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Case Report

Spontaneous hemothorax in pregnant patient with pulmonary arteriovenous malformation ☆

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ABSTRACT

We present a case of a 34-year-old pregnant patient at 26 weeks' gestation by in vitro fertilization with past medical history of hypertension and infertility who presented to the hospital with abdominal pain. The patient stated her pain was in her left upper quadrant. The morning before arriving to the hospital the patient stated she woke up at 0300 with increasingly severe pain in the same area.

A computed tomography angiogram of the chest demonstrated a left-sided pulmonary arteriovenous malformation with adjacent complex left effusion on chest suspicious for a hemothorax. The hemothorax was thought to be brought about by rupture of the arteriovenous malformation with likely intermittent small volume hemorrhages into the pleural space. Thoracic Surgery and Interventional radiology (IR) were each consulted for management of the arteriovenous malformation. Due to the patient's stable hemodynamic status and concern that an invasive procedure might enable a larger rupture and more substantial hemorrhage, the decision was made for embolization of the arteriovenous malformation.

Introduction

Pulmonary arteriovenous malformations (PAVMs) are an abnormal connection between pulmonary arteries and pulmonary veins without a concordant capillary network bridging the 2. Approximately 50% of cases of PAVMs are asymptomatic and the majority are incidentally detected on imaging. However, with significant pressures generated in the pulmonary arterial circuit a shunt can develop [1]. This abherent connection can lead to critical sequalae such as stroke, transient ischemic attacks, cerebral abscesses, or massive hemoptysis.

Hereditary hemorrhagic telangiectasia (HHT), also known as Osler-Weber-Rendu syndrome, is an autosomal dominant disorder of blood vessels characterized by epistaxis, telang-

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iectasias, and arteriovenous malformations [2]. Nearly 70% of cases of PAVMs are associated with HHT, making it the predominant association [2]. In a 1974 study from the Mayo Clinic, symptoms were more common in PAVM patients with HHT than those without [3]. While the other 30% of PAVMs unassociated with HHT are thought to be sporadic, PAVMs are known to have a female predominance and to be associated with pregnancy as a major risk factor [1]. Due to the increase in circulating blood volume of 40% and increased cardiac output, the risk for hemoptysis in pregnant patients with PAVMs is greater in theory than those non-pregnant PAVM patients. According to Ference and Shannon [4], 8 of the 28 cases (29%) of hemothorax associated with PAVM reported in the literature at the time occurred during pregnancy [4]. A separate study from Bevelaqua et al. cited that 30%-50% of HHT patients with AVM bleeding were pregnant [5].

Traditionally, treatment approach for bleeding AVM consists of a surgical or procedural approach. For hemodynamically unstable patients, a thoracotomy and surgical ligation of the bleeding vessels is preferred. However, for patients who are hemodynamically stable, a more conservative approach with endovascular embolization or non-operative management is indicated [6]. Mager et al. [7] conducted a study in 112 patients who received embolotherapy of PAVMs with mean follow-up of 62.2 months to evaluate long-term outcomes, defined as decreased right-to-left shunting and increased P_aO_2. They found that the long-term outcomes of embolotherapy were successful in 83% of patients overall and in 96% of patients in whom all angiographically visible PAVMs were embolized [7].

Case description

Presentation

We present a case of a 34-year-old G4P0030 patient at 26 weeks' gestation by in vitro fertilization with past medical history of hypertension (treated with oral labetalol) and infertility who presented to the hospital with abdominal pain. The patient stated her pain was in her left upper quadrant/left back. The pain was worse with deep breaths. The pain began a week prior and went away with hot compresses. The morning before arriving to the hospital the patient stated she woke up at 0300 with increasingly severe pain in the same area.

A computed tomography angiogram of the chest demonstrated a left-sided pulmonary arteriovenous malformation with adjacent complex left effusion on chest suspicious for a hemothorax (Fig. 1). The hemothorax was thought to be brought about by rupture of the arteriovenous malformation with likely intermittent small volume hemorrhages into the pleural space. Thoracic Surgery and Interventional radiology (IR) were each consulted for management of the arteriovenous malformation. Due to the patient's stable hemodynamic status and concern that an invasive procedure might enable a larger rupture and more substantial hemorrhage, the decision was made for embolization of the arteriovenous malformation.

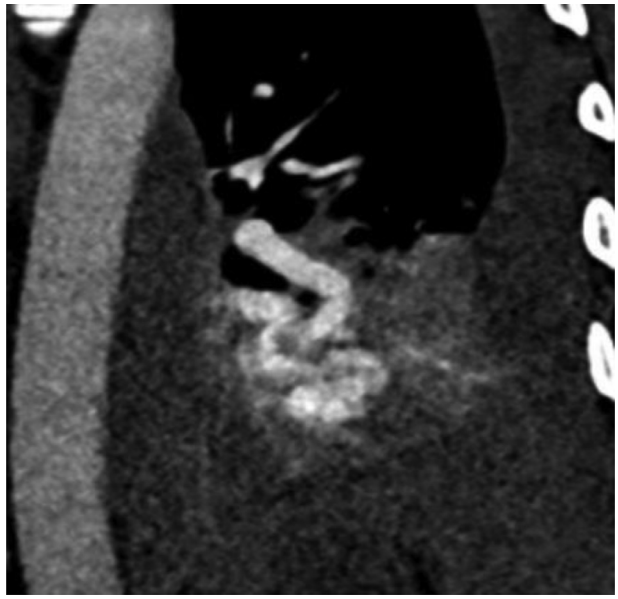

Fig. 1 – CT angiogram demonstrating left lower lobe pulmonary AVM with adjacent hemothorax.

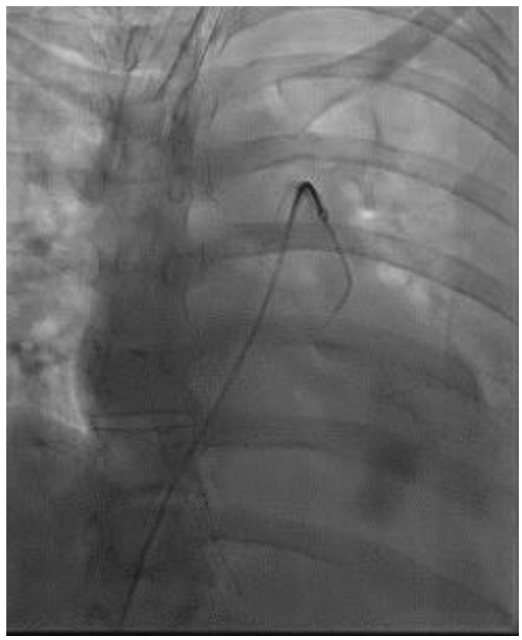

Fig. 2 – Selection of the left main pulmonary artery and left lower lobe pulmonary artery.

Technique

The patient was intubated and placed under anesthesia. Right femoral venous access was obtained using ultrasound-guidance. Care was taken to minimize fetal radiation. Main pulmonary artery access was obtained using a 7 French 70 cm sheath, 5 French flush pigtail catheter, and a 0.035 guidewire, and left lower lobe pulmonary artery access was obtained using angled glide catheter and hydrophilic wire (Fig. 2). Left lower lobe pulmonary artery angiography demonstrating the

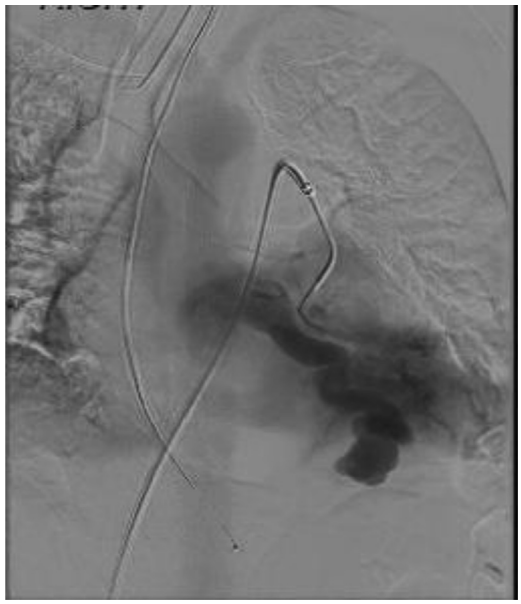

Fig. 3 – Angiography showing arterio-venous malformation.

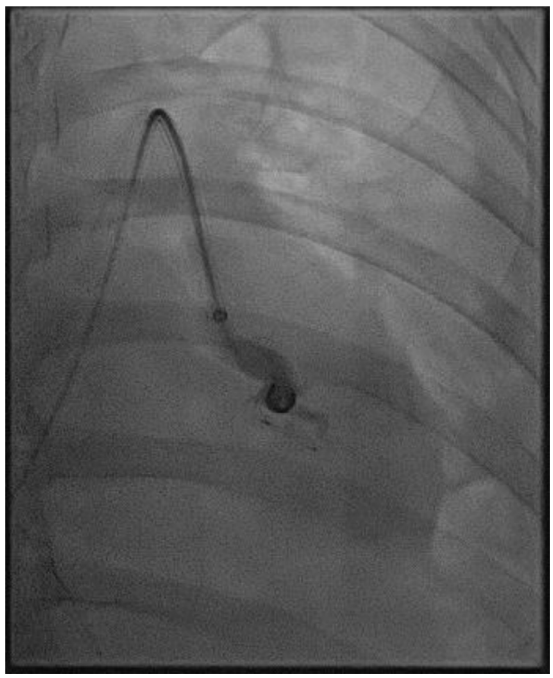

Fig. 4 – Angiography demonstrating AVM filling distal to 6 mm Amplatzer-IV plug.

known left lower lobe arteriovenous malformation with a dominant, enlarged, tortuous inflow artery with direct outflow into a dominant, enlarged, tortuous outflow vein (Fig. 3).

The angled glide catheter was used to select the segmental and subsequently the subsegmental pulmonary artery branch supplying the arteriovenous malformation, confirmed with repeat angiography. The inflow vessel was embolized with a 6 mm Amplatzer-IV plug (St. Jude Medical, St. Paul, MN). Repeat angiography demonstrated sluggish but persistent filling of the arteriovenous malformation (Fig. 4).

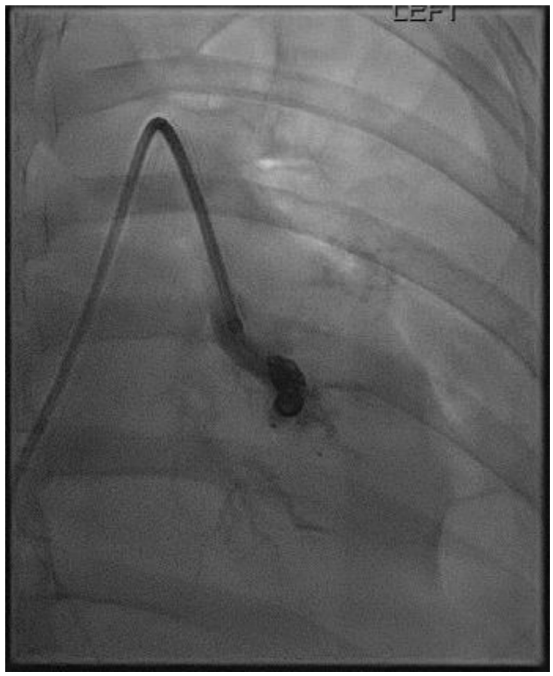

Fig. 5 – Angiography demonstrating additional 6 mm Tornado coil and 6 mm Nester coil with persistent outflow venous filling through the coil pack as well as a second inflow vessel.

Additional embolization was performed using a 6 mm Tornado coil and a 6 mm Nester coil (Cook Medical, Bloomington, Indiana, USA). Repeat angiography demonstrated persistent flow through the coil pack into the AVM nidus as well as a second inflow vessel which had been obscured by the larger, dominant inflow vessel (Fig. 5).

Additional coils were placed at the first coil pack, and the second inflow vessel was selected with the glide catheter and hydrophilic wire and embolized with additional Tornado and Nester coils (Fig. 6). Angiography at the second inflow vessel demonstrated stasis through that portion of the malformation with no flow into the arteriovenous malformation (Fig. 7).

Angiography at the segmental pulmonary artery demonstrated delayed filling of the pulmonary venous outflow of the malformation, suspected to reflect filling via flow through normal parenchyma. To confirm, this, the angled glide catheter was exchanged for a 5.5 French Fogarty balloon catheter (Teleflex Inc, Wayne, PA, USA). The balloon was gently inflated and an occlusion angiogram was performed, demonstrating delayed filling of the outflow vessel after parenchymal enhancement but no filling of the nidus (Fig. 8). A final angiogram from the left pulmonary artery demonstrated delayed filling of the venous outflow after parenchymal enhancement but no filling of the nidus (Fig. 9).

Recovery and follow-up

The patient had some improvement in her pain symptoms overnight after the procedure. She underwent thoracostomy tube placement with thoracic surgery on post-

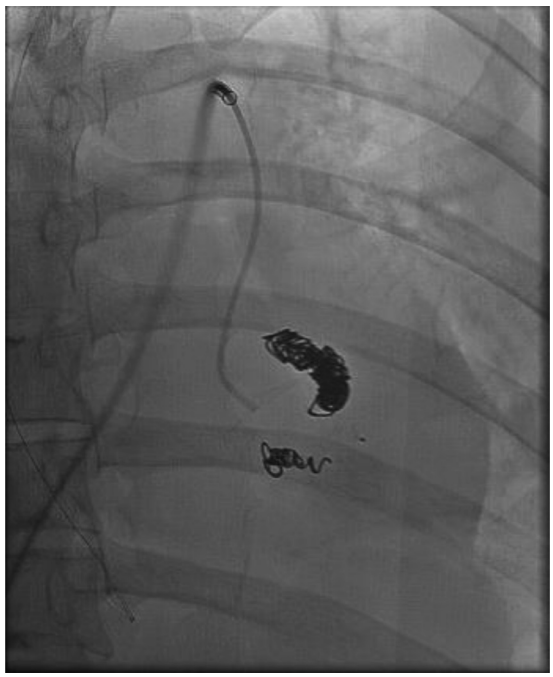

Fig. 6 – Fluoroscopy confirms additional Tornado and Nester coil placement in the dominant inflow vessel and the second inflow vessel.

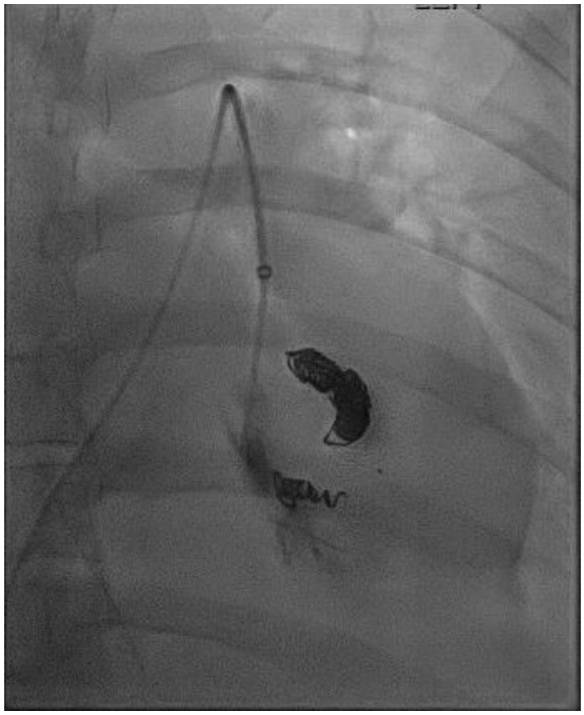

Fig. 7 – Angiography demonstrating no filling of AVM through the second inflow vessel after embolization with Nester coil.

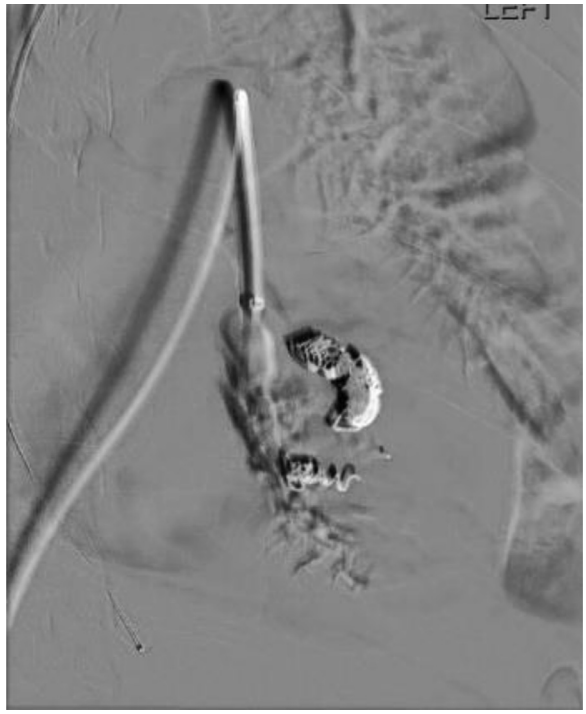

Fig. 8 – Occlusion angiogram with Fogarty balloon.

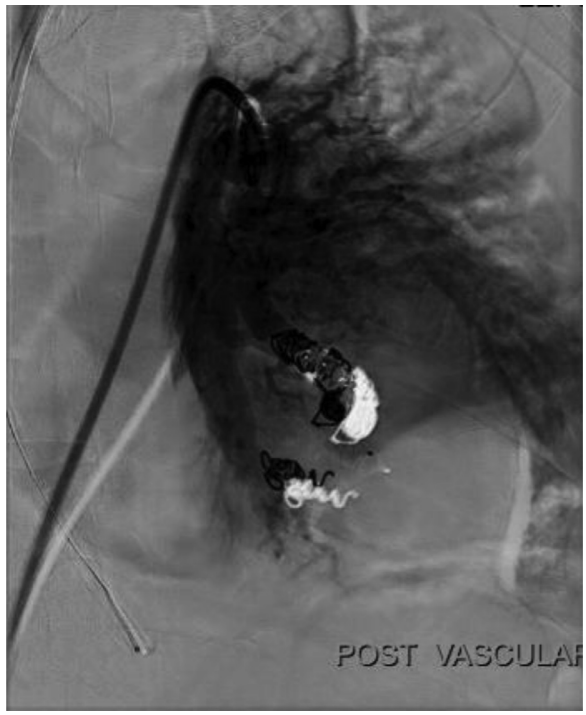

Fig. 9 – Final angiogram demonstrating no filling of the nidus.

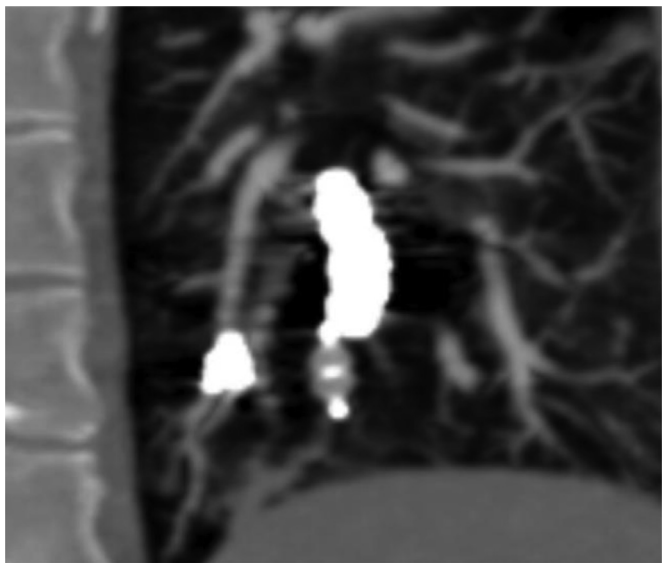

Fig. 10 – Four-month follow-up CT angiogram demonstrating involution of AVM nidus.

procedural day 1 with evacuation of 700 mL of old blood. She experienced some focal incisional pain at the thoracostomy site which was successfully treated with a thoracic epidural and subsequently with lidocaine patch and oral acetaminophen and cyclobenzaprine. Her recovery was otherwise uneventful and she was discharged home 3 days after embolization.

The pregnancy was delivered via Caesarian section at 39 weeks' gestation due to breech presentation and the patient's history of arteriovenous malformation. The delivery was uneventful aside from some postpartum hemorrhage successfully treated with uterotonics. Mother and infant were discharged home 2 days after delivery.

Due to high clinical suspicion of the patient having HHT, the patient was referred to Pediatric Genetics' clinic. Genetic testing demonstrated a variant of unknown significance in ACVRL1, a mutation associated with HHT and pulmonary arterial hypertension. A lip lesion was identified as a possible cutaneous arteriovenous malformation. Additional workup for HHT including abdominal ultrasound and brain MRI demonstrated no apparent additional arteriovenous malformations. Follow-up CT angiography at 4 months demonstrated involution the arteriovenous malformation nidus and no new arteriovenous malformation (Fig. 10). Agitated saline "bubble study" echocardiogram demonstrated no apparent right-to-left shunting. A plan was initiated for follow-up with Pulmonology and Genetics clinic and surveillance CT angiography in 3-5 years or prior to any additional planned pregnancies.

Discussion

PAVMs can affect multiple areas of the lungs with the lower lobes being most commonly affected. They can also be bilateral with the incidence ranging from 8% to 20%. In the general population it develops in 2-3 per 100,000 people [8]. Pregnancy is a period of high-risk for PAVM complications especially during the second and third trimester as there is an increase in cardiac output and a 40% increase in blood volume. In addition to hemodynamic changes, hormonal changes during pregnancy have been shown to play a role in the growth of arteriovenous malformations [9,10].

Shovlin et al. reported one of the chief conditions that can exacerbate PAVMs is iron deficiency, which is a common disease process in pregnant patients as the global prevalence is 41.8% [11,12]. As mentioned above, untreated PAVM can lead to multiple complications including massive hemoptysis, stroke, and hemothorax such as in this patient. Hemothorax incidence secondary to AVM, according to Shovlin et al., is 1.4%.

Given that severe complications can arise, it is important to diagnose PAVMs early. With the advantage of early diagnosis, early monitoring and potential treatment can be administered. Shovlin et al. found that there was a statistically significant increase in survival of women diagnosed with PAVMs previously.

The symptoms a patient describes can initially seem obscure, especially in those who were asymptomatic previously, such as this patient. Symptom onset to hemothorax can be rapid, particularly in pregnancy, as blood volume expansion can exacerbate the AVM. A high clinical suspicion for HHT is necessary for patients such as this, so a referral to a geneticist and proper monitoring as well as treatment can be conducted once the patient has recovered from their presenting illness [12].

Patient consent

Written, informed consent was obtained from the patient represented in this case report for publication and use in teaching by Dr. William R. Winter. Spontaneous hemothorax in pregnant patient with pulmonary arteriovenous malformation.

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