Progression of Pulmonary Arteriovenous Malformation During Pregnancy: Case Report and Review of the Literature

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Pulmonary arteriovenous malformations (PAVM) expand during pregnancy because of increases in blood volume, cardiac output, and venous distensibility. More than half of the cases reported during pregnancy are associated with hereditary telangiectasia. In this case a 36-year-old primigravida presented at 24 weeks of gestation with new onset hemoptysis and dyspnea. A PAVM was noted in the right lower lobe during angiography and was successfully treated with embolization. Recurrence of symptoms occurred at 36 weeks of gestation after recanalization of the PAVM. Cesarean delivery was performed because of both this recurrence and breech presentation. The patient's symptoms subsequently resolved after delivery. The patient underwent a segmentectomy without complication 6 months after delivery. Thus, women with known PAVM or a history of hereditary telangiectasia should be followed with serial chest roentgenograms and arterial blood gases to detect acute progression of the PAVM. Embolization can be used during pregnancy if the PAVM is symptomatic.

Pulmonary arteriovenous malformation (PAVM) is a rare, but potentially life-threatening cause of hemoptysis during pregnancy. The increased blood volume and cardiac output, reorganization of collagen, and increased venous distensibility associated with pregnancy predispose women with PAVM to an increased likelihood of bleeding. We describe a case of PAVM initially presenting in pregnancy and review the available literature.

CASE REPORT

A 36-year-old primigravida presented at 24 weeks of gestation with the new onset of hemoptysis and Reprint requests to: Michael W. Varner, MD, Department of Obstetrics and Gynecology, University of Utah School of Medicine, 50 North Medical Drive, Salt Lake City, UT 84132.

Authors whose names are accompanied by an asterisk (*) have indicated, in accordance with the Accreditation Council for Continuing Medical Education (ACCME) Standards, that they have a relationship which could be perceived by some people as a real or apparent conflict of interest, but do not feel it has influenced their participation.
At 36 weeks of gestation, the patient was readmitted with a second episode of hemoptysis of 50 ml of bright red blood in a 5-minute period. The patient's hematocrit on the second admission was 41.9 percent. Repeat bronchoscopy verified the persistence of the right lower lobe AVM. Because of continued hemoptysis, the patient was transferred to the intensive care unit and orally intubated. A Fogarty catheter was placed into her right lower lobe bronchus and inflated with 0.75 ml of normal saline to tamponade the AVM. After approximately 6 hours, the balloon was deflated, and the patient immediately produced 10 to 20 ml of bright red blood. The balloon was re-inflated for an additional 24 hours and then removed without further hemoptysis.

Repeat angiography identified a persistent AVM with recanalization of the previous embolization site. Attempts at embolization were unsuccessful because the previously placed coil precluded vascular access. Arterial O₂ saturation remained above 95 percent. Antepartum fetal heart rate monitoring showed no sign of fetal compromise. Other obstetric parameters were normal except for the incidental finding of breech presentation.

Because of the failed re-embolization plus breech presentation, the patient underwent primary low transverse cesarean delivery, resulting in a healthy 3450-gm female infant with Apgar scores of 8 and 9 at 1 and 5 minutes, respectively.

The postoperative course was unremarkable and the patient had no further hemoptysis. She was discharged on postoperative day 4 and continued to do well. She underwent elective segmentectomy with removal of the AVM at 6 months postpartum.

**REVIEW OF THE LITERATURE**

Pulmonary AVM is a relatively rare condition, but over 200 cases have been described in nonpregnant patients. The etiology of PAVMs may be either congenital or acquired (1). Approximately half of all PAVMs are associated with the syndrome of hereditary telangiectasia or Osler-Weber-Rendu disease (2, 3). Acquired PAVM can result from trauma, schistosomiasis, hepatic cirrhosis, and carcinoma (1).

**PAVM DURING PREGNANCY**

Nine PAVMs associated with pregnancy have been previously reported (2, 4–6) and are listed in Table 1. In many of these cases, PAVM symptoms progressed rapidly during pregnancy, and several had documented radiographic progression of their lesions (5, 7).

**ETIOLOGY OF PAVM PROGRESSION**

In 1966 Pritchard showed that the average increase in blood volume during a normal singleton pregnancy is approximately 1570 ml, occurring most rapidly during the second trimester (8). Clapp et al. (9) found that by 11 weeks of gestation blood volume has increased by as much as 11 percent. He also noted an increase of 30 to 50 percent in cardiac output in pregnant women. The increase in blood volume and cardiac output results in increased pulmonary blood flow, with preferential flow across the low resistance of the PAVM. The augmentation of flow across the PAVM causes dilatation of the AVM (4, 6).

Venous distensibility increases by 150 percent during pregnancy with a return to normal by 8 to 12 weeks' postpartum (10). Progesterone causes smooth muscle relaxation and leads to arterial and venous distention, and subsequent decreasing resistance across the AVM, which leads to progression in size and shunt fraction (6, 10, 11).

The pregnancy-associated increase in steroid hormones results in an increased incidence of AVM rupture. The risk of rupture of splenic artery, renal, and cerebral AVMs are all increased during pregnancy (2). The data in Table 1 suggest a similar trend with PAVMs during pregnancy.

**PRESENTATION**

In the nonpregnant patient, usually PAVMs are diagnosed at an average age of 41 years (12). The reported cases of pregnancy-associated PAVM involve women between the ages of 20 and 37 years old, with the majority of cases presenting in the late second trimester (23–28 weeks' gestation, see Table 1).

PAVM, in the asymptomatic, nonpregnant patient, is diagnosed most commonly as an incidental finding on a routine chest x-ray. However, a classic triad of signs and symptoms of PAVM has been reported, which consists of dyspnea, cyanosis, and clubbing (7, 14, 15). Chronic hypoxia associated with PAVM can result in cyanosis and clubbing. A shunt of at least 20 percent is required to produce hypoxia severe enough to result in these findings (13, 14).

The most common presenting symptoms associated with PAVM in pregnancy are dyspnea and chest pain. Eight of the nine pregnant women with PAVM...
TABLE 1  
Summary of previous case reports of pulmonary arteriovenous malformations in pregnancy

<table>
<thead>
<tr>
<th>Author</th>
<th>Patient age (years)</th>
<th>Parity*</th>
<th>EGA presentation† (weeks)</th>
<th>Signs and symptoms</th>
<th>Course</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chanatry (2) 1992</td>
<td>23</td>
<td>2-0-0-2</td>
<td>35</td>
<td>Acute onset chest pain, dyspnea, hypoxemia and hypertension.</td>
<td>Hemothorax required chest tube and intubation C delivery at 32 weeks viable fetus. Subsequent resection of pulmonary tissue.</td>
</tr>
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<tr>
<td>Laroche et al. (11) 1992</td>
<td>37</td>
<td>0-0-2-0</td>
<td>29</td>
<td>Increased telangiectasia, dyspnea, cough, pleuritic chest pain, history of cyanosis and clubbing.</td>
<td>Continued massive hemothorax severe dyspnea and circulatory collapse. Emergent lobectomy and subsequent cesarean delivery 2 months later.</td>
</tr>
<tr>
<td></td>
<td>(with a history of hereditary telangiectasia)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Baumgardner and Kroll (16) 1993</td>
<td>25</td>
<td>4-0-1-4</td>
<td>23</td>
<td>Dyspnea, chest pain, cough, respiratory distress.</td>
<td>Treated with oxygen supplementation, refused embolization, subsequent spontaneous vaginal delivery at 36 weeks.</td>
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<td></td>
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<tr>
<td>Bevelaqua et al. (19) 1992</td>
<td>24</td>
<td>24-26</td>
<td>24-26</td>
<td>Acute onset left chest pain (pleuritic), dyspnea, slight cyanosis with clubbing, decreased breath sounds on left chest, hemothorax.</td>
<td>Cui embolization, normal spontaneous vaginal delivery at term, with no further bleeding except for epistaxis.</td>
</tr>
<tr>
<td></td>
<td>(with family history of Osler-Weber-Rendu)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gammon (5) 1990</td>
<td>27</td>
<td>24</td>
<td></td>
<td>Left chest pain, dyspnea, hemoptysis, bilateral pleural effusions, PO₂ 54 mmHg telangiectasia on lips and mucous membranes, clubbing and hemothorax.</td>
<td>Treated with diuresis, oxygen supplementation, and afterload reduction. Worsening hypoxia required intubation and embolization. Induced labor at 30 weeks resulting in normal vaginal delivery. Postpartum shunt reduced from 23% to 7%.</td>
</tr>
<tr>
<td>Steinberg and McClanahan (15) 1955</td>
<td>31</td>
<td></td>
<td>Postpartum</td>
<td>Dyspnea on exertion, fatigue, 6 previous normal spontaneous vaginal deliveries, anemia.</td>
<td>Developed dyspnea, clubbing, and cyanosis. Operation offered but denied, no follow-up.</td>
</tr>
<tr>
<td>Steinberg and McClanahan (15) 1955</td>
<td>34</td>
<td>6-0-0-6</td>
<td></td>
<td></td>
<td>Treated with iron, had normal spontaneous vaginal delivery. Subsequent recurrent episodes of fatigue, dyspnea, heart attack at 50 years old undergoes left lobectomy.</td>
</tr>
</tbody>
</table>

* Parity: term-premature-abortion-living children (as written numerically).
† EGA, estimated gestational age when signs and symptoms first occurred.
presented with a complaint of dyspnea, whereas five reported chest pain, usually pleuritic in nature. These symptoms are very similar to those associated with pulmonary embolus. In two of the cases reported, pulmonary embolus was felt to be the most likely diagnosis before identification of the PAVM. Four of the previously reported PAVMs in pregnancy were noted to have significant clubbing and cyanosis. It should be emphasized that PAVM may initially present in pregnancy with nothing more than the common complaints of fatigue, dyspnea, and musculoskeletal discomfort seen in late pregnancy.

Five of the nine previous reports were associated with hemotherax and/or hemoptysis. These symptoms are usually associated with acute rupture of the AVM leading to intrapulmonnic and intrapleural hemorrhage. In the nonpregnant population, such complications can be expected in approximately 10 percent of patients (13, 14). The apparent increased incidence of rupture noted in the pregnant patient is consistent with the tendency noted toward rupture of cerebral, renal, and other AVMs during pregnancy.

Half of the women with reported PAVM during pregnancy also had a history of familial hereditary telangiectasia. This is consistent with the known association of PAVM with hereditary telangiectasia in the general population. Therefore, it is important to inquire about family history and the presence of oral or other telangiectasia in women who present with signs and symptoms consistent with PAVM. These women are more likely to have multiple lesions, symptoms that progress more rapidly, and a higher rate of complications (12).

An effect of progressive maternal parity on PAVM symptomatology has been proposed (2, 6). Several of the cases reported involve women with a parity of 2 or greater. Steinberg and McClenaham (15) reported a case of a woman who had six uncomplicated pregnancies and presented with dyspnea and fatigue during her seventh pregnancy. After this final pregnancy, she suffered from recurrent episodes of dyspnea on exertion, fatigue, and headache and ultimately required a lobectomy.

**DIAGNOSIS**

PAVM should be considered in all pregnant women who present with acute or chronic pulmonary compromise. Women who present with hemoptysis warrant prompt evaluation including chest roentgenogram, sputum culture, and microscopic analysis, tuberculin skin testing, bronchoscopy, and possibly pulmonary angiography. Bronchoscopy can be used to identify the site of hemorrhage.

The differential diagnosis for hemoptysis during pregnancy is similar to that in the nonpregnant patient. Infection, neoplasm, and vascular disease all must be considered. A complete differential diagnosis for hemoptysis during pregnancy is found in Table 3.

Physical examination should be used to detect any sign of chronic hypoxia, such as clubbing or cyanosis. Careful pulmonary examination is required to rule out significant hemotherax or effusion. Auscultation of the chest may reveal the presence of a vascular bruit (15, 22). Any mucocutaneous lesions should also be noted as these would be consistent with the diagnosis of hereditary telangiectasia.

Dines et al. (12) found that in the nonpregnant patient radiographic evidence is present in almost all
cases of significant PAVM. Therefore, the chest roentgenogram is a useful diagnostic tool. Two-dimensional echocardiography with the administration of agitated saline has been used by some to screen for vascular malformations (16, 17). However, pulmonary angiography is necessary for definitive diagnosis (14). The angiogram will aid in the determination of number, location, and vascular origin of all PAVM present.

**MANAGEMENT OF PAVM DURING PREGNANCY**

Management of the patient with a PAVM depends on the severity of the individual lesion(s). Initial efforts should be directed to the stabilization of the patient using respiratory support, consisting of supplemental oxygen and bedrest as necessary. Once a source of bleeding has been identified in the patient with hemoptysis, endoscopic placement of a Fogarty catheter can be used to stop ongoing hemorrhage (18). Patients with severe symptoms or continued bleeding may require more definitive therapy including embolization or surgical removal of the AVM.

Little has been published regarding the appropriate management of PAVM during pregnancy. In the present case, attempts were made to embolize the PAVM. Embolization has become an accepted method of treatment for PAVM in the nonpregnant patient (19–22). Keller et al. (20) states that embolization is a safe alternative to surgical treatment whereby a significant amount of normal lung parenchyma is spared (20). Initial attempts at embolization were successful in our patient, but recanalization and recurrent hemoptysis occurred as the pregnancy progressed. However, in two of the nine case reports, patients were treated successfully with coil embolization without further progression or recanalization of the PAVM (5, 19).

Lobectomy or segmental resection of the PAVM has also been used as a definitive treatment. One patient with a significant PAVM underwent emergent lobectomy at 29 weeks’ gestation with subsequent resolution of all symptoms and eventual cesarean delivery of a healthy male at approximately 37 weeks’ gestation (11).

The optimum method of delivery in the patient with PAVM remains unclear. Vaginal deliveries were accomplished in seven of the cases reported, whereas two required cesarean delivery. One patient with significant hemothorax was treated conservatively with a chest tube and oxygen supplementation and eventually delivered vaginally (16). Three of the most symptomatic patients (i.e., those that presented with hemothorax or severe hypoxia) were treated with coil embolization or resection while pregnant and eventually delivered vaginally.

Two cases of worsening PAVM eventually required operative abdominal delivery. One patient who presented at 23 weeks of gestation with severe hypoxia required a cesarean delivery at 32 weeks’ gestation after progression of her hypoxia (4). A similar case resulted in a cesarean delivery at 32 weeks’ gestation (2).

Vaginal delivery is possible in those cases where no significant symptoms exist or when there is no progression of the disease process. Both of the cases that were delivered abdominally involved women with worsening PaO₂ or progressive symptoms of rupture, such as hemoptysis or hemothorax. In our case, an abdominal delivery was felt to be most appropriate secondary to the breech presentation of the fetus and to avoid the possibility of continued or additional rupture of the PAVM. However, a vaginal delivery can be accomplished when no signs or symptoms of progression or significant compromise are present.
REFERENCES