Pregnancy with hypoplastic left lung complicated by pneumothorax and pulmonary embolism

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Abstract
We report a case of a 34-year-old lady with past history of asthma and pulmonary tuberculosis, who presented 5 weeks pregnant with acute dyspnea. Her chest X-ray showed left-sided complete lung collapse and concomitant right-sided pneumothorax. The pneumothorax was initially managed conservatively with a chest tube but due to its persistence despite suction, was subsequently changed to a Pneumostat™, with which she was later discharged. She had a normal echocardiography (ejection fraction [EF] 67%) at 5 weeks of gestation but developed pulmonary hypertension (EF 55%, pulmonary arterial pressure 40.7 mmHg) as the pregnancy progressed. She delivered a healthy baby at 35 weeks via elective lower section caesarean section with spinal anesthesia. We followed her up post-natally and noted the presence of left-sided pulmonary embolism, hypoplastic left lung, and left pulmonary artery. The management of this complex case involved a multidisciplinary effort between general medical, respiratory, obstetric, and cardiothoracic teams.

Introduction
We describe a case of a lady in early pregnancy who presented with acute dyspnea, with total collapse of left lung and concurrent right-sided pneumothorax. The case posed multiple clinical challenges and dilemmas during her pregnancy.

Case Report
A 34-year-old lady, 5 weeks pregnant (gravida 5, para 4) presented with acute dyspnea. Apart from well-controlled asthma, she also had pulmonary tuberculosis (pTB) at the age of 18 and was successfully treated with 9 months of anti-TB medication. She did not smoke, consume alcohol, or use recreational drugs. She was a housewife, but worked in a fabric factory during her earlier years. She had four children, aged 9, 7, 5, and 4, and had been independently well. She was of slim build, weighing only 30 kg. On examination, she was tachypneic at 34 breaths per minute; reduced breath sounds were noted on the left side of the chest with trachea deviation to the left. Her oxygen saturations (SpO₂) were 97% on room air with a normal arterial blood gas.

The admission chest X-ray showed a pneumothorax in the right lung with contralateral total lung collapse. Due to worsening dyspnea, a chest tube was inserted but remained bubbling 4 days later despite suction. The chest tube was subsequently changed to a Pneumostat™. It was in for 19 days before eventually being removed when the right lung had fully expanded (Fig. 1). During her stay, investigations for pTB by serial sputum AFB were negative.

A multidisciplinary meeting (MDT) was held to discuss further management. Based on the patient’s choice, normal echocardiogram findings (ejection fraction [EF] 67%), with no pulmonary hypertension [pH]), normal oxygenation, and a good pre-morbid history, the decision was made to proceed with the pregnancy. Furthermore, termination of pregnancy at that stage was deemed high risk in view of only a single functioning lung.

She was re-admitted at 31 weeks of gestation as her exercise capacity had deteriorated from a modified medical research council scale of 2 to 4. A repeat echocardiogram showed an ejection fraction of 55% and pulmonary arterial pressure of 40.7 mmHg. In view of the deterioration in symptoms and echocardiogram findings, another MDT was...
carried out to formulate a birth plan where input from both neonatologists and intensivists was sought. A decision was made for an elective LSCS.

She delivered a healthy baby girl weighing 2.24 kg at 35 weeks of gestation through spinal anesthesia. The baby’s Apgar score was recorded as 9 at 1 min and 10 at 5 min. Postnatally, spirometry showed a mixed obstructive and restrictive picture (FEV1/FVC = 79%, FEV1 = 47%, FVC = 59% predicted) and computer tomography pulmonary angiogram (CTPA) showed the presence of a left-sided filling defect, most likely representing a pulmonary embolus (PE) with complete left-sided lung collapse (Fig. 2). The scan also showed hypoplastic left lung and left pulmonary artery. Oral anti-coagulant was initiated for this lady and a referral to the transplant team was also made.

Discussion

The challenges in managing this case came from not just the hemodynamic changes of pregnancy but also due to complications of the pre-existing chronic lung disease. Our patient had underlying hypoplastic left lung made worse by previous pTB, although she had coped very well prior to this pregnancy and admission.

According to the British Thoracic Society (BTS) guidelines, management for secondary pneumothorax in pregnant patients and in patients who are unfit for surgery will involve medical pleurodesis, or alternatively, ambulatory management with a Heimlich valve, as in this case. The use of a Pneumostat would allow patients limited by a conventional chest tube to ambulate earlier. In a systematic review on ambulatory treatment for pneumothorax by Brims et al., the success rates related to the use of Heimlich valve were reported to be as high as 85%, and recurrence rates vary from 7 to 24% at follow-up [1]. Unfortunately, studies looking at the successful use of Heimlich valve for pneumothorax in pregnancy are sparse, most likely due to the rarity of the incidence. In fact, we found only one case in the literature of the use of a one-way valve for pneumothorax in pregnancy [2].

Ideally, our patient would have been a candidate for surgical pleurodesis with recurrence rates for open thoracostomy and pleurectomy described to be 1% or 5% for video-assisted thoracoscopic surgery (VATS) with pleurectomy [3]. Unfortunately, due to the severity of the existing pulmonary disease, one lung ventilation surgery employed during VATS procedures was contra-indicated in her case [4]. Therefore, medical pleurodesis with 10–20% recurrence rate instead should be explored.
The decision to proceed with the pregnancy was also appropriate as there were no contraindications to successful pregnancy to both mother and fetus. Nonetheless, at second admission PH had developed and it is likely that this was related to the PE. The mother was concerned of potential radiation to the fetus; hence, earlier CTPA was not performed. Had it been diagnosed earlier, she would have benefited from earlier anticoagulation. The decision for elective LSCS and bilateral tubal ligation was carried out following a second MDT meeting with input from the obstetricians and intensivists. Although vaginal delivery was not contraindicated, LSCS as the delivery option was chosen based on concerns about inability of the mother to increase cardiac output and manage hemodynamic changes during contractions and pushing [5]. Due to concerns related to “splinting of the diaphragm” from a growing fetus and the risk of going into spontaneous labor, delivery was planned at around 35 weeks of gestation. The BTS guidelines also advocate elective-assisted delivery with regional anesthesia near or at term in pregnancies in order to reduce the risk of recurrence [3].

The long-term management involves maximization of bronchodilation and perhaps lifelong anticoagulation in view of the elevated pulmonary pressure, along with assessment by the lung transplant team [6]. The main learning point from this case with multiple needs should center on the involvement of multidisciplinary approach. High-risk pregnancy needs input and care given by various disciplines to ensure the optimum management for both mother and baby. Cases like this require a management plan that is specifically tailored to the needs of both mother and fetus.

Disclosure Statements

No conflict of interest declared.

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

References