Spontaneous Recovery of Multiple Hepatic Artery Aneurysms with Segmental Arterial Mediolysis

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Abstract
A 50-year-old woman with liver dysfunction complained of back pain. Computed tomography showed multiple fusiform aneurysms in the right and left hepatic arteries. As she was hemodynamically stable, antihypertensive therapy was selected to reduce the risk of rupture. During hospitalization, spontaneous and progressive thrombosis formation of multiple hepatic artery aneurysms was observed. She was diagnosed with segmental arterial mediolysis based on her clinical course and imaging findings, and she was discharged after 48 days. One year following discharge, computed tomography showed complete recovery and patent, normal hepatic arterial branches. Segmental arterial mediolysis should be considered as a condition that can cause multiple hepatic artery aneurysms, which can be treated successfully with antihypertensive therapy and careful follow-up observation with imaging when the patient’s hemodynamic state is stable.

Key words: case reports, segmental arterial mediolysis, multiple hepatic artery aneurysms, treatment

Introduction
Segmental arterial mediolysis (SAM) is characterized by multiple aneurysmal dilatations and stenosis induced by idiopathic dissections, and it can potentially cause spontaneous hemorrhage [1, 2]. SAM is also known as the “one-shot disease” because it sometimes results in spontaneous recovery; however, it can be fatal, and interventional or surgical treatment is required if rupture occurs [1, 3-5]. Therefore, the treatment strategy for SAM remains controversial. Herein, we describe a patient who spontaneously recovered from multiple hepatic artery aneurysms and was eventually clinically diagnosed with SAM.

Case report
A 50-year-old woman with a history of hypertension and Sjögren syndrome complained of general fatigue and fever. She was previously diagnosed with sepsis, disseminated intravascular coagulation, and meropenem hydrate-induced acute liver dysfunction, and she received conservative care at a local hospital for 2 weeks. However, she complained of acute, severe back pain and left flank pain 1 day after discharge. A contrast-enhanced computed tomography (CT) scan showed multiple fusiform aneurysms (maximum diameter, 25 mm) in the left and right hepatic arteries (Figures 1a, b), and soft tissues were adjacent to the aneurysms or portal vein, showing hyperattenuation in non-contrast-enhanced CT and hypoattenuation in contrast-enhanced CT, indicating hematoma or thrombosis of aneurysms (Figures 1c, d, e). Extravasation of contrast media was not observed. She was transferred to our hospital’s emergency room. Upon admission, she had mild fever (37.4°C-37.7°C), but was not tachycardic (heart rate, 82-92 bpm). Her blood pressure was slightly high (145/57 mmHg). Blood test results showed in-
From day 2 of hospitalization, the medication was administered intravenously to prevent rupture because aneurysm rupture was not excluded based on the CT findings. In addition, 10 mg of nicardipine hydrochloride was administered from the day of admission to day 7 of hospitalization considering the infection, 1 g ceftriaxone sodium hydrate was administered to prevent hyperattenuation on non-contrast-enhanced CT, indicating hematoma or thrombosis of an aneurysm (arrows).

Anemia was not observed (hemoglobin, 12.9 mg/dL). Concomitant findings included inflammatory reactions (white blood cell count, 13,500 × 10^3/μL; C-reactive protein concentration, 12.15 mg/dL), liver dysfunction (aspartate aminotransferase, 45 IU/L; alanine transaminase, 75 IU/L; lactate dehydrogenase, 236 IU/L), and coagulation disorders (fibrinogen, 549 mg/dL; fibrin degradation products, 5.3 μg/mL; D-dimer, 1.7 μg/mL), but anemia was not observed (hemoglobin, 12.9 mg/dL). Considering the infection, 1 g ceftriaxone sodium hydrate was administered from the day of admission to day 7 of hospitalization. In addition, 10 mg of nicardipine hydrochloride was administered intravenously to prevent rupture because aneurysm rupture was not excluded based on the CT findings. From day 2 of hospitalization, the medication was changed to 80 mg telmisartan and 12.5 mg atenolol. Her blood pressure decreased to 126/72 mmHg and 105/50 mmHg on days 2 and 3, respectively. The aneurysms did not enlarge; however, one aneurysm thrombosed on follow-up CT on day 3 (5 days after CT shown in Figure 1) of hospitalization (Figure 2). Several causes of hepatic artery aneurysm, such as vasculitis, SAM, fibromuscular dysplasia, and mycotic aneurysm, were suspected. Among them, mycotic aneurysm was less suspected because her vital signs were stable despite severe infectious disease, and procalcitonin test result (0.42 ng/mL) on day 3 of hospitalization was negative. However, we were unable to determine a definite diagnosis. Angiography was performed for differential diag-

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**Figure 1.** Computed tomography imaging at symptom presentation  
a, b: Contrast-enhanced computed tomography (CT) showing multiple fusiform aneurysms (maximum diameter, 25 mm) (arrows) in the left and right hepatic arteries.  
c, e: Soft tissues are noted adjacent to the aneurysms or the portal vein showing hyperattenuation on non-contrast-enhanced CT, indicating hematoma or thrombosis of an aneurysm (arrows).  
d: Soft tissues are noted adjacent to the portal vein showing hypoattenuation on contrast-enhanced CT, indicating hematoma or thrombosis of aneurysm (arrows).

**Figure 2.** Follow-up computed tomography on day 3 of hospitalization (5 days after the CT shown in Figure 1)  
a, b, d: Contrast-enhanced CT reveals no enlargement of the aneurysms or extravasation and demonstrates thrombosis of one of the aneurysms (arrows).  
c, e: Non-contrast-enhanced CT demonstrates thrombosis of one of the aneurysms (arrows).
Hepatic arteriography on day 12 of hospitalization. Hepatic arteriography showed multiple fusiform aneurysms in the left and right hepatic arteries, which had a “beaded appearance” in the left hepatic artery (Figure 3). No abnormality was observed in the splenic artery, superior mesenteric artery, and bilateral renal arteries, and extravasation of contrast media was not observed. We considered endovascular treatment, such as embolization and stent graft placement, for multiple hepatic artery aneurysms; however, SAM was strongly suspected from these imaging findings in addition to her clinical course. In addition, she was hemodynamically stable throughout her hospitalization. Therefore, instead of treating the aneurysms, we decided to continue antihypertensive therapy to reduce the risk of rupture. Furthermore, careful follow-up imaging was performed to monitor the size of the hepatic artery aneurysms.

The patient’s perception of pain improved 22 days after hospitalization, and magnetic resonance imaging (MRI) showed spontaneous and progressive thrombosis formation of the aneurysms in the right and left hepatic arteries compared with previous CT shown in Figure 2 (Figure 4). Contrast-enhanced CT scan performed on day 40 of hospitalization showed complete thrombotic occlusion accompanied with a reduction in size of the aneurysms in the left hepatic artery and partial thrombosis formation of the aneurysms in the right hepatic artery (Figures 5a, b). We diagnosed these lesions as SAM because 1) they developed in a middle-aged patient; 2) arteriography showed multiple fusiform aneurysms and a beaded appearance; and 3) aneurysms spontaneously thrombosed and reduced in size without interventional or surgical treatment.

The patient was discharged 48 days after hospitalization because abdominal pain completely improved, and we considered that the threat of rupture was minimal. In addition, her blood pressure was well controlled (122/91 mmHg) through oral administration of 0.5 mg amiodipine besylate alone. Contrast-enhanced CT scan performed 6 months after discharge showed complete recovery from multiple hepatic aneurysms and patent, normal hepatic arterial branches (Figures 5c, d), and the same results were observed on follow-up contrast-enhanced CT, 1 year after discharge (not shown); therefore, we finally diagnosed her with SAM.

Discussion

The pathological characteristics of SAM include mediolysis, which is the vacuolization and lysis of media, leading to arterial gap formation [1, 2]. These arterial gaps lead to intimal collapse, followed by arterial wall dissection and dissecting hematoma formation [1, 2, 6, 7]. The remaining adventitia becomes distended, leading to dissecting aneurysm formation [2, 6]. Sudden massive hemorrhage can occur if the aneurysms of the adventitia rupture [1, 5]. After the initial arterial injury (mediolysis), the restoration phase begins with growth of the granulation tissue in the arterial gaps. Subsequently, the granulation tissue is replaced with fibrosis to heal the arterial wall and help restore its shape [1, 2]. Complete or partial resolution of vascular abnormality or stabilization of SAM could be achieved over the natural disease course [4]. In our case, multiple intrahepatic aneurysms showed spontaneous and progressive thrombosis and reduction in size, which eventually disappeared completely in 6 months.

Abbas et al. reported that most hepatic artery aneurysms are single (92%) and extrahepatic (78%) [8]. Therefore, multiple and intrahepatic artery aneurysms are extremely rare. The differential diagnosis for spontaneous intrahepatic artery aneurysm includes atherosclerosis, infection, SAM,
arterial fibrodysplasia, vasculitis, polyarteritis nodosa, systemic lupus erythematosus, Wegener granulomatosis, Kawasaki disease, and neurofibromatosis [5, 8, 9]. The imaging features of SAM include arterial dissection, fusiform aneurysm, beaded appearance, dissecting hematoma, arterial wall thickening, arterial stenosis, and arterial occlusion [1, 2, 4]. Our case showed multiple fusiform intrahepatic artery aneurysms and a beaded appearance on initial CT and/or angiography. Based on these findings, the diagnosis of SAM was considered compatible.

To the best of our knowledge, only four cases of SAM involving the intrahepatic arteries have been previously reported [3, 5, 7, 9]. As the occurrence of SAM in the intrahepatic artery is extremely rare, physicians would likely not
consider SAM when assessing patients with hepatic artery aneurysm. For example, the first reported patient with SAM died because of hemorrhagic shock [7]. Of the three remaining patients, two underwent coil embolization for bleeding from the aneurysm and painful symptoms, or rupture of the aneurysm with fistulous connection to the portal vein [3, 9]. In contrast, Tomonaga et al. reported a patient with ruptured SAM who successfully underwent conservative treatment [5].

There is no standardized treatment strategy for SAM. There are several treatment options: surgery; endovascular treatments, such as transcatheter arterial embolization (TAE), angioplasty, and stent placement; and conservative treatment [1, 3, 4]. TAE is indicated if active bleeding due to rupture of the aneurysm occurs [1, 3]. However, TAE in patients with stable hemodynamic status should be carefully considered because new arterial wall dissection and aneurysm rupture may be caused by catheter manipulation [1]. Stent placement for SAM is reportedly extremely rare [3], and organ ischemia may be a potential indication. However, definite indication of stent placement is not established. Theoretically, stent graft placement could be another treatment option among endovascular treatments for ruptured SAM. To the best of our knowledge, however, stent graft placement had not been reported to date. Generally, placement of thicker device is necessary for stent graft than placement of bare stent, and the stent (graft) must be placed at least between an angiographically "healthy to healthy" segment. However, it might worsen (new dissection and rupture) SAM. Thus, we consider that stent graft placement has lesser advantage than TAE for the treatment of ruptured SAM.

Our patient had multiple hepatic artery aneurysms in both liver lobes. We considered endovascular treatment, such as embolization and stent graft placement; however, SAM was strongly suspected from the imaging findings, in addition to her clinical course. Moreover, her hemodynamic status was stable. Therefore, instead of treating the aneurysms, we decided to continue antihypertensive therapy and perform careful follow-up observation with imaging. With regard to conservative treatment, the usefulness of antihypertensive therapy has been noted [1, 4, 10]. Pillai et al. reported that patients with SAM who presented with concurrent abdominal pain without intra-abdominal hemorrhage should be managed conservatively [1]. Approximately 40% of patients with SAM will have new lesions during the natural disease course; thus, careful follow-up observation with imaging is necessary [4].

In conclusion, SAM should be considered as one of the conditions that can cause multiple hepatic artery aneurysms. SAM can be successfully treated with antihypertensive therapy and careful follow-up observation with imaging when the patient’s hemodynamic state is stable.

Conflict of Interest: Ryo Morita, Daisuke Abe, Takeshi Soyama, Yusuke Sakuhara, Masayoshi Kajiyama, Kohsuke Kudo declares that they have no conflicts of interest.

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References