Segmental arterial mediolysis mimics systemic vasculitis

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Abstract

Segmental arterial mediolysis (SAM) is a rare, nonarteriosclerotic, noninflammatory vascular disease and mostly affects medium-to-large sized abdominal arteries as well as presents with hemorrhages in the abdominal cavity. We report the case of a patient with SAM of the celiac, right renal, jejunal branch of the superior mesenteric, left gastric, and splenic arteries who was diagnosed by excluding other causes and in whom transcatheter embolization was performed in two different sessions, but he died because of an undefined reason. SAM mimics systemic vasculitis and causes abdominal pain; it should be considered because abdominal hemorrhage or arterial infarction can result in death.

Keywords: Pseudoaneurysm, segmental arterial mediolysis, vasculitis

Introduction

Segmental arterial mediolysis (SAM) is a rare, nonarteriosclerotic, noninflammatory vascular disease that mostly affects medium-sized abdominal arteries and presents with hemorrhages in the abdominal cavity. It is characterized by vacuolization and lysis of the arterial smooth muscle cells (1).

The lesions are mostly seen in the branches of the celiac artery and superior mesenteric artery (2). The main clinical symptom is abdominal pain caused by abdominal hemorrhage or arterial infarction (3). Aneurysm rupture or bowel infarction requires surgical or interventional radiological therapy. Recently, transcatheter embolization has been performed following angiography because it is less invasive if the source of the hemorrhage can be detected by arteriography (4).

Herein, we describe the case of a patient with SAM of celiac, right renal, jejunal branch of the superior mesenteric, left gastric, and splenic arteries who was diagnosed by excluding other causes and in whom transcatheter embolization was performed.

Case presentation

A 37-year-old man with a 6-day history of abdominal pain was admitted to the emergency department wherein widespread fluid collection was revealed by abdominal ultrasound. On admission, he was hemodynamically stable. Physical examination revealed normal vital signs with full and symmetric pulses throughout. Abdominal examination revealed abnormal mild epigastric tenderness without rebound tenderness and guarding. Laboratory data on admission showed elevated C-reactive protein (CRP: 9.56 mg/dL) levels. The complete blood count and other serum chemistry profiles were within normal limits. Computed tomography angiography (CTA) of the abdomen demonstrated widespread intra- and retroperitoneal fluid collection. Arterial images showed 50% stenosis of the celiac artery orifice and pseudoaneurysm (14 mm) immediately below the stenosis (Figure 1); this appearance was believed to occur as a consequence of dissection, but at that stage, flap dissection could not be detected. The intensive fluid collection was assumed to be hematoma secondary to hemorrhage from this probable dissection. During the procedure, active extravasation could not be distinguished. In addition, portal vein was under the pressure of the hematoma. Because of the hematoma and the suspicion of dissection and acute bleeding, the patient underwent digital subtraction angiography (DSA). DSA demonstrated web-like stenosis of the right interlobar renal artery secondary to SAM, dissectant saccular aneurysm of the proximal celiac trunk, and fusiform aneurysm of the jejunal branch of the superior mesenteric artery. Transarterial detachable hydrocoil and pushable coil embolization of the saccular aneurysm (celiac artery aneurysm) as well as pushable coil embolization of the fusiform aneurysm (aneurysm of the jejunal branch of the superior mesenteric artery) were performed without complications. For the follow-up, the patient was transferred to the intensive care unit of the department of general surgery where he was observed for 6 days. During the hospital stay, his hemodynamic status continued to be stable. When he was discharged, he had no complaints and had normal laboratory
levels. The nonspecific mildly elevated CRP level resolved during the hospital course without specific treatment.

Three weeks after his first admission, he was re-admitted to a rheumatology clinic for further evaluation. In his previous medical history, he had a 15 pack-year smoking history. He had no previous medical history of vasculitis (no history of oral or genital ulcers and constitutional symptoms). With regard to his family history, both his parents were diabetic and hypertensive. On physical examination, the patient was stable and had normal findings with no abdominal symptom.

Antineutrophilic cytoplasmic and antinuclear antibodies were negative; serum complement levels were normal. Other laboratory parameters, including urine analysis and echocardiography, were also normal; there was no laboratory evidence suggesting vasculitis or endocarditis. Viral infections and mycotic aneurysms were excluded with laboratory and radiological findings. Pathergy test that was performed to determine the possibility of Behçet’s disease was also negative. The most recent serum CRP level was 0.73 mg/dL. CTA of the thorax was performed for the etiology, and no signs supporting vasculitis or infection were reported. One month after the first procedure, abdominal CTA was repeated to monitor changes in arterial pathology. An informed consent form was filled before the treatment.

Upon performing control CTA after 1 month, the patient was discharged. We called the patient when he cancelled his appointment and learned that he died. We understood that this was a sudden death possibly because of massive hemorrhage.

Discussion

Herein, we report a case of SAM, which is a rare disease. Although we could not make a histological diagnosis, the exclusion of all other possibilities and the typical appearance of angiographic findings led us to make this diagnosis. In line with this result, many cases reported in the literature do not have histological confirmation (2, 4-6).

SAM is characterized by the vacuolization and lysis of the arterial smooth muscle cells (1). The lesions are mostly observed in the branches of the celiac artery and superior mesenteric artery (2). Slavin et al. (1) showed that it involved the branches of the celiac artery in 60%, superior mesenteric artery in 17%, renal arteries in 14%, and inferior mesenteric artery in 9% of the patients. Inada et al. (7) showed that the middle colic artery was most frequently involved followed by gastric and gastroepiploic arteries in a review of 52 cases.

Although SAM is not a systemic disease, two or more arterial segments, i.e., most frequently abdominal arteries and the branches of the celiac axis, may be affected (8). In our case, two different CTAs performed over 1 month showed the aneurysms of multiple abdominal arteries. We planned close control and frequent CTA to detect the new aneurysms that may develop with time.

The pathogenesis of SAM is unknown, but it has been suggested that the arterial lesions, i.e., lysis in vessel walls, develop as a consequence of an inappropriate response by the vessel endothelium to vasospasm such as hypoxia, hypotension, or sepsis (9). The main clinical symptom is abdominal pain caused by abdominal hemorrhage or arterial infarction. Patients may present with life-threatening hemorrhages, generally in the abdominal cavity, in the retroperitoneum, or at the base of the brain. However, angiography is not usually performed for the evaluation of patients with abdominal pain; hence, the diagnosis of SAM is probably underestimated.

Typical lesions of SAM occur in a skip pattern within the large abdominal arteries (8). The most common radiological findings are aneurysms, arterial dilatation at acute onset, and occlusions, while dissections and stenotic lesions were observed in delayed cases. Of the
81 patients with reported survival outcomes, there was a total SAM-related mortality of 26% (21 patients). Of the 21 patients who died from SAM-related causes, 13 were reported to have died before any intervention was attempted (i.e., death on arrival at hospital) or owing to no intervention; 5 patients died despite attempts of nonoperative conservative management. All but two of the SAM-related mortalities were within 30 days of the initial presentation (3). The reason of our patient’s death may be hemorrhage because of the rupture of a newly developed artery aneurysm.

The causes for this arteriopathy are not yet known. Atherosclerosis, fibromuscular dysplasia, various forms of systemic vasculitis, and neurofibromatosis should be considered in the differential diagnosis. The characteristic clinical features including recurrent aphthous stomatitis and genital ulcerous lesions can help to distinguish this inflammatory disease; moreover, laboratory screening tests, including nonspecific acute phase reactants and specific auto-antibodies, can be helpful in differentiating patients with the abovementioned diseases from patients with SAM. Congenital structural vascular disease such as Ehler–Danlos syndrome should be included in the differential diagnosis; however, this syndrome usually presents with skin laxity, joint hypermobility, and lens subluxation. Our patient neither had any medical history of symptoms resembling vasculitis, connective tissue diseases, and infections nor did he have a family history of dysplasia and aneurysms.

Vascular symptoms are related to dissection or rupture of the aorta and its branches. Michael et al. emphasized that the diagnosis of SAM should be made using DSA or CTA based on the characteristic pattern of arterial involvement and morphologic changes after excluding vasculitis by clinical and laboratory findings. In our case, the diagnosis was made by excluding vasculitis, connective tissue diseases, congenital structural vascular diseases, and infections by clinical and extensive laboratory analysis as well as the results of CTA and DSA. High level of CRP was controversial with the nature of the disease being noninflammatory. Without any specific treatment other than embolization and supportive medications for hemodynamic stability and homeostasis, CRP normalized over days. Therefore, we associated this increased CRP level with intra-abdominal hemorrhage.

The optimal treatment of SAM is crucial. The misdiagnosis of inflammatory vasculitis may lead to patients being incorrectly treated with immunosuppressive or corticosteroid therapies (9). Supportive medications for hemodynamic stability and homeostasis are fundamental. Previously, surgical treatment was indicated. Recently, transcatheter embolization has been performed following angiography because it is less invasive if the source of hemorrhage is detected by arteriography (4). Surgical treatment is required in patients with recurrent bleeding or in whom embolization has failed. Many authors recommend to monitor the patients with SAM who undergo arterial embolization at short intervals to evaluate the changes in the lesions (4, 10).

In summary, we have described the case of a patient with SAM of celiac, right renal, jejunal branch of the superior mesenteric, left gastric, and splenic arteries who was diagnosed by excluding other causes and in whom transcatheter embolization was performed in two different sessions, but he died because of an undefined reason. The most dramatic presentation of SMA is a sudden life-threatening hemorrhage that might be the reason for the death of our patient. Patients should be followed up closely after transcatheter embolization.

In conclusion, SAM is a cause of abdominal pain and should be considered because abdominal hemorrhage or arterial infarction is the reason for the pain. At present, with the frequent use of angiography in the evaluation of the complicated abdominal pain, awareness of the disease increases with more presented cases diagnosed by CTA. The disease may be progressive and lethal despite successful treatment with endovascular interventions. Patients should be followed up closely after transcatheter embolization.

Ethics Committee Approval: N/A.

Informed Consent: Written informed consent was obtained from the patient who participated in this case.

Peer-review: Externally peer-reviewed.

References