Case study

Pleural Fibroma Presenting as Spontaneous Pneumothorax in Late Pregnancy, an Exceedingly Rare Co-occurrence: A Case Report

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ABSTRACT

Introduction: Pleural fibroma or solitary fibrous tumor of the pleura (SFTP), a pleural tumor with malignancy risk presenting as pneumothorax is rare. In addition, spontaneous pneumothorax occurring in pregnancy is an uncommonly encountered scenario. We report a rare co-occurrence of a pleural fibroma causing spontaneous pneumothorax in a pregnant patient and review literature pertaining to these uncommon conditions.
Case Presentation: This is a 42-year-old lady in her third trimester with gestational diabetes mellitus (GDM) and hypertension who presents with sudden-onset dyspnea. Clinical examination was suggestive of a right-sided pneumothorax which was confirmed with a chest X-Ray. Chest X-Ray also revealed supradiaphragmatic right pleural base lesion which was well-circumscribed. A chest tube was inserted and continued to drain for another 11 days and was finally removed. A CT Thorax done due to persistent air leak after 3 days of chest drainage revealed a homogenously enhancing soft tissue density at the right pleural base which was suggestive of a pleural fibroma. This patient delivered a healthy term baby girl. She was offered right thoracoscopic surgery, but she declined. At one-year follow-up, she was keeping well.
Discussion & Conclusion: Due to the scarcity of such cases, high-level evidence is lacking. The authors review previously published literature and discuss the indications and timing of surgical intervention in both these conditions. We outline general principles of management for spontaneous pneumothorax in pregnancy and SFTP. We place emphasis that every case should be managed in a multidisciplinary, individualized fashion with decision-making shared by the well-informed patient.

Keywords: Spontaneous pneumothorax, Pregnancy, Pleural Fibroma, Chest Tube)

1. INTRODUCTION

Spontaneous pneumothorax is rare in pregnancy, although the actual occurrence may be underreported. The largest study to date on the subject is a review of 87 case reports [1]. Due to the scarcity of pneumothorax in pregnancy, no high-level evidence can be used as a guide to decide on the indications of definitive surgery for pneumothorax, the timing of surgery, the best mode of delivery and the safety of a prolonged chest drain should surgery be deferred until delivery. In addition, pleural fibroma, or solitary fibrous tumor of the pleura (SFTP), an uncommon tumor of the pleura presenting as pneumothorax is rare. The importance of the SFTP diagnosis lies in its malignant potential as well as its potential to recur post-resection. However, due to rarity, no large prospective study exists to support routine resection for all SFTP.

The authors discuss a lady in her third trimester who presented with a spontaneous right-sided pneumothorax from a right base of pleura SFTP. We reviewed literature on spontaneous pneumothorax in pregnancy and SFTP with the aim of establishing sound guides on the principles of managing these rare and complex cases.

2. PRESENTATION OF CASE

A 42-year-old lady who was Gravida 3, Para 1+1 at 33 weeks 6 days of gestation presented to our center with irregular uterine contractions. This pregnancy was complicated with gestational diabetes mellitus (GDM) and gestational hypertension which were well-controlled. She has a history of a complete miscarriage at 9 weeks gestation and one previous lower-segment Caesarean section performed in the emergency setting for fetal distress, and fortunately, this child has thrived well to date. Admission physical assessment of the patient revealed she was not in labor; however, she was kept for observation and was planned for discharge after 24 hours of observation. She did not require tocolysis as there were no uterine contractions observed in the ward. Admission antenatal fetal ultrasound scan was unremarkable.

Unfortunately, the patient developed sudden onset dyspnea on the morning of planned discharge. Examination revealed normal vital signs and pulse oximetry reading however there were reduced breath sounds and hyperresonance from percussion on the right chest. A chest X-ray (CXR) done with the use of an abdominal shield showed a large right-sided pneumothorax with a supradiaphragmatic right base of pleura well-defined lesion (Fig. 1). A chest drain was inserted and connected to a Pneumostat™ drain which has a one-way flutter valve also known as a Heimlich valve. We planned for a CT of the Thorax postpartum to confirm the nature of the right pleural lesion and as an adjunct to plan for surgery. However, due to persistent air leak seen on the Pneumostat™ drain, we proceeded with a contrast-enhanced CT (CECT) of the thorax on the third day post drainage in anticipation of possible early surgery for persistent pneumothorax.

The CECT Thorax demonstrated a well-circumscribed, homogenous, and mildly enhancing soft tissue density at the right pleural base measuring 2.4cm in widest dimension and it causes atelectasis of the adjacent segment (Fig. 2). We proceeded with a multidisciplinary team discussion between the obstetrician, respiratory physician, radiologist, and thoracic surgeon. Consensus made was that this right pleural lesion is likely benign and is most probably a pleural fibroma based on its CT features. Furthermore, there were no features to suggest bullae, blebs, bronchopulmonary fistula, or diaphragmatic hernia. Hence, we decided to manage this patient conservatively as close to term as possible given her comorbidities of GDM and gestational hypertension unless physiological disturbance to the patient or her pregnancy occurs despite the chest drain. We explained the risk of empyema from having a prolonged chest tube in place to the patient and we expectantly informed her the possibility of needing thoracic surgery during pregnancy should the pneumothorax become persistent, recurrent or cause adversities during labor.

We discharged the patient home with the chest drain and successfully removed it 11 days after insertion. She was readmitted electively at term for delivery, and it was decided that she was to be delivered via elective lower segment Caesarean section in view of her age with GDM and a previous scar. She underwent elective Caesarean section with bilateral tubal ligation under spinal anaesthesia at 38 weeks 2 days gestation and delivered a healthy baby girl of 2.87kg with Apgar score of 9/10. Postoperatively, she was well and did not require any oxygen therapy. She was discharged home on post Caesarean section Day 3 with her newborn. She was offered right thoracoscopic surgery for excision of the pleural fibroma however she declined surgery. She has been seen at one-year post index

pneumothorax and remains well. A chest X-ray done at this visit showed no interval change of the pleural lesion.

3. DISCUSSION

Spontaneous pneumothorax in pregnancy is rare, with less than 100 cases reported to date [1]. Most spontaneous pneumothorax in pregnancy are related to bullae and blebs and tend to occur during the last trimester or in labor [2,3]. Most patients were young and of low gravidity [3]. Physiological changes in pregnancy are thought to heighten the risk of rupture of blebs or bullae, i.e., increased oxygen demands with increased respiratory rates, and tidal volume increase [4]. Rarer causes of spontaneous pneumothorax in pregnancy are catamenial pneumothorax [5]. Fortunately, most of these pneumothoraces were not associated with severe physiological disturbances, although recurrence is a common feature estimated to occur at a rate of around 40% after the initial episode [4].

A few points of discussion on the management of spontaneous pneumothorax are mentioned below:

1. Are radiological investigations harmful to a pregnant patient or her fetus?
   Chest radiograph is safe in pregnancy when a shield is used [6]. In fact, fetal radiation dose associated with a CT of the chest ranges 0.01 to 0.66mGy which is a very low risk for teratogenesis, and the use of a pelvic shield may only mitigate that risk slightly [7,8].
2. Should pneumothorax be drained via aspiration or a tube thoracostomy/chest drain?
   Although small pneumothorax (<2cm or <20%) can be managed conservatively without the need of a drain or aspiration, the risk of expansion exists. Hence, pregnant patients with small spontaneous pneumothorax should be admitted for observation. Frequently enough, aspiration may fail to adequately drain the air, requiring a tube thoracostomy [9]. A more recent review reported that more than 60% of cases require a chest drain [1]. Whether this reflects the severity of initial clinical presentation of these cases or a progression from a small pneumothorax after conservative management or aspiration is unclear. Regardless, physiological compromise is an indication for a tube thoracostomy.
3. Is there a need for surgical intervention?
   Since most of these cases occurred because of a ruptured bulla, surgery is logically the best option to reduce recurrence risk. Although it is intuitive to presume the risk of recurrence is reduced with corrective surgery, a more recent review of literature looking into 87 cases reported that half of all patients had resolution without interventions be it thoracotomy, video-assisted thoracoscopic surgery (VATS) or pleurodesis [1]. Although dated literature of a series of recurrent pneumothorax treated surgically demonstrated that surgery can be safely performed during pregnancy, surgery nonetheless entails risks to both the patient and her fetus [10]. These risks include intraoperative bleeding, single-lung ventilation, placental hypoperfusion, premature labor and postoperative pain. Postoperative uterine contractions have been reported despite intraoperative prophylactic tocolysis and can be successfully mitigated [11,12]. Interestingly, overall fetal complications were not increased compared with the general population regardless of type of intervention [1]. In summary, surgery should be offered when patients have a persistent air leak or recurrence and this decision for surgery should be an individualized multidisciplinary shared decision [6,11]. For example, patients living in remote areas with difficult access to emergency care should be considered for surgery even after the first episode [13].

4. When is the best time for surgical intervention? Can intervention be deferred until postpartum?
   The optimal time for surgery is during the second trimester, which is after organogenesis and before pregnancy becomes advanced [12]. However, often, these cases occurred in the third trimester and labor on its own can cause a recurrent pneumothorax owed to the repeated physiological Valsalva maneuver [4]. Proceeding with surgery in the third trimester carries the risk of premature delivery of the fetus. Hence the question begs, in a patient with a persistent air leak, does a prolonged chest tube carry an increased morbidity for the patient and the fetus? Theoretically, a prolonged chest drain carries the risk of infection and empyema and that would lead to both maternal and fetal complications. However, some texts document a long term thoracostomy for up to 7 weeks for a persistent pneumothorax in a third trimester patient followed by definitive surgery after term delivery [2]. In fact, various case reports and series indicate to us that waiting for delivery of the baby for definitive imaging and surgery may not be inferior to emergent management in cases of a persistent air leak [1,3,4]. We recommend an individualized approach for each patient. If a patient has indications for surgery (recurrence or persistence), in the first or second trimester, surgery can be safely pursued as it outweighs the risk of an infected chest drain. However, if a patient is already in the third trimester, we recommend deferment of definitive surgery until term delivery with attentiveness to the care of the long-term drain.
5. What is the optimal mode of delivery in patients with spontaneous pneumothorax?
   Theoretical risk of recurrent pneumothorax potentiated by the labor process has sparked debate. Various authors recommend assisted delivery to reduce Valsalva maneuvers and hence reduce pneumothorax risk [4]. In such texts, assisted delivery with forceps is preferred over vacuum extraction which still requires maternal effort. Furthermore, Caesarean section is preferred above spontaneous vaginal delivery although one must bear in mind that Caesarean section is heralded by its own set of risks. Where Caesarean section becomes necessary, regional anaesthesia is preferred over general anaesthesia with positive pressure ventilation. Nitric oxide is considered a relative contraindication for these patients [11]. Newer summation of these cases shows that half of all these patients undergo spontaneous vaginal delivery safely [1]. Hence, decision on the optimal mode of delivery should primarily be driven by obstetrical indications rather than the theoretical risks of a recurrent pneumothorax as demonstrated in our patient.

One interesting aspect of the case we present is the absence of a bleb or bulla causing pneumothorax. Our radiographic imaging did not find features to suggest the pneumothorax is caused by a bleb or a bulla. We did not find any texts indicating pleural fibroma could present with pneumothorax. One other differential diagnosis we considered was a supradiaphragmatic accessory liver tissue as demonstrated in a case report, although these are extremely rare [14]. Pleural fibromas or solitary fibrous tumors of the pleura (SFTP) are rare tumors arising from the areolar tissue subjacent to the mesothelial lined pleura and account for around 5-10% of all primary pleural tumors [15,16,17]. Most are benign and asymptomatic although some may present with respiratory symptoms and paraneoplastic syndrome [13,16,17]. Some of these lesions present as large obstructing lesions and can cause mediastinal shift [18]. A retrospective review on 45 patients reported that majority of patients were females and were around the age of 60 years old with lesions under 10cm [19]. This series reported a malignancy rate of around 10% of all SFTP with good oncological outcomes if resection margins are clear. Most SFTPs occur at the lower third in the chest with a small percentage of these tumors (6%) presenting adjacent to the

hemidiaphragm [20]. Although these tumors run a benign course, recurrence and malignant transformation are known features, which is why most literatures cite the necessity of surgery and close follow up after resection. Needle biopsy and tumor spillage during surgery should be avoided for these reasons as well although needle biopsy was found to be safe in a small group of patients studied retrospectively [19]. Furthermore, confirming the diagnosis of SFTP is not easy before surgery [17]. CT scan generally shows an isolated mass with well-defined boundaries with mild to moderate enhancement. Although the “wait-and -see” is generally reserved for patients with poor performance status, no long-term prospective follow-up studies can support routine surgery for all benign SFTPs. Our patient declined surgical intervention but agreed to be followed up which is reasonable given the benign features on CT scan.

4. CONCLUSION

Due to the scarcity of spontaneous pneumothorax in pregnancy, no randomized trials can be conducted to draw high-level evidence to form guidelines. In addition, the scarcity of literature on SFTP make long-term decision making more challenging in this patient. Hence, our decision-making was an individualized multidisciplinary decision shared with a well-informed patient.

CONSENT

All authors declare that written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the informed consent is available and has been submitted together with this case report.

ETHICAL APPROVAL (WHERE EVER APPLICABLE)

Not applicable

REFERENCES

1. Agrafiotis AC, Assouad J, Lardinois I, Markou GA. Pneumothorax and pregnancy: a systematic review of the current literature and proposal of treatment recommendations. The Thoracic and Cardiovascular Surgeon. 2021 Jan;69(01):095-100.
2. Levine AJ, Collins FJ. Treatment of pneumothorax during pregnancy. Thorax. 1996 Mar;51(3):338.
3. Tanase Y, Yamada T, Kawaryu Y, Yoshida M, Kawai S. A case of spontaneous pneumothorax during pregnancy and review of the literature. Kobe J Med Sci. 2007 Jan 1;53(5):251-5.
4. Lal A, Anderson G, Cowen M, Lindow S, Arnold AG. Pneumothorax and pregnancy. Chest. 2007 Sep 1;132(3):1044-8.
5. Yoshioka H, Fukui T, Mori S, Usami N, Nagasaka T, Yokoi K. Catamenial pneumothorax in a pregnant patient. The Japanese Journal of Thoracic and Cardiovascular Surgery. 2005 May;53(5):280-2.
6. Garg R, Sanjay VD, Usman K, Rungta S, Prasad R. Spontaneous pneumothorax: An unusual complication of pregnancy-A case report and review of literature. Annals of Thoracic Medicine. 2008 Jul;3(3):104.

7. Mathur S, Pillenahalli Maheshwarappa R, Fouladirad S, Metwally O, Mukherjee P, Lin AW, Bharatha A, Nicolaou S, Ditkofsky NG. Emergency imaging in pregnancy and lactation. Canadian Association of Radiologists Journal. 2020 Aug;71(3):396-402.
8. Jain C. ACOG Committee Opinion No. 723: guidelines for diagnostic imaging during pregnancy and lactation. Obstetrics & Gynecology. 2019 Jan 1;133(1):186.
9. Annaiah TK, Reynolds SF. Spontaneous pneumothorax—a rare complication of pregnancy. Journal of Obstetrics and Gynaecology. 2011 Jan 1;31(1):80-2.
10. Dhalla SS, Teskey JM. Surgical management of recurrent spontaneous pneumothorax during pregnancy. Chest. 1985 Aug 1;88(2):301-2.
11. Reid CJ, Burgin GA. Video-assisted thoracoscopic surgical pleurodesis for persistent spontaneous pneumothorax in late pregnancy. Anaesthesia and intensive care. 2000 Apr;28(2):208-10.
12. Van Winter JT, Nichols III FC, Pairolero PC, Ney JA, Ogburn Jr PL. Management of spontaneous pneumothorax during pregnancy: case report and review of the literature. InMayo clinic proceedings 1996 Mar 1 (Vol. 71, No. 3, pp. 249-252). Elsevier.
13. Kocher GJ, Schmid RA. Pleural Pathologies and Malignant Effusion. InTips and Tricks in Thoracic Surgery 2018 (pp. 199-213). Springer, London.
14. Fang CC, Neerudu M, Hernandez LA. Supradiaphragmatic accessory liver tissue mimicking pleural tumour: excision by transdiaphragmatic uniportal video-assisted thoracoscopic surgery (U-VATS). Journal of Surgical Case Reports. 2021 Aug;2021(8):rjab338.
15. Jaramillo FA, Gutierrez F, Bhalla S. Pleural tumours and tumour-like lesions. Clinical radiology. 2018 Dec;73(12):1014-24.
16. Singh D. Imaging of chest wall and pleura. InThoracic Imaging 2019 (pp. 325-360). Springer, Singapore.
17. Sun N, Wang J, Cheng Z, Han W, Li G, Tong L. Solitary fibrous tumor of the pleura in a 22-year-old woman: a case report. Journal of International Medical Research. 2020 Sep;48(9):0300060520959495.
18. Fatimi SH, Inam H, Chagan FK, Choudry UK. Solitary fibrous pleural tumor. A rare and challenging case. International Journal of Surgery Case Reports. 2020 Jan 1;66:346-9.
19. Savu C, Melinte A, Posea R, Galie N, Balescu I, Diaconu C, Cretoiu D, Dima S, Filipescu A, Balalau C, Bacalbasa N. Pleural Solitary Fibrous Tumors—A Retrospective Study on 45 Patients. Medicina. 2020 Apr;56(4):185.
20. Verschakelen JA, Gleeson F, Tsakok M. The chest wall, pleura, diaphragm and intervention. Grainger & Allison's Diagnostic Radiology, 2 Volume Set E-Book. 2020 May 25:38.

Figures

Fig. 1. Chest X-Ray at presentation of dyspnea. There is large right pneumothorax with a small well-defined right pleural base lesion seen which is fully surrounded by air suggestive that this lesion is separate from the right hemidiaphragm.

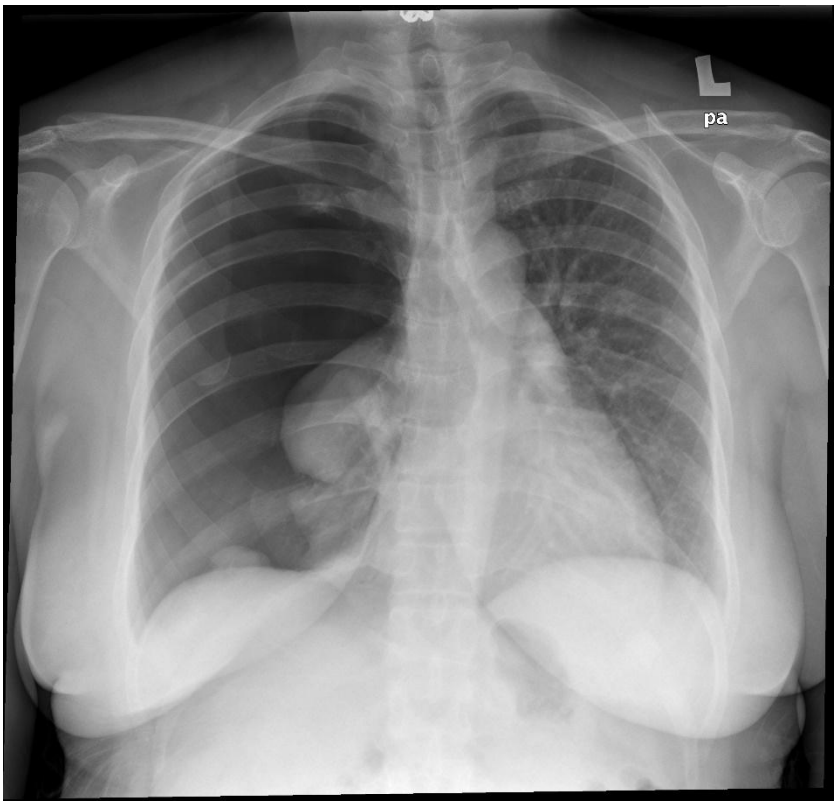

Fig. 2. Contrast-enhanced CT Thorax in coronal view (both lung and soft tissue windows) showing the well-circumscribed soft tissue density with mild enhancement at the right pleural base suggestive of a pleural fibroma. Right chest tube is seen lateral to this lesion.

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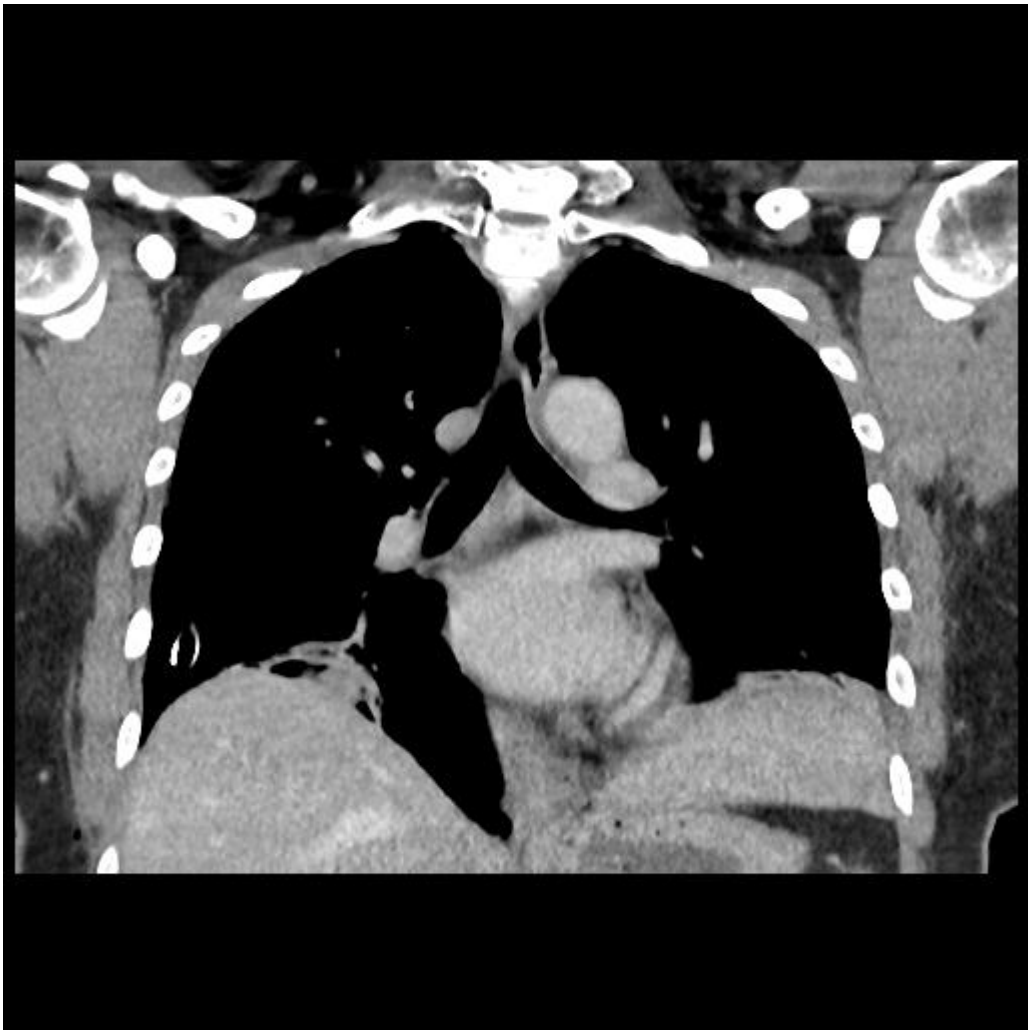

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