Case report

Contained aortic rupture in a term pregnant patient during the COVID-19 pandemic

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SUMMARY
Aortic dissection and rupture is a rare occurrence in pregnant and postpartum patients. This case discusses the presentation and diagnosis of a patient with an acute contained thoracic aortic aneurysm rupture at 38 weeks of gestation, after presenting with throat pain and syncope during the COVID-19 pandemic. The patient underwent emergent caesarean delivery for non-reassuring fetal heart tracing, following which continued syncope workup revealed an aortic aneurysm and pericardial effusion. Diagnosis in this case was finalised with multimodality imaging, including transthoracic echocardiogram, and the patient underwent surgical aortic repair.

BACKGROUND
Aortic dissection and rupture are rare but life-threatening conditions, particularly during pregnancy. Most commonly, these conditions arise in the third trimester or postpartum period, with the most common cause of death being cardiac tamponade.1 2 Aortic dissections are stratified into two categories based on the Stanford Criteria, with Type A involving the ascending aorta and Type B not involving the ascending aorta.3 Type A aortic dissections are more common during pregnancy and postpartum, and can lead to a mortality rate as high as 50% if not treated surgically within 48 hours.4-6 Risk factors for aortic dissection include connective tissue disorders, such as Marfan syndrome and Loeys-Dietz syndrome, congenital cardiovascular anomalies, such as bicuspid aortic valve and aortic coarctation, familial history of aortic dissection and chronic hypertension.1 3 7

We present the case of a patient diagnosed with an acute aortic rupture at term during the COVID-19 pandemic. While previous case reports have described the diagnosis and management of aortic dissection and rupture during pregnancy, this case uniquely describes the challenges of diagnosing aortic dissection and rupture during the COVID-19 pandemic as well as the use of echocardiogram in determining this diagnosis.

CASE PRESENTATION
A 33-year-old female gravida 1 presented at 38 weeks of gestation to a hospital in Queens, New York, after a syncopal episode. The patient had received routine prenatal care with a private physician, and her pregnancy thus far had been uncomplicated. She reported feeling well the day prior to presentation, but woke from sleep around 04:00 on the day of presentation with shortness of breath, throat pain and ‘clamminess’. The patient’s husband woke to her falling off the bed, having lost consciousness and subsequently hitting her head and abdomen. On waking, the patient endorsed dizziness, mild shortness of breath and throat pain. On arrival to labour and delivery triage, the patient was noted to have oligohydramnios on pelvic ultrasound with an amniotic fluid index of 1, and fetal tachycardia with minimal to moderate variability. The patient herself was tachycardic to 109 beats/min with a blood pressure of 88/66 mm Hg, vital signs that are considered within normal limits for a pregnant patient. The patient was given a bolus of intravenous fluids, and a COVID-19 swab was sent in addition to standard inflammatory markers recommended in the work-up of COVID-19.

Figure 1 Chest X-ray on hospital day 1 with calcification 3.5 cm below the carina (arrow).

Figure 2 Coronal view of CT angiogram of the chest, moderate pericardial effusion and no pulmonary embolus.
Unusual association of diseases/symptoms

INVESTIGATIONS

An ECG demonstrated sinus tachycardia with non-specific T-wave inversions in lead III. A portable chest X-ray was obtained and demonstrated a calcification 3.5 cm below the carina with some suggestion of mediastinal widening, which was not commented on in the report (figure 1).

Given the non-reassuring fetal status and lack of improvement with intratrue resuscitation with intravenous fluids and positioning, the decision was made to proceed with caesarean delivery under spinal anaesthetic as the patient was asymptomatic and hemodynamically stable. The patient underwent an uncomplicated primary caesarean section using spinal anaesthetic, delivering a viable female infant with Apgar scores of 8 and 9 at 1 min and 5 min, respectively. After completion of the case, the patient’s COVID-19 swab had returned negative. Repeat COVID-19 swab was performed given the high false-negative rate. The patient’s laboratory results were otherwise significant for a troponin of 0.056 ng/mL and a D-dimer of 2301 ng/mL.

Given the patient’s history of a syncopal episode with an elevated D-dimer and troponin level, there was concern for a pulmonary embolism with right heart strain as the aetiology for her presentation. Subsequently, a CT angiogram of the chest was obtained and demonstrated moderate pericardial effusion and an ascending thoracic aortic aneurysm measuring 4.5 cm in anteroposterior (AP) diameter without dissection; no pulmonary embolus was present (figures 2 and 3).

The cardiology team was paged and performed a transthoracic echocardiogram (TTE) at bedside with findings of a large pericardial effusion and apparent clotted blood within the pericardial space, as well as an ascending aortic root aneurysm measuring 5.2 cm without obvious dissection (figures 4 and 5). The team then discovered that the patient’s father had died at age 40 from an acute aortic event.

TREATMENT

Due to concern for a concealed dissection and possible need for surgical intervention, the patient was urgently transferred to a cardiothoracic surgery team at a quaternary centre. There she underwent urgent valve-sparing aortic root replacement. Intraoperatively, the surgical team found significant hemopericardium with clotted blood, as noted on the TTE, with a ~1 cm contained ascending thoracic aortic rupture tamponaded by the pulmonary artery in the setting of a bicuspid aortic valve and ascending aortic aneurysm.

OUTCOME AND FOLLOW-UP

The patient’s post-operative course was uncomplicated from both an obstetric and cardiothoracic standpoint and she was discharged home on postoperative day 5. As an outpatient, she subsequently underwent genetic testing and was diagnosed with Loeys-Dietz syndrome secondary to an SMAD3 mutation.

DISCUSSION

Aortic dissection or rupture is a rare occurrence in pregnancy and postpartum. Aortic dissection during pregnancy makes up only 29 cases out of 9707 total cases documented by the International Registry of Acute Aortic Dissections over the past 20 years. Despite the rare occurrence of this disease, pregnancy itself is a risk factor for aortic pathology. In women under 40 years old, up to 50% of aortic dissections occur during pregnancy or postpartum.
The incidence of aortic dissection or rupture is 5.5 per million patients during pregnancy and postpartum, as compared with 1.7 per million among non-pregnant patients. In patients with aortic dissection during pregnancy, up to 62% are diagnosed with an underlying predisposing pathology. The most commonly inherited and congenital conditions include Marfan syndrome, bicuspid aortic valve and Loeys-Dietz syndrome. Obtaining an accurate family history can be essential in eliciting a patient’s risk for this condition during pregnancy. Our patient noted history of sudden death in her father around age 40 from an aortic dissection or rupture and was subsequently diagnosed with Loeys-Dietz syndrome. Loeys-Dietz syndrome is an autosomal dominant connective-tissue disorder due to genetic mutations, such as in the SMAD3 gene, as in our patient. It confers higher risk to patients during pregnancy, including an increased risk of aortic dissection in the third trimester. A thorough family history not only aids in making this rare diagnosis, but is also essential for preconception counselling in these patients.

Diagnosis of aortic dissection and rupture requires reliance on history, clinical examination and imaging. While the classic presentation of aortic dissection includes sharp chest pain radiating to the back and dyspnoea, syncope can be present in up to 13% of cases. In case reports of aortic dissection in pregnancy, symptoms have included tearing chest pain, back pain, dyspnoea, hypertension, nausea/vomiting, syncope and seizure. The broad overlap between these symptoms and other more common complaints of pregnancy such as labour, pre-eclampsia and peripartum cardiomyopathy may confuse the clinician in diagnosing an aortic dissection. The gold standard for diagnosing aortic aneurysm and dissection is the CT angiogram. If aortic disease is suspected antepartum, MRI can also be used, but the use is limited to hemodynamically stable patients. Studies have additionally shown the utility of the echocardiogram as a tool for diagnosis given its high sensitivity and specificity. Transesophageal echocardiogram has a higher sensitivity and specificity for diagnosing aortic dissection than TTE, but is an invasive procedure and less available in some places. TTE is a cost-efficient diagnostic modality that may aid in diagnosing hemopericardium, increasing diagnostic concern for aortic dissection or rupture, and allow for assessment of the aortic root size and the presence of bicuspid aortic valve. The TTE findings seen in this case, along with the patient’s history of syncope, were the predominant clinical clues for the cardiology team to determine the diagnosis of aortic aneurysm rupture.

At the time of our patient’s presentation, the COVID-19 pandemic both further complicated and aided in her diagnosis. Up to 37% of pregnant patients with COVID-19 have presented with dyspnoea. During the work-up of the patient’s syncope and dyspnoea, a D-dimer test was sent, a laboratory test that is frequently excluded from a dyspnoea work-up in pregnant patients given baseline elevations in D-dimer levels for healthy pregnant women. During the COVID-19 pandemic, however, D-dimer elevation has been associated with severe pneumonia in pregnancy and is included in the standard COVID-19 panel at our hospital. The elevated D-dimer and negative COVID-19 testing in our patient spurred further work-up for pulmonary embolism, despite her improved symptoms, and ultimately led to her diagnosis.

Aortic dissection and rupture are rare occurrences in pregnant patients, and require a high index of clinical suspicion given the similarity of symptoms with other pathologies during pregnancy, particularly during the COVID-19 pandemic. This case presents a rare pathology in a 33-year-old pregnant woman, in which COVID-19 testing and echocardiography served to clinch the diagnosis and facilitate transfer for urgent surgical intervention.

Learning points

- While the gold standard for diagnosing aortic aneurysm and dissection is CT angiogram, echocardiogram has the potential to quickly aid in diagnosis given its high sensitivity and specificity.
- Maintaining a broad differential is essential to patient care during the COVID-19 pandemic, as unrelated diagnoses with similar presentations to COVID-19 may still affect patients.
- In pregnancy, a patient presenting with aortic dissection may have similar symptoms to more common complaints such as labour, pre-eclampsia and peripartum cardiomyopathy.
- More than half of pregnant patients with aortic dissection in pregnancy have an underlying predisposing pathology; eliciting a thorough family history is essential to care of these women.

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