LARGE SPONTANEOUS RIGHT CATAMENIAL PNEUMOTHORAX WITH DIAPHRAGMATIC DEFECT AND LIVER HERNIATION

Venkata Kishore Mukku, M.D.,PGY-III, Emily Cassidy, M.D., Catalina Negulescu, M.D., Tonya Jagneaux, M.D., John Godke M.D.

Baton Rouge General Internal Medicine Residency Program, affiliate of Tulane University School of Medicine

Introduction

Catamenial pneumothorax (CP) is a rare cause of spontaneous pneumothorax that affects predominantly in women of child bearing age (1). In the literature, only a few cases of CP with diaphragmatic defect and liver herniation are reported (2). CP is most common in premenopausal women age 30–50 years with peak incidence between 30-35 years (3). CP typically occurs within 72 to 96 hours of onset of menstruation (4). CP is associated with pelvic endometriosis in 30-50% of cases, and almost always involving the right side of thorax (5,6). Etiology of CP is poorly understood but several proposed theories such as migration of air from peritoneal cavity into the pleural space through pre-existing diaphragmatic defects, retrograde implantation of endometrial tissue through diaphragmatic defects, lymphatic or hematogenous spread of endometrial tissue to visceral pleura and spontaneous rupture of blebs during hormonal changes (7).

Case Description

A 40 year old nulliparous woman with medical history significant only for endometriosis presented with severe chest tightness of 1 day duration. One day prior to presentation, she became symptomatic while cycling for 10 minutes. The following day, she developed a severe cough associated with recurrent chest tightness and disorientation. This prompted her to the emergency department. The patient is a pharmacist, nonsmoker, and denies illicit drug use or recent travel. She had endometrial laser ablation with myomectomy in 2006. Hormonal contraceptives have been used since 2004 but were stopped 3 months before with the hopes of becoming pregnant. Her last menstrual period was 4 days before the onset of symptoms. On physical examination, she had diminished breath sounds on the right. All the routine laboratory work was unremarkable. A Chest radiography (CXR) showed a large right spontaneous pneumothorax with what was thought to be, a 5.6 cm pleural mass at the right lung base (image 1). Following the pneumothorax diagnosis, the patient underwent emergent right thoracostomy with pigtail catheter placement. A repeat CXR revealed marked reexpansion of the lung but persistence of the right pleural mass. A follow up computed tomography (CT) scan of chest showed 33mm diaphragmatic defect with a 5.8 x 4.6 x 3.9 cm area of herniated liver corresponding to presumed pleural mass (image 2). Following complete thoracic imaging the patient underwent video-assisted thoracoscopic surgery (VATS), mechanical pleurodesis, and open repair of the right diaphragmatic defect (images 4, 5, 6). Intraoperatively, an endometrial implant was noted on chest wall (image 3). On post-operative day-3, she began her menstrual cycle and was evaluated by a gynecologist consultant who recommended hormonal therapy to reduce the risk of recurrent pneumothorax. Due to a persistent air leak, the chest tube was transitioned to a Heimlich valve to facilitate home discharge. The patient was discharged on postoperative day 8. She was subsequently seen by cardiovascular thoracic surgeon as an outpatient with resolution of air leak and removal of chest tube.

Images

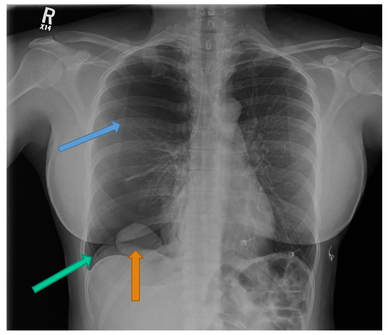

Image 1: CXR show pneumothorax, diaphragmatic air and large 56mm mass at base of lung confirmed as liver on CT scan

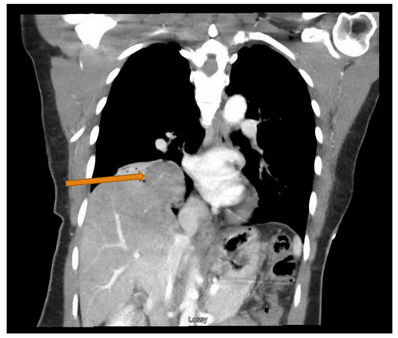

Image 2: Coronal view of CT show 33mm diaphragmatic defect with liver herniation, 5.8 x 4.6 x 3.9cm

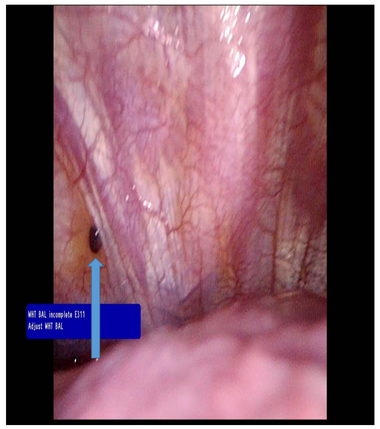

Image 3: Endometrial implant on chest wall

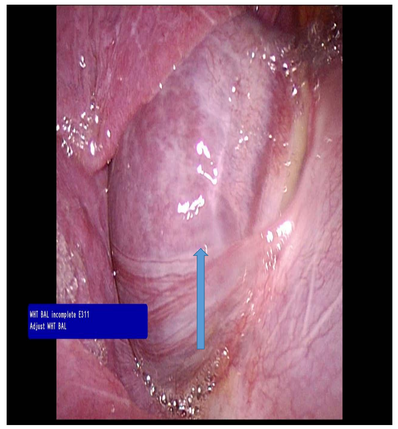

Image 4: Diaphragmatic defect with herniation of liver

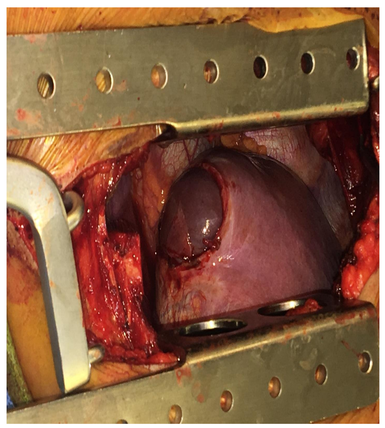

Image 5: Diaphragmatic defect with herniation of liver before repair

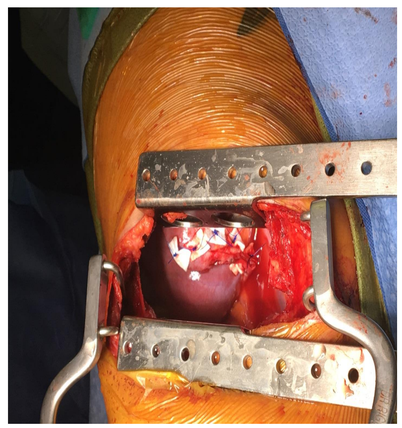

Image 6: Repair of diaphragmatic defect and liver herniation

Discussion

Though uncommon this case describes a woman with endometriosis who presented with a pneumothorax and a large diaphragmatic defect. Despite no histological diagnosis, her clinical course, intraoperative finding of an endometrial implant, diaphragmatic defect with liver herniation and diaphragmatic air are consistent with CP. She underwent VATS, mechanical pleurodesis with open diaphragmatic repair followed by hormonal therapy. VATS allows direct visualization of implants, nodules throughout thoracic cavity, ability to resect apical blebs, parenchymal, and diaphragmatic implants. VATS should be timed around beginning of menstrual flow to allow maximum visibility of endometriosis implants. Diaphragm, visceral and parietal pleura needs to be explored for endometriosis implants and all accessible lesions need to be excised (8). It's important to block the hormonal support from the ovary to the existing endometrial tissue, to prevent further seeding. Hormonal therapy such Gonadotropin-releasing hormone (GnRH) analogs are first line agents administered for a period of 6 to 12 months post-operatively can result in endometrial hypotrophy. Other alternatives are oral contraceptive pills, progestins, danazol or aromatase inhibitors. Combination of surgery and hormonal treatment reduces recurrence (1, 9 and 10).

Conclusion

Catamenial pneumothorax have high rate of recurrence nearly 30 to 40%. Due to high rate of recurrence, these patients require a more aggressive approach of pleurodesis and hormonal treatment postoperatively for a period of minimum 6 months. Early diagnosis of catamenial pneumothorax is imperative to minimize morbidity/mortality. Therefore, in any young female who presents with pneumothorax during menses, catamenial pneumothorax should be suspected.

References

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