A case of ruptured segmental arterial mediolysis of the hepatic artery : Report of a Case

Tetsuo Tomonaga[1],Susumu Eguchi[1], Masaaki Hidaka[1], Mitsuhisa Takatsuki[1], Akihiko Soyama[1], Izumi Muraoka[1], Akimasa Mizutani[2], Hiroshi Fukui[2], and Takashi Kanematsu[1]

[1]Department of Surgery, Nagasaki University Graduate School of Biomedical Sciences, 1-7-1 Sakamoto, Nagasaki 852-8501, Japan and [2]Nagasaki Memorial Hospital, Nagasaki, Japan

Address for correspondence:

Susumu Eguchi, Department of Surgery, Nagasaki University Graduate School of Biomedical Sciences, 1-7-1 Sakamoto, Nagasaki 852-8501, Japan

Telephone number: +81-95-819-7316

Facsimile number: +81-95-819-7319

E-mail: sueguchi@nagasaki-u.ac.jp

Runnning title: Segmental arterial mediolysis of the hepatic artery

Type of the paper: Case report

Key words: segmental arterial mediolysis, hepatic artery, rupture

Abbreviations: segmental arterial mediolysis (SAM); alpha-fetoprotein (AFP); protein induced by vitamin-K absence (PIVKA-II); computed tomography (CT)

ABSTRACT

This article reports a patient with intra-abdominal hemorrhage secondary to a rare vascular disease, segmental arterial mediolysis (SAM). The patient was a 68-year-old male who presented with chilling and severe abdominal pain. Abdominal computed tomography results suggested the presence of an intra-abdominal hemorrhage. Although visceral angiography illustrated multiple aneurysms in the branches of the hepatic artery, active bleeding was not evident. Conservative therapy including transfusion was performed, and re-angiography revealed the disappearance of multiple hepatic arterial aneurysms 8 months later. This is the first case of intra-abdominal hemorrhage related to SAM of the hepatic artery in which natural history of SAM was seen.

Key words: Segmental arterial mediolysis; Hepatic aneurysm; spontaneous intra-abdominal hemorrhage

INTRODUCTION

Segmental arterial mediolysis (SAM) is a rare nonarteriosclerotic, noninflammatory vascular disease of unknown origin that involves the visceral arteries of the abdomen. Lesions typically occur in a skip pattern within the large abdominal arteries and have no predilection for bifurcations. SAM primarily affects the outer layer of the media, leading to smooth muscle cell vacuolar degeneration. The disruption of vacuoles and concomitant loss of their fluid contents ultimately results in disruption of the media, intramural hemorrhage, and periadventitial fibrin deposition. Gaps may be filled with fibrin, thrombi, or granulation tissue and can lead to saccular aneurysms, dissecting aneurysms, or thrombosis. The intima is spared from these lytic changes, and there is minimal inflammation (1).

SAM was first described in the literature by Slavin in 1976.This disease can involve a variety of arterial systems in adults and most commonly affects the abdominal vasculature. When the splanchnic vasculature is involved, the typical clinical manifestation is an acute process

including intestinal ischemia, infarction, and/or intra-abdominal hemorrhage. During the past 33 years, approximately 35 cases of abdominal SAM involving adults have been reported in the literature (2). To our knowledge, the present case is the first treated case of SAM affecting the hepatic artery in which the natural history of SAM could be observed.

REPORT OF A CASE

A 68-year-old previously healthy Japanese male presented to our referral hospital with chills and severe abdominal pain. The sharp abdominal pain began gradually and was focused in the right upper abdomen. Physical examination revealed an uncomfortable male with mild tachycardia, and right upper quadrant abdominal pain with guarding. Abnormal laboratory studies at presentation included a white blood cell count of 24,770/mm^3 with a differential of 82% neutrophils. Hb 9.7 g/dl Plt 278,000/mm^3 AST 2421 IU/l ALT 4607 IU/l BUN 37.7 mg/dl Cre. 3.8 mg/dl, CRP 17.8 mg/dl.

An abdominal computed tomography (CT) scan (Fig.1) showed a large loculated fluid collection extending around the liver, spleen, and Douglas pouch. A high-density area appearing to be hematoma was observed in the post area of the right liver lobe. The mass-like structure was not uniformly observed on the front side of the right lobe under the dome of the right liver lobe. The primary diagnosis was therefore ruptured hepatocellular carcinoma. Because the renal

function was deteriorated, the CT was refrained from contrast media. The patient's hemodynamics became stable, and he was admitted for fluid-infusion.

The next day, his anemia was aggravated (Hb 9.7→8.7 g/dl). An abdominal CT scan showed increased fluid collection, which led to a transfusion with 4 units of the erythrocyte. As for the tumor markers, alpha-fetoprotein (AFP):1.8 ng/ml(＜20)・protein induced by vitamin-K absence (PIVKA-II):60ng/ml(＜20). Hepatitis virus markers were all negative. He did not have the polydipsia of alcohol. Because the cause was undetected, he was referred to our hospital for further treatment.

Given these findings, the patient underwent visceral angiography (Fig. 2), which showed normal celiac and superior mesenteric arteries but abnormal hepatic artery. The hepatic artery itself contained regions of aneurismal dilation interspersed with focal and long segmental stenosis. Active bleeding, however, was not present. Since the patient's hemoglobin decreased considerably shortly after admission, he was kept hemodynamically stable with blood products

and intravenous fluid resuscitation. Conservative treatment was performed with blood transfusion, prescription of a vasopressor, and respiratory-care by endotracheal-intubation because aneurysms and stenosis were distributed at a hole liver.

On the 5th hospital day, the decrease in Hb ceased, and abdominal paracentesis was carried out under the guide of ultra sonography for drainage of intra-abdominal blood. On the 9th hospital day, the endotracheal tube was extubated. On the 15th hospital day, abdominal CT was performed after we observed an improvement in renal function in the blood sample.

Because there were no signs of fever or abdominal pain, the patient was transferred to a referral hospital on the 29th hospital day for the purpose of follow-up observation. After the patient transfer, the patient was discharged on the 42nd hospital day. Approximately 8 months later, a follow-up abdominal CT was performed. Hematoma in the hepatic right lobe had decreased, and the hepatic subcapsular hematoma had been obscured. Intense expansive change in the hepatic artery could no longer be observed in the CT (Fig. 3). Currently, the patient has no

specific symptoms and enjoys good health.

DISCISSION

The two leading causes of intra-abdominal hemorrhage are blunt trauma and benign gynecologic disease (3). Less frequent etiologies of intra-abdominal hemorrhage include neoplastic, inflammatory, and vascular lesions (4). The vascular disorders include variceal rupture in patients with portal hypertension, inflammatory vasculitides such as polyarteritis nodosum and rupture of aortic or mesenteric aneurysms. Our patient case exhibits an exceedingly rare etiology of intra-abdominal hemorrhage due to SAM.

The initial autopsy and case reports by Slavin and others have described “segmental mediolytic arteritis” as a previously unknown disease involving peculiar lesions within the large abdominal arteries (5-7). However, Gruenwald defined a similar lesion in 1949 in the coronary arteries of neonates (8). In 1992, the name of the disease was changed to segmental arterial

mediolysis (SAM) because of the apparent lack of an inflammatory process (6,9).

In the beginning, rupture of hepatocellular carcinoma or metastatic liver cancer was suspected in the present case from the diagnostic imaging (plain abdominal CT, echo). Possibly being affected by hypotensive shock after intraperitoneal hemorrhage, the patient already had renal dysfunction on initial presentation. After patient transfer, CT of the abdominal artery using carbon dioxide was performed immediately due to his impaired renal function. Multiple aneurysms were then observed in the bifurcation of the hepatic artery, and the patient was diagnosed with SAM due to its multiple and beaded appearance. Embolization was not performed for there was no further active hemorrhage in the angiography. We investigated the possibility of excising the hepatic right lobe where coagulation had been detected. However, a conservative treatment of blood transfusion and administration of vasopressor was followed because of the following factors: (1) no active hemorrhage was observed; (2) the patient's general condition (vital signs); (3) there was an increased risk of hepatic failure if aneurysms ruptured in the

remaining (left) lobe after excising the right lobe, since aneurysms were detected in both hepatic lobes. Erythrocyte transfusion was appropriately performed until the 4th hospital day. We inferred that hemostasis on hematoma was complete because the progress of anemia had stopped, as was shown in the blood sample results on the 5th hospital day. A closed drain was therefore placed into the peritoneal cavity from the right lower quadrant to initiate continuous suction of hematoma because of the fear of abdominal infection (the drain was extubated on the 25th hospital day).

The hepatic function improved gradually, while the renal function improved within a few days after the initiation of strict circulation control with blood transfusion after patient transfer. However, CT was performed on the 15th hospital day and the remaining aneurysms were detected in both hepatic lobes. There was no active hemorrhage. Aneurysms were still observed in both hepatic lobes, but hepatic excision was not performed. On the 29th hospital day, the patient was transferred on foot. After the treatment at the new hospital, the patient was discharged on the 42nd

hospital day.

Approximately 6 months after symptom onset, the patient had almost no subjective symptoms and returned to his work as a fisherman. A blood sample indicated a reference interval of hepatic function. Abdominal CT indicated a small number of remaining hematomas and irregularity of the superficial liver, although aneurysms previously observed in both hepatic lobes had disappeared.

In conclusion we experienced a case of SAM that was specific to the hepatic artery. The patient in this case was able to survive a case of the disease where aneurysms had spread extensively in both hepatic lobes in response to conservative treatment with strict circulation control using transfusion and vasopressor and abdominal drainage at an appropriate timing. SAM is called a 'one shot disease' and can be survived once the patient overcomes the acute phase. However, it is still a dangerous pathology with high fatality, and further accumulation and investigation of the case are thought to be essential.

REFERENCES

1. Maren Michael,Urs widemer,Thomas Pfammatter et al.Segmental Areterial Mediolysis:CTA Findings at Presentation and follow-Up.AJR:187,December 2006;1463-1469.

2. Daniel S. Rengstorff,Edward L. Baker,Laurerence F. Yee et al.Intra-abdominal Hemorrhage Caused by Segnebtal Arteial Medyolysis of the Inferior Mesenteric Artery:Report of a Case.Dis Colon rectum,May 2004;769-772.

3. Ellis H, Griffiths PW, McIntyre A. Haemoperitoneum: a record of 129 consecutive patients with notes on some unusual cases. Br J Surg 1958;45:606-10.

4. Akriviadis EA. Haemoperitoneum in patients with ascites. Am J Gastroenterol 1997;92:567-75.

5. Heritz DM, Butany J, Johnston KW, Sniderman KW. Intraabdominal hemorrhage as a result of segmental mediolytic arteritis of an omental artery: case report. J vasc Surg 1990;12:561-5.

6. Leu HJ. Cerebrovascular accidents resulting from segmental mediolytic arteriopathy of the cerebral arteries in young adults. Cardiovasc Surg 1994;2:350-3.

7. Slavin RE, Cafferty L, Cartwright J. Segmental mediolytic arteritis, a clinicopathologic and ultrastructural study of two cases. Am J Surg Pathol 1989;13:558-68.

8. Guenwald P. Necrosis in the coronary arteries of newborn patients. Am Heart J 1949;38:889-97.

9. Armas OA, Donovan DC. Segmental mediolytic arteritis involving hepatic arteries. Arch Pathol Lab Med 1992;116:531-4.

FIGURE LEGENDS

Figure 1

Computed tomography showing the hemorrhage in the right lobe of the liver.

Figure 2

Angiographic view showing massive segmental arterial aneurysma in all the hepatic arteries (left).

The superior mesenteric artery showed no abnormality (right).

Figure 3

Computed tomographic view and arterial reconstruction. Hematoma in the right lobe of the liver was shrunken (right). Hepatic arteriogram showed a disappearance of segmental arterial aneurysma in the hepatic arteries (left).

Fig.1

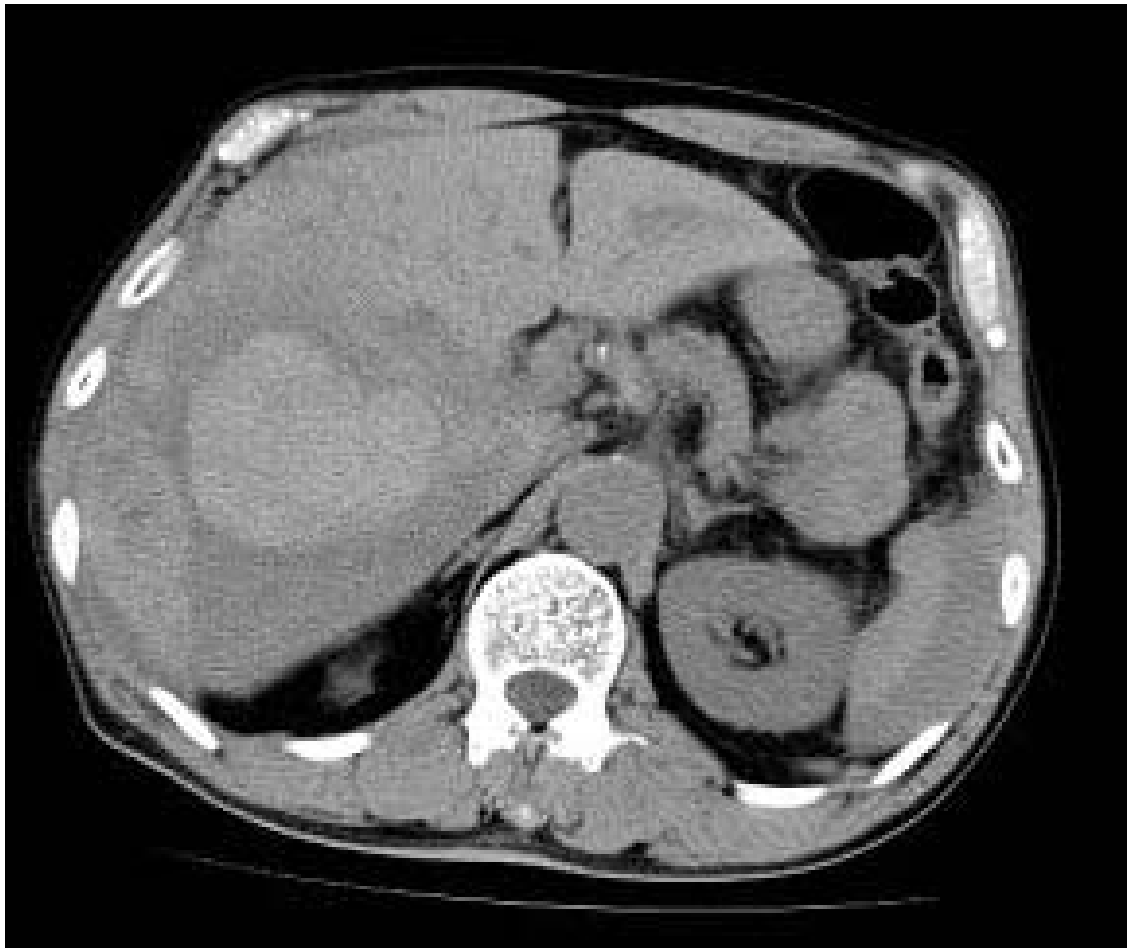
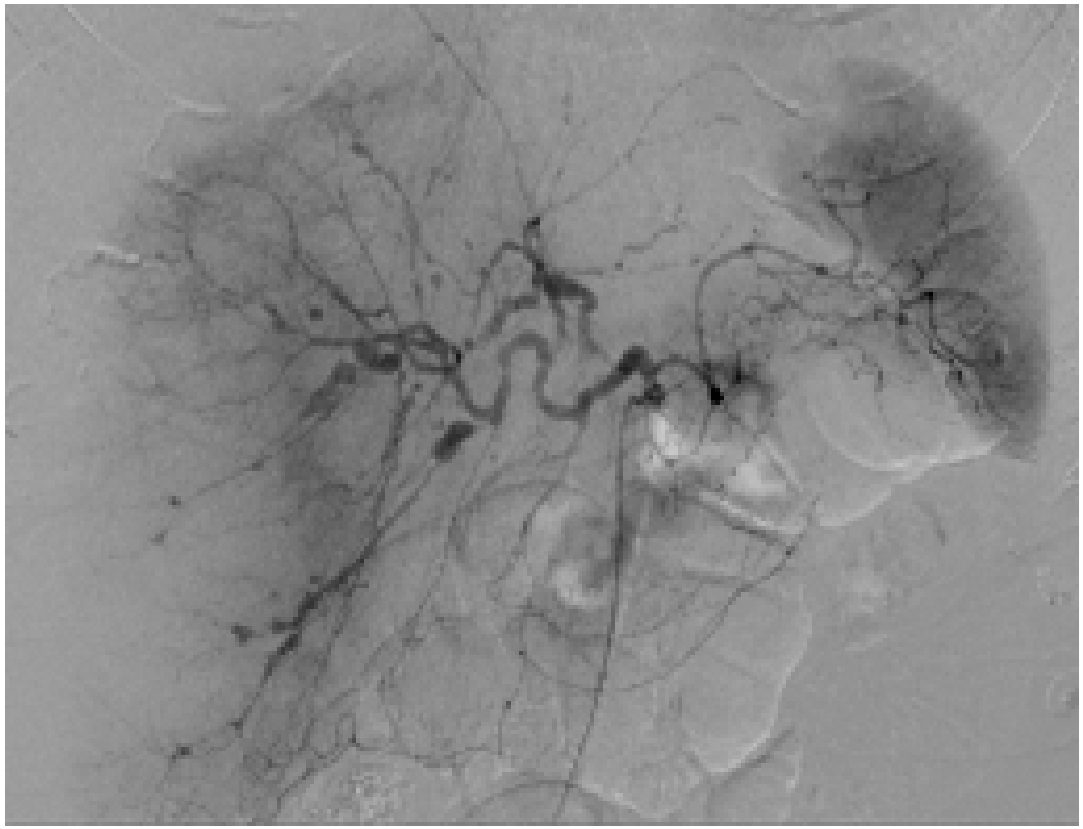


Fig.2



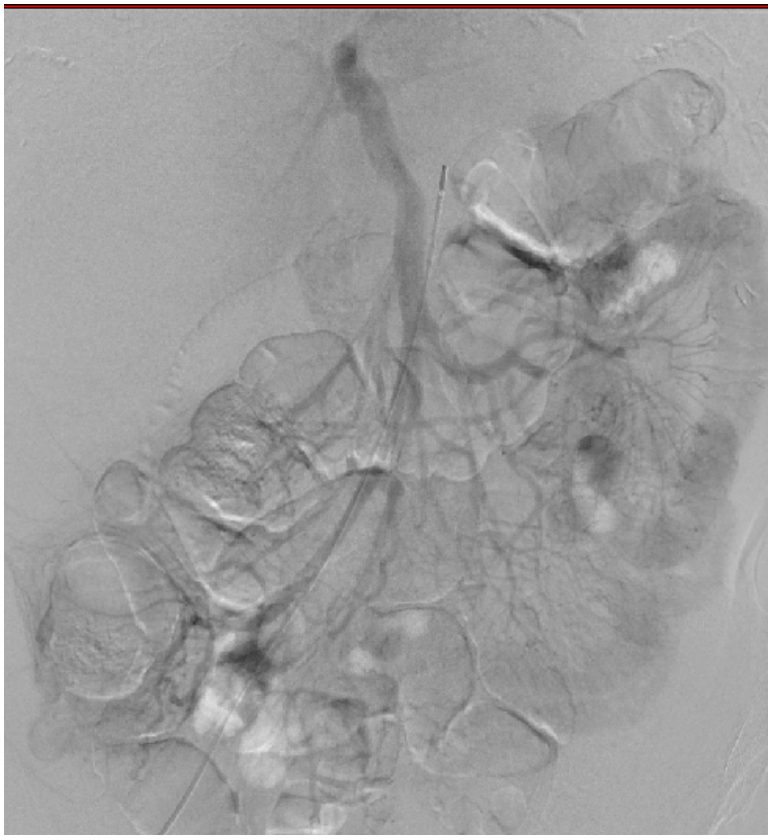


Fig. 3　8 months after the rupture