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CASE REPORT | LIVER

An Unusual Presentation of Segmental Arterial Mediolysis

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ABSTRACT

Segmental arterial mediolysis (SAM) is a rare nonatherosclerotic and noninflammatory disease that often affects medium to large-sized arteries. We report a case of SAM involving bilateral hepatic arteries in an elderly woman. Although her initial presentation mimicked vasculitis, the clinical course and imaging led to the diagnosis of SAM. She was treated with coil embolization and stenting of the involved hepatic vessel leading to dramatic clinical improvement. It should be differentiated from vasculitis because there is no role of steroids in the management of SAM.

KEYWORDS: Hepatic aneurysms, Segmental arterial mediolysis, Vasculitis

INTRODUCTION

Segmental arterial mediolysis (SAM) is a rare disease that affects medium to large-sized arteries and often mesenteric arteries. Although a nonatherosclerotic and noninflammatory process, the disease can mimic vasculitis.[1] It can lead to hemorrhage or infarction and typically presents with severe abdominal pain.[2] It most commonly affects the celiac artery and superior mesenteric artery.[1] The natural history of the disease is unclear, but vessel wall degeneration can lead to dissection, aneurysm, stenosis, and vessel rupture in affected patients. Given its severity, early detection by using computed tomogram angiography is imperative. In this report, we describe a unique presentation of SAM in an elderly woman.

CASE REPORT

A 65-year-old woman with history of smoking half pack a day for 10 years presented to the emergency room with 3-day history of sudden-onset dull epigastric pain, radiating to the back, without any exacerbating or relieving factors. The abdominal pain was associated with nausea, vomiting, and decreased oral intake. A review of the system was negative for fever, chest pain, palpitations, shortness of breath, cough, rashes, nodules, oral ulcers, genital ulcers, dizziness, motor or sensory weakness, and changes in bowel or bladder habits. She denied any history of alcohol use, intravenous drug use, over-the-counter medications, or herbal products.

Her vital signs on presentation were as follows: blood pressure 140/87 mm Hg, heart rate 92 beats per minute, respiratory rate 16 per minute, and oxygen saturation of 96% on room air. Physical examination revealed epigastric tenderness without rebound tenderness or guarding. Her initial laboratory test results were significant for the white blood cell count 15,200/μL with neutrophil predominance, C-reactive protein 9.4 mg/L, and erythrocyte sedimentation rate 92 mm/hour. Her liver function tests showed aspartate transaminase 2,173 U/L, alanine transaminase 2,013 U/L, total bilirubin 0.6 mg/dL, albumin 3.7, and international normalized ratio 1.1. Further laboratory test results ruled out infectious etiology for her presentation. Rheumatologic workup was negative for antinuclear antibodies, anti-neutrophil cytoplasmic antibodies, abnormalities in complement proteins, human immunodeficiency virus, and hepatitis B or C.

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Axial contrast-enhanced computed tomography of the abdomen demonstrated multiple large aneurysms involving bilateral hepatic arteries with diffuse irregularity and ectasia of the hepatic arterial system and blood products surrounding the left hepatic artery aneurysm (Figure 1). Celiac artery angiogram demonstrated aneurysm of both left and right hepatic arteries (Figure 2). She subsequently underwent coil embolization of the left hepatic artery (Figure 3). The procedure was complicated by complete occlusion of the left hepatic artery and spontaneous thrombosis of the right hepatic artery on catheterization. computed tomography angiogram (CTA) of the abdomen after the procedure showed reconstitution of flow within hepatic arterial supply by collaterals. Postprocedure investigations revealed downward trending inflammatory markers and transaminases, with levels returning to baseline within a week.

DISCUSSION

In this study, we report a rare case of SAM with an unusual pattern of vasculature involvement. Although, in our case, histologic confirmation was not provided, clinical presentation, laboratory findings, and radiologic pattern led to the diagnosis of SAM.

SAM often affects medium-to-large splanchnic arteries and is characterized by lysis of the tunica media.[2] The natural course of SAM is unpredictable, and it can quickly spread to involve other vascular segments.[3] It can often lead to detrimental complications such as dissection, aneurysm, stenosis, and vessel rupture.[3,4]

Although SAM from a pathophysiological nature is non-inflammatory, the clinical presentation can mimic vasculitis.[1] Inflammatory markers are not elevated in SAM.[3,5,6] Our patient had a high erythrocyte sedimentation rate and C-reactive protein on presentation, suggesting vasculitis as the etiology of the aneurysms.

Notably, the decrease in inflammatory markers during her hospital stay without administration of steroids favored SAM over a vasculitis process. Her high inflammatory markers were attributed to extravasation of blood from left hepatic artery aneurysm. Rapid normalization of inflammatory markers after coil embolization and stenting further confirmed the hypothesis. It is imperative that patients undergo autoimmune workup to rule out vasculitis due to overlap in clinical presentation and difference in the management of SAM and vasculitis.

Early detection is key in the prognosis of SAM because the clinical course tends to be unpredictable and complications of vascular injury including stenosis, dissection, aneurysm, or rupture may occur. CTA is the best modality for radiographic diagnosis of SAM. Typical findings on CTA are non-atherosclerotic arterial mural wall thickening with a multifocal

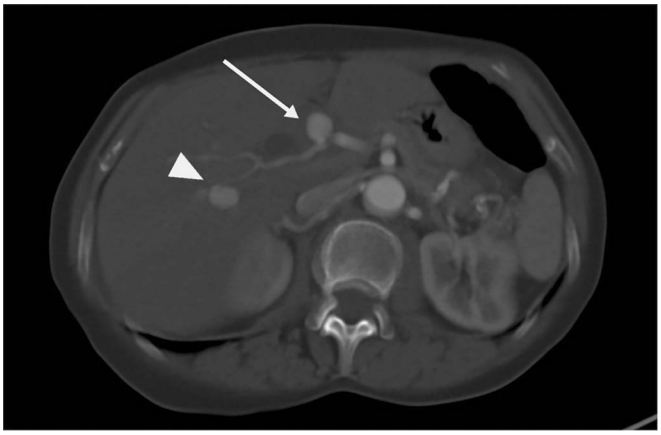

Figure 1. Axial contrast-enhanced computed tomography shows left hepatic artery aneurysm (arrow) and right hepatic artery aneurysm (arrowhead).

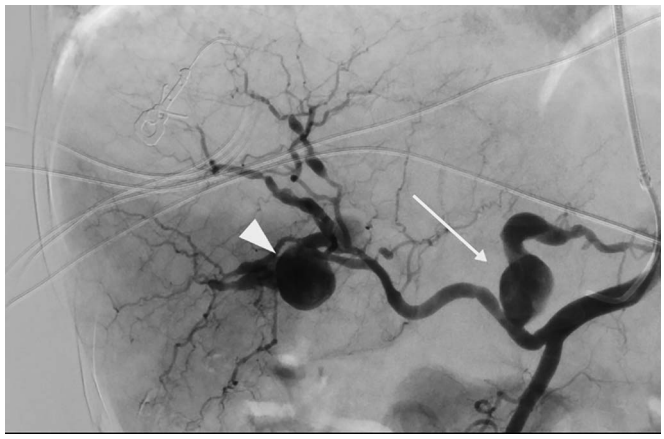

Figure 2. Celiac artery angiogram image showing the pre-embolization image of left hepatic artery aneurysm (arrow) and right hepatic artery aneurysm (arrowhead).

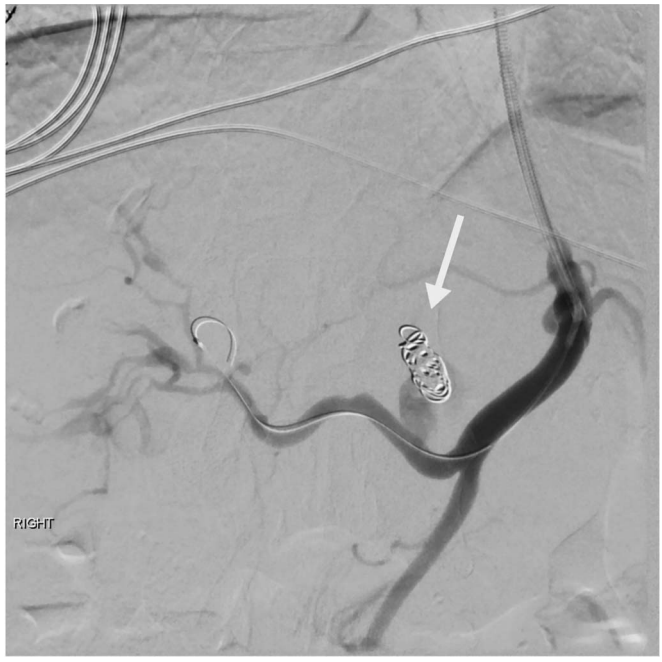

Figure 3. Celiac artery angiogram image of left hepatic artery aneurysm after coiling (arrow).

skip pattern of luminal strictures and poststenotic aneurysmal dilatation of arteries.[7] SAM without clinical manifestations does not require therapy.[1] Patients with hemodynamic instability or end-organ ischemia require endovascular therapy with coil embolization, as in our case.[7]

Being a rare disease, there is a lack of adequate literature on SAM. To the best of our knowledge, this is the first reported case with multiple aneurysms involving both hepatic arteries. Further research is warranted to better understand the clinical presentation, natural history, and management of SAM.

DISCLOSURES

Author contributions: R. Karna: concept of the study, drafting the work, revising for important intellectual content, and final approval of the version to be published. D. Kaye, B. Jagdish: drafting the work, revising for important intellectual content, and final approval of the version to be published. A. Johnston and D. Venkat: revising for important intellectual content and final approval of the version to be published. All authors: Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. Article guarantor: R. Karna is the article guarantor.

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