A Case of Perimesencephalic Subarachnoid Hemorrhage with Cerebral Venous Sinus Thrombosis due to Stenosis of the Junction of the Vein of Galen and Rectus Sinus

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Abstract
Perimesencephalic subarachnoid hemorrhage (PMSAH) is a group of diseases characterized by bleeding around the brainstem. In recent years, it has been suggested that PMSAH is associated with the venous system. We report a case of PMSAH caused by stenosis of the junction of the vein of Galen (VG) and the rectus sinus (RS). A 39-year-old man presented with restlessness at work. He was administered diazepam, and a few minutes later, his consciousness became clear. Imaging showed subarachnoid hemorrhage (SAH) around the right midbrain, occlusion of the right sigmoid sinus and the right transverse sinus, stenosis of the junction of the VG and RS, T2 hyperintensity in the right middle temporal gyrus, and obstruction of the right vein of Labbe. The location of the SAH coincided with stenosis of the junction of the VG and RS. PMSAH was diagnosed due to the increase in intracranial venous pressure caused by the patient’s sinus obstruction in addition to the stenosis of the junction of the VG and RS. It is necessary to pay attention to venous return when PMSAH is observed.
Introduction

Perimesencephalic subarachnoid hemorrhage (PMSAH) is a subarachnoid hemorrhage (SAH) characterized by bleeding around the brainstem [1]. PMSAH is a disease with unknown sources of bleeding. PMSAH is found in two-thirds of SAH cases with unknown causes [2]. In recent years, it has been reported that the venous system and increased intracranial venous pressure are involved as sources of bleeding in addition to bleeding of arterial origin, including microaneurysms and arterial dissections [3]. We report a case of PMSAH in which deep vein perfusion disorder and sinus occlusion were considered the causes.

Case Report/Case Presentation

A 39-year-old man had become aware of a strong headache 3 days prior to admission. At the hospital, the patient complained of illness while performing construction work and collapsed on the spot with the support of his colleagues. Subsequently, he became restless, and communication was difficult. Following this, hospital emergency code blue was declared. At the time of contact with the doctor, the patient's Glasgow Coma Scale score was 13 (E4V4M5). He was in a state of restlessness and did not follow instructions. He was moved to our critical care center and injected with 5 mg of diazepam. A few minutes later, his consciousness became clear. The patient showed no neurological abnormalities. No factors contributing to an increase in intracranial pressure, such as the Valsalva maneuver, were noted immediately before the onset of restlessness.

Noncontrast computed tomography (CT) revealed a SAH around the right midbrain (Fig. 1a, b). CT angiography (CTA) and venography (CTV) revealed no aneurysm and no abnormal findings in the arterial system, including dural arteriovenous fistula (AVF). Occlusion of the right sigmoid sinus (SS) and right transverse sinus (TS), stenosis of the junction of the vein of Galen (VG) and rectus sinus (RS), and proximal vasodilation of the stenosis were observed. No abnormal outflow tract was observed in the basal vein system (Fig. 1c, d).

Magnetic resonance imaging revealed hyperintensities on the T2-weighted images of the right middle temporal gyrus (Fig. 2a). T2 star-weighted imaging (T2*) showed hypointensity along the sulcus of the right temporo-occipital lobe. This finding suggested stagnation of venous return and a thrombotic obstruction (Fig. 2b). Time-of-flight magnetic resonance angiography showed a mottled hypointensity in the right SS and TS, suggesting cerebral venous thrombosis (Fig. 2c).

Digital subtraction angiography (DSA) revealed occlusion of the right SS and right TS, as well as stenosis of the junction of the VG and RS. The basal vein flowed into the VG, and visualization of the RS was slightly delayed. In addition, obstruction of the right vein of Labbe and pooling of the contrast medium were observed (Fig. 3a, b).

Laboratory examinations revealed hypercholesterolemia. Fibrinogen levels were slightly elevated in the coagulation and fibrinolytic systems, but no other abnormal findings were observed. The findings included triglyceride 354 mg/dL, total cholesterol 275 mg/dL, high density lipoprotein 51 mg/dL, low density lipoprotein 143 mg/dL, antithrombin III 129%, prothrombin time-international normalized ratio 0.95; activated partial thromboplastin time 21.6 s, fibrinogen 449 mg/dL, fibrin/fibrinogen degradation products <2.0 μg/mL, D-dimer 0.9 μg/mL; protein S 117%, protein C 118%, homocysteine 10.3 nmol/mL, anti-double stranded DNA immunoglobulin G <10 IU/mL; anticardiolipin antibodies and anti-beta2 glycoprotein I antibodies <1.2 U/mL, and anti-cardiolipin immunoglobulin G <8 U/mL.
Fig. 1. Noncontrast head CT: right perimesencephalic subarachnoid hemorrhage (a, axial; b, coronal). Sagittal head CTA/V image: stenosis of the junction of the vein of Galen and rectus sinus as well as the proximal vasodilation of the stenosis (c). Head 3D-CTV: occlusion of the right sigmoid sinus and right transverse sinus (d).

Fig. 2. a Head MRI (FLAIR image): T2-weighted image hyperintensity in the right middle temporal gyrus. b Head MRI (T2 star-weighted imaging): hypointensity along the sulcus of the right temporo-occipital lobe. c Head TOF-MRA: mottled hypointensity in the right SS and right TS.
The patient was admitted to the intensive care unit with a diagnosis of PMSAH, cerebral edema due to intracranial venous sinus obstruction, and stenosis of the VG and RS junction. Therapy was started with heparin and antiepileptic drugs (levetiracetam 1,000 mg/day). Since there was no increase in bleeding and no relapse of symptoms, he was discharged from the intensive care unit the next day and transferred to the general ward. The patient began to transition from heparin to warfarin 3 days after admission. During the course, no increase in a hematoma or neurological abnormalities was observed on imaging, and there was no recurrence of epilepsy-like symptoms at the time of admission. The patient’s prothrombin time-international normalized ratio was confirmed to be stable, and he left the hospital 12 days after admission with a modified Rankin Scale score of 0. The cause of the venous sinus thrombosis could not be identified. His magnetic resonance imaging 2 months after onset showed no change in the sinus obstruction findings; however, the edema of the right temporal lobe had improved, and the hypointensity along the sulcus of the right temporo-occipital lobe on T2* disappeared. In addition, the patient’s PMSAH has not relapsed.

Discussion/Conclusion

Here, we investigated the cause of PMSAH and its association with cerebral venous thrombosis. Many reports have suggested the involvement of the venous system as a cause of PMSAH. In this case, the findings of stenosis of the junction of the VG and RS and the findings of thrombosis of the SS and TS coexisted. It has been reported that factors that increase intracranial pressure, such as the Valsalva maneuver, are triggers of PMSAH [4]. Abnormalities in the intracranial venous system, especially those that may exhibit elevated intracranial pressure, are suggested to increase the risk of PMSAH.

Abnormalities of the basal vein of Rosenthal (BVR) have been reported to be related to PMSAH. Some studies have reported that patients with nonaneurysmal PMSAH have a higher prevalence of BVR abnormalities than patients with aneurysmal SAH [3, 5–7]. In addition, one report showed that the side of the PMSAH relates to the side of BVR abnormalities. This report supports the hypothesis that PMSAH is related to the venous system [3].

Fig. 3. DSA (right ICAG): occlusion of the right SS and the right TS, stenosis of the junction of VG and RS, slightly delayed the visualization of the RS, obstruction of the right vein of Labbe, and pooling of contrast medium (a, A–P; b, lateral).
There are some studies on BVR, but there are few reports on VG [8–10]. In a case report by Mathews et al. [8], retrograde flow from the inflow into the VG through the superior vermian vein was present. PMSAH due to TS obstruction has also been reported [11]. Fu et al. [11] proposed a mechanism in which intracranial venous hypertension due to TS thrombosis caused a rupture of the basal vein system around the midbrain.

In this case, the stenosis of the junction of the VG, RS, and TS thromboses was mixed. Risk factors for venous sinus thrombosis include trauma, infections, prothrombotic conditions, malignant tumors, postoperative catheterization, and genetic abnormalities. However, the association between venous sinus thrombosis and anatomical abnormalities of the intracranial venous system is not clear [12, 13]. In children, there was a report investigating the association between anatomical abnormalities of the venous system and intracranial sinus thrombosis; however, no significant association was found [14]. There is a report that hypoplasia of the TS has been reported to be associated with venous sinus thrombosis [15], and it is thought that an abnormal venous system, especially one exhibiting perfusion stasis, may cause venous sinus thrombosis. However, in cases of congenital stenosis of the junction of the VG and RS, the cause of sinus thrombosis remains questionable. In this case, the obvious cause of sinus thrombosis could not be identified.

We have not found any case reports of PMSAH with abnormal findings in the basal vein system in addition to sinus thrombosis. In this case, there were no abnormalities in the BVR; however, the findings of stenosis of the junction of the VG and RS could be confirmed by CTV, magnetic resonance venography (MRV), and DSA. DSA showed that the inflow from the VG into the RS was slightly delayed. The findings of the right SS and TS thrombosis were well defined. The hypointensity along the sulcus of the right temporo-occipital lobe on T2*, obstruction of the right vein of Labbe, and pooling of contrast medium suggested conditions for increased intracranial venous pressure. In addition, the localization of the SAH was consistent with stenosis of the junction of the VG and RS. There are no reports investigating whether the stenosis of the junction of the VG and RS is congenital. The obvious cause of venous sinus thrombosis cannot be determined, and in this case, it was thought that the PMSAH occurred due to increased intracranial venous pressure due to obstruction of the right SS and TS in addition to the stenosis of the junction of the VG and RS.

During the process leading to this diagnosis, PMSAH was confirmed by non-contrast CT; then, CTA was employed to identify the presence of an aneurysm, AVF, or arteriovenous malformations. Since intracranial venous thrombosis was confirmed in the venous phase, we carefully reviewed the maximum intensity projection-CTA/V images because AVF may be complicated by intracranial venous thrombosis. In this process, we confirmed the findings of stenosis at the junction of the VG and RS. Since the MRV and DSA showed the same findings, we diagnosed the patient with PMSAH due to a venous return disorder. When diagnosing PMSAH, modalities such as CTV and MRV may be useful to evaluate the intracranial venous system.

We report a case of PMSAH with SS and TS thrombosis and stenosis of the VG and RS junction. When diagnosing PMSAH, it is necessary to observe the arterial system but also identify the causes of increases in intracranial venous pressure, including venous return disorders and drainage patterns.

Statement of Ethics

Our patient gave his written informed consent for the publication of the article (including imaging results). Ethical approval is not required for this study in accordance with local or national guidelines.
Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Kyoya Sakashita (the principal researcher) examined the patient, contributed to data collection and interpretation, and wrote the manuscript. Kei Miyata examined the patient together with Dr. Sakashita and reviewed the manuscript. Ryohei Saito and Ryota Sato took care of the patient. Sangnyon Kim and Nobuhiro Mikuni reviewed the manuscript.

Data Availability Statement

All data that support the findings of this study are included in this article. Further inquiries can be directed to the corresponding author.

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