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Case Report

A Case of Segmental Arterial Mediolysis: Hemoperitoneum with Hemorrhagic Shock due to Rupture of a Visceral Artery Aneurysm Following Subarachnoid Hemorrhage

Keisuke Ota[1] and Noriaki Matsubara[2]

Objective: Segmental arterial mediolysis (SAM) is a non-inflammatory and non-atherosclerotic vascular disease characterized by segmental medial defect/necrosis of muscular arteries as a result of mediolysis. SAM affects the visceral and intracranial arteries, and causes arterial dissection and aneurysm. We report a case of aneurysmal subarachnoid hemorrhage (SAH) followed by hemoperitoneum due to a ruptured visceral artery aneurysm.

Case Presentation: A 54-year-old man developed SAH from a ruptured anterior communicating artery aneurysm, which was clipped on the same day. Thereafter, he was treated to prevent cerebral vasospasm. Six days after onset, he developed acute anemia and his blood pressure decreased, suggesting hemorrhagic shock. Hemoperitoneum was detected on computed tomography and abdominal angiography was performed. Irregular and stenotic arterial findings, and an unusual aneurysm with contrast stasis were found in a branch vessel from the right gastroepiploic artery. SAM was diagnosed based on the clinical course and angiographic characteristics. Endovascular treatment consisted of embolization of the visceral artery aneurysm with liquid embolic material. after embolization, the vital signs stabilized and he recovered from shock. Acute treatment for SAH was continued. Although the patient did not develop vasospasm-related sequelae, he was transferred to the rehabilitation hospital 7 weeks after onset and his modified Rankin Scale score at 3 months after onset was 2.

Conclusion: Visceral artery aneurysm associated with SAM should be considered as the cause of hemoperitoneum with hemorrhagic shock during the acute phase of SAH.

Keywords segmental arterial mediolysis, subarachnoid hemorrhage, hemoperitoneum, intracranial aneurysm, visceral artery aneurysm

Introduction

Segmental arterial mediolysis (SAM) is a non-inflammatory non-arteriosclerotic disease in which segmental lysis of the arterial media induces arterial dissection and aneurysm.[1,2)]

Patients with SAM often present hemoperitoneum caused by the rupture of a visceral artery aneurysm, and also present subarachnoid hemorrhage (SAH) due to rupture of an intracranial aneurysm. Moreover, induction of subarachnoid and hemoperitoneum by the rupture of SAM-induced intracranial and visceral artery aneurysms within a short time has been reported, although it is rare.[2–12)]

We report a patient in whom an aneurysm ruptured in a visceral artery 6 days after the onset of SAH induced by rupture of an anterior communicating artery aneurysm, causing hemorrhagic shock.

Case Presentation

The patient was a 54-year-old man. He had a past medical history of hypertension and diabetes, but no history of hemorrhagic disease or hemorrhagic event. Headache and nausea suddenly developed, and the patient visited the

[1]*Department of Neurosurgery, Anjo Kosei Hospital, Anjo, Aichi, Japan*
[2]*Department of Neurosurgery and Neuroendovascular Therapy, Osaka Medical College, Takatsuki, Osaka, Japan*

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Corresponding author: Noriaki Matsubara. Department of Neurosurgery and Neuroendovascular Therapy, Osaka Medical College, 2-7, Daigakumachi, Takatsuki, Osaka 569-8686, Japan
Email: neu147@osaka-med.ac.jp

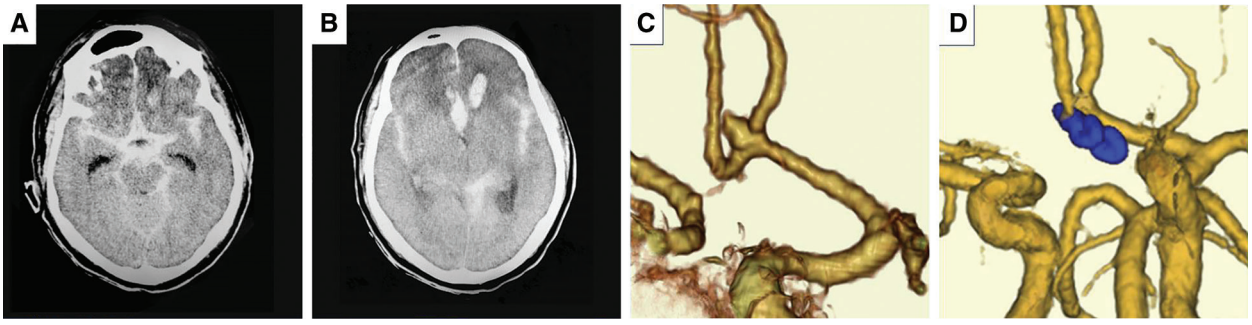

Fig. 1 (**A**, **B**) Diffuse SAH was observed on head CT acquired at onset. (**C**) An approximately 3-mm anterior communicating artery aneurysm was present on preoperative 3D-CT angiography. (**D**) Occlusion of the aneurysm by clipping was confirmed by postoperative 3D-CT angiography. 3D-CT: three-dimensional computed tomography; SAH: subarachnoid hemorrhage

outpatient emergency service. The state of consciousness deteriorated to somnolence after vising the emergency service. The consciousness level was Japan Coma Scale II-10. Mild paralysis was noted in the right upper and lower limbs. On head computed tomography (CT), SAH was present accompanied by hematoma in the left frontal lobe (**Fig. 1A** and **1B**) and was diagnosed as Hunt and Hess Grade 3 SAH. No abnormality was detected by blood count or biochemical examination. Three-dimensional CT angiography was performed and an irregularly shaped aneurysm with a maximum diameter of 2.9 mm was found in the anterior communicating artery (**Fig. 1C**).

On the same day, clipping of the anterior communicating artery aneurysm was performed through the interhemispheric approach under general anesthesia. The aneurysm was small and irregularly shaped although there were no microscopic findings suggesting a dissecting aneurysm and a saccular aneurysm appearance was observed. The surgical clipping and postoperative courses were uneventful without complications (**Fig. 1D**). The consciousness level improved to Japan Coma Scale I-10 within several days after surgery. Subsequently, preventive treatment for cerebral vasospasm following SAH was performed. No intentional blood pressure increase or active loading of infusion was performed, but the systolic blood pressure was higher and circulation was managed to prevent dehydration (normovolemia). Administration of fasudil, ozagrel, and cilostazol was initiated 2 days after the onset as drug therapy.

The blood pressure suddenly decreased 6 days after the onset of SAH. The systolic blood pressure decreased to the 70s, suggesting shock. No hematemesis or melena was observed, but mild abdominal distension was noted on physical examination. Severe anemia was detected on blood testing. The blood hemoglobin level decreased from 11.0 to 5.6 g/dL. Emergency blood transfusion of red blood cells and fresh frozen plasma preparations was performed. Plain abdominal CT revealed massive hemoperitoneum (**Fig. 2**). On abdominal contrast-enhanced CT, enhanced mass lesion continuous to blood vessel was observed below the stomach. The condition was diagnosed as abdominal visceral aneurysm rupture-induced hemoperitoneum. Abdominal angiography was immediately performed. Irregular findings with a beaded appearance were observed in blood vessels branching from the right gastroepiploic artery to the great omentum on celiac arteriography. Moreover, an approximately 15-mm aneurysm was present in the affected vessel, demonstrating stagnation of contrast medium in the aneurysm, which is a characteristic angiography finding suggesting SAM (**Fig. 3A** and **3B**). Autoantibodies (anti-nuclear antibody, lupus anticoagulant antibodies, anti-cardiolipin antibody, and antineutrophil cytoplasmic antibodies) were not positive on blood testing performed on a later day.

Embolization of the visceral artery aneurysm was performed following abdominal angiography under local anesthesia. A 4Fr shepherd hook catheter was placed in the celiac artery through a sheath inserted into the right femoral artery. A Marathon (Medtronic, Minneapolis, MN, USA) microcatheter was guided to the right gastroepiploic artery (**Fig. 3C** and **3D**). Using n-butyl 2-cyanoacrylate, the aneurysm was embolized and occluded together with the affected vessel located proximal and distal to the aneurysm (**Fig. 4**). After embolization of the visceral artery aneurysm, the blood pressure recovered and shock resolved with stabilizing vital signs. No re-aggravation of anemia was noted. Acute-phase treatment of SAH was continued thereafter, including treatment for cerebral vasospasm. Vasospasm-induced cerebral ischemia did not develop. The patient was transferred to a hospital for rehabilitation of right paresis 7 weeks after onset. The modified Rankin

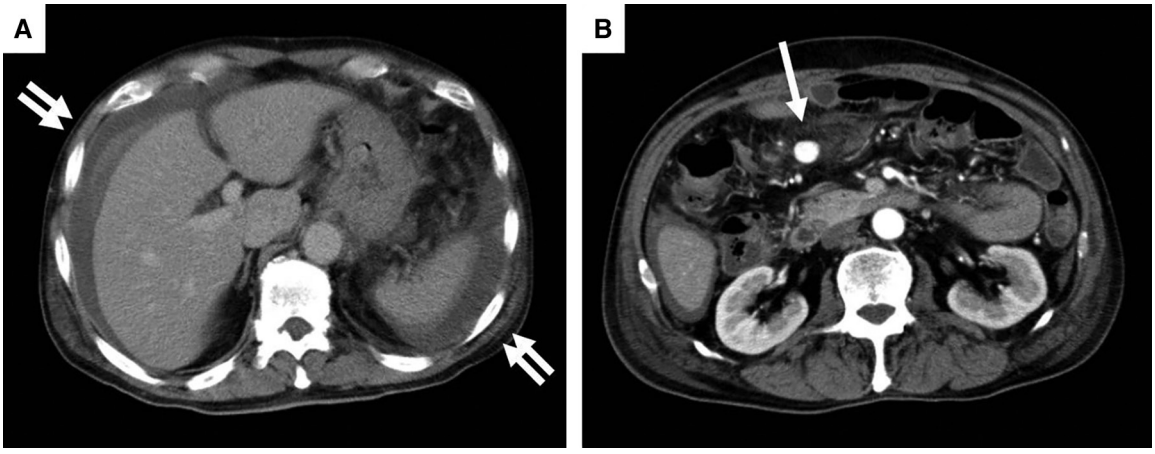

Fig. 2 (**A**) Massive hemoperitoneum was observed on abdominal plain CT (double arrow). (**B**) Enhanced lesion continuous to blood vessel was observed on contrast-enhanced abdominal CT (arrow). CT: computed tomography

Scale score at 3 months after onset was 2. No recurrence of the cerebral aneurysm was observed on follow-up after 1 year and no new SAM-induced event was noted.

Discussion

SAM is a disease inducing dissection and aneurysm through segmental lysis of the arterial media. The pathological characteristics of SAM are island-shaped residues (medial island) of the media observed on the arterial wall, collapse of the intima while retaining the dilated adventitia (medial gap), and the absence of inflammatory and atherosclerotic findings in the wall. The histological changes start with vacuolar degeneration of smooth muscle cells in the media followed by lysis of the media from the outer side, forming a space accompanied by exudation and fibrin deposition. When the intima tears and the arterial wall are dissected in this state, the remaining adventitia dilates, forming an aneurysm.[1,2] SAM is a rare pathology and no epidemiological data, such as the prevalence per population or incidence, are available. On the other hand, according to a review by Shenouda et al., the mean onset age of SAM is 57 years old, demonstrating a higher incidence in the middle-aged and elderly, with a male: female ratio of 1.5:1, being higher in males. The disease develops with abdominal pain in 66%, accounting for a high percentage, and hemoperitoneum-induced shock develops in approximately one-third. The disease develops with neurological manifestations in 14%, which is not rare.[13] It may develop with arterial dissection-induced cerebral ischemia and headache, but intracranial aneurysm rupture-induced SAH is more frequently reported. SAM is definitively diagnosed based on the pathological findings of the affected vascular lesions, but lesions may be discovered in an asymptomatic state and endovascular treatment has become more frequently performed for ruptured visceral artery aneurysm, making collection of pathological tissue difficult in many cases. Moreover, no SAM-specific autoantibody or inflammation marker has been reported. Therefore, it is difficult to diagnose it or evaluate its activity based on blood examination results. When pathological tissue cannot be collected, the diagnosis of SAM is made based on the clinical course and angiography findings.[14] In the present patient, SAM was diagnosed by the exclusion of other diseases, the presence of hemoperitoneum from the right gastroepiploic artery, which is a rare lesion site, angiography findings, such as irregular vascular dilation and narrowing in the beaded area, and aneurysm formation.

SAM affects multiple blood vessels in the body and lesions most frequently develop in the visceral arteries, with the main trunk of the celiac artery and its branches, the splenic and hepatic arteries being common sites.[13] In this patient, the lesion was present in the right gastroepiploic artery. Rupture of visceral artery aneurysms causes hemoperitoneum and that may induce hemorrhagic shock requiring urgent treatment.[1] For the treatment of SAM-induced visceral artery aneurysms, surgery had been mainly performed, but selection of the treatment method has shifted to endovascular treatment due to its recent advances.[1,13,15] In the present patient, the visceral artery aneurysm was embolized by endovascular treatment and a favorable outcome was achieved.

SAM induces intracranial arterial lesions second to those in the visceral arteries. According to a review, SAM was associated with intracranial aneurysms in 13% of all cases.[13] The common sites of cerebral aneurysms

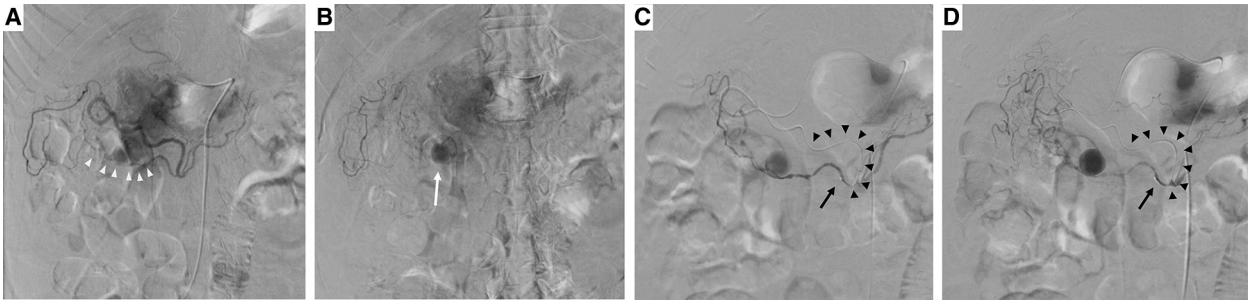

Fig. 3 Celiac arteriography (**A**: arterial phase, **B**: capillary phase). Irregular findings with a beaded appearance were noted in the blood vessel branching from the right gastroepiploic artery to the great omentum (arrow heads). An approximately 15-mm aneurysm was present in the affected vessel and contrast medium stagnated in the aneurysm on angiography (arrow). These findings are suggesting angiographic appearance in SAM. Right gastroepiploic arteriography (**C**: arterial phase, **D**: capillary phase). A microcatheter was selectively guided to the proximity of the involved vessel. The arrowhead and arrow indicate the distribution and tip of the catheter, respectively. SAM: segmental arterial mediolysis

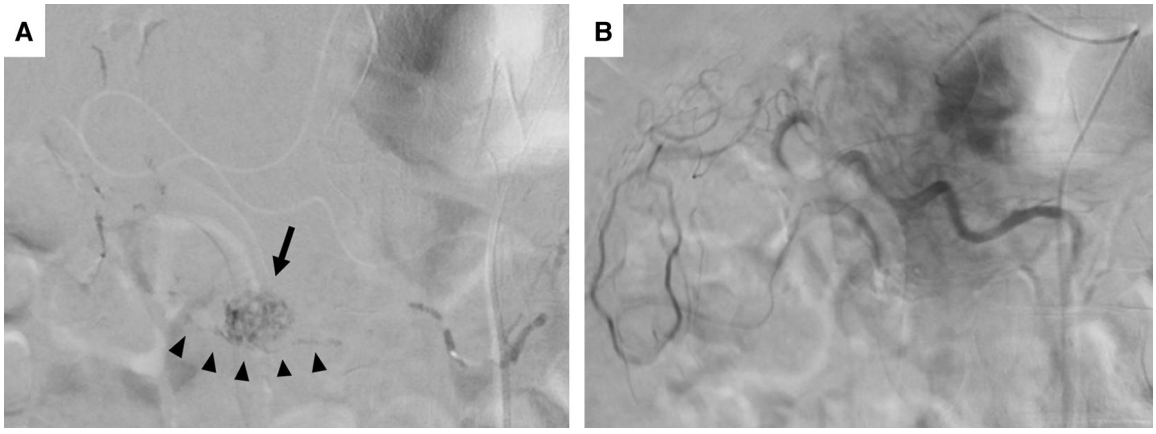

Fig. 4 (**A**) NBCA was injected through the microcatheter, and the affected vessel and aneurysm were embolized. The arrow and arrowhead indicate NBCA casts in the aneurysm and affected vessel, respectively. (**B**) The aneurysm was not visualized by right gastroepiploic arteriography after embolization. NBCA: n-butyl 2-cyanoacrylate

are the internal carotid artery and vertebral artery. Regarding aneurysm morphology, dissecting aneurysms are frequently reported, but saccular aneurysms may also be formed.[3,4,11] The anterior communicating artery aneurysm in this patient was small and irregularly shaped and no finding clearly suggested dissection in the intraoperative observation. Moreover, as SAM was not originally considered, histopathological tissue was not evaluated. The intracranial aneurysm in this patient was considered to be associated with SAM based on the clinical course, but it is also possible that usual saccular cerebral aneurysm rupture was incidentally complicated by SAM-induced visceral artery aneurysm rupture.

Several SAM cases with hemoperitoneum that developed in the acute phase of SAH from intracranial aneurysm rupture were recently reported.[3–12] Case reports of SAM with hemoperitoneum that developed in the acute phase of SAH are presented in **Table 1**. Based on analysis of the present case and previously reported cases, hemoperitoneum developed 7.9 days after the onset of SAH on average. On the other hand, to our knowledge, no case of SAH following hemoperitoneum has been reported, suggesting that stress and increased blood pressure in the acute phase of SAH are involved in rupture of SAM-induced visceral artery aneurysms. Although the hormone level in acute-phase SAH was not measured in this patient, the involvement of endogenous catecholamine in the pathogenesis of SAM was suggested in a previous study.[16] Other than these, management of the systemic circulation and drug therapy for the treatment of cerebral vasospasm are suspected to have an influence. In our patient, the water balance was set to normovolemia, but blood pressure was controlled at a higher level and several antithrombotic drugs were used. These may have been a cause of visceral artery aneurysm rupture or promoted hemoperitoneum. Many SAM cases in which hemoperitoneum developed in the acute phase of SAH were reported from Japan,[3,4,6–10,12] suggesting a racial difference concerning onset. However,

Table 1 Case reports of SAM that caused hemoperitoneum in the acute phase of subarachnoid hemorrhage

Authors	Authors' country	Age	Sex	Intracranial aneurysm	Morphology	Rupture status	Treatment for intracranial aneurysm	Drug therapy for vasospasm	Visceral artery aneurysm	Rupture status	Treatment for visceral artery aneurysm	Interval (SAH) hemoperitoneum)	Diagnosis	Clinical outcome
Fuse et al. 1996	Japan	56	F	Lt ICA C1	Saccular	Ruptured	Surgery (clipping)	Ozagrel	Gastroepiploic artery	Ruptured	Surgery	16 days	Clinical and angiographical	No deficit
				Rt MCA	Saccular	Unruptured	None		Bil Gastric arteries	Unruptured	Surgery			
Stetler et al. 2012	United States	59	F	Rt IC-PC	Saccular	Ruptured	Endovascular (coiling)	NA	Rt Hepatic artery	Ruptured	Endovasualar	3 days	Clinical and angiographical	Neurologically improved
Shinoda et al. 2016	Japan	47	M	Lt VA	Dissecting	Ruptured	Endovascular (coiling/PAO)	NA	Middle colic artery	Ruptured	Envovascular→ Surgery	8 days	Pathology	mRS 3
Ro et al. 2018	Japan	NA	M	VA	Dissecting	Ruptured	None	NA	Middle colic artery	Rpture	None	NA	Pathology (autopsy)	Death
									Bil gastric arteries	NA	None			
									Bil gastroepiploic arterises	NA	None			
									Inferior pancreaticoduodenal artery	NA	None			
									Rt renal artery	NA	None			
Hellstern et al. 2018	Germany	30	M	BA trunk	BBA	Ruptured	Endovascular (coiling→flow diverter)	NA	Splenic artery	Ruptured	Surgery	0 day	Pathology	Barthel Index 90
				Bil ICA petrous	Dissecting	Unruptured	None							
Hayashi et al. 2018	Japan	49	F	Lt ICA C1	Saccular	Ruptured	Surgery (clipping)	NA	Splenic artery	Ruptured	Endovascular	4 days	Clinical and angiographical	Mild aphasia
									Gastroduodenal artery	Unruptured	None			
									Common hepatic artery	Unruptured	None			
									Superior mesenteric artery	Unruptured	None			
Isaji et al. 2018	Japan	45	M	Rt VA	Dissecting	Ruptured	Endovascular (coiling/PAO)	Fasudil, Ozagrel, Clopidgrel, Argatroban	Accessory middle colic artery	Ruptured	Endovascular	8 days	Clinical and angiographical	No deficits
									Superior mesenteric artery	Unruptured	Endovascular			
Inazuka et al. 2019	Japan	77	F	Rt ICA	BBA	Ruptured	Surgery (clipping)	NA	Celliac artery	Ruptured	None	8 days	Pathology (autopsy)	Death
									Splenic artery	Unruptured	None			
Ohara et al. 2019	Japan	82	F	Lt VA-PICA	Fusiform	Ruptured	Endovascular (coiling)	Fasudil, Ozagrel, Aspirin	Rt Gastroepiploic artery	Ruptured	Endovascular	14 days	Clinical and angiographical	Bedridden
Tanaka et al. 2020	Japan	60s	M	Rt ICA C1	BBA	Ruptured	Surgery (trapping with bypass)	Fasudil, Cilostazol	Posterior inferior pancreaticoduodenal artery	Ruptured	Endovascular	12 days	Pathology (autopsy)	Death
Present case. 2020	Japan	54	M	AcomA	Saccular	Ruptured	Surgery (clipping)	Fasudil, Ozagrel, Cilostazol	Rt Gastroepiploic artery	Ruptured	Endovascular	6 days	Clinical and angiographical	mRS 2

BA: basilar artery; BBA: blood blister-like aneurysm; Bil: bilateral; F: female; ICA: internal carotid artery; IC-PC: internal carotid artery-posterior communicating artery; Lt: left; M: male; MCA: middle cerebral artery; mRS: modified Rankin Scale; NA: not available; PAO: parent artery occlusion; PICA: posterior inferior cerebellar artery; SAM: segmental arterial mediolysis; VA: vertebral artery

the accumulation and investigation of patients are necessary. When rapid hemorrhagic shock develops in the acute phase of SAH (especially dissecting intracranial aneurysm rupture), it is necessary to take SAM into consideration.

Conclusion

We reported a patient in whom hemorrhagic shock developed due to hemoperitoneum following the onset of SAH. When hemorrhagic shock develops in the acute phase of SAH, hemoperitoneum from a ruptured SAM-associated visceral artery aneurysm must be considered.

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Disclosure Statement

The authors declare no conflict of interest.

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