CASE REPORT

Incidental finding of an arteriovenous malformation originating from the diaphragm: Case report and literature review

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ABSTRACT

Arteriovenous malformations (AVMs) are defined as aberrant linkages between arteries and veins in the absence of a capillary network. This case report will discuss the incidental finding of an AVM likely originating from the diaphragm. A 65-year-old female presents for an umbilical hernia repair. Her past medical history is significant for HTN, type 2 diabetes mellitus, gastroesophageal reflux disease, and class 1 obesity. She has a past surgical history of hysteroscopy in 2020 and laparoscopy in 1996. She has no history of tobacco use, alcohol use, or recreational drug use. She is allergic to contrast. A computed tomography (CT) scan was ordered and incidentally showed a soft tissue mass in the left upper quadrant adjacent to the stomach and diaphragm, possibly a gastrointestinal stromal tumor (GIST) or splenule, in addition to diastasis of the abdominal wall. Biopsies of the esophagus, stomach, and duodenum from the esophagogastroduodenoscopy (EGD) were all benign. A nuclear medicine scan of the liver and spleen found an estimated 2.3 cm soft tissue nodule located between the fundus of the stomach and the left hemidiaphragm that does not demonstrate sulfa colloid uptake and is therefore not consistent with a splenule. This is the only incidence cited of an AVM originating from the diaphragm. We suspect that the origin of this AVM is the inferior phrenic artery. The decision for management for this case was to get a CT scan of the abdomen and chest to trace the path of the AVM and manage with surveillance.

Key Words: Arteriovenous malformation, Diaphragm, Hernia, Inferior phrenic artery

Arteriovenous malformations (AVMs) are defined as aberrant linkages between arteries and veins in the absence of a capillary network. Due to the differences in flow states between arteries and veins, the direct surge of elevated arterial flow into venous structures can indefinitely lead to the rupture of venous walls.[1] The likelihood of mortality from the rupture of cerebral AVMs is around 20%, with a 45% chance of encountering minor or major deficits, and a 35% chance of complete recovery.[2] While AVMs are typically found in the brain and spinal cord, their occurrence in other locations is less common. The occurrence of identified AVMs in the brain is less than 10.3 per 100,000, but the specific epidemiology of other AVMs is unknown.[3] Depending on the location of the AVM, different imaging modalities can be used. For example, cerebral AVMs can be diagnosed using computed tomography, magnetic resonance imaging, and

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angiography. On the other hand, peripheral AVMs can be identified using a doppler or duplex ultrasound or magnetic resonance/computed tomographic angiography.[4]

Symptoms and treatments of AVMs vary based on presentation and location. Cerebral AVMs are classified using the Spetzler-Martin grading scale.[1] This scale also estimates surgical risk in candidates. Treatment includes surgical resection, embolization, stereotactic radiosurgery, or even a combination of all three. Peripheral AVMs can be classified into six types by angiography: type I, type IIa, type IIb, type IIc, type IIIa, and type IIIb. The treatment of each classification results in embolizing the given AVM, but the method to do so varies. For instance, type II AVMs are initially managed by decreasing blood flow and subsequently treated with ethanol embolotherapy, while type I AVMs are directly addressed through the use of coils to embolize the connection between the artery and vein.[5] Examples of agents used to embolize AVMs are silastic spheres, gelfoam, silk suture, dehydrated ethanol, etc. Presently, n-butyl cyanoacrylate, ethylene-vinyl alcohol copolymer or platinum coils and polyvinyl alcohol particles are used.[6]

This case report will discuss the incidental finding of an AVM likely originating from the diaphragm as shown in Figure 1.

There was an incidence of a spontaneous aortocaval fistula that emptied into the right atrium and traversed the hemidiaphragm as well as multiple reports of AVMs of the lung that are single or multiple, usually below the diaphragm, but not involving the diaphragm.[7,8]

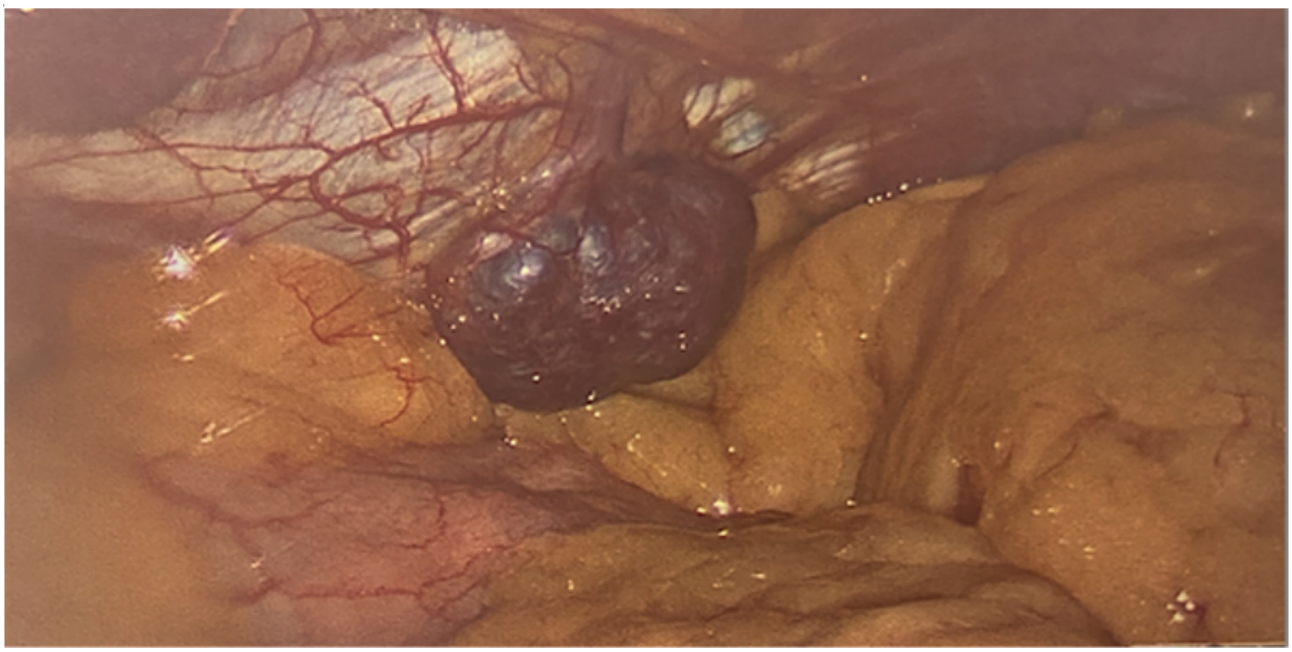

Figure 1. Laparoscopic view demonstrating the AV malformation

1. Case presentation

A 65-year-old female presents to the office as referred by her primary care physician for an umbilical hernia and intermittently painful masses surrounding the umbilicus. Her past medical history is significant for hypertension, type 2 diabetes mellitus, GERD, and class 1 obesity. She has a past surgical history of hysteroscopy in 2020 and laparoscopy in 1996. She has no history of tobacco use, alcohol use, or recreational drug use. She is allergic to contrast.

She began complaining of the hernia in 2/2023. A computed tomography (CT) scan was ordered and incidentally showed a soft tissue mass in the left upper quadrant adjacent to the stomach and diaphragm, possibly a gastrointestinal stromal tumor (GIST) or splenule, in addition to diastasis of the abdominal wall as shown in Figure 2.

Following this, an esophagogastroduodenoscopy (EGD) was ordered and confirmed a firm extrinsic impression noted in the fundus, consistent with the CT findings of mass in the proximal lesser sac as shown in Figure 3.

Biopsies of the esophagus, stomach, and duodenum from the EGD were all benign. Then, an endoscopy showed a 29 mm lesion, not consistent with GIST, but possibly a splenule or other abdominal lesion. Finally, a nuclear medicine scan of the liver and spleen found an estimated 2.3 cm soft tissue nodule located between the fundus of the stomach and the left hemidiaphragm that does not demonstrate sulfa colloid uptake and is therefore not consistent with a splenule as shown in Figure 4.

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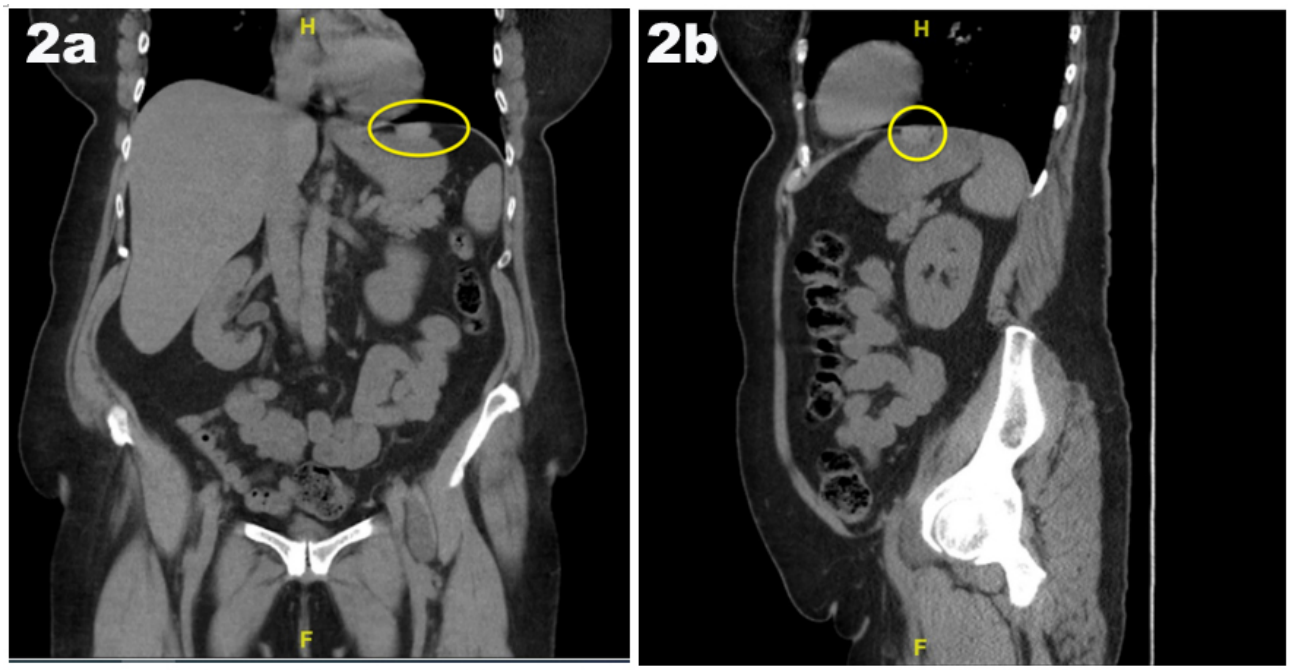

Figure 2. Single coronal and sagittal view of computed tomography scan demonstrating the AV malformation, respectively. The AVM was described as a well-circumscribed soft tissue mass measuring 2.4 cm × 1.9 cm × 1.2 cm present adjacent to the stomach and diaphragm

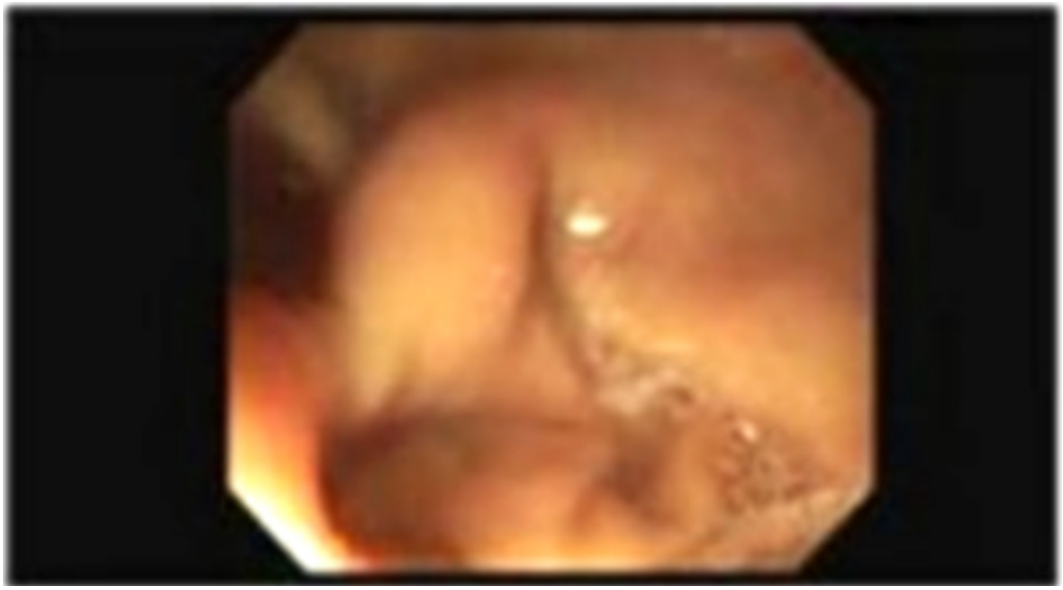

Figure 3. Esophagogastroduodenoscopy demonstrating the AV malformation. The AVM was described as a 29 mm × 15 mm homogenous hypoechoic lesion, isoechoic to spleen and liver. The lesion appeared to arise outside of the stomach wall

At this time, 10/2023, the decision was made to operate. The planned procedures were an exploratory laparoscopy, primary umbilical hernia repair, excision of two abdominal subcutaneous masses, and transversus abdominis plane block with Exparel. A moderately-sized arteriovenous malformation along the fundus that was stable with no signs of hemorrhage was found. Neither biopsy nor excision was conducted due to the benign and vascular nature of AVMs. A small umbilical hernia defect 1 cm in size was also discovered, as well as two abdominal subcutaneous masses that were excised and soft in palpation. The hernia was repaired using a 0-Vicryl suture. The two subcutaneous masses were excised

using Bovie electrocautery. The trocar sites and transverse incisions were approximated using 4-0 Monocryl in subcuticular fashion before being approximated with Dermabond. Biopsies of the abdominal masses showed homogenous adipose tissue without necrosis or hemorrhage. The patient was discharged home from the PACU with an uncomplicated recovery. Follow up abdominal CT scan was limited due the patient's allergy to contrast but did not show any pulmonary vasculature component. In 02/2024, post-op visit showed that the patient had no abdominal pain or tenderness to palpation, a well-healing hernia, and was tolerating a diet.

Further care options of surveillance, resection, and embolization were presented as options to the patient, who elected abdominal CT scan every 3 years to follow any changes in the AVM.

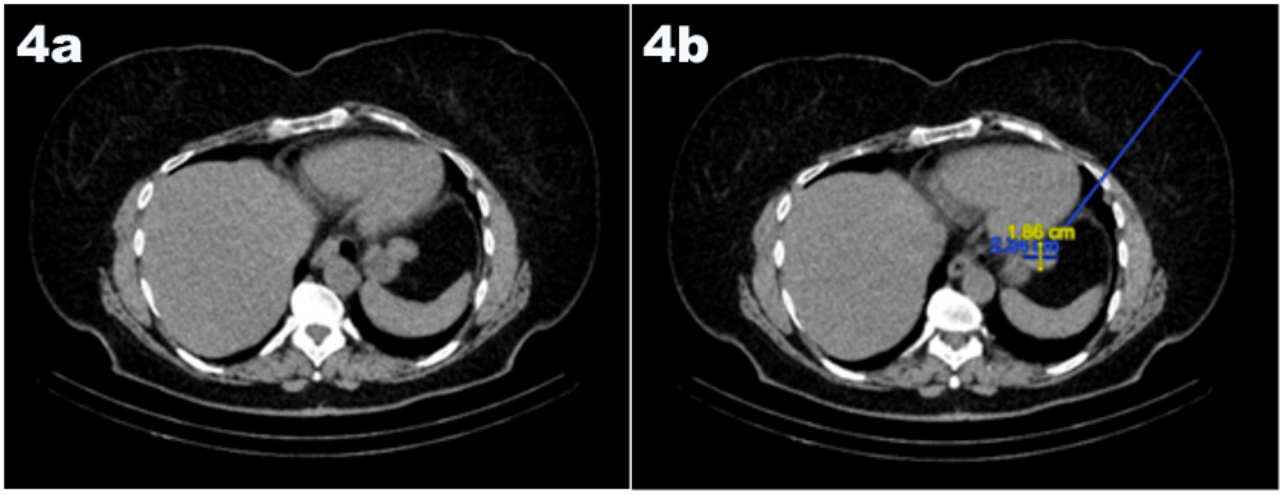

Figure 4. Nuclear medicine scan demonstrating the AV malformation. The AVM was described as an ovoid exophytic soft tissue density nodule located between the fundus of the stomach and the left hemidiaphragm. It measures approximately 2.3 cm transverse $\times$ 1.9 cm AP $\times$ 1.4 cm craniocaudal

2. Discussion

This is the first reported case cited of an AVM originating from the diaphragm. We suspect that the origin of this AVM is the inferior phrenic artery. While surgical debulking and embolization by interventional radiology have been the mainstay of managing AVMs, a complete cure is rare, as resection is difficult to determine and recurrence is common.[9] The decision for management for this case was to get a CT scan of the chest and abdomen to trace the path of the AVM and manage with surveillance. Over time, there can be a gradual increase in blood shunting, leading to the enlargement of the AVM and the development of localized venous hypertension.[9] This, in turn, results in decreased perfusion pressure to the surrounding tissues, causing tissue ischemia, pain, and subsequent tissue ulceration with bleeding.[9] Patients with small AVMs may experience periods of stability; however, the expansion of the AVM, accompanied by increased AV shunting, often manifests with heightened pulsation and bruit, can progress to emerge to a large AVM; patients are at risk of cardiac involvement and decompensation.[9] Once these symptoms and signs of progression occur within a large AVM, patients are often at risk of cardiac involvement and decompensation.[9] Surgical intervention is usually reserved for AVMs that are symptomatic or that will lead to end-organ dysfunction.[9] Until the patient experiences symptoms or imaging shows that the AVM is growing, surveillance is planned for further care.[9]

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Authors Contributions

Student Doctor Gose and Student Doctor Patel were responsible for data collection, literature review, and writing the manuscript. Dr. Posch and Dr. Bowling were responsible for editing and revising the final manuscript.

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DATA SHARING STATEMENT

No additional data are available.

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CONFLICTS OF INTEREST DISCLOSURE

The authors declare they have no conflicts of interest.

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