A 53-year-old female, hypertensive, former smoker presented to our hospital with an exacerbation of chronic obstructive pulmonary disease with pneumonia. Laboratory investigation revealed elevated white blood cell count (12.1x10^9/L). Imaging of the patient involved computed tomography (CT) of the chest, revealing bilateral diffuse reticulo nodular infiltrates in keeping with her diagnosis. Imaging included limited views of the upper abdomen significant for prominent vascular structures. Subsequently, she underwent CT angiography of her abdomen/pelvis, confirming a 2.4x1.6 cm enhancing lesion abutting the right hepatic artery near the liver hilum, consistent with an aneurysm, and an additional 8x3-mm aneurysm (Figure 1). Spleen, gallbladder, pancreas, adrenal glands, and kidneys were unremarkable. There was no lymphadenopathy. Further laboratory investigations showed normal erythrocyte sedimentation rate (4 mm/h) and serum protein electrophoresis. Hepatitis and autoimmune (anti-nuclear antibody, extractable nuclear antigen antibody panel [anti-SSA, anti-SSB, anti-SmRNP, Scl-70, anti–Jo-1], rheumatoid factor CCP antibody, C3-C4 total hemolytic complement, anti-neutrophil cytoplasmic antibodies [PR3 and MPO] antibodies) panels were negative. In a vascular surgical consultation a month later, repeat CT angiography revealed an additional 8x4-mm left hepatic aneurysm, a smaller right hepatic aneurysm now enlarged to 10 mm, and fistulous communication and extensive shunting to the portal vein with the larger 2.5-cm aneurysm. Her infectious episode had resolved, and she underwent coil embolization of the hepatic artery aneurysms with Penumbra Ruby coils (Penumbra, Inc, Alameda, CA) sized 10 mm×35 cm, 8 mm×40 cm, 4 mm×10 cm, and 3 mm×12 cm (Figure 2). The larger aneurysm was cannulated with a 0.016-in Fathom Steerable guide wire (Boston Scientific) and a HI-FLO Renegade microcatheter (Boston Scientific). The microcatheter was then exchanged for the Penumbra PX Slim microcatheter (Penumbra, Inc) to cannulate the smaller aneurysm. This was advanced with a 0.014-in Transcend EX wire (Boston Scientific). Postintervention imaging showed a marked decrease in the filling of the portal vein. No aneurysms were noted in other visceral vessels. At the end of the procedure, a liver biopsy was undertaken to aid in establishing a diagnosis. Pathology revealed disruption of the tunica media (arrows) with mucoid intimal changes (Figure 3A, hematoxylin and eosin, ×200). Elastic tissue staining showed focal disruption of the elastic lamina of the arteries (Figure 3B, between arrowheads, van Gieson, ×400) and disordered smooth muscle in the media (Figure 3C, smooth muscle actin immunohistochemical stain, ×200), consistent with segmental arterial mediolysis (SAM). The patient was started on propranolol 60 mg daily and losartan 50 mg daily for aggressive blood pressure control. At 4 months of follow up, the patient’s blood pressure was under control, and no new aneurysms were found on serial imaging.

SAM is an extremely rare vasculitis of unknown origin. 1 It occurs in all age groups, affecting mainly adults and elderly individuals, with a male predominance. The disease is characterized by the occurrence of dissecting aneurysms. Arterial rupture, resulting in hemoperitoneum or retroperitoneal bleeding, occurs in <30% of cases and is associated with high mortality. 2 SAM targets mainly medium-sized arteries within the abdominal cavity. The aorta is almost never involved in SAM, but the celiac trunk, the superior and inferior mesenteric arteries, are involved in >80% of cases. 1 Differential diagnoses include vasculitides (mainly polyarteritis nodosa, Behcet disease, and Takayasu arteritis), mycotic aneurysms, and non-inflammatory vasculopathies such as fibromuscular dysplasia and cystic medial necrosis.3,4 No serum or genetic biomarker is available for the diagnosis of SAM. The treatment of SAM depends on the clinical manifestations. Many presentations resolve spontaneously without major consequences and can be managed conservatively. For cases of arterial rupture and hemorrhagic shock, either transarterial (eg, transarterial coil embolization) or surgical interventions are indicated, depending on the gravity of the case. 5,6

Disclosures

None.

References


Figure 1. Computed tomography angiography depicting a 2.4×1.6-cm right hepatic artery aneurysm and a smaller 8×3-mm aneurysm.

Figure 2. Computed tomography angiography after coil embolization with Penumbra Ruby coils (Penumbra, Inc, Alameda, CA) sized 10 mm×35 cm, 8 mm×40 cm, 4 mm×10 cm, and 3 mm×12 cm.

Figure 3. A. Disruption of the smooth muscle in the media (arrows) with associated mucoid intimal changes. Note the lack of significant inflammation (hematoxylin and eosin, ×200). B. Loss of the internal elastic lamina (between arrowheads) of the same small hepatic artery (van Gieson, ×400). C. Disordered smooth muscle in the media (smooth muscle actin immunohistochemical stain, ×200).
Biopsy Proven Hepatic Segmental Arteriolar Mediolysis Successfully Treated With Coil Embolization
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