Intraperitoneal hemorrhage due to segmental arterial mediolysis associated with cerebral vasospasm after subarachnoid hemorrhage

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Abstract

A man in his 50s with no significant past medical history developed subarachnoid hemorrhage due to ruptured left middle cerebral artery aneurysm. 9th hospital day, he experienced a ruptured visceral aneurysm with segmental arterial mediolysis, and we successfully treated with transarterial embolization using metallic coils.

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CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

CM: wrote and drafted the manuscript. CM, YF, ST, KK, NH and NS: helped draft the manuscript. All authors read and approved the final manuscript.

ETHICAL APPROVAL

All subjects enrolled in this research have given their informed consent, which has been approved by the institutional committee on human and/or animal research, and this protocol has been found acceptable by them.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

KEY CLINICAL MESSAGE

We should always consider the possibility of segmental arterial mediolysis (SAM) in case of shock after subarachnoid hemorrhage (SAH). In addition, SAH with cerebral vasospasm may be associated with SAM, suggesting that intra-abdominal hemorrhage due to SAM may occur during cerebral vasospasm.

ABSTRACT

A man in his 50s with no significant past medical history developed subarachnoid hemorrhage due to ruptured left middle cerebral artery aneurysm. 9^{th} hospital day, he experienced a ruptured visceral aneurysm with segmental arterial mediolysis, and we successfully treated with transarterial embolization using metallic coils.

KEYWORDS

segmental arterial mediolysis, subarachnoid hemorrhage**,**cerebral vasospasm, visceral artery aneurysm, intracranial aneurysm, intraabdominal hemorrhage

1 INTRODUCTION

Segmental arterial mediolysis (SAM) is a noninflammatory, nonatherosclerotic disease that causes segmental lysis of the outer arterial media. [1] It can result in separation of the media from the adventitia leading to dissecting aneurysms.[1] Most commonly,an aneurysm forms in the large abdominal aortic branches, and its rupture causes intra-abdominal hemorrhage. [2] The cerebral arteries can also be involved, but SAM is not yet well known in the neurological field. In rare cases, SAM has been reported to occur in conjunction with subarachnoid hemorrhage (SAH). We report a case of intra-abdominal bleeding due to SAM during the cerebral spasm after SAH, which was successfully treated with coil embolization of an intraperitoneal aneurysm.

2 CASE HISTORY

A 59-year-old man with no significant past medical history suddenly lost consciousness after onset of a severe headache. He presented to the emergency department at our hospital. On admission, his Glasgow Coma Scale was E3V2M5. A clinical diagnosis of grade IV was made on the World Federation of Neurological Surgeons [WFNS] scale. Computed tomography (CT) scan showed SAH from the basal cistern to the left Sylvian fissure (Fisher group 3). Three-dimensional CT angiography revealed an aneurysm (6.7 mm x 6.2mm x 4.9 mm) with blebs was found in the middle cerebral artery (M2) bifurcation of the left MCA. (Figure 1) Non-contrast enhanced CT of the chest and abdomen for screening showed no abnormalities. We diagnosed SAH due to ruptured left MCA aneurysm and performed clipping for the aneurysm on the same day. External decompression was also added because of the elevated cerebral pressure. Intraoperative findings indicated that the aneurysm was a saccular appearance. (Figure 2) To prevent cerebral vasospasm, spinal drainage was started postoperatively and administration of ozagrel sodium 80 mg/day and fasudil hydrochloride hydrate 30 mg x 3/day was started. 8 days after onset, the cerebral angiography showed narrowing at the left M2 due to cerebral vasospasm and delayed blood flow in peripheral vessels. (Figure 3) Therefore, low-molecular-weight dextran was added to maintain cerebral blood flow, and blood pressure was maintained at a higher level. (Figure 4) 9^{th} hospital day, when we performed tracheostomy for respiratory management due to prolonged disturbance of consciousness, he suddenly lapsed into hypovolemic shock state. The head CT showed no new intracranial hemorrhage or infarction. However, non-contrast and contrast-enhanced CT of

the chest and abdomen showed bloody ascites and abnormal dilatation of the superior mesenteric artery and anterior superior pancreatoduodenal artery (ASPDA), suspected intraperitoneal hemorrhage from the same site. (Figure 5) We determined that the patient was in hemorrhagic shock due to abdominal hemorrhage. He was administered catecholamine, blood transfusion, and insertion of an aortic balloon pumping to stabilize his circulation. An emergency abdominal angiography revealed a beaded appearance of the ASPDA. (Figure 6) The ruptured ASPDA pseudoaneurysm was successfully treated by transarterial coil embolization. (Figure 7) The immunological tests performed to investigate the cause of the disease showed that proteinase-3 antineutrophil cytoplasmic antibody (PR3ANCA) was mildly elevated. Antinuclear antibodies, anticardiolipin antibodies, and myeloperoxidase-anti-neutrophil cytoplasmic antibodies (MPOANCA) were negative, and complement titers, C3 and C4, were not elevated. 10 days after the SAH, the CT revealed severe left MCA territory infarction due to cerebral vasospasm and we performed conservative treatment for the patient. On the 44th day, the patient had good cerebral pressure control and underwent cranioplasty. The patient was transferred to a rehabilitation hospital on the 77 days, and was discharged home with a modified Rankin Scale 3.

3 DISCUSSION

SAM is first reported by Slavin in 1976, based on pathological examination of cases of intra-abdominal hemorrhage due to abdominal aneurysms. [3] There are two main types of arteries: the elastic arteries, and the muscular arteries. Elastic arteries consist of elastic tissue in the tunica media and include the aorta and common carotid artery. Muscular arteries contain smooth muscle cells in the tunica media and include cerebral arteries and visceral arteries. [4] SAM occurs in muscular arteries because of segmental lysis of the arterial tunica media smooth muscle cells. [2]It often shows findings of a spindle aneurysm or dissection.[5-10] The definitive diagnosis of SAM is based on pathological findings, the clinical diagnosis depends on the exclusion of other vasculitis and the finding of beaded vessel irregularities, dissection, or aneurysm on angiography. Specifically, the criteria are (1) middle-aged and elderly patients, (2) exclusion of underlying diseases such as inflammatory changes or atherosclerotic changes, (3) sudden onset of intra-abdominal bleeding, and (4) presence of bead-like changes in blood vessels on angiography. [11]Differential diseases include atherosclerotic disease, systemic vasculitis (Behcet's disease, polyarteritis nodosa), fibromuscular dysplasia (FMD), Ehlers–Danlos disease type IV, mycotic aneurysms, cystic medial necrosis and Marfan syndrome. [1]

In this case, the patient was a middle-aged man in his 50s with no history of collagen disease or other inflammatory or atherosclerotic diseases. Laboratory blood samples showed no elevated inflammatory response, and MPOANCA and complement titers were normal. Only PR3ANCA was elevated in immunological tests, which required differentiation from granulomatous polyangiitis. It was not characteristic because of no findings of upper respiratory tract symptoms, pulmonary symptoms, or nephritis. 9^{th}hospital day, the patient developed sudden intra-abdominal hemorrhage, and abdominal angiography showed beaded vasodilatation confined to the ASPDA. Based on the above, SAM was diagnosed because it met the criteria for clinical diagnosis.

The etiology of SAM is still unclear, but some authors believe that vasoactive substances such as norepinephrine are involved and cause vasoconstriction. [12,13] When SAH occurs, the activation of the sympathetic nervous system causes a surge of catecholamines, which stimulate α1 receptors widely distributed in smooth muscle cells of the tunica media.[12] Because systemic norepinephrine increases approximately threefold within 48 hours of onset, an intra-abdominal aneurysm may have formed at an early stage after the onset of SAH in this case. [14] The blood pressure was maintained at a high level during the management of cerebral vasospasm and the blood pressure further increased due to stimulation associated with tracheostomy. Therefore, it may have contributed to the aneurysm rupture.

Including this case, 14 cases of intraperitoneal hemorrhage due to SAM after SAH have been reported. [2, 5-10, 15-20] (Table 1) There are many reports from Japan. This is because of the high incidence of SAH in Japan compared to the worldwide.[21] Intracranial cerebral aneurysms included 6 dissecting aneurysms, 2 blistering aneurysms, and 6 saccular aneurysms. In addition including this case, 11 patients had intraperitoneal hemorrhage during the cerebral vasospasm period after SAH. The cerebral vasospasm often begins

3-4 days after onset, peaks at 7-10 days, and lasts until about 14-21 days.[22] This is the period when SAM may occur. The blood pressure is often maintained high and antiplatelet medications are used for cerebral vasospasm management. It may contribute to intraperitoneal hemorrhage.

The cerebral infarction caused by cerebral vasospasm was reported in 5 cases. SAH with cerebral vasospasm may be prone to intra-abdominal vasospasm as well, which may lead to aneurysm formation. Therefore, SAM should be considered when cerebral vasospasm occurs after SAH. In addition, in many cases, the patients had a good level of consciousness before the intra-abdominal hemorrhage and were able to report abdominal symptoms. Therefore, they suspected abdominal disease from the beginning and were able to examine the patients. On the other hand, our patient was unconsciousness due to severe SAH, making it difficult to differentiate the cause of shock. Recognizing SAM can lead to early treatment and save their lives.

4 CONCLUSION

SAH with cerebral vasospasm may be associated with SAM, suggesting that intra-abdominal hemorrhage due to SAM may occur during cerebral vasospasm. SAM should be considered in the treatment of SAH in patients with impaired consciousness and inability to complain of abdominal pain.

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FIGURES AND TABLES

Figure 1. Three-dimensional computed tomography (CT) angiography showing an aneurysm (arrow) with blebs at the M2 bifurcation of the left middle cerebral artery.

Figure 2. Intraoperative photograph showing a saccular aneurysm (arrow) at the M2 bifurcation of the left middle cerebral artery.

Figure 3. The cerebral angiography indicating narrowing at the left M2 (arrow) due to cerebral vasospasm.

Figure 4. This chart explaining the clinical course of a patient with various anticonvulsant treatments and the development of segmental arterial mediolysis (SAM).

Figure 5. Axial view of the abdominal contrast-enhanced computed tomography (CT) scan revealing abnormal band dilatation of the anterior superior pancreatoduodenal artery (ASPDA) (arrow).

Figure 6. Anterior superior pancreatoduodenal artery (ASPDA) angiography revealing

pseudoaneurysm in the ASPDA. (arrow).

Figure 7. Anterior superior pancreatoduodenal artery (ASPDA) angiography illustrating coil embolization (arrow) of the pseudoaneurysm.

Figure 1

Figure 2

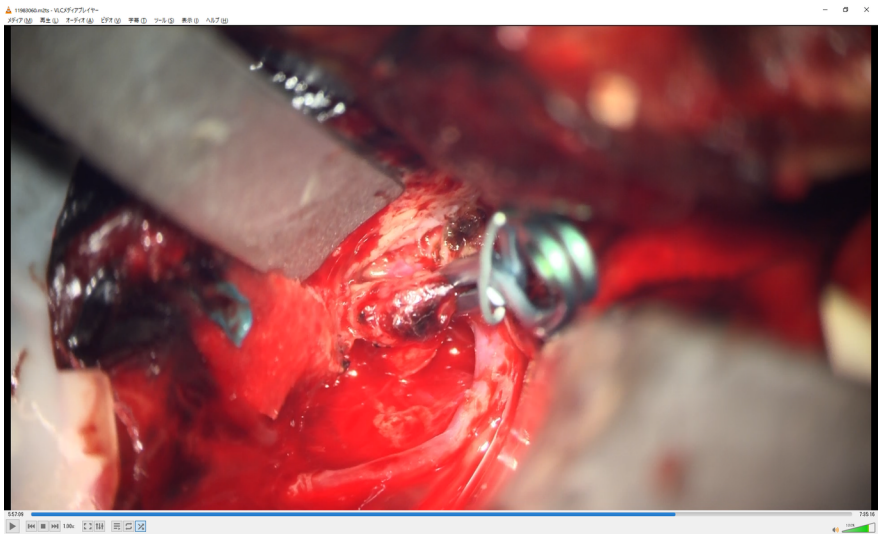

Figure 3

Figure 4

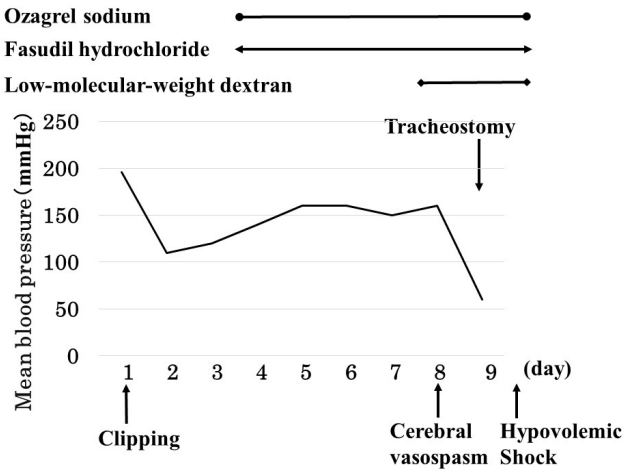

Figure 5

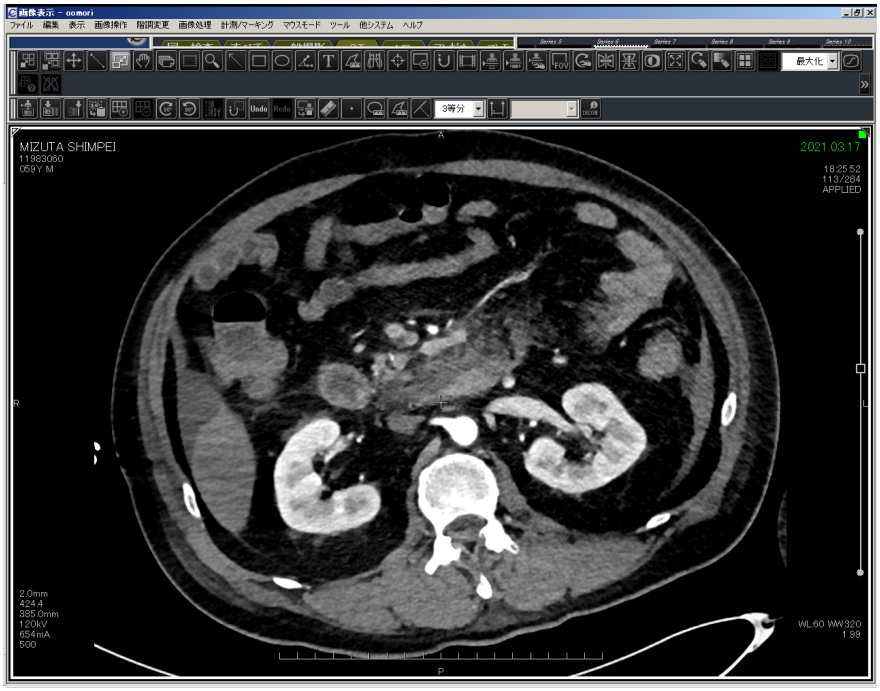

Figure 6

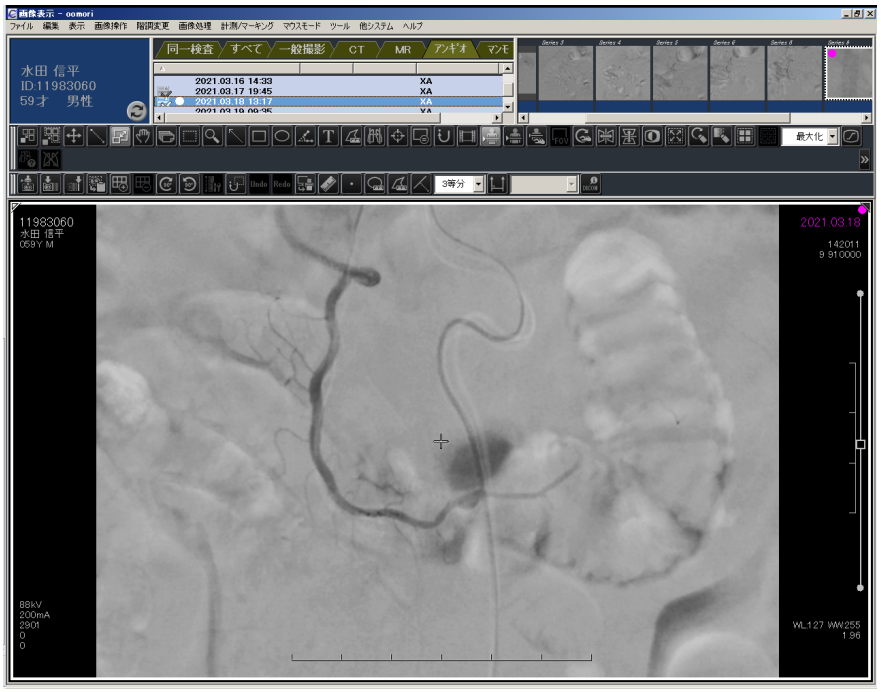

Figure 7

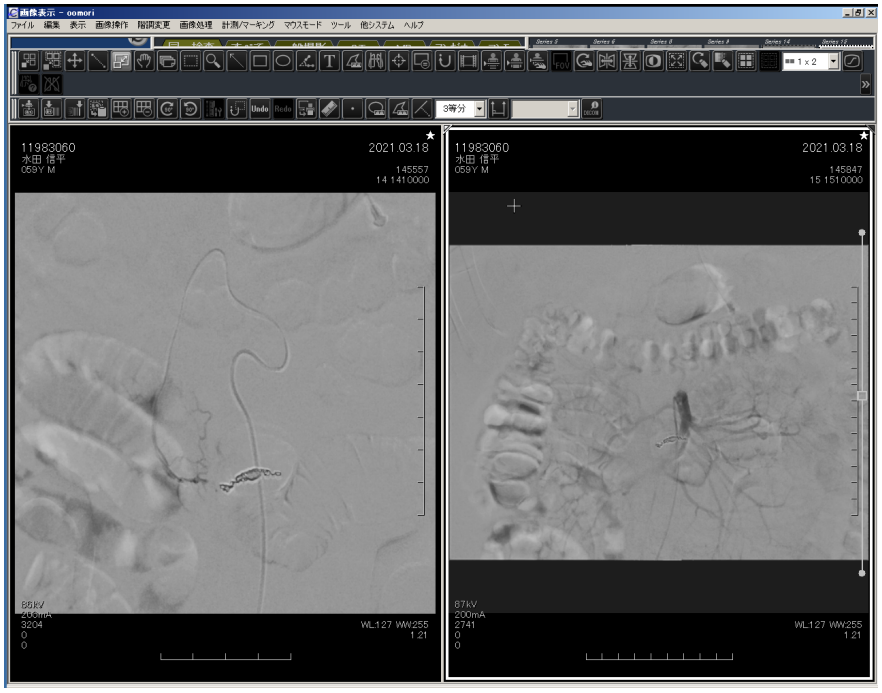

Table 1: Reported cases of subarachnoid hemorrhage associated with intra-abdominal hemorrhage owing to Segmental arterial mediolysis (SAM).

No.	Author, year	Age, sex	Intracranial aneurysm	Aneurysm type	Site	WFNS grade	Treatment intracranial aneurysm	Symptoms before in-of traperitoneal hemorrhage	Visceral aneurysm	Duration of intraperitoneal hemorrhage after SAH (days)	Cerebral infraction due to cerebral vasospasm
1	Fuse et al., 1996 [15]	56, F	Left ICA	Saccular	II	Clipping	Abdominal pain	Gastroepiploic, gastric artery	16		N.A.
2	Sakata et al.,2002 [5]	48,M	RightVA, Left ICA	Dissection	V	None	N.A.	Superior mesenteric artery, left external iliac artery	N.A.		N.A.
3	Stetler et al., 2012 [16]	59, F	Right IC-PC	Saccular	III	IVR	Abdominal pain	Hepatic artery	3		None
4	Shinoda et al., 2016 [6]	47, M	Left VA	Fusiform	V	IVR	Abdominal pain	Middle colic artery	8		N.A.
5	Welch et al., 2017 [7]	61, M	PSA	Fusiform	N.A.	IVR	Anemia	Splenic artery	0		N.A.
6	Hellstern et al., 2017 [8]	30,M	Both ICA	Dissection	IV	IVR	Shock	Splenic artery	0		N.A.
7	Hayashi et al.,2018 [2]	49, F	Left ICA	Saccular	II	Clipping	Abdominal pain	Splenic artery	4		Left temporal lobe infraction
8	Isaji et al., 2018 [9]	45, M	Right VA	Dissection	II	IVR	Shock	Superior mesenteric artery	8		None
9	Ohara et al., 2019 [10]	82, F	Left VA-PICA	Fusiform	II	IVR	Shock	Hepatic artery	14		Cerebellar infraction

No.	Author, year	Age, sex	Intracranial aneurysm	Aneurysm type	Site	WFNS grade	Treatment intracranial aneurysm	Symptoms before in-of traperitoneal hemorrhage	Visceral aneurysm	Duration of intraperitoneal hemorrhage after SAH (days)	Cerebral infraction due to cerebral vasospasm
10	Inazuka et al., 2019 [17]	77, F	Right ICA	BBA	II	Clipping	Anemia	Celiac artery, splenic artery	8	None	
11	Hori et al., 2020 [18]	67, F	Right IC-PC	Saccular	II	Clipping	Abdominal pain	Splenic artery	14	Right frontal lobe infraction	
12	Tanaka et al., 2020 [19]	78, M	Right ICA	BBA	III	Trapping with bypass	Shock	Posterior inferior pancreaticoduodenal artery	12	Right ICA territory infraction	
13	Ota et al., 2020 [20]	54,M	Acom	Saccular	IIorIII	Clipping	Shock	Right gastroepiploic artery	6	None	
14	Present case, 2021	59, M	Left MCA	Saccular	IV	Clipping	Shock	anterior superior pancreatoduodenal artery	9	Left MCA territory infraction	

Table legends

Acom: anterior communicating artery, BBA: blood blister-like aneurysm, F: female, ICA: internal carotid artery, IC-PC: internal carotid-posterior communicating artery, IVR: interventional radiology, M: male, MCA: middle cerebral artery, mRS: modified Rankin Scale, NA: not available, PICA: posterior inferior cerebellar artery, PSA: posterior spinal artery, SAH: subarachnoid hemorrhage; VA: vertebral artery, WFNS: World Federation of Neurological Surgeons.