Segmental arterial mediolysis is a rare nonarteriosclerotic and noninflammatory vascular disease first described by Slavin and Gonzalez-Vitale in 1976. In most cases, patients with SAM may present with intraabdominal hemorrhage caused by the rupture of visceral aneurysms. To date, only 3 cases of intracranial and intraabdominal aneurysms coexisting in the same patient have been reported. This is a rare case report of SAH caused by rupture of an ACA aneurysm associated with SAM.

**Case report**

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The authors report the rare case of a 58-year-old man with segmental arterial mediolysis (SAM) with associated intracranial and intraabdominal aneurysms, who suffered subarachnoid hemorrhage (SAH) due to rupture of an intracranial aneurysm. This disease primarily involves the intraabdominal arterial system, resulting in intraabdominal and retroperitoneal hemorrhage in most cases. The patient presented with severe headache and vomiting. The CT scans of the head revealed SAH. Cerebral angiography revealed 3 aneurysms: 1 in the right distal anterior cerebral artery (ACA), 1 in the distal portion of the A1 segment of the right ACA, and 1 in the left vertebral artery. The patient had a history of multiple intracranial aneurysms involving the splenic, gastroepiploic, gastroduodenal, and bilateral renal arteries. He underwent a right frontotemporal craniotomy and fibrin coating of the dissecting aneurysm in the distal portion of the A1 segment of the right ACA, which was the cause of the hemorrhage. Follow-up revealed no significant changes in the residual intracranial and intraabdominal aneurysms. An SAH due to SAM with associated multiple intraabdominal aneurysms is extremely rare. The authors describe their particular case and review the literature pertaining to SAM with associated intracranial and intraabdominal aneurysms.

(key words: • subarachnoid hemorrhage • segmental arterial mediolysis • intracranial aneurysm • intraabdominal aneurysm • anterior cerebral artery • vascular disorders)

SEGMENTAL arterial mediolysis is a rare nonarteriosclerotic and noninflammatory vascular disease first described by Slavin and Gonzalez-Vitale in 1976. In most cases, patients with SAM may present with intraabdominal hemorrhage caused by the rupture of visceral aneurysms.

To date, only 3 cases of intracranial and intraabdominal aneurysms coexisting in the same patient have been reported. This is a rare case report of SAH caused by rupture of an ACA aneurysm associated with SAM.

Abbreviations used in this paper: ACA = anterior cerebral artery; ACoA = anterior communicating artery; CCA = common carotid artery; SAH = subarachnoid hemorrhage; SAM = segmental arterial mediolysis; VA = vertebral artery.

Case Report

**History.** This 58-year-old man with a history of hypertension presented with chronic gastritis at a local clinic. He was referred to the Department of Radiology at our hospital after abdominal ultrasonography revealed evidence of a splenic artery aneurysm. Digital subtraction angiography of the abdomen revealed multiple intraabdominal aneurysms involving the splenic, gastroepiploic, gastroduodenal, and bilateral renal arteries (Fig. 1). Laboratory screening ruled out the presence of vasculitis, after
Intracranial and intraabdominal aneurysms coexisting with SAM

which SAM was considered as a possible diagnosis for his condition. The patient, however, refused endovascular treatment for the largest aneurysm involving the splenic artery, and returned to the local clinic to continue his previous treatment. Three months after the abdominal angiography study performed at our hospital, he returned to the local clinic suffering from a severe headache and vomiting. A CT scan of the head revealed an SAH that was dominant in the basal cistern and left sylvian fissure (Fig. 2). However, the physician at the local clinic overlooked these findings and prescribed NSAIDs without diagnosing the SAH. The patient returned home with the medication; however, the headache persisted and he consulted another hospital, where MR angiography of the head revealed multiple intracranial aneurysms in the right distal ACA and left VA. He was eventually transferred to our hospital for further treatment.

Examination. On examination, no neurological deficits were observed. Cerebral angiography revealed an aneurysm in the distal portion of the A1 segment of the right ACA in addition to those in the right distal ACA and left VA (Fig. 3). The patient subsequently received a diagnosis of SAH caused by rupture of the aneurysm in the A1 segment of the right ACA.

Operation. The patient underwent a right frontotemporal craniotomy to prevent rerupture of the aneurysm.

During surgery, the aneurysm was observed to be dissecting rather than saccular (Fig. 4). Intraoperative indocyanine green videography demonstrated no collateral flow via the ACoAs when we temporarily occluded the proximal segment of the right A1. We could not trap the dissecting aneurysm; therefore, coating with fibrin glue (Bolheal) and oxidized cellulose (Surgicel) was performed.

Postoperative Course. The patient subsequently resumed his normal life with no neurological deficits. Follow-up MRI studies of the head obtained 12 months after surgery revealed no brain damage due to microsurgical intervention (Fig. 5), and MR angiography of the head revealed no significant changes in the residual intracranial aneurysms in this period (Fig. 6). The patient has been observed for 12 months and has no neurological deficits.

Discussion

A rare nonarteriosclerotic and noninflammatory vascular disease, SAM was first described by Slavin and
This disease is characterized by necrosis of the outer tunica media–adventitia junction and replacement of the lysed muscle fibers by fibrin, erythrocytes, and granulation tissue. Transmural mediolyis results in arterial wall defects that can lead to focal dissection, aneurysm formation, stenosis, or hemorrhage caused by rupture of the artery. In most cases, patients with SAM present with intraabdominal hemorrhage caused by rupture of a visceral aneurysm. The most common sites of visceral arterial aneurysms are the splenic, hepatic, superior mesenteric, celiac, gastric, gastroepiploic, jejunal, ileal, and colonic arteries and their tributaries. Inada et al. reported 27 cases of SAM in Japan, of which 33.3% had multiple intraabdominal aneurysms similar to our case.

To date there are only 3 reported cases of intracranial and intraabdominal aneurysms coexisting in the same patient. Sakata et al. reported an autopsy case of SAM involving the right VA, left ICA, renal, superior mesenteric, and left external iliac arteries. The patient died of SAH as a result of a ruptured intracranial arterial dissection, and there was an accessory finding of intraabdominal arterial dissection. Obara et al. reported a case of a cervical ICA aneurysm with mural thrombosis resulting in cerebral infarction, which eventually led to hemiparesis. This patient also had multiple visceral aneurysms involving the celiac, hepatic, and superior mesenteric arteries. He was successfully treated with reconstructive surgery by using autologous vein grafts for each aneurysm. The third case, an autopsy case of SAM involving the right gastroepiploic, left gastric, and right VA, was reported by Ro et al.

The differential diagnosis of SAM includes various systemic vasculitis entities, such as polyarteritis nodosa, Takayasu disease, Behçet disease and Henoch-Schönlein purpura, congenital structural vascular disease such as Ehler-Danlos syndrome, and mycotic aneurysms. In our case, laboratory screening and the patient’s general condition excluded systemic vasculitis and congenital structural vascular disease. Moreover, regarding mycotic aneurysms, the patient had no previous infectious disease. Our clinical and radiological findings are compatible with SAM, although we have no pathological examination available, such as previously reported.

The natural history of unruptured aneurysms is poorly understood, and treating such aneurysms is controversial. Sakano et al. reported that follow-up angiography at 6 months revealed an absence of unruptured intraabdominal aneurysms. In our patient, there were 2 unruptured intracranial aneurysms (1 each in the distal ACA and left VA) at the time of discharge. We routinely monitor these unruptured aneurysms every 3 months, and have seen no interval changes to date.

**Disclosure**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Matsuda, Hironaka. Acquisition of data: Matsuda. Analysis and interpretation of data: Matsuda. Drafting the article: Matsuda, Hironaka. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Matsuda.

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