

## Spontaneous Celiac and Splenic Artery Dissection

Takahiro ZENDA<sup>1,\*</sup>, Ichiro ARAKI<sup>1</sup>, Naomichi HAMANO<sup>1</sup>, Hiroto NISHIDA<sup>2</sup>,  
 Masatoshi IKEDA<sup>3</sup>, Hisashi BUNKO<sup>4</sup> and Tetsuya AOKI<sup>5</sup>

1) Department of Internal Medicine, Asanogawa General Hospital, Kanazawa, Japan

2) Department of Radiology, Asanogawa General Hospital, Kanazawa, Japan

3) Department of Cardiology, Kanazawa Cardiovascular Hospital, Kanazawa, Japan

4) Department of Radiology, Kanazawa Cardiovascular Hospital, Kanazawa, Japan

5) Department of Internal Medicine, Naruwa Clinic, Kanazawa, Japan

### ABSTRACT

Dissection of the splanchnic artery unrelated to an aortic lesion is extremely rare. We describe a patient with dissection of the celiac and splenic arteries causing splenic circulatory impairment. A 55-year-old Japanese man was referred to our hospital for left back pain that suddenly occurred 3 days previously and spread to the left flank. He had complicated sleep apnea syndrome well controlled with continuous positive airway pressure, and had been prophylactically taking aspirin (100 mg/day) because of asymptomatic cerebral lacunar infarcts. Contrast-enhanced computed tomography (CT) in the arterial phase revealed dissection from the celiac root extending to the entire splenic artery, the caliber of which was irregularly narrowed, causing malperfusion in the spleen. Because of hemodynamic stability and lack of impending sequelae, the patient was carefully observed with rest, strict blood pressure control, and aspirin administration. One month later, CT revealed restoration of the caliber of the dissected arteries and regression of the organizing false lumen, which confirmed the patient's recovery. Despite the extreme rarity or nonspecific symptoms, splanchnic artery dissection should be considered a potentially life-threatening emergency. This case supports the possible benefit of starting antithrombotic treatment early to prevent thrombotic sequelae such as organ infarction and aneurysmal formation.

**Key words:** Splenic artery dissection, Contrast-enhanced computed tomography, Antithrombotic therapy, Splenic infarction

Spontaneous dissection of a splanchnic artery unrelated to the aorta is an extremely rare condition<sup>1,3,4,6,7,11,12,15-18</sup>; however, it has been increasingly reported especially in the last decade because of the development of diagnostic imaging modalities and their widespread use<sup>1,5-8,10-12,15-17</sup>. Among the splanchnic arteries, spontaneous dissection occurs most commonly in the superior mesenteric artery (SMA) followed by celiac artery; however, it is much rarer in the common hepatic artery or splenic artery<sup>7,16</sup>. To our knowledge, only 23 cases of splenic artery dissection have been appropriately reported<sup>1,4-6,8,10,11,16-18</sup>. Although computed tomography (CT) with contrast medium allows diagnosing splenic artery dissection dramatically easily<sup>4</sup>, the diagnosis remains difficult without a high index of suspicion<sup>4</sup> and misdiagnosis as other acute abdominal diseases may occur<sup>17</sup>. This is because of its rarity, unpredictable natural history<sup>12,17</sup>, absent or nonspecific symptoms<sup>5,6,8</sup> and acute fulminant

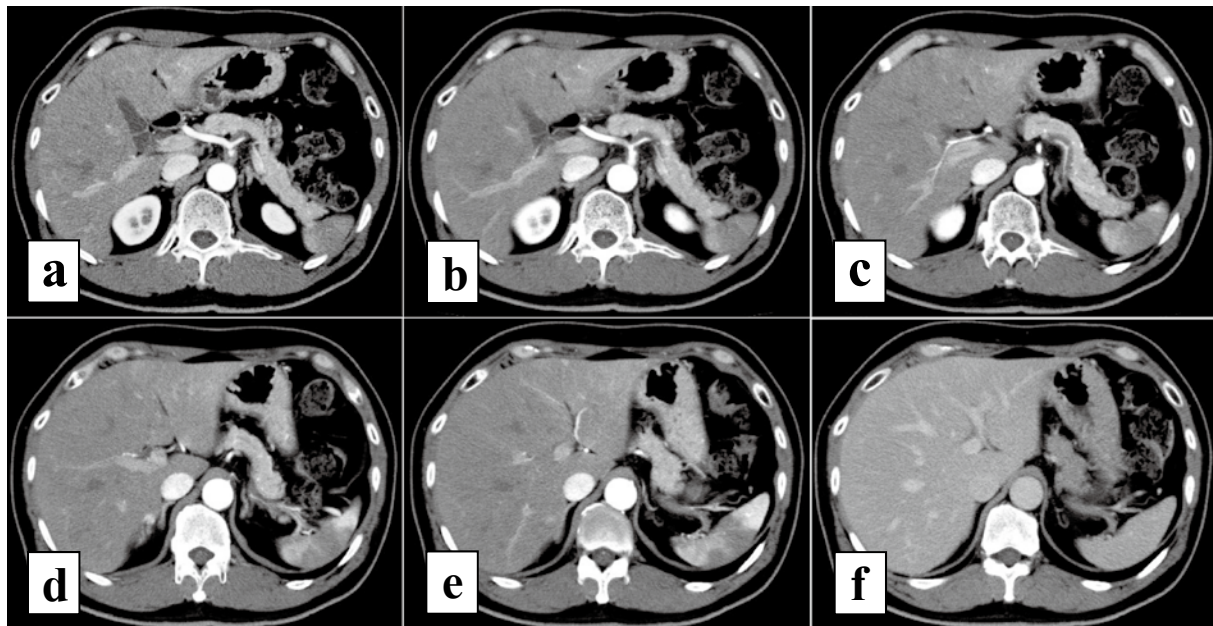
manifestation as sudden death<sup>1</sup>.

Herein, we describe an extremely rare and diagnostically confusing case of spontaneous dissection from the celiac trunk to the entire splenic artery causing circulatory insufficiency in the spleen. In addition, the possible beneficial effect of anti-thrombotic treatment in the acute phase of arterial dissection is discussed.

### CASE REPORT

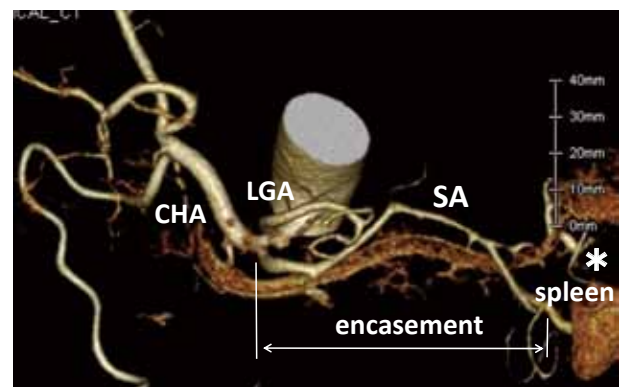
A 55-year-old Japanese man was first referred to our emergency room because of a sudden onset of middle and left-sided back pain that occurred while he was lying on his back when receiving dental care. He had no history of intense exercise or abdominal bruising before the back pain occurred. He was a non-smoker at the time of presentation but had smoked up to 20 cigarettes a day for 34 years until 1 year prior. He had been receiving

\* Address correspondence to: Takahiro Zenda, MD, PhD  
 Department of Internal Medicine, Asanogawa General Hospital, 83 Kosaka-machi-naka, Kanazawa 920-8621, Japan  
 Tel: +81-76-252-2101, Fax: +81-76-252-2102, E-mail: zenda@asanogawa-gh.or.jp



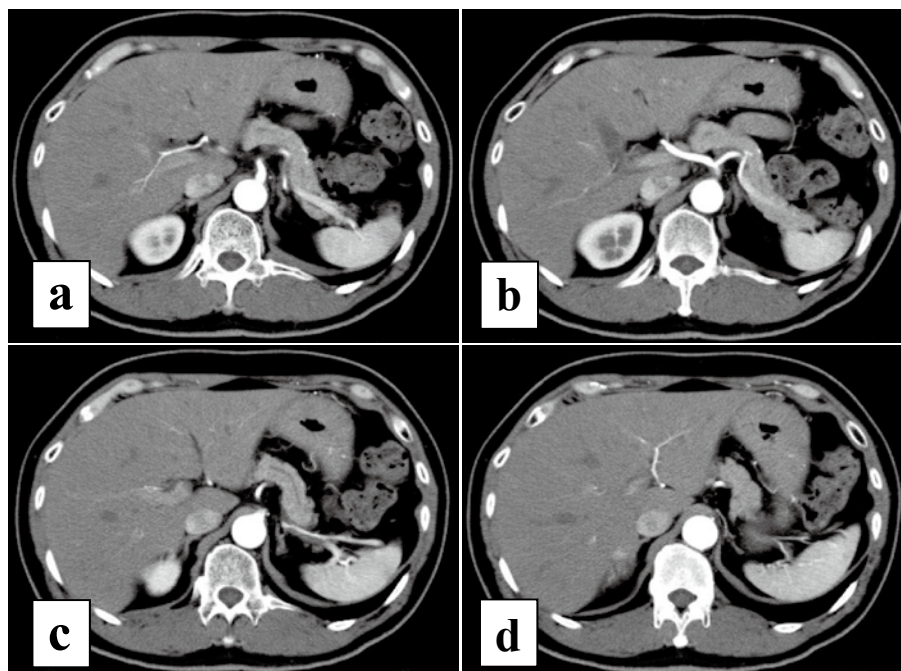
**Fig. 1.** Contrast-enhanced computed tomography in the dynamic arterial phase revealed dissection from the root of the celiac trunk (a, b) and the entire splenic artery to the hilum of the spleen (c-e). The true lumen was narrowed by encasement of a low-density area, which probably reflected the organized false lumen due to thrombus. Either the common hepatic artery or the left gastric artery seemed to be free from the dissection. In the dynamic arterial phase, malperfusion regions were clearly seen in the spleen; however, these ischemic changes were restored in the equilibrium phase (f), suggesting that the spleen did not become infarcted.

treatment for sleep apnea syndrome (SAS) for 10 years, and was in good condition owing to continuous positive airway pressure. In addition, he had been prophylactically taking aspirin (100 mg/day) for 3 years before because of cerebral lacunar infarctions that were asymptomatic and detected with magnetic resonance imaging during a medical health check. At presentation, his general condition was good, with blood pressure of 142/84 mmHg and heart rate of 76 beats/min. Physical examination, electrocardiography, chest and abdominal roentgenography, or abdominal ultrasonography without Doppler analysis all revealed no findings to explain his back pain. A peripheral blood cell count did not indicate anemia but showed mild leukocytosis (10,200/ $\mu$ l) with normal differentiation. Except for a 206 mg/dl increase in low-density lipoprotein cholesterol, blood biochemistry showed no other abnormalities including inflammatory reactions related to C-reactive protein, renal function, liver enzymes, serum amylase, and troponin T. Urinalysis including sediment analysis was normal. On the basis of the patient's condition and the results of the examinations performed, he was served with a prescription of acetaminophen for the back pain. Acetaminophen transiently relieved the back pain; however, the pain gradually intensified and spread to the left flank region. Three days later, the patient underwent upper gastrointestinal endoscopy and abdominal ultrasonography performed by another physician; however, no lesion accounting for the pain was detected. Then, he was introduced to our hospital again for a closer



**Fig. 2.** Computed tomography angiogram demonstrating the irregularly narrowed caliber of the splenic artery from the celiac trunk to the splenic hilum. Malperfusion in the spleen is shown as a defect (asterisk). An ostium of the intimal laceration causing the dissection or an intimal flap was not identified. The common hepatic artery and left gastric artery seemed to be unaffected. CHA, common hepatic artery; CT, celiac trunk; LGA, left gastric artery; SA, splenic artery.

examination for the back and left-side flank pain. Contrast-enhanced CT in the dynamic arterial phase revealed entire splenic artery dissection from the root of the celiac trunk (Fig. 1a,b) to the hilum of the spleen (Fig. 1c-e). The true lumen of the splenic artery was narrowed by encasement of a low-density area, which probably reflected the organized false lumen due to a thrombus. Both the common hepatic artery and the left gastric artery seemed to be free from the dissection. In the dy-



**Fig. 3.** Contrast-enhanced computed tomography in the dynamic arterial phase 1 month later revealed enlargement of the caliber of the dissected splenic artery and regression of the encasing thrombotic pseudo lumen (a-c). The spleen was homogeneously enhanced from the arterial phase (c, d). No sequelae associated with arterial dissection such as aneurysmal formation were found.

dynamic arterial phase, malperfusion regions were revealed clearly in the spleen (Fig. 1d,e); however, these ischemic changes were restored in the equilibrium phase (Fig. 1f), suggesting that the spleen did not become infarcted. There was no obvious increase of the fat tissue density around the affected artery, named the “dirty fat sign”<sup>16</sup>. A CT angiogram demonstrated the irregularly narrowed caliber of the entire splenic artery (Fig. 2). Malperfusion in the spleen was shown as a defect (Fig. 2, asterisk). Neither an ostium of the intimal laceration causing the dissection nor an intimal flap was identified. The common hepatic artery and left gastric artery were considered to be unaffected. The patient’s condition was finally diagnosed as dissection of the celiac and splenic arteries complicated with circulatory insufficiency of the spleen.

Although the patient’s general condition, including vital signs, was stable and there were no impending complications such as splenic infarction and aneurysmal formation, he was hospitalized for careful observation with treatment including resting, strict control of blood pressure, and continuation of aspirin intake. Additional blood examinations for syphilis (*Treponema pallidum* hemagglutination and rapid plasma regain methods), antinuclear antibody, or hyperglobulinemia for the screening of autoimmune disease were negative.

Approximately 1 week later, the pain in the left back and flank region subsided completely. One month later, on the contrast-enhanced CT in the dynamic arterial phase, the caliber of the splenic

artery became larger concomitant with regression of the encasing thrombotic pseudo lumen (Fig. 3a-c). The spleen was homogeneously enhanced from the arterial phase (Fig. 3c,d). The patient recovered with conservative treatment and without any sequelae associated with arterial dissection, such as aneurysmal formation.

## DISCUSSION

According to the result of a large number of autopsy evaluations covering 47 years at the Mayo Clinic, only two cases of splenic artery dissection were detected from 28,512 autopsy cases (0.007%)<sup>14</sup>. In a recent national inpatient database in Japan named the Japanese Diagnosis Procedure Combination, only 23 cases of splenic artery dissection were recorded in 18.3 million inpatients (0.000001%)<sup>7</sup>. These results suggest that splenic artery dissection unrelated to the aorta is an extraordinarily rare condition<sup>1,4,8</sup>. However, as celiac artery dissection frequently involves the hepatic and splenic arteries<sup>1,3-5,10-13,15-18</sup>, and recent diagnostic imaging modalities demonstrated that more than half of patients with splenic artery dissection were asymptomatic<sup>3,5,6,8,11,14,18</sup>, it is likely that the incidence of splenic artery dissection is underestimated<sup>11,16</sup>.

Before contrast-enhanced dynamic CT, which is the gold standard imaging modality for diagnosing arterial dissection<sup>3-5,8,10,12,15</sup>, became available, diagnosis of splenic artery dissection was exclusively made postmortem<sup>1,4,5,8,15</sup>. In the present case, it took approximately 4 days to arrive at the correct

diagnosis of dissection of the celiac and splenic arteries, owing to the rarity of the disease, nonspecific clinical features<sup>1)</sup> of persisting back and left flank pain, lack of significant physical examination or laboratory data, and partial response to a non-steroidal anti-inflammatory agent<sup>1,4)</sup>. Retrospectively, the left flank pain, which is presumably related to splenic circulatory insufficiency secondary to splenic artery dissection<sup>12)</sup>, might have provided an alarming clue to diagnosing splenic artery dissection in the present case. Thus, this case shows the diagnostic difficulty of splenic artery dissection<sup>4)</sup>, in the absence of suspicion for this disease entity. Although the present patient recovered with conservative treatments despite the delayed diagnosis, splanchnic artery dissection should be considered a potentially life-threatening condition<sup>1,4,7,10,11,16,17)</sup> that needs to be recognized early.

In the various clinical backgrounds of splanchnic artery dissection<sup>1,3-6,8,10-19)</sup>, hypertension<sup>1,4,8,10,12,15,19)</sup>, being a male patient in the fifth or sixth decade of life<sup>1,4-6,8,15,17)</sup>, smoking habit<sup>4-6,8,10,15)</sup>, atherosclerosis<sup>3,5,8,10-12,15-18)</sup>, and iatrogenic causes<sup>3,8,11,12,17,18)</sup> are relatively common. On pathology, fibromuscular dysplasia<sup>3,4,6,10,12,15-18)</sup> and cystic median degeneration<sup>1,3,4,11,12,15-18)</sup> of the arterial wall, which is defined as an accumulation of basophilic mucopolysaccharide substance with cyst-like formations in the tunica media causing loss or vulnerability of elastic and muscle fibers<sup>1,20)</sup> and with a strong relationship to hypertension<sup>4)</sup>, are considered the most common findings (found in up to 83%) of patients with splenic artery dissection<sup>4)</sup>. In the present case, except for the male sex, age in the fifth decade, and past smoking habit of the patient, no particular predisposition to splenic artery dissection could be identified, suggesting an idiopathic dissection similar to many other reported cases<sup>3,5,10-12,15,16,18)</sup>. Further, we believe the concurrent well-controlled SAS scarcely influenced the splenic artery dissection, although the relationship between SAS and dissection of the aorta is exclusively described in several articles<sup>2,9,19)</sup>.

In this case, the dissection was severe, extending from the celiac trunk to the entire splenic artery, and the true lumen of the dissected splenic artery was narrowed causing partial malperfusion in the spleen on enhanced CT. However, thromboembolic sequelae such as infarction in the spleen or aneurysmal formation were not present as complications. This favorable consequence may be related to the administration of the antiplatelet agent aspirin. As intramural hematoma or thrombus, which is formed by blood flowing via the entry of disruption of the intima into two layers of the arterial wall (i.e. between subintimal and medial layers or medial and adventitial layers)<sup>3,5,11,18)</sup>, plays a critical role in deterioration of arterial cleavage, luminal occlusion, or aneurysmal formation<sup>1,3,12)</sup>, it is likely that the antithrombotic treatment might

have exerted a protective effect against thromboembolic sequelae. This speculation is supported by a report in which massive splenic infarction subsequent to splenic artery dissection was exaggerated in a patient with coagulopathy by mutation of factor V Leiden<sup>6)</sup>. Considering these observations, an immediate start of antithrombotic therapy may be effective in preventing the progression of arterial dissection and thromboembolic complications, although some authors do not recommend antithrombotic therapy for splanchnic artery dissection<sup>15)</sup>.

The optimal treatment for celiac or splenic artery dissecting remains inconsistent<sup>5,11,12,15,16)</sup>, because of its extreme rarity and variations in the involved site or extension, luminal potency, and the background comorbidity in a case-by-case basis. Besides strict blood pressure control<sup>5,6,8,11,15-17)</sup>, several treatments such as antiplatelet or anticoagulant therapy are available to prevent thromboembolic sequelae<sup>5,6,8,11,12,15,17)</sup>. Moreover, surgical resection of an aneurysm or infarcted organ<sup>5,8,11,16,18)</sup> or endovascular intervention of the affected portion of the artery<sup>4,5,8,10,17,18)</sup> can be applied. Nevertheless, we believe that the early diagnosis with a high index of suspicion of splanchnic artery dissection leads to the appropriate treatment.

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