

Ruptured Gastric Aneurysm in α -1 Antitrypsin Deficiency

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ABSTRACT

We present a unique vascular complication of α -1 antitrypsin deficiency (AATD) in a patient with an acute onset of epigastric pain and hemodynamic instability. Abdominal computed tomography angiography detected hemoperitoneum and hematoma within the gastrohepatic ligament with active extravasation. Abdominal angiography revealed left gastric aneurysms. An association between AATD and vascular aneurysms has been suggested to be secondary to unopposed proteolytic activity against arterial structural proteins. The aneurysm formation in aortic, superior mesenteric, inferior mesenteric, and splenic arteries has been reported. We report the first case with ruptured gastric artery aneurysm as a complication of AATD.

INTRODUCTION

α -1 Antitrypsin deficiency (AATD) is an autosomal codominant disease. The protease inhibitor gene, which encodes the α -1 antitrypsin protein, is highly pleomorphic with more than 120 identified alleles.¹ Protease inhibitor-MM (Pi-MM) is the normal phenotype, and other allelic variants are associated with reduced expression of α -1 antitrypsin at different levels. The Pi-ZZ variant accounts for 95% of AATD patients in whom α -1 antitrypsin activity is severely suppressed.^{2,3} α -1 Antitrypsin is the circulatory serine protease inhibitor that maintains the integrity of connective tissue. Deficiency of α -1 antitrypsin results in a range of clinical consequences, including lung disease (eg, emphysema, bronchiectasis), liver disease (eg, chronic hepatitis, cirrhosis), skin disease (eg, panniculitis), and vasculitis (eg, granulomatosis with polyangiitis) due to the imbalance between protease and antiprotease activity.³ However, nonatherosclerotic vascular complications have been infrequently reported and include dissection, aneurysm, and fibromuscular dysplasia.⁴

There are reports regarding the association between AATD and aneurysm formation in various sites particularly aortic, superior mesenteric, inferior mesenteric, splenic, and cerebral arteries.⁵⁻⁹ These complications could be explained by the deficiency of α -1 antitrypsin resulting in the inadequate protection against proteolytic effects of elastase and collagenase leading to the degradation of connective tissue of arterial wall. However, to the best of our knowledge, there is no report thus far regarding gastric artery aneurysm in patients with AATD.

CASE REPORT

A 47-year-old Caucasian female with a history of homozygous (ZZ genotype) AATD, liver cirrhosis, emphysema on long-term oxygen therapy presented with sudden onset of stabbing epigastric pain radiating to the left shoulder accompanied by dizziness. On admission, she had a blood pressure of 79/58 mm Hg and heart rate of 130/min. Her abdomen was diffusely tender to palpation but was without rebound tenderness. Laboratory tests revealed a hemoglobin of 9.2 g/dL from a prior baseline of 11.5 g/dL. After 2 units of packed red blood cells were transfused, her hemoglobin was found to have decreased further to 8.8 g/dL. Coagulation studies, platelet count, and other laboratory findings were within normal range. Abdominal computed tomography angiography detected small amount of hemoperitoneum and abdominal hyperattenuation within the gastrohepatic ligament, suggestive of hematoma and concerning for active extravasation (Figure 1). Abdominal angiography demonstrated 2 moderate sized foci of

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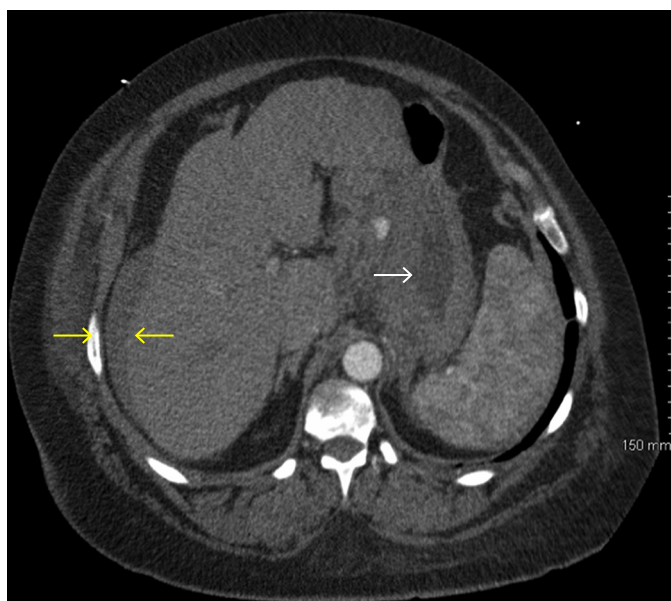


Figure 1. Abdominal computed tomography angiography revealed abdominal hyperattenuation within the gastrohepatic ligament (white arrow) and small amount of hemoperitoneum (yellow arrows).

aneurysmal dilatation arising from the proximal to mid left gastric artery (Figure 2). Transcatheter arterial embolization was successfully performed on the proximal left gastric artery (Figure 3). The angiography further noted moderate dilatation of the splenic artery and underlying moderate dilatation involving portions of the distal right main renal artery and segments of the distal left main renal artery. Upon 6-month follow-up, she had no recurrent bleeding, and repeat computed tomography angiography abdomen showed stable postprocedural changes related to coiling of the left gastric artery and interval decrease in hematoma size at the gastrohepatic ligament.

DISCUSSION

The gastric artery is a rare aneurysm site. Most patients remain asymptomatic until aneurysm rupture. Thus, the

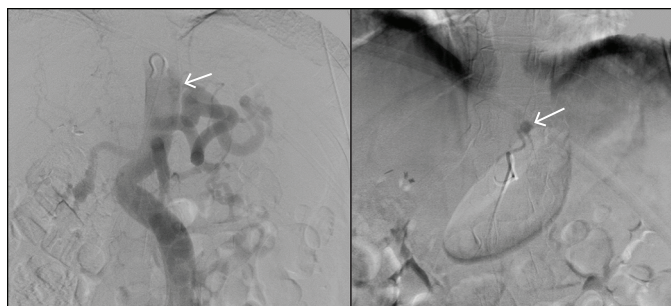


Figure 2. Abdominal angiography demonstrated 2 moderate sized foci of aneurysmal dilatation arising from the proximal to mid left gastric artery (arrows).

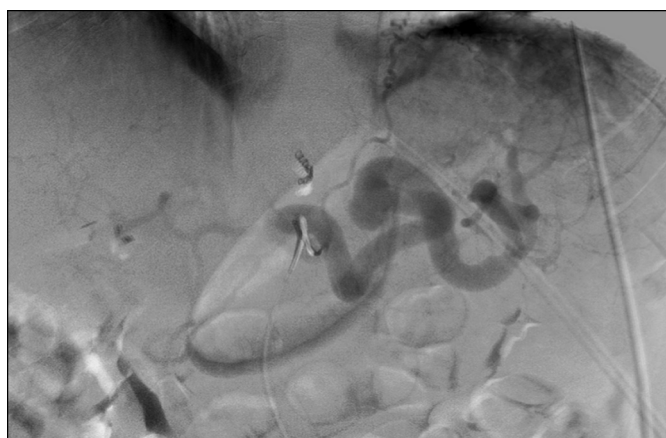


Figure 3. Transcatheter arterial embolization was performed upon the proximal left gastric artery with no further evidence of filling or the aneurysms.

majority of afflicted patients present to the hospital with acute abdominal pain and hemorrhagic shock requiring urgent intervention, including ligation or embolization by surgical or radiologic approaches, as in this patient. Transcatheter embolization is the preferred method in patients with high surgical risk and has a success rate of 85% from a previous study.¹⁰

Several studies regarding the association of AATD and arterial aneurysms showed contradictory results. Schardey et al.¹¹ reported that Pi-allelic variants are associated with the pathogenesis of aortic aneurysm. Schievink et al.⁷ supported this association in intracranial aneurysm with an odds ratio of 2.56 compared with the general population. In contrast, Elzouki et al.⁵ found no significant association between the Pi-ZZ variant and abdominal aortic aneurysm. St Jean et al.¹² identified that Pi-phenotype had no effect on abdominal aortic or intracranial aneurysm formation.

In our patient, we also observed aneurysms at other sites, including renal and splenic arteries, indicating the systemic nature of aneurysm formation. After careful review, AATD most likely contributed to the ruptured gastric aneurysm in this patient.

DISCLOSURES

Author contributions: All authors wrote the manuscript. V. Jaruvongvanich is the article guarantor.

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Informed consent was obtained for this case report.

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