid pressure. The prognosis of patients with this disease was previously considered favorable, but many patients with permanent loss of vision have recently been reported, highlighting the importance of early detection and treatment.[2)] The pathophysiology of IIH remains unclear, but previous studies[2,3)] reported association with impaired cerebral venous return due to stenosis of transverse sinus (TS). Recent studies suggested that endovascular intervention, including stenting, is useful for improving venous return in IIH patients with stenosis of the TS.[4–12)] However, in Japan, neither the disease nor its pathogenesis is well understood, and few studies have reported on endovascular treatment for IIH.

In this study, we report a patient with headache, visual disorder, and papilledema due to IIH with the right TS stenosis and the left TS occlusion, successfully treated with right TS stenting.

Case Presentation

A 37-year-old woman referred to our hospital because of headache, dizziness, blurred vision and diplopia. Two years prior to the initial presentation, she developed headache and it became chronic. Four months prior to the presentation, she developed dizziness and vomiting, and they exacerbated

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upon waking over the past 2 months. She developed blurred vision, transient visual obscurity, and diplopia. She consulted a local ophthalmology clinic. Bilateral papilledema was detected, and she was referred to our hospital. As her past medical history, minimal change nephrotic syndrome developed at age of 22, and corticosteroid therapy led to remission. At the age of 34, a benign left ovarian tumor was resected.

Ophthalmological examination disclosed impaired abduction in the left eye and marked papilledema in both eyes (**Fig. 1A**), suggesting increased intracranial pressure. Lumbar puncture revealed an increased cerebrospinal fluid pressure of 40 cmH_2O. Magnetic resonance venography (MRV) suggested stenosis of the right TS and occlusion of the left TS (**Fig. 2A**). There were no other significant findings on brain magnetic resonance imaging (MRI) such as cerebral parenchymal/ventricular enlargement. Therefore, a diagnosis of IIH was made. Treatment using oral acetazolamide was started, but headache and papilledema exacerbated, blurred vision of the left eye was progressive. Cerebral angiography (right internal carotid angiography) showed occlusion of the left TS and severe stenosis of the right TS. Venous return from the superior sagittal sinus (SSS) and straight sinus stagnated at the stenotic site of the right TS. There was a marked delay in the venous phase involving the entire cerebrum (**Fig. 2B**). In the venous phase on left internal carotid angiography, the Labbe vein to the left sigmoid sinus was visualized. Left vertebral angiography revealed that the venous return of posterior fossa was not mediated by the TS and draining to the other connections such as the pterygoid plexus. Angiography suggested that intracranial hypertension and delay of the venous return arise from occlusion of the left TS and severe stenosis of the right TS. The dose of acetazolamide was increased to 1000 mg and headache slightly reduced. However, there was no improvement in papilledema. Furthermore, visual impairment progressed, and the patient was admitted to our department for endovascular treatment to prevent loss of the vision in both eyes. On admission, the height and body weight were 151 cm and 51.8 kg, respectively. Her consciousness was alert. Pulsatile headache and transient visual obscurity in the bilateral eyes, impaired abduction of the left eye were present. There were no other abnormal neurological findings including anisocoria and meningeal irritation sign. In the laboratory tests, the blood count, blood biochemistry, coagulation and fibrinolytic system data including protein S antigen/activity, protein C antigen/activity, autoantibodies including anti-phospholipid antibody, pituitary/thyroid/adrenal hormones, and vitamins were normal. Brain MRI revealed no abnormalities in the parenchyma and the ventricles.

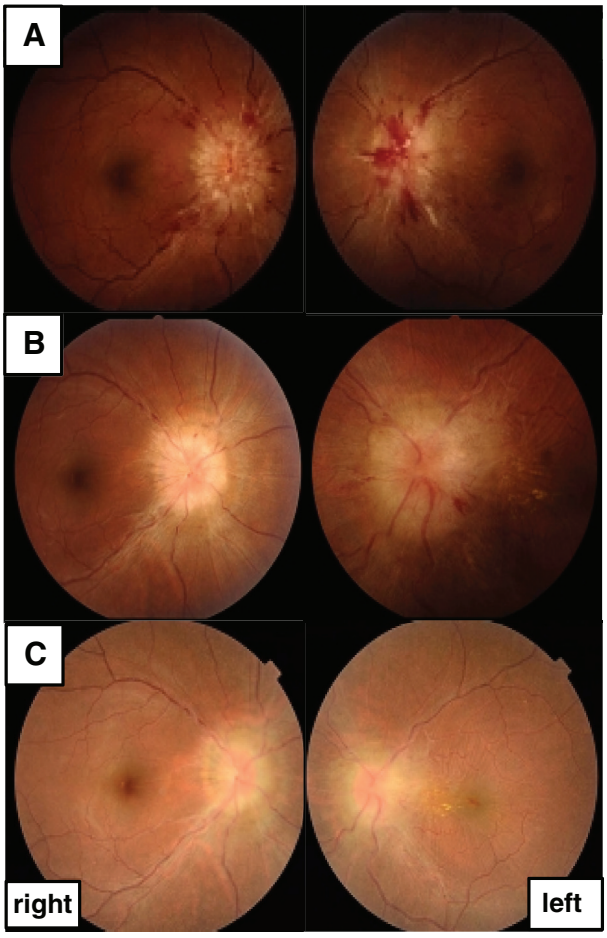

Fig. 1 Photograph of the ocular fundus reveals severe papilledema. (**A**) Postoperative image of the ocular fundus at 7 days reveals marked improvement of papilledema. (**B**) Two-month follow-up photograph of the ocular fundus reveals the disappearance of papilledema.

Prior to endovascular treatment, the off-label use of a stent was approved by the ethics review board of our hospital. Dual antiplatelet therapy (DAPT) using aspirin and clopidogrel was started 1 week before procedure. Under general anesthesia, a 7-Fr Shuttle guiding sheath (Cook Medica, Bloomington, USA) was inserted into the right femoral vein and a 5-Fr sheath introducer was inserted into the left femoral artery. Subsequently, 5000 units of heparin was administered for systemic heparinization. A 5-Fr catheter for angiography was inserted into the right internal carotid artery, and the stenotic site of the right TS was evaluated using digital subtraction angiography (DSA) and three-dimensional DSA (3D-DSA). As a venous-side system, we used a coaxial system consisting of a 7-Fr Shuttle

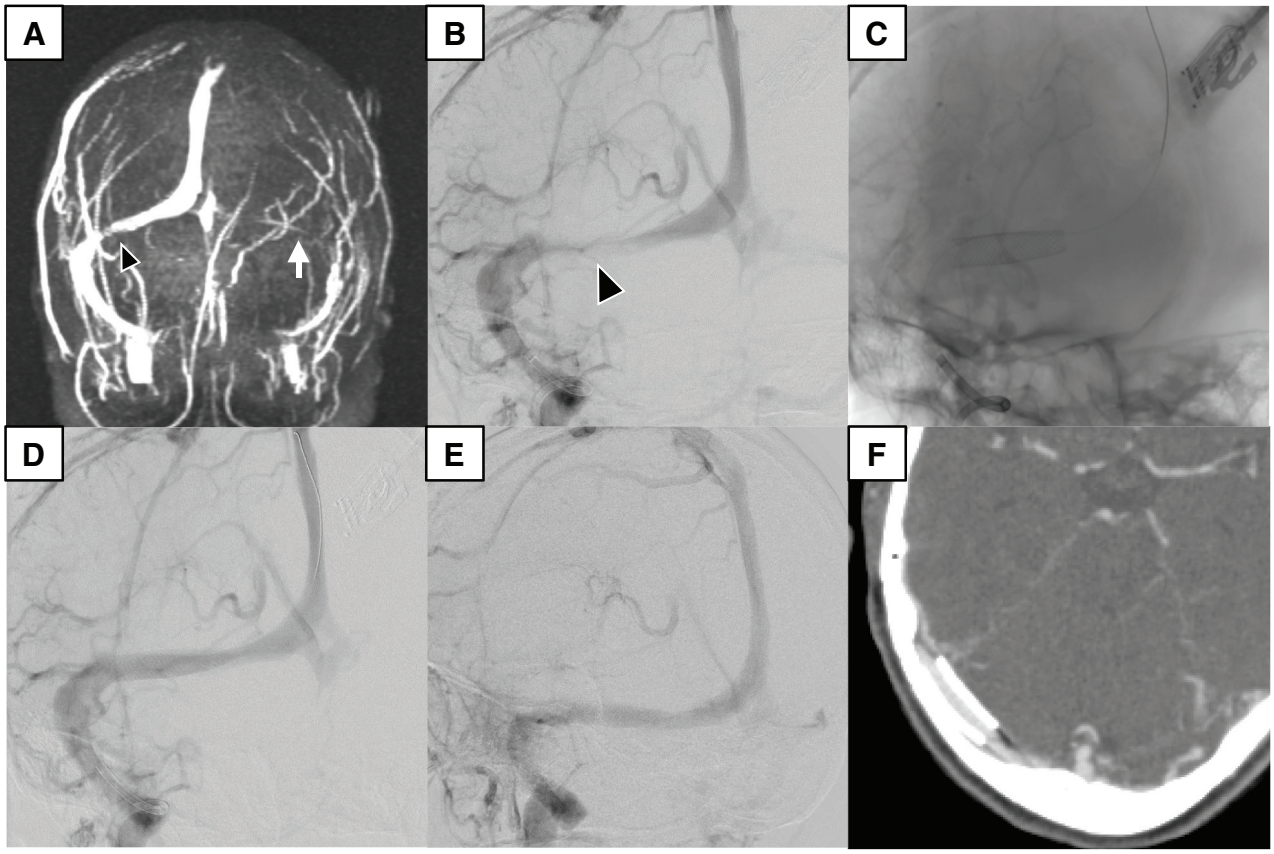

Fig. 2 (**A**) MRV demonstrates stenosis of the right TS (TS: ▲arrowhead) and occlusion of the left TS (↑arrow) (**B**) Preoperative DSA of the right internal cerebral artery shows severe stenosis of the proximal side of the TS (▲arrowhead). The outflow from the SSS to the right transverse and occipital sinuses was delayed. (**C**) Native angiography after stent deployment. (**D**) Postoperative DSA after stent deployment shows the normalized TS. (**E**) Twelve-month follow-up angiography shows no restenosis in the right TS. (**F**) Thirty-month follow-up CT angiography shows good patency of the stent. DSA: digital subtraction angiography; MRV: magnetic resonance venography; SSS: superior sagittal sinus; TS: transverse sinus

guiding sheath, 7-Fr FUBUKI 100 cm guiding catheter (ASAHI INTECC, Aichi, Japan), and 5-Fr Cerulean catheter (Medikit, Tokyo, Japan). Using a Radifocus guidewire 0.035 (Terumo, Tokyo, Japan), the Cerulean was guided to the right TS, and lesion crossing at the stenotic site of the right TS was performed using an Echelon 10 (Medtronic, Irvine, CA, USA) and CHIKAI 0.014 200 cm (ASAHI INTECC) through the Cerulean. The SSS, sinus confluence, and left TS were reached, and respective intravenous pressures were serially measured. The right sigmoid sinus venous pressure was 10 mmHg, whereas the medial cranial venous pressure at the SSS to stenotic site of the TS was 22 mmHg; the pressure gradient in the venous sinus was thought to lead impaired venous return and intracranial hypertension. Using a CHIKAI 0.014 300 cm (ASAHI INTECC), the Echelon 10 and Cerulean were removed. After evaluating the shape of the stenotic site using intravascular ultrasound (IVUS), the FUBUKI guiding catheter was guided to the right sigmoid sinus. Predilatation was not conducted at the right TS and a Carotid WALLSTENT 8 mm × 21 mm (Boston Scientific, Natick, MA, USA) was inserted. The directly inserted stent was uniformly dilated to approximately 6 mm, but only the stent tip show slight dilation failure and postdilatation was conducted using a Sterling 6.0 × 20 mm (Boston Scientific). After stenting, IVUS confirmed favorable stent dilation (**Fig. 2C**). DSA and 3D-DSA also confirmed sufficient dilation at the stenotic site (**Fig. 2D**). The medial cranial venous pressure at the site of right TS stenting was decreased to 12 mmHg. The pressure gradient in the venous sinus almost disappeared (≤1 to 2 mmHg) after stenting and the procedure was completed. Heparin was not reversed and postoperative heparinization was not continued.

After stenting, headache promptly disappeared. Visual impairment and diplopia disappeared 7 days after treatment. A marked improvement in papilledema was noted (**Fig. 1B**). At this point, the cerebrospinal fluid pressure on lumbar puncture was decreased to 26 cmH_2O. Two months

after stenting, papilledema disappeared (**Fig. 1C**). After 6 months, DAPT was switched to monotherapy by discontinuing aspirin. After 12 months, monotherapy was finished. There were no abnormal findings, such as in-stent stenosis, on cerebral angiography after 11 months (**Fig. 2E**). Contrast-enhanced computed tomography (CT) 30 months after stenting confirmed the patency of the right TS (**Fig. 2F**).

Discussion

Diagnostic criteria for IIH were prepared by Friedman et al.[1,13]: there are no symptoms other than those suggestive of intracranial hypertension, such as papilledema; the composition of cerebrospinal fluid are normal despite an increase in the cerebrospinal fluid pressure on a spinal tap; and other diseases that induce intracranial hypertension can be excluded by imaging study such as MRI.

Epidemiologically, IIH is frequent in young obese females. The symptoms are chronic headache, transient visual obscurity, pulsatile tinnitus, diplopia, posterior dorsal pain, and abducens palsy, eventually leading to blindness in some cases. Anemia, vitamin A deficiency, and adrenal dysfunction may be risk factors for IIH.[2] In the present case, corticosteroids were administered to treat nephrotic syndrome, but the administration period was not long and many years passed since then, so association is unclear. However, a previous study suggested that IIH developed after adrenalectomy for Cushing's syndrome[14]; abnormal changes in steroid hormone balance may have led to IIH. In addition, a recent study reported some patients with polycystic ovary syndrome (PCOS) developed IIH and the testosterone levels in the blood/cerebrospinal fluid were high even in non-PCOS patients with IIH.[15] Therefore, such androgenic hormone may be associated with the pathogenesis of IIH. In the present case, she was not obese, but she underwent unilateral ovariectomy. Imbalance of the sexual hormone may also have played a role to have IIH.

The physiopathology of progressive intracranial hypertension remains unclear, but IIH can be classified into two types: IIH characterized by spontaneous stenosis of the TS, which is not a secondary condition due to cerebral sinus thrombosis or dural arteriovenous fistulae, and stenosis-free IIH. Farb et al.[3] reported that stenosis of the bilateral sinuses was observed in 4 (7%) of 59 controls who underwent contrast-enhanced MRV with gadolinium, whereas stenosis of the bilateral TSs was identified on MRV in 27 (93%) of 29 patients diagnosed with IIH. In IIH patients with stenosis of the TS, impaired venous return related to stenosis may give rise to intracranial hypertension, leading to the progression of sinus stenosis through degeneration of the sinus wall associated with sinus deformity or increased intra-sinus pressure resulting topical microhernia.[2,5]

The pressure gradient after sinus stenosis is measured[4] to confirm that stenosis of the TS is responsible for the venous stasis. In the present case, there was a pressure gradient of 12 mmHg at the stenotic site of the right TS. After stenting, the pressure gradient disappeared, suggesting an association between stenosis and venous stasis.

Recent studies reported the efficacy of sinus stenting for IIH with venous stenosis. The primary points of these studies are summarized in **Table 1.**[4–12] In the present case, chronic progressive headache, papilledema, and abducens nerve paralysis, suggestive of intracranial hypertension were observed, as previously reported. In addition, lumbar puncture revealed markedly increased cerebrospinal fluid pressure. On the other hand, obesity, which was described in many studies, was absent in the present case. Concerning the stent type, self-expanding carotid artery stents were used in many studies. In almost all studies, a symptom improvement rating of ≥90% was achieved and subdural hematoma was noted in three patients as an intracranial complication of the intervention. The maximum follow-up period was 136 months and there was no acute occlusion of a sinus stent.

Many studies reported that plain balloon angioplasty led to restenosis at a high rate, finally requiring stenting[4,5,16]; therefore, in the present case, treatment was performed under the assumption that stenting may be required. As the mechanism of sinus stenosis progression, it may be caused by degeneration of the TS wall related to external pressure-associated sinus deformity or increased intra-sinus pressure,[2,5] differing from thrombotic or atherosclerotic occlusion, as described above. In the present case, when performing stenting, self-expansion of a stent was relatively readily achieved in the absence of predilatation, suggesting occlusion related to a soft substance such as a thin septum structure. However, stenting may prevent disease deterioration related to the progression of sinus stenosis.

Other surgical procedures for IIH include shunting and optic nerve sheath fenestration (ONSF). In Japan, there is no health insurance that covers stenting for the sinus stenosis and shunting is selected as a first-choice procedure in many cases. Indeed, in Japan, only two reports on venous sinus stenting for IIH have been published.[16,17] Shunting is the symptomatic therapy for achieving decompression, and infection or shunt occlusion may develop. A meta-analysis comparing the three methods reported that the improvement

Table 1 Summary of previous reports of TS stenting for IIH

	Case	Gender (F/M)	Mean age	Mean BMI	Sympton		CSF pressure		Mean pressure gradient		Follow-up (months)
					Headache	PAP	Before	After	Before	After	
Higgins et al.	12	12/0	33	36.9	7 (58)	5 (42)	34	NR	19	6	14.2
Owler et al.	4	3/1	27	30	4 (100)	4 (100)	29	NR	19	0.25	9.8
Donnet et al.	10	8/2	41	27.3	8 (80)	10 (100)	40.2	19	19	NR	17.2
Bussiere et al.	10	10/0	34	35.9	10 (100)	9 (90)	NR	NR	28	11	20.1
Ahmed et al.	52	47/5	34	30 >in 47	40 (77)	46 (88)	32.9	24	19	1	24
Albuquerque et al.	15	12/3	32.3	NR	15 (100)	NR	NR	NR	NR	NR	20
Kumpe et al.	18	12/6	37.9	31.6	10 (56)	16 (89)	40	NR	21	3	43.7
Fields et al.	15	15/0	34	39	10 (67)	15 (100)	NR	NR	24	4	14
Radvany et al.	12	11/1	39	33	7 (58)	11 (92)	40	NR	12	1	12
Yamaguchi et al.	1	0/1	33	NR	1	1	25	NR	15	2.7	12
Miyachi et al.	2	2/0	22	19	2 (100)	2 (100)	44	11	23	15	3.5
Our case	1	1/0	37	22.7	1	1	40	26	12	2	30

BMI: body mass index; CSF: cerebrospinal fluid; IIH: intracranial hypertension; PAP: papilledema; TS: transvers sinus

rates for papilledema and visual field defect after shunting, which is the most conventional method, were lower than those after stenting, confirming the limitations of shunting such as the high rate of additional surgery and high incidence of complications.[18] Thus, venous sinus stenting should be performed in IIH patients with stenosis of the TS to recover anterograde venous return, which may be physiologically rational.

The use of antithrombotic agents for sinus stenting has been controversial. In the present case, antiplatelet drugs were administered for 1 year and there was no in-stent restenosis during the ≥1-year follow-up after discontinuation. Many previous studies adopted two antiplatelet drugs in accordance with carotid artery stenting and discontinued them in the long term. It remains to be clarified whether there is intimal change of a stent placed in the cerebral sinus. However, according to previous studies, restenosis occurs at proximal to the stent in many cases, but not in the stent.[5,8,10] This may be associated with sinus deformity or degeneration of the sinus wall, differing from intimal outgrowth after standard carotid artery stenting or in-stent thrombosis. Based on the present case, after the use of antiplatelet drugs for a specific period, they may be discontinued in the long term, as previously reported.

Conclusion

We performed right TS stenting for IIH with stenosis of the TS, promptly reducing intracranial hypertension and papilledema. Many studies reported that venous return-improving treatment by stenting was effective for intracranial hypertension related to sinus stenosis. This treatment should be attempted as an option of surgical treatment for refractory cases to medical treatment, at the risk of restenosis.

Disclosure Statement

The authors completed self-reporting of conflicts of interest (COI) to the Japanese Society of Internal Medicine and Japan Neurosurgical Society. The authors declare no conflicts of interest regarding the publication of this article.

References

1) Friedman DI, Jacobson DM: Diagnostic criteria for idiopathic intracranial hypertension. *Neurology* 2002; 59: 1492–1495.
2) Markey KA, Mollan SP, Jensen RH, et al: Understanding idiopathic intracranial hypertension: mechanisms, management, and future directions. *Lancet Neurol* 2016; 15: 78–91.

3) Farb RI, Vanek I, Scott JN, et al: Idiopathic intracranial hypertension: the prevalence and morphology of sinovenous stenosis. *Neurology* 2003; 60: 1418–1424.
4) Higgins JNP, Cousins C, Owler BK, et al: Idiopathic intracranial hypertension: 12 cases treated by venous sinus stenting. *J Neurol Neurosurg Psychiatry* 2003; 74: 1662–1666.
5) Owler BK, Parker G, Halmagyi GM, et al: Pseudotumor cerebri syndrome: venous sinus obstruction and its treatment with stent placement. *J Neurosurg* 2003; 98: 1045–1055.
6) Donnet A, Metellus P, Levrier O, et al: Endovascular treatment of idiopathic intracranial hypertension: clinical and radiologic outcome of 10 consecutive patients. *Neurology* 2008; 70: 641–647.
7) Bussière M, Falero R, Nicolle D, et al: Unilateral transverse sinus stenting of patients with idiopathic intracranial hypertension. *AJNR Am J Neuroradiol* 2010; 31: 645–650.
8) Ahmed RM, Wilkinson M, Parker GD, et al: Transverse sinus stenting for idiopathic intracranial hypertension: a review of 52 patients and of model predictions. *AJNR Am J Neuroradiol* 2011; 32: 1408–1414.
9) Albuquerque FC, Dashti SR, Hu YC, et al: Intracranial venous sinus stenting for benign intracranial hypertension: clinical indications, technique, and preliminary results. *World Neurosurg* 2011; 75: 648–652; discussion 592–595.
10) Kumpe DA, Bennett JL, Seinfeld J, et al: Dural sinus stent placement for idiopathic intracranial hypertension. *J Neurosurg* 2012; 116: 538–548.
11) Fields JD, Javedani PP, Falardeau J, et al: Dural venous sinus angioplasty and stenting for the treatment of idiopathic intracranial hypertension. *J Neurointerv Surg* 2013; 5: 62–68.
12) Radvany MG, Solomon D, Nijjar S, et al: Visual and neurological outcomes following endovascular stenting for pseudotumor cerebri associated with transverse sinus stenosis. *J Neuroophthalmol* 2013; 33: 117–122.
13) Friedman DI, Liu GT, Digre KB: Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. *Neurology* 2013; 81: 1159–1165.
14) Wagner J, Fleseriu CM, Ibrahim A, et al: Idiopathic intracranial hypertension after surgical treatment of cushing disease: case report and review of management strategies. *World Neurosurg* 2016; 96: 611.e15–611.e18.
15) O'Reilly MW, Westgate CS, Hornby C, et al: A unique androgen excess signature in idiopathic intracranial hypertension is linked to cerebrospinal fluid dynamics. *JCI Insight* 2019; 4: e125348.
16) Miyachi S, Hiramatsu R, Ohnishi H, et al: Endovascular treatment of idiopathic intracranial hypertension with stenting of the transverse sinus stenosis. *Neurointervention* 2018; 13: 138–143.
17) Yamaguchi R, Sato K, Fujimaki H, et al: A case of stent placement for intracranial hypertension associated with venous sinus stenosis. *JNET J Neuroendovasc Ther* 2017; 11: 203–208.
18) Satti SR, Leishangthem L, Chaudry MI: Meta-analysis of CSF diversion procedures and dural venous sinus stenting in the setting of medically refractory idiopathic intracranial hypertension. *AJNR Am J Neuroradiol* 2015; 36: 1899–1904.