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Case Report

Emergent embolization of a ruptured splenic artery aneurysm complicating Menkes disease

John F. Olivieri, BS[a], *Arthie Jeyakumar, MD*[a], *Giridhar M. Shivaram, MD*[a,b], *Kevin S.H. Koo, MD*[a,b], *Eric J. Monroe, MD*[a,b,*]

[a] *Department of Radiology, Division of Interventional Radiology, University of Washington, Seattle, WA, USA*
[b] *Department of Radiology, Division of Interventional Radiology, Seattle Children's Hospital, Seattle, WA, USA*

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ABSTRACT

We report a 7-year-old boy with Menkes disease complicated by rupture of a large splenic artery aneurysm. The aneurysm was successfully embolized with microcoils and n-butyl cyanoacrylate. Further angiographic evaluation revealed marked tortuosity of mesenteric and lower extremity vasculature, including the femoral arteries bilaterally, without aneurysm formation. The patient has since been evaluated annually with computed tomography angiography and there have been no additional vascular complications of his disease during 3-year follow up.

Introduction

Menkes disease, a rare X-linked disorder due to copper maldistribution, results in decreased elastin and collagen cross-linking [1,2]. This multisystemic disease is characterized by neurodegenerative, dermatologic, and connective tissue complications and is frequently fatal in the first several years of life. Vascular abnormalities have been previously described in the disease, rarely resulting in abdominal cavity aneurysms [1,3,4]. Here, we present a 7-year-old male child with known Menkes disease complicated by ruptured splenic artery aneurysm. The aneurysm was effectively embolized using microcoils and n-butyl cyanoacrylate glue.

Case

The patient initially presented in the first year of life with seizures and developmental delay. Menkes disease was suspected based on reduced copper and ceruloplasmin levels as well as classic phenotypic features of hypotnoia, brittle hair, lax skin, and bladder diverticula. DNA testing confirmed mutation of the ATP7A gene. At the age of 7, he presented acutely to his pediatrician with bilious emesis in the setting of abdominal guarding and lethargy. A CT of the abdomen prior to arriving to our institution revealed a 4.1 × 3.8 cm splenic aneurysm and large surrounding hematoma (Fig. 1). Upon transfer to our institution, he was hemodynamically stable but labs demonstrated a decrease in hematocrit to 34 from 40.

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* *Corresponding author*.
E-mail addresses: ejmonroe@uw.edu, eric.monroe@seattlechildrens.org (E.J. Monroe).

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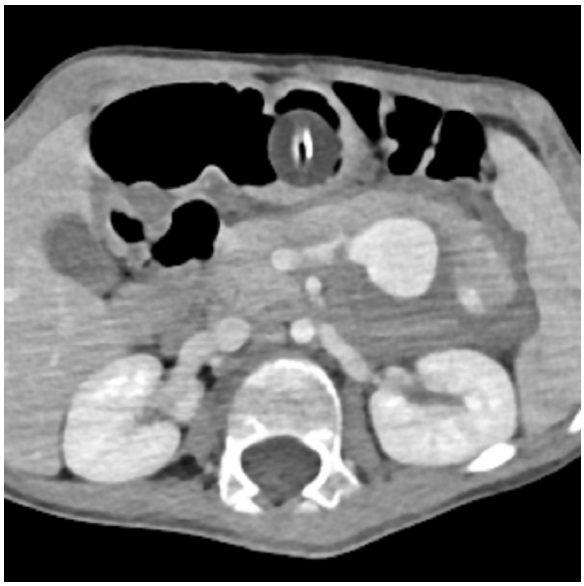

Fig. 1 – Contrast enhanced CT of the abdomen. Axial image through the upper abdomen demonstrates a 4.1 x 3.8 cm splenic aneurysm and large surrounding hematoma.

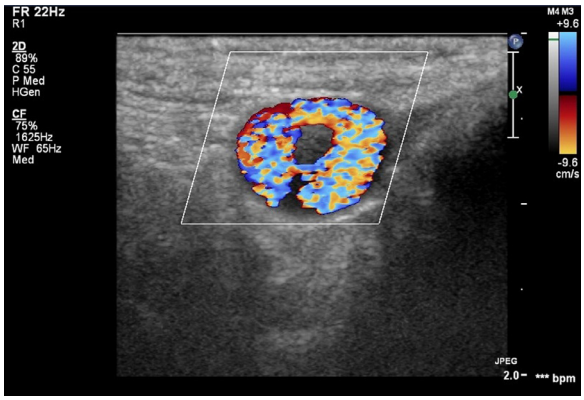

Fig. 2 – Access planning ultrasound. Color Doppler ultrasound image of the left common femoral artery in transverse view demonstrates marked "corkscrew" tortuosity.

The patient was brought emergently to interventional radiology. At the time of vascular access using ultrasound, markedly tortuous iliofemoral vessels were observed bilaterally (Fig. 2). Following micropuncture access of the right common femoral artery, a 0.018-inch wire was carefully negotiated through the iliac arteries and into the aorta. Following exchange for a 0.035-inch wire, a 4 French vascular sheath was placed. Celiac angiography demonstrated a large aneurysm splaying the superior and inferior divisions of the splenic artery (Fig. 3a). Following coaxial microcatheter introduction, the aneurysm sac was selected and embolized with microcoils. Persistent opacification of the neck resulted in additional embolization using a 1:3 mixture of n-butyl cyanoacrylate and ethiodized oil (Fig. 3b). Splenic parenchymal perfusion was preserved via pancreatic and short gastric artery collateral flow. Following the embolization, pelvic aortogram and bilateral lower extremity run-off angiograms were acquired (Fig. 4). These images demonstrated markedly tortuous iliac and femoral arteries bilaterally. The patient progressed appropriately and was discharged on postprocedure day 3. He is now 3 years postintervention with no further vascular complications of his disease. Yearly abdominal computed tomography angiographies have been acquired for surveillance of his abdominal and pelvic vasculature, demonstrating stability of multifocal subcentimeter mesenteric aneurysms and no evidence of splenic infarction.

Discussion

Although the exact mechanisms are unclear for the wide range of clinical features of Menkes disease, an abnormality in the cellular utilization of copper and its incorporation into multiple enzymes, namely lysyl oxidase, is thought to be responsible [2–4]. Severe consequences manifest early in life, with death frequently occurring before the age of 5 [1]. Treatment with parenteral copper has been shown to only improve neurologic symptoms and death but not affect the consequences of the abnormal connective tissue [5]. A series complied by Godwin et al [6] catalogued the reported cases of arterial aneurysms in Menkes disease. Several cases of mesenteric arterial involvement resulted in fatal hemorrhage, highlighting that the need to identify and treat the vascular anomalies in patients with the disease is significant. For those undergoing interventional treatment for splenic arterial aneurysms, only splenectomy has been described.

The impairment of elastin and collagen cross-linking has been proposed to be responsible for the vascular anomalies that have been found with those with Menkes disease as the copper-dependent lysyl oxidase enzyme is responsible for this process [2]. Increased tortuosity of arteries and changes in the diameter and kinking of vascular vessels have been demonstrated using magnetic resonance angiography (MRA) imaging of both intracranial and extracranial structures [7]. Additionally, pathological analysis of vessels from those with Menkes has demonstrated disintegration of the internal elastic lamina of arteries and abnormalities in the wall thickness and sizes of the lumen present in these vessels [3]. Pathologic examination of an iliac artery aneurysm found in a patient with Menkes disease performed by de Figueiredo Borges et al [8] showed that the disappearance of elastic fibers and presence of abnormal collagen accompanies the destruction of smooth muscle cells, mucoid degeneration, and extracellular matrix breakdown that is seen in aneurysm formation in other diseases processes.

In our case, endovascular treatment of the splenic artery aneurysm was chosen to avoid the risks associated with surgery. Adaletli et al [9] described effective stent graft treatment of an aneurysms complicating Menkes disease. While a stent graft may have been considered in the case presented, stent graft introduction would have required a significantly larger femoral access sheath, of concern given the iliofemoral tortuosity and irregularity present on planning ultrasound as well as vascular fragility associated with this condition.