Pulmonary arteriovenous malformations (PAVMs) is a rare condition characterized by abnormal vascular connections between pulmonary arteries and veins. Increased pulmonary blood flow during pregnancy can exacerbate intrapulmonary shunt, which can cause stroke or pulmonary hemorrhage and lead to maternal death, miscarriage, and fetal growth restriction. PAVMs may be misdiagnosed, as their prominent symptoms are similar to those of pulmonary embolism (PE). We report herein a case of a 20-year-old woman diagnosed with PAVMs, who was initially suspected to have PE. She developed hypoxemia and deep vein thrombosis immediately after delivery that resulted in fetal growth restriction and asymptomatic acute cerebral infarction. Undiagnosed chronic hypoxemia was considered as a differential diagnosis of PE, since the patient showed signs of chronic hypoxia such as clubbing and cyanosis. Our findings underscore the importance of thorough systemic assessment during the first visit of patients with suspected PE.

Introduction

Pulmonary arteriovenous malformations (PAVMs) are abnormal vascular connections between pulmonary arteries and veins, diagnosed in approximately two to three persons per 100,000 population. Women are more likely to develop PAVMs than men (roughly 1.5 times higher incidence). PAVMs are mostly congenital and are underlying causes of hereditary hemorrhagic telangiectasia (HHT), and can lead to chronic hypoxemia and death due to stroke or brain abscess. Age at diagnosis of PAVMs reportedly ranges from 10 to 79 years, indicating that PAVMs may be undiagnosed in women of reproductive age. Previous studies reported that 18% of PAVMs tend to enlarge gradually, and pregnancy in particular is an exacerbating factor. However, there have only been a few reports on PAVMs during pregnancy and puerperium.

Pregnant and puerperal women are almost five times more likely to develop venous thromboembolism compared to those who are not. While the clinical presentation of PAVMs resembles that of pulmonary embolism (PE), it is important to avoid misdiagnosis as treatment strategies differ entirely, with the latter being an acute disease that still remains the leading cause of maternal death in developed countries. We herein report a case of PAVMs in a pregnant woman who was originally suspected of having PE.

Case presentation

A 20-year-old primigravida with no known past medical history, smoking history, or family history had fetal growth restriction (FGR) of −1.5 SD since the 30th week of gestation. No maternal condition which would contribute to FGR was identified. She delivered a healthy...
male infant weighing 2,260 g via vaginal delivery at 39 + 1 weeks in another hospital. Apgar score was 9 at both 1 and 5 minutes, and umbilical arterial blood pH was 7.37. She vomited for 1.5 hours after delivery, and although she did not complain of dyspnea, her oxygen saturation was 82% in room air. She had swelling in her left leg with accompanying pain. As PE was suspected, computed tomography pulmonary angiography (CTPA) was performed. Although results were not indicative of PE, deep vein thrombosis (DVT) involving the left common iliac vein and the left femoral vein, and a 5-mm nodule connected to blood vessels in the left pulmonary apex, were noted. The patient was transferred to our hospital for further evaluation and management.

On examination, the patient was not in acute distress and had no complaint of dyspnea. Her heart rate was 69 beats per minute, respiratory rate was 26 breaths per minute, oxygen saturation was 91% on O2 therapy at 10 L/min via a non-rebreather mask, and PaO2 was 47 mmHg on arterial blood gas analysis. Her left leg was swollen, with pain on palpation. Clubbing of the fingers and cyanosis of the lips were observed during physical examination (Figure 1). Respiratory examination was normal, and jugular venous distention was absent. Chest radiograph revealed no abnormal findings, and there were no T-wave changes or an S1Q3T3 pattern on electrocardiogram. As laboratory findings included a D-dimer level of 1,090 ng/ml, CTPA was repeated; findings did not suggest PE. However, CTPA scans revealed the presence of a 5.5-mm PAVM in the left upper lobe and multiple PAVMs in both lower lobes (Figure 2). A temporary inferior vena cava filter was placed, and anticoagulant therapy (low-molecular-weight heparin) was initiated. Pulmonary scintigraphy (ventilation/perfusion scanning) revealed a pulmonary-to-systemic shunt rate of 40% (4-6% normal). Magnetic resonance imaging of the brain was performed to rule out the possibility of cerebral AVM, revealing that the patient suffered an asymptomatic acute cerebral infarction in the left occipital lobe (Figure 2).

On repeat history taking, the patient reported fatigue when she was a student and multiple episodes of epistaxis and cyanosis of nail beds since childhood. During hospitalization, her oxygen saturation was 70% on room air at rest, which decreased to 60% during low-level activities such as talking and breastfeeding. She had no complaint of dyspnea. She was discharged on postpartum day 10 and referred to another hospital for further management. She was explained that transcatheter embolization (TCE) had been performed for the PAVM in the left upper lobe, which relieved her hypoxia, and that genetic analysis led to a definitive diagnosis of HHT.

Discussion

The present case highlights two important clinical issues. First, undiagnosed chronic hypoxemia should be considered as a differential diagnosis of PE during pregnancy and peripartum. That is, while PE should first be suspected when dyspnea occurs during pregnancy, undiagnosed chronic disease, such as cyanotic congenital heart disease caused by right-to-left shunting of blood, should also be considered as an underlying cause of hypoxemia.

PE is common in pregnancy and accounts for >20% of all causes of maternal deaths in the United States.6) Most cases of severe PE occur during the postpartum period.7) There are no established predictive criteria for PE, and D-dimer should not be used to exclude PE in pregnancy.7) In our patient, DVT and hypoxemia led us to suspect PE. Since the clinical symptoms of PAVMs in pregnant women (e.g., dyspnea, chest pain, hemoptyisis) are similar.
to those of PE, PAVMs are often misdiagnosed as PE.8)

Second, our observations underscore the importance of thorough general physical examination and interviews during the first prenatal visit. Episodes of epistaxis and signs of chronic hypoxia, such as clubbing and cyanosis, may remain unnoticed until adverse events occur. PAVMs tend to rapidly enlarge during the second and third trimesters due to increased blood volume, cardiac output, and venous distensibility.9) PAVMs are associated with life-threatening complications, such as brain abscess, hemothorax, and stroke, and 1% of pregnancies with PAVMs result in maternal death.10) Furthermore, PAVMs can cause miscarriage or FGR due to chronic hypoxia.9) Physicians—especially obstetricians—need to identify PAVMs before they exacerbate during pregnancy. When clubbing and cyanosis of the fingers or an episode of epistaxis are noted, a diagnosis of PAVMs should be considered early on in order to prevent complications such as FGR and stroke with timely treatment.

PAVMs can be obliterated with appropriate treatment, such as TCE, i.e., the standard treatment. Indications for the treatment are a feeding artery diameter ≥ 3 mm or symptomatic PAVMs.1) Although no management protocols have been established for pregnant women with PAVMs, TCE in the third trimester is feasible.9) In the present case, hypoxemia completely disappeared after TCE in the puerperal period with no complications.

Approximately 90% of PAVMs are associated with HHT (Rendu–Osler–Weber syndrome).1) The Curacao criteria for the diagnosis of HHT include (1) epistaxis, (2) telangiectasia, (3) gastrointestinal telangiectasia and arteriovenous malformation, and (4) a positive family history. HHT is definitively diagnosed if at least three of the above criteria are present. If PAVMs are present before or during pregnancy, this is considered a high-risk pregnancy and TCE should be considered. Even if TCE is deemed unnecessary, the patient should be carefully observed for dyspnea, chest pain, and hemoptysis.

There is no consensus on opting for a cesarean section in pregnant women with PAVMs. Vaginal delivery is possible, as in the present case. Prophylactic antibiotics should be administered for the risk of bacteremia. A prolonged second stage of labor should be avoided, as chronic hypertension may cause hemodynamic stress, which in turn can exacerbate PAVMs. If cesarean section is performed, ruling out spinal AVM by magnetic resonance imaging should be considered before regional analgesia use.10)

In conclusion, although PE is one of the most life-threatening diseases in pregnant women, physicians should also be aware of the possibility of undiagnosed chronic hypoxia, such as that caused by PAVMs, which can significantly influence perinatal outcomes. In order to detect undiagnosed chronic diseases before they become exacerbated by pregnancy, sonography and laboratory tests during pregnancy should be supplemented with general physical examination and careful history taking during the first prenatal visit.

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Disclosure
There is no competing interest to declare.

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