

Vanishing lung syndrome in a pregnant woman: case report and literature review

Síndrome dos pulmões evanescentes em uma gestante: relato de caso e revisão de literatura

Síndrome del pulmón evanescente en una mujer embarazada: aporte de un caso y revisión de la literatura

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Abstract

The presence of an underlying lung disease in pregnant women can present as a challenging case. The Vanishing Lung Syndrome is considered a rare condition, in which pulmonary emphysema is usually observed. We describe a unique case of Vanishing Lung Syndrome in pregnant patient. Female patient, 29 years old, started having progressive dyspnea on exertion in 2019, without other associated symptoms. She was hospitalized and a chest imaging study showed the presence of a large bulla in the upper right hemithorax with “hypertensive” behavior. The patient was lost to follow-up and returned two years later, pregnant, with a gestational age of 17 weeks, with an important restrictive component described in the spirometry. A bullectomy and right middle lobectomy were performed, and the patient showed significant symptom and pulmonary evaluation improvement. There are no documented cases of this rare condition in a pregnant patient, with an emphasis on seeking to maintain maternal and fetal well-being. The management of Vanishing Lung Syndrome in pregnant women is challenging, especially when lung function is impaired, as in the described case.

Keywords: Pregnancy; Vanishing lung; Bullectomy.

Resumo

A presença de doença pulmonar subjacente em gestantes pode apresentar-se como um caso desafiador. A Síndrome dos Pulmões Evanescentes é considerada como uma condição rara em que se observa costumeiramente o acometimento enfisematoso pulmonar. Descrevemos um caso único de Síndrome do Pulmão Evanescentes em uma paciente grávida. Paciente do sexo feminino, 29 anos iniciou em 2019 quadro de dispneia aos esforços, progressiva, sem outros sintomas associados. Foi internada e evidenciado em estudo de imagem de tórax a presença de bolha volumosa em campo superior de hemitórax direito de comportamento "hipertensivo". A paciente perdeu seguimento e retornou após dois anos, gestante, com idade gestacional de 17 semanas, com importante componente restritivo, descrito na espirometria, sendo optado pela realização de bulectomia e lobectomia média a direita, evoluindo com melhora importante dos sintomas e da avaliação pulmonar. Não há casos documentados dessa rara condição em paciente grávida, com ênfase na busca pela manutenção do bem-estar materno e fetal. O manejo da Síndrome dos

Pulmões Evanescentes em gestantes é algo desafiador, sobretudo quando existe comprometimento da funcionalidade pulmonar, como no caso descrito.

Palavras-chave: Gestante; Pulmão evanescente; Bullectomia.

Resumen

La presencia de una enfermedad pulmonar subyacente en mujeres embarazadas puede presentar un caso desafiante. El Síndrome del Pulmón Evanescente se considera una entidad poco frecuente en la que suele observarse afectación enfisematosa pulmonar. Describimos un caso único de Síndrome del Pulmón Evanescente en una paciente embarazada. Paciente mujer de 29 años que inició en 2019 con disnea de esfuerzo progresiva, sin otra sintomatología asociada. Fue hospitalizada y estudio imagenológico de tórax evidenció la presencia de ampolla voluminosa en campo superior de hemitórax derecho con comportamiento “hipertensivo”, descrita en espirometría, optándose por bullectomía y lobectomía media derecha, evolucionando con importante mejoría de síntomas y evaluación pulmonar. No hay casos documentados de esta rara condición en pacientes embarazadas, con énfasis en la búsqueda del mantenimiento del bienestar materno. El manejo del síndrome del pulmón evanescente en mujeres embarazadas es un desafío, especialmente cuando la funcionalidad pulmonar está comprometida, como en el caso descrito.

Palabras clave: Embarazo; Pulmón que desaparece; Bullectomía.

1. Introduction

The presence of underlying pulmonary disease in pregnant women can present as a challenging case. The management of the underlying disease, seeking to recognize and prevent the worsening of this disease, is extremely important. Moreover, managing and minimizing the side effects of the different therapies during these phases requires the expertise of a multidisciplinary team, involving the collaboration between intensive care physicians, pulmonologists, and obstetricians, in an effort to minimize morbidity and mortality to the fetus and the mother. (Budey et al., 2005; Sharma et al., 2009; Vij et al., 2014)

The Vanishing Lung Syndrome (VLS) was initially described in 1937 by Burke, who reported the history of a 35-year-old female smoker who developed progressive dyspnea and acute respiratory failure secondary to a giant lung bulla (Burke, 1937). It is considered a rare condition, in which pulmonary emphysema is usually observed, especially in the upper lobes, occupying more than one-third of one or both pulmonary fields. Its pathophysiology is still not completely understood, but it is known that alveolar wall destruction occurs and, consequently, it results in the formation of subpleural bullae, which agglutinate to form a giant bulla (Shah et al., 2007; Mohammad et al., 2013; Gao et al., 2015). This condition is often underdiagnosed or mistaken by pneumothorax, making it essential to perform an adequate anamnesis, associated with the indicative radiological exams (MacDuff et al., 2010; Lai et al., 2013).

Some cases of this syndrome have been reported in the literature. However, there are no documented cases of this rare condition in a pregnant patient, with an emphasis on seeking to maintain maternal and fetal well-being. Therefore, this case illustrates the approach to this rare syndrome in a pregnant patient in the second trimester of gestation, who already had compromised pulmonary function.

2. Methodology

The present study is a case study report (Estrela, 2018), carried out by collecting data from medical records and review of literature, whose search was performed in PubMed, Scielo and Lilacs. The literature was searched using keywords of Pregnancy; Vanishing lung; Bullectomy. Case reports, case series and observational studies describing the incidence, clinical features, and outcomes of The Vanishing Lung Syndrome patients were included in the review. Demographic features, risk factors, clinical features, diagnostic methods, treatment and outcome were analyzed. All the authors approved this selection process. Written informed consent was obtained from the patient to publish this case report and the project was approved by the Research Ethics Committee of Hospital Geral Dr Cesar Cals, under number: 5422022.

3. Case Report

A 29-year-old female patient started having progressive dyspnea on exertion in 2019, with no other associated symptoms. She sought medical care at a tertiary hospital in the city of Fortaleza, state of Ceará, Brazil, where hospitalization was indicated. A chest imaging study was performed (Figure 1), which disclosed the presence of a large-volume bulla in the upper right hemithorax, measuring 15 x 11 cm with “hypertensive” behavior, herniating to the opposite side, displacing mediastinal structures and determining segmental atelectasis, in addition to important dextroconvex scoliotic deviation of the dorsolumbar vertebral axis. A surgical approach was then proposed to perform a bullectomy. However, the patient was lost to follow-up and did not undergo the procedure.

Figure 1 - Chest imaging study.



Caption: Left - High Resolution Computed Tomography of the Chest. Right - Chest X-ray. Source: Authors.

The patient returned to the hospital two years later, while pregnant (Gestational Age: 17 weeks) due to dyspnea progression. She was then transferred to Hospital Geral Dr César Cals, for joint follow-up with the Pulmonology, Thoracic Surgery and Obstetrics teams. On the admission physical examination, the presence of significant scoliosis was observed in the thoracic region; on palpation, there was reduced chest expansion on the right and on auscultation, a vesicular murmur was present on the left and abolished in the lower two-thirds of the right hemithorax. The other organs and systems showed no alterations on physical examination. A spirometry test was performed, which showed severe restrictive pulmonary disorder [pre-bronchodilator: FVC 0.96 (34.4%), FEV1 0.79 (32.7%) and FEV1/FVC 82.27; post-bronchodilator: FVC 0.98 (35.4%), FEV1 0.85 (35.4%) and FEV1/FVC 86.46]. The echocardiogram showed an ejection fraction (EF) of 69% and pulmonary artery systolic pressure of 30 mmHg. She underwent an obstetric ultrasonography, which showed fetal heartbeat (158 bpm) and active fetal movements, with no evidence of intrauterine growth restriction (fetal weight = 181 grams). She also had a normal alpha-1-antitrypsin level (194 mg/dL).

In this context, the case was discussed among the specialties and the surgical approach was indicated, which occurred when the patient had a gestational age of 21 weeks and 3 days, with the patient being submitted to a bullectomy and middle lobectomy on the right (Figure 2).

Figure 2 - Images of the pulmonary bulla in the intraoperative period.



Caption: Left - Bullectomy and middle lobectomy surgical specimen. Right - Thoracotomy showing a pulmonary bulla. Source: Authors.

She developed hemodynamic instability in the immediate postoperative period, which was associated with high-output drainage with a bloody appearance in a chest tube, and developed hemorrhagic shock, requiring the use of vasoactive drugs and packed red blood cell transfusion. An exploratory thoracotomy was indicated, which revealed bleeding from the intercostal artery in the 1st intercostal space, followed by hemostasis and hemodynamic control.

After the procedure, the patient showed clinical improvement and was weaned from the vasoactive medication. Extubation was performed on the third postoperative day and the patient was discharged from the intensive care unit on the fifth postoperative day for follow-up in a pulmonology ward. A new obstetric ultrasonography was performed, which showed fetal heartbeats (153 bpm) and active intrauterine fetal movement, with no evidence of intrauterine growth restriction (fetal weight = 534 grams). A new spirometry was also performed, which showed moderate restrictive respiratory disorder [FVC 1.43 (51.4%), FEV1 1.26 (52.5%) and FEV1/FVC 88.28]. During her stay in the pulmonology ward, the patient showed clinical improvement, with maintenance of the pregnancy. She was discharged from the hospital for outpatient follow-up with the pulmonology, thoracic surgery and high-risk prenatal teams.

4. Results and Discussion

The vanishing lung syndrome is a condition characterized by the presence of giant pulmonary bullae, which may be idiopathic or secondary to other conditions. When secondary, it can be associated with conditions such as smoking, illicit drug abuse, pneumoconiosis, alpha-1-antitrypsin deficiency and chronic obstructive pulmonary disease (Sunanda et al., 2010; Muhamad et al., 2020). It usually affects young men, but it can occur in both sexes. Patients may present with a variety of symptoms, such as cough, chest pain, dyspnea and others (Tsao et al., 2012; Mohammad et al., 2013; Im et al., 2016).

The present study describes a challenging case of a pregnant woman at 17 weeks of gestation, who presented with a giant lung bulla, also known as Vanishing Lung Syndrome. There are no reports of similar cases in the literature in this specific group of patients, which makes the study extremely valuable to guide physicians regarding the best way to treat these patients. In agreement with the cases described in the literature, the patient had mild symptoms of dyspnea on exertion, which

progressively worsened over the years. Moreover, etiologically, no initial causal factor was found to justify the origin of the bulla, since the patient did not have any risk factors and her alpha-1-antitrypsin levels were within the normal range.

Pregnancy is a period in which several adaptations occur in the woman's body. Respiratory function is significantly affected, as the gravid uterus progressively results in an elevation in the resting position of the diaphragm and a change in the chest configuration, which enlarges in its anteroposterior diameter. In this context, in relation to respiratory mechanics, an increase in minute volume due to an increase in tidal volume is observed in the first trimester, whereas functional residual capacity and expiratory reserve volume decrease (Hirnle et al., 2013). The values obtained by forced spirometry, including forced vital capacity (FVC), forced expiratory volume in 1 second (FEV1), and peak expiratory flow (PEF) remain mostly unchanged during pregnancy. Therefore, the dyspnea complaints may occur during pregnancy. Moreover, risks to the health of the pregnant woman and the fetus can occur when associated with previous pulmonary conditions (Grindheim et al., 2012).

The main concern regarding this patient was the fact that she was a pregnant woman in the second trimester of pregnancy, whose pulmonary function was already altered, as shown by the spirometry (severe restrictive pulmonary disorder). In addition to the baseline condition, she also had significant scoliosis, which worsens the pulmonary function impairment. In this context, according to the pulmonary mechanics of the pregnant woman, it would be expected that the patient's pulmonary function would deteriorate over the months, which could generate significant risks for the mother and the fetus. Therefore, after a comprehensive analysis and discussion of the case, the surgical approach was chosen.

The surgical approach is indicated, above all, for patients who are symptomatic or who already have complications secondary to the underlying disease. Different techniques have been reported, with the choice being made according to clinical status and size of the lung bulla. Among these techniques, we can mention those performed via thoracoscopy, such as intracavitary drainage and laser ablation (Kim et al., 2016). There are also reports of progressive drainage through a Foley catheter. In the last decade, the bullectomy carried out through endoscopic resection emerged as an option, which has shown to be a safe approach. The resection must be accompanied by appropriate pharmacological therapies and effective respiratory physical therapy (Marchetti et al., 2015; Kadowaki et al., 2018)).

Due to the size of the bulla in this patient and the uncertainty of viable lung parenchyma, the open thoracoscopy approach was chosen, in which surgical excision was adequately performed, with satisfactory re-expansion of the trapped lung parenchyma. In the postoperative period, physical therapy was extremely important to help in the rehabilitation of adequate pulmonary function. Furthermore, a substantial improvement was observed in pulmonary function tests after the bullectomy, as shown by the second spirometry.

Post-bullectomy surgical mortality rates in patients with lung bullae with preserved lung function are as low as 0 to 2.5%. On the other hand, the overall mortality and morbidity rates are high in patients with compromised lung function (Schipper et al., 2004). Schipper et al. (2004) described that the main complications described are mainly related to prolonged air leak (53%), atrial fibrillation (12%), and mechanical ventilator-associated pneumonia (9%) (Schipper et al., 2004).

The patient required surgical reapproach due to complications associated with the procedure, i.e., bleeding. The reapproach was carried out satisfactorily, with no sequelae for the mother or the fetus. In contrast to what has been described in the literature, despite the fact that the patient initially showed compromised pulmonary function, the surgical procedure was a success, allowing the maintenance of the pregnancy and improvement in the pulmonary function of our patient.

A long-term clinical follow-up of this patient is extremely important, as several studies in the literature have described improvements in lung function, lung volume and clinical complaints. However, it was observed that these improvements decreased over the years (Schipper et al., 2004; Palla et al., 2005).

5. Conclusion

This report describes a successfully conducted case of a woman in the second trimester of pregnancy with Vanishing Lung Syndrome. The management of this condition in pregnant women is challenging, especially when lung function is compromised, as the case described herein. Future research should be carried out to help professionals better manage this rare syndrome and we expected that the reported case can be used as a guide for the best way to manage this disease in this group of patients.

Conflicts of interest

The authors declare no conflicts of interest.

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