Right-sided spontaneous, massive hemothorax in a 27-week pregnant lady: a case report

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Abstract

Spontaneous, massive hemothorax due to pulmonary varix is an uncommon, yet catastrophic cause of respiratory distress during pregnancy and postpartum. Presentation is often confused with pulmonary embolism, the treatment of which, worsens the condition. Diagnosis is challenging because of the elusive abnormalities on radiographic and bronchoscopic examination. Currently, there are no treatment guidelines for pulmonary varix. A 23-year-old pregnant lady in her second trimester, with intra-uterine fetal demise, presented to our emergency with dyspnoea, fever, and right-sided chest pain for three days. Chest X-ray and therapeutic thoracocentesis revealed massive right-sided hemothorax and computed tomography suggested pulmonary varix as the cause which was managed with emergency chest tube drainage. Massive, spontaneous hemothorax due to pulmonary varix in pregnancy is a rare occurrence, but it requires timely, multidisciplinary assessment and management for the betterment of the mother and the fetus.

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Introduction

Spontaneous, massive hemothorax is a rare, but catastrophic emergency condition. It is an uncommon cause of respiratory distress during pregnancy and postpartum. Hemothorax is defined as pleural fluid with a hematocrit greater than 50%.[1] Such patients may be asymptomatic or present with chest pain, dyspnea, shortness of breath, hemoptysis, epistaxis, or chest pain. The definitive diagnosis is made with contrast-enhanced computed tomography (CECT) and pulmonary angiography. Once a diagnosis of a hemothorax is established, almost always, a chest thoracostomy is required to evacuate the blood and clots inside the chest cavity and transfusion of blood products is often necessary. It may result from a variety of etiologies and in some patients, they may even remain unknown despite exploratory thoracotomy.[2,3] In any pregnant or postpartum patient with hemothorax, choriocarcinoma must be excluded.[4] Video-assisted Thoracoscopic Surgery (VATS) is the standard treatment but specific treatment depends on the cause.[5] There are currently no treatment guidelines for pulmonary varix.[6]

Case Report

A 23-year-old pregnant lady presented to the emergency department (ED) at KIST Medical College and Teaching Hospital (MCTH) in June 2023 from Sarlahi, a remote area of Terai, Nepal. She came with chief complaints of sudden onset of shortness of breath, fever, and chest pain for three days. Shortness of breath was gradual in onset and progressive in nature without any positional or diurnal variation. The severity increased from Modified Medical Research Council (MRC) grade 0 to grade 4 in the past three days. It was associated with right-sided chest pain which was dull aching, pressing type, non-radiating, and aggravated by lying down. She gave a history of fever with a maximum temperature recorded at 100.4°F, associated with chills and rigor, continuous type, relieved by taking antipyretic oral medication. She was in her 27th week of gestation, with uneventful antenatal visits. Her past medical and surgical history were insignificant. According to gravida (G), parity (P), and living (L) status, she was a multigravida lady, G_4, with 3 living issues, P_3L_3, without any complications in the past pregnancies. On arrival, she was dyspneic and appeared pale and cyanosed. Her initial vitals were pulse rate (PR) of 126

beats per minute (bpm), blood pressure (BP) of 110/70 mm of Hg, respiratory rate (RR) of 30 per minute, and saturation (SpO2) of 70% in the room atmosphere. She was categorized as triage II[7] and kept in the resuscitation bay, Monitors were attached and high-flow oxygen (O_2) was given via face mask at the rate of 15 liters per minute (L/min). Intravenous access with 18 gauge was gained and a pint of normal saline (NS) 500 ml was initiated.

On her systemic review of the respiratory system, the chest wall was asymmetrical, decreased movement on the right side and the trachea was shifted to the left. On percussion, the dull note was observed and auscultation revealed decreased air entry on the right side. On abdominal examination, her gravid uterus size was corresponding to 26 weeks of gestation (WOG), no tenderness, however, fetal heart sound (FHS) could not be heard. Her cardiovascular and central nervous system examinations were grossly intact.

Laboratory investigations sent were complete blood count (CBC), random blood sugar (RBS), renal function test (RFT), prothrombin time/international normalized ratio (PT/INR), blood grouping, and d-dimer. Meanwhile, extreme tachycardia at the rate of 185 beats per minute was revealed in bedside electrocardiogram (ECG), and severe metabolic acidosis with pH 7.2, HCO_3 8.7 mmol/L, pCO_2 20.3 mmHg, pO_2 67 mmHg and a lactate of 6.7 mmol/L in arterial blood gas analysis (ABG). Following management of the abnormal physiologies, in view of suspected septic shock, the empirical injectable broad-spectrum antibiotic of a combination of Piperacillin and Tazobactam, 4.5 grams and sodium bicarbonate ($NaHCO_3$) 50 mEq (1 mEq/kg) intravenous were given. On reassessment after 30 minutes of arrival, her BP dropped to 90/50 mmHg. Thus, second wide bore IV access was secured and inotrope (Inj. Noradrenaline) was started at 0.1mcg/kg/minute via syringe pump. Foley's catheterization was done with an initial output of 200 ml. Inj. Paracetamol 1gm was given for her pain.

Ultrasonography of the abdomen and pelvis (USG) showed a single intrauterine pregnancy of 27^{+3} WOG with no cardiac activity, right-sided pleural effusion, and minimal pericardial effusion with no intraperitoneal collection. Therapeutic tapping revealed bloody pleural fluid suggestive of hemothorax. Further, a portable chest X-ray (CXR) was done which showed homogenous opacity throughout the right lung field as shown in Figure 1. On-duty consultants of obstetrics and gynecology (OBG), cardiothoracic and vascular surgery (CTVS), and internal medicine departments were consulted in view of $G_4P_3L_3$ at 27^{+3} WOG with intrauterine fetal demise (IUFD) with massive right-sided effusion with severe metabolic acidosis in septic shock with severe anemia with suspected amniotic fluid (pulmonary) embolism.

A 32-French chest tube was inserted after consent, connected to a water seal drain and placement was confirmed with air column and drainage of bloody pleural fluid. The initial drain was 500 ml in less than 2 minutes and hence the tube was clamped. Thereafter, there was an improvement in her saturation of 94% with 12 L/min O2 via face mask and respiratory rate of 24 per minute. Post-procedural portable CXR was done to confirm the placement of the tube which was observed slightly high up.

The patient and her relatives were counseled about the possible need for immediate surgical thoracotomy for the management of the condition. However, due to financial limitations, they denied the surgical approach and opted for conservative management. Therefore, the patient was admitted to the surgical intensive care unit (SICU) and the night of admission was quite uneventful.

On the second day of admission, the chest tube was manipulated to locate the tip in the lower zone, and a CT Chest was done which revealed a tortuous and tubular dilated peripheral vein draining to the right superior pulmonary vein (maximum diameter 11 mm) suggestive of pulmonary varix. Also, it showed right-sided gross hemopneumothorax (approx. 680 ccs) with a chest tube in situ, adjacent parenchymal collapse, and features of mediastinal shift towards the left side (Figure 2).

The same day, a fresh stillbirth (male baby) was expelled spontaneously and the placenta was removed manually with a total blood loss of 100 ml. The patient was shifted to the surgical ward on 4th day of admission. Her vitals were stable and CXR was done in series to note improvement with a subsequent decrease in the hemothorax but poor lung expansion was observed with the development of pneumothorax (Figure 3).

A total of 2150 mL of old blood was drained from the right pleural cavity during her entire stay in the hospital. There was no subsequent clinical or imaging evidence of further blood loss. She was advised for bronchoscopy and VATS but refused due to financial issues and on the 12th day of admission she was discharged on request with a chest tube in situ as they finally agreed to visit a government hospital for further management. The patient was called via phone multiple times to know about her progress but she was unreachable thereafter.

Discussion

Spontaneous, non-traumatic hemothorax could be caused by numerous etiology of which pneumothorax is the most common cause, followed by vascular abnormalities, coagulopathies, neoplasia, thoracic diseases such as pulmonary infarction due to thromboembolism, infections or sequestrations.[8] Pulmonary arteriovenous malformation (AVM) is a disease with quite similar findings, clinically and radiographically. Pulmonary varix should be distinguished from it because their treatment strategies are completely different.[6] In pregnancy, diaphragmatic ectopic pregnancy, gestational trophoblastic disease (GTD), diaphragmatic endometriosis or rupture of subcapsular hematoma could also lead to spontaneous hemothorax. Idiopathic spontaneous hemothorax is a diagnosis of exclusion when every known cause has been investigated.[2,5] In our case, CT

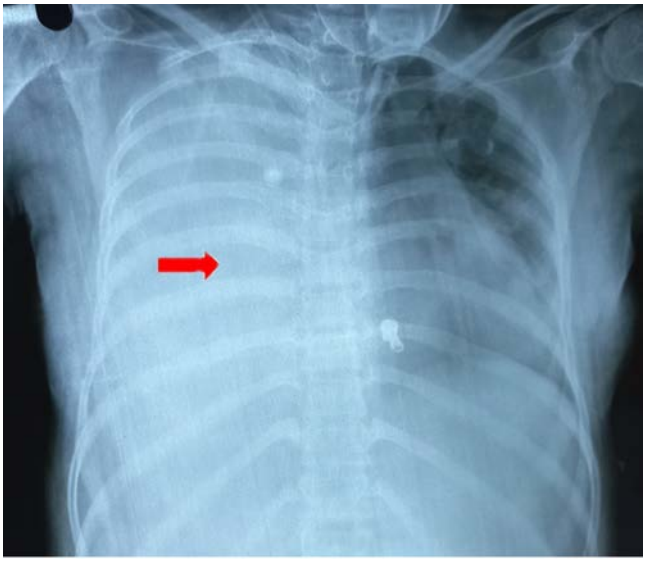

Figure 1. CXR showing homogenous opacity throughout the right lung field and mediastinal shift to the left.

chest was useful to construct the possible cause of the massive hemothorax, *i.e.* pulmonary varix. However, due to financial constraints, neither CECT chest nor pulmonary angiography could be performed on our patient to rule out thromboembolism/ AVM.

A pulmonary varix is as a localized aneurysmal dilatation of a segment of a pulmonary vein, also known as a pulmonary venous aneurysm, present at any age, without gender predominance, occurring in isolation or associated with obstruction of the pulmonary veins.[9–11] It was originally described in 1843 by Arnett and Patton.[12] Only 71 cases have been reported as of 2011 and the incidence remains unknown as it has been reported infrequently.[13]

Pulmonary varices are a rare pulmonary venous disorder whose etiology remains uncertain but can be either congenital or acquired and isolated or associated with varices in other organs as well.[14,15] Congenital varices develop during the embryonic period and may coexist with various heart diseases such as pulmonary vein atresia and anomalous pulmonary vein that were not present in our case. Acquired forms are associated with diseases with increased pulmonary vein pressure like mitral valve disease or distal occlusion of the pulmonary veins, liver cirrhosis, or emphysema.[15,16] In our case, bedside echocardiography was performed but did not reveal significant findings. It morphologically can be classified into three types: saccular type, tortuous type, and confluent type. The present case had a tortuous dilated vasculature in the right lung and thus was diagnosed with a tortuous type.[6]

Pulmonary varices are usually asymptomatic and are mostly incidental findings during health check-ups. They may not be visible on CXR or may present as solitary pulmonary nodules. CT chest may show tortuous, serpiginous vessels without a vascular nidus and normal-appearing pulmonary arteries, allowing varices to be distinguished from pulmonary AVM, as in our case. Catheter pulmonary angiography remains the gold standard but due to its

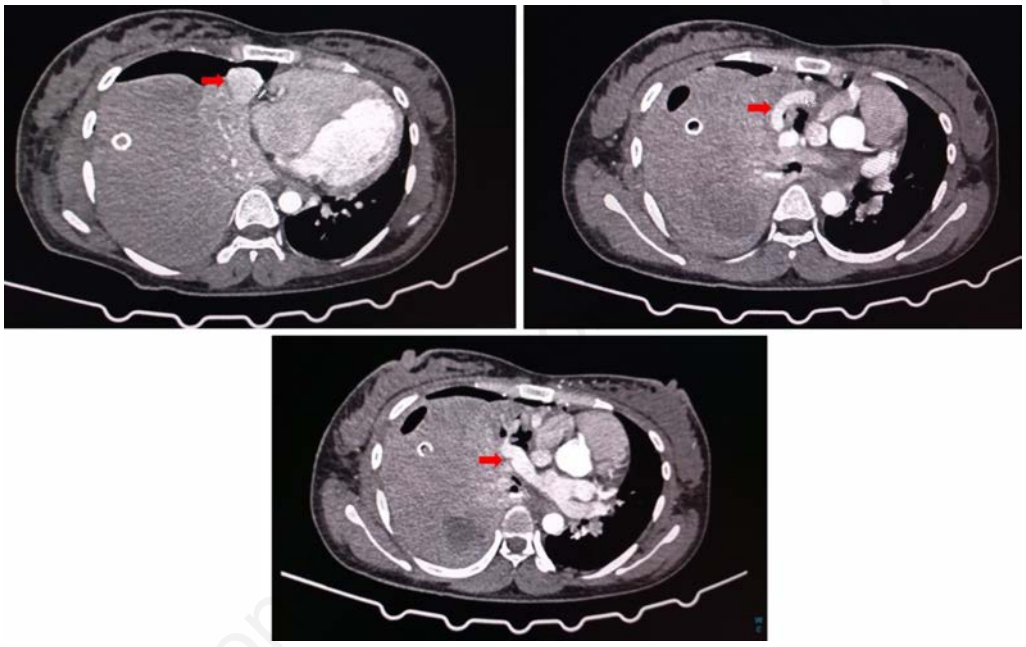

Figure 2. CT Chest coronal sections, showing pulmonary varix denoted by red arrows.

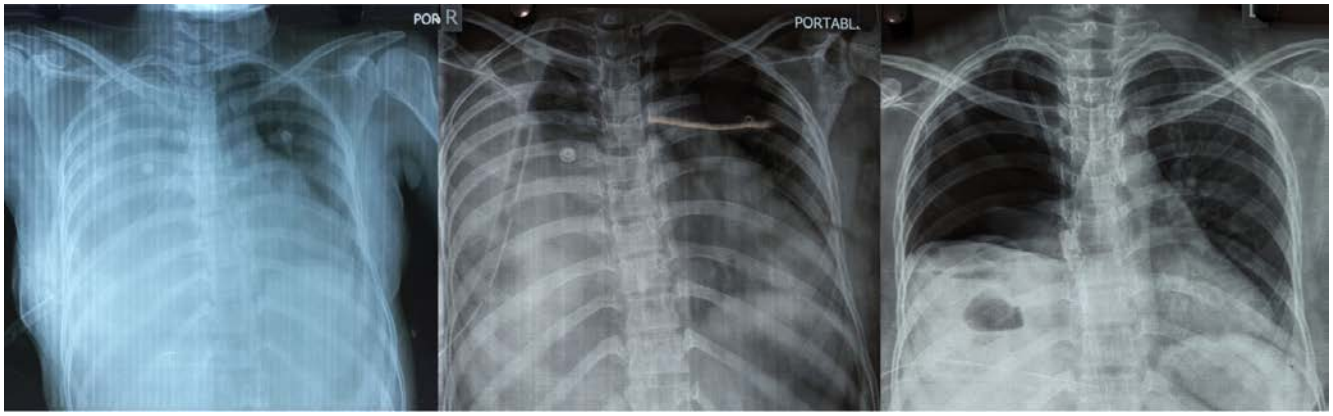

Figure 3. CXRs of days 2, 5, and 10 of admission showing improvement.

invasiveness, in recent years, the diagnosis of these diseases has been established using CECT chest and magnetic resonance imaging (MRI). Three-dimensional CT has also proven to show similar results.[6] The size criteria for diagnosis of a pulmonary vein varix have not yet been established, but an aneurysm is usually defined as an increase in 50% of the normal diameter.[10]

In hemothorax during pregnancy without evidence of fetal distress, management should be centered on the mother's pulmonary and hemodynamic status. If there is extensive hemorrhage with mediastinum shifts, it requires emergency treatment with termination of pregnancy. However, in our case, there was fetal demise already and hence management was focused on the mother.[16]

Treatment is usually unnecessary unless varices rapidly increase in size or complications such as hemoptysis, thromboembolic disease, or rupture occurs. They can be treated conservatively with chest tube insertion in most patients if the bleeding persists for less than 24 hours. Surgical intervention is reserved for patients with massive hemorrhage with hemodynamic instability, persistent bleeding for more than 24 hours, or retained blood clot refractory to tube drainage, all of which were present in our case. Before 1990, the surgical approach was usually through a thoracotomy, which has now been gradually replaced by the VATS approach with or without axillary mini-thoracotomy which is an easy, accessible, and safe procedure. Furthermore, this minimally invasive procedure is advantageous due to less wound pain, better wound cosmetics, and less post-operative respiratory impairment. Since VATS was unavailable in our hospital setting, conservative management with bed rest, oxygen supplementation, manual aspiration, and chest tube drainage were considered.[17,18] However, despite proper chest tube insertion and drainage in our case, there was no lung expansion and she developed pneumothorax. A possible cause of lung non-expansion could be a condition called "trapped lung" which is the sequela of remote pleural space inflammation resulting in the development of a mature, fibrous membrane that prevents lung expansion during fluid removal. Thus, there could be defective pleural healing even after inflammation in the pleural space has resolved which is considered as a complication of active pleural inflammation, or hemothorax as in our case.[19] Follow-up CECT chest could not be performed to support this theory or rule out other causes of non-expandable lungs such as fistula, endobronchial obstruction, or pleural disease.

This is, to the best of our knowledge, the first case reported of pulmonary varix leading to massive, spontaneous hemothorax in pregnancy with IUFD, in our hospital. On presumptive diagnosis of pulmonary embolism, many are treated accordingly which could be catastrophic. Hence, it requires timely, multidisciplinary assessment and management for the betterment of the mother and the fetus.

References

1. Martinez FJ, Villanueva AG, Pickering R, et al. Spontaneous hemothorax. Report of 6 cases and review of the literature. Medicine (Baltimore) 1992;71:354-68.
2. Doshi HM, Robinson S, Chalhoub T, et al. Massive spontaneous hemothorax during the immediate postpartum period. Tex Heart Inst J 2009;36:247-9.
3. Yung CM, Bessen SC, Hingorani V, et al. Idiopathic hemothorax. Chest 1993;103:638-9.
4. KR Graettinger, Avery DM Jr. Spontaneous hemothorax during pregnancy: a comprehensive review of the literature. Available from: https://jfmo.cchs.ua.edu/files/2014/04/MASSIVE-SPONTANEOUS-HEMOTHORAX.pdf
5. Ali HA, Lippmann M, Mundathaje U, Khaleeq G. Spontaneous Hemothorax: A Comprehensive Review. Chest 2008;134:1056-65.
6. Yamaguchi B, Kodama Y, Watanabe K, et al. Pulmonary varix clearly demonstrated by 3D-CT before pulmonary angiography. Radiol Case Rep 2022;17:4183-7.
7. Ebrahimi M, Heydari A, Mazlom R, Mirhaghi A. The reliability of the Australasian Triage Scale: a meta-analysis. World J Emerg Med 2015;6:94–9.
8. Patrini D, Panagiotopoulos N, Pararajasingham J, et al. Etiology and management of spontaneous haemothorax. J Thorac Dis 2015;7:520-6.
9. Bartram C, Strickland B. Pulmonary varices. Br J Radiol 1971;44:927-35.
10. Kim YH, Marom EM, Herndon JE, McAdams HP. Pulmonary vein diameter, cross-sectional area, and shape: CT analysis. Radiology. 2005;235:43-9; discussion 49-50.
11. Haddad J, Badran A, Pavão R, et al. Pulmonary varix: A case report. Rev Port Cardiol Engl Ed 2016;35:443.e1-443.e4.
12. Arnett JC, Patton RM. Pulmonary varix. Thorax 1976;31:107-12.
13. Berecova Z, Neuschl V, Boruta P, et al E. A Complex Pulmonary Vein Varix - Diagnosis with ECG gated MDCT, MRI and Invasive Pulmonary Angiography. J Radiol Case Rep 2012;6:9-16.
14. Agustin M, Shay S, Gonzalez J, et al. Massive pulmonary hemorrhage from bronchial varix. Case Rep Pulmonol 2020;2020:9175785.
15. Gleason JB, Shekar SP, Hernandez F, et al. Pulmonary varix: an uncommon pulmonary vascular anomaly. Clin Pulm Med 2017;24:87.
16. Lee HN, Hyun D. Pulmonary arteriovenous malformation and its vascular mimickers. Korean J Radiol 2022;23:202-17.
17. Luh SP, Tsao TCY. Video-assisted thoracic surgery for spontaneous haemopneumothorax. Respirology 2007;12:443-7.
18. Luh SP. Diagnosis and treatment of primary spontaneous pneumothorax. J Zhejiang Univ Sci B 2010;11:735-44.
19. Huggins JT, Doelken P, Sahn SA. The unexpandable lung. F1000 Med Reports 2010;2:77.