

Catamenial Pneumothorax: A Rare Disease among Menstruating Women

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Abstract: *Catamenial Pneumothorax (CP) is an extremely rare, but very dangerous condition. While it only comprises 3% - 6% of all spontaneous pneumothorax cases among menstruating women, its unique presentation makes CP an important subject for research. The main criteria for the diagnosis of catamenial pneumothorax are known as the sine qua non criteria, which is spontaneous pneumothorax within 72 hours before or after the start of a menstrual period. In addition to this, CP often presents with concomitant endometriosis, right - sided location, and pleural lesions; however, these criteria do not have to be met for diagnosis. While the exact cause of CP has not been determined there are four main theories that explain its possible etiology.*

Keywords: hemopneumothorax, catamenial pneumothorax, spontaneous pneumothorax, spontaneous, menstrual, endometriosis, pulmonology, pneumothorax, catamenial

1. Introduction

The term catamenial is derived from Greek and means "monthly". The term catamenial pneumothorax was first used in the medical literature in 1972, although the disorder had been reported in the medical literature decades earlier.

History and etymology

It was initially described in 1958 by Maurer. The term 'catamenial' was named by Lillington in 1972. It is derived from the Greek words pertaining to and monthly.

Other Terms

Catamenial pneumothorax (CP) is the most common form of thoracic endometriosis syndrome, which also includes catamenial hemothorax, catamenial haemoptysis, and catamenial hemopneumothorax and endometriosis lung nodules.

Definition

Catamenial pneumothorax is an extremely rare disease that affects women. **Pneumothorax** is the medical term to collapsed lung, a condition in which air or gas is trapped in the space surrounding the lungs causing the lungs to collapse. **Women with catamenial pneumothorax** have recurrent episodes of pneumothorax that occur within 72 hours on and off to the start of menstruation.

Catamenial pneumothorax occurs in women of child - bearing age and, by occurs within 72 hours before or after the onset of menstruation. Pneumothorax may be recurrent and there may be a history of pelvic endometriosis.

Incidence

Catamenial pneumothorax affects women during their reproductive years, most often during their thirties or forties. The exact incidence of the disorder is unknown. Many researchers believe that catamenial pneumothorax often goes undiagnosed or misdiagnosed, making it difficult to determine its true frequency in the general population.

Causes

The exact cause of catamenial pneumothorax is unknown. Some cases are associated with the abnormal development of endometrial tissue outside of the uterus (endometriosis), although the exact nature of this relationship in these cases is unknown.

Several different theories have been proposed involving **metastatic, hormonal and anatomical abnormalities**. It is possible that catamenial pneumothorax may have different causes in different cases.

Metastatic Model

In the metastatic model, catamenial pneumothorax caused by the abnormal migration of endometrial tissue from the lining of the uterus (endometrium) to other areas of the body such as the diaphragm or the space in between the membranes lining the chest cavity wall and the lungs (pleural space). When endometrial tissue is found outside of the uterus, the term endometriosis is used. Most of the women with catamenial pneumothorax have endometriosis. Endometriosis can cause small holes or openings (fenestrations) in the diaphragm, which would allow air and fluid to pass through into the pleural space. Many women with catamenial pneumothorax have endometrial tissue in the lungs, a condition called thoracic endometriosis. However, some women with catamenial pneumothorax do not have diaphragmatic fenestrations or endometrial tissue in the lungs suggesting that other factors may play a role in the development of the disorder or that other causes of the disorder (apart from endometriosis) exist.

Hormonal Model

In the hormonal model, researchers believe that a hormone known as prostaglandin F₂, which is elevated during ovulation, causes narrowing (constriction) of the small tubes within the lungs (bronchioles). Bronchiolar narrowing may cause the small air sacs (alveoli) of the lungs to rupture, allowing air to become trapped in the pleural space.

Anatomic Model

In the anatomical model, researchers believe that the absence of the cervical mucous plug, a normal occurrence during the menstrual cycle, allows air to pass from the genital tract into the pleural space through small holes or openings (fenestrations) in the diaphragm.

Pathology

Other theories have been proposed to explain catamenial pneumothorax:

- passage of air from the uterus into the peritoneal cavity and through diaphragmatic fenestrations
- alveolar rupture precipitated by broncho constriction due to prostaglandin F2
- Vascular micro embolism of endometrial tissue.

Another Theory

Another theory that has been proposed as a cause of catamenial pneumothorax is the spontaneous ruptures of blebs. Blebs are small blisters or pustules that may be filled with fluid or air and can develop on the lungs. Some researchers speculate that hormonal changes during the menstrual cycle may cause blebs to rupture, which in turn can result in pneumothorax.

Signs & Symptoms

The symptoms and severity of catamenial pneumothorax can vary from one episode to another and from one person to another. In the majority of affected women, the right lung is affected.

Symptoms that may occur with catamenial pneumothorax include monthly episodes of chest pain that can radiate to the shoulder blades, shortness of breath or difficulty breathing (dyspnea), dizziness, fatigue, and a dry cough. Some women have reported a “crackling” sound upon inhaling during an

episode. Chest pain associated with a collapsed lung is often severe and the condition often requires prompt medical attention.

Diagnosis

The diagnosis can be made in women of reproductive age who present with pneumothorax within 72 hours of the onset of menstruation.

A diagnosis of catamenial pneumothorax is made based upon a detailed patient history, a thorough clinical evaluation and identification of characteristic symptoms (i. e., repeated episodes of pneumothorax in conjunction with the onset of menses).

A variety of tests may be required to rule out other conditions and to identify associated conditions such as thoracic or pelvic endometriosis or damage to the diaphragm.

A minimally invasive procedure known as video - assisted thoracoscopy (VATS) may be used as a diagnostic aid. During a VATS procedure, a 1 - cm rigid tube (thoracoscope) with a tiny, fiber - optic camera at the end is passed through a small incision in the chest. This allows physicians to examine the lungs, chest cavity and diaphragm.

CT is a more sensitive technique to demonstrate the above findings, including small nodules and defects. Occasionally part of the liver may herniate through a larger diaphragmatic defect (collar sign). MRI may suggest pleural - based masses attributable to endometrial implants.

Clinical picture

Pneumothorax typically presents with chest pain, sometimes with dyspnoea. Around 90% occur on the right.

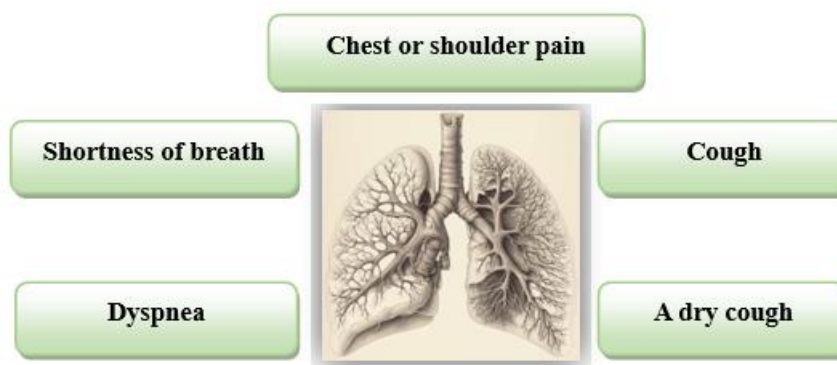


Figure 1: Clinical Manifestations of Catamenial Pneumothorax (CP)

Treatment Modalities

Most catamenial pneumothoraces are small and self - resolving.

Video - assisted thoracoscopic surgery (VATS)

Video - assisted thoracoscopic surgery, a minimally invasive surgical technique for diagnosing and treating chest conditions

VATS treatment of thoracic endometriosis includes partial diaphragmatic resection and repair, excision of all visceral pleural implants and talc pleurodesis. This is followed by hormone therapy to lessen the chance of recurrence which

can be as high as 40%. Continuous administration of the oral contraceptive pill causes atrophy of the endometrium and avoids cyclic proliferation and necrosis of endometrial deposits.

An episode of pneumothorax may be treated with oxygen followed by observation and rest if the collapse is small. Serious episodes of pneumothorax may require the insertion of a chest tube to release trapped air and/or blood, thereby allowing the lungs to re - expand. Both surgery and hormonal therapy, either separately or in combination, have been used to treat women with catamenial pneumothorax.

No specific guidelines exist for the optimal treatment of catamenial pneumothorax. Specific therapies may depend upon the exact cause of pneumothorax, an individual's age and general health, personal preference, and/or other factors.

Surgery may be performed to remove (excise) all suspected areas of endometrial tissue in the lungs and pleural space and to repair any damage or holes within the diaphragm. Surgery may also be used to remove small blisters located on the top of the lungs (apical blebs).

In addition, the artificial destruction of the pleural space (pleurodesis) may also be used to treat women with catamenial pneumothorax. Chemicals or drugs may be used to cause inflammation of the two layers of the pleura. This inflammation causes the pleurae to stick together (adhere) eliminating the pleural space. Another procedure, called pleural abrasion, can also be used to cause inflammation and adhesion of the pleurae. During pleural abrasion, the pleurae are inflamed through friction by wearing down or rubbing away (abrading) the pleurae.

Another surgical procedure that has been used to treat some affected women involves a mesh made from specialized material. During this procedure, a mesh is placed over the diaphragm in order to block any tiny holes that may have been missed during surgery. The mesh is absorbed over time and the resultant scar tissue eliminates any remaining holes in the diaphragm. This procedure is recommended even in women who have undergone pleurodesis.

Hormonal Therapy

Hormonal therapy may also be used to treat women with catamenial pneumothorax, usually as an adjunct to surgical therapy. Gonadotropin releasing hormone agonists are drugs that suppress ovulation and prevent the release of other hormones including estrogen or progesterone. These drugs are commonly used to treat women with endometriosis, but have been effective in some women with catamenial pneumothorax, even those who do not have signs of endometriosis.

2. Conclusions

Surgery is the treatment of choice of catamenial pneumothorax. It should aim to complete management of all lesions. The most common complication is recurrence. Early diagnosis and multidisciplinary treatment including hormonal therapy may be beneficial in high risk patients. Recurrent spontaneous pneumothorax and pleural effusions in young females due to endometriosis is extremely rare but should be suspected in absence of other secondary causes.

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