Catamenial pneumothorax: optimal hormonal and surgical management


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Abstract

Objective: To provide further information addressing the etiology, optimal hormonal management and surgical management in catamenial pneumothorax (CP).

Methods: We retrospectively analyzed records of all female patients operated on for spontaneous pneumothorax at a university hospital between January 1993 and March 2002.

Results: In eight of 24 patients, pneumothoraces were timed with menses. Seven patients were on hormonal medications pre-operatively and six post-operatively. All six patients taking estrogen/progesterone replacement had recurrences pre-operatively and two of three had recurrences post-operatively while on these medications. No patient suffered a pneumothorax either pre- or post-operatively while taking a gonadotropin releasing hormone agonist (two and three patients, respectively). Intraoperative findings included diaphragmatic implants [5] diaphragmatic fenestrations [4], apical blebs [2] and visceral pleural implants [2]. All pathology was specifically addressed at the time of surgery. Pleural space management included mechanical pleurodesis in seven and pleurectomy with talc insufflation in 1. Follow-up ranged from 27 to 63 months with a mean of 48 months. Three patients developed post-operative recurrences. One was managed without intervention and two required additional procedures.

Conclusion: Catamenial pneumothorax is under appreciated, representing up to one-third of women with spontaneous pneumothorax. Hormonal agents that allow for menses are ineffective. Gonadotropin releasing hormone agonists should be considered as part of the pre-operative or post-operative management in high risk patients. Our findings suggest that an additional intervention to augment pleural symphysis at the level of the diaphragm should be performed.

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1. Introduction

Catamenial pneumothorax (CP) is a rare condition which exclusively affects women during their reproductive years [1]. The term catamenial is derived from the Greek word meaning monthly. This entity, first reported in the 1950s, exhibits a temporal relationship between the onset of pneumothorax symptoms and menses [2]. Endometriosis has been a frequently implicated factor, although it is not universally documented. The currently held hypotheses that explain endometriosis may also be applied to CP; however, no single hypothesis explains all of the findings in CP.

The treatment for CP is directed towards the management of the pneumothorax and the prevention of its recurrence. As many of the patients have a diagnosis of pelvic endometriosis, hormonal therapy may also have a role in the management of CP as well. The impact of various hormonal therapies on CP is, however, unknown.

In order to evaluate the prevalence, presentation, effectiveness of hormonal therapy and results of surgical therapy, we performed a retrospective review of female patients presenting with recurrent pneumothorax at a single institution.

2. Materials and methods

We performed a retrospective review from January 1993 to March 2002 of all female patients undergoing surgery for spontaneous pneumothorax. Twenty-four women with recurrent spontaneous pneumothorax were selected for surgical management. Of these, eight were classified as having catamenial pneumothorax. The diagnosis was made pre-operatively in five patients who reported a history of symptoms timed with their menses. The remainder had classic pathologic findings discovered intraoperatively and subsequently had a confirmatory clinical history on additional questioning. Operative procedures were performed by three different thoracic surgeons. Operative approach was thorascopic with a mechanical pleurodesis representing the standard approach to recurrent pneumothorax at our institution. Follow-up data was obtained from...
3. Results

All eight patients were pre-menopausal, and their ages ranged from 23 to 41 with a mean of 35 years. Pneumothoraces occurred within 24 h of the onset of menses in two patients, within 48 h in 3, within 72 h in two and 96 h in 1. Presenting symptoms included chest pain in all eight patients, cough in three and shortness of breath in 6. The number of pre-operative episodes varied between two and almost monthly, depending on when the patients were referred for intervention. The right pleural space was involved in all eight patients. One patient had a history of left recurrent pneumothoraces previously managed by surgical intervention at another institution. Three patients underwent tube thoracostomy initially and five were managed conservatively. Associated co-morbid conditions included Ehlers Danlos syndrome Type III in one, asthma in three, and Hepatitis C in one patient. Four patients had previously documented pelvic endometriosis and one had a previous negative laparoscopy. Unless the patients had symptoms of pelvic endometriosis, no additional diagnostic studies were performed looking for asymptomatic disease.

3.1. Hormonal manipulation

All eight patients received some form of hormonal treatment at some time. Patients were placed on hormonal therapy by their gynecologist, prior to being referred to a thoracic surgeon. All six patients taking hormonal replacement pre-operatively (five oral and one transdermal), had recurrent pneumothoraces while on this therapy. Patients were on this therapy for the management of associated symptoms of endometriosis (1), for contraception (2) or for the prevention of catamenial pneumothorax (3). Two patients, one with almost monthly episodes of CP, were placed on a gonadotropin-releasing hormone (GnRH) agonist pre-operatively specifically to manage their CP. They did not have a recurrence while on this drug. Three patients were on hormonal replacement post-operatively and two of these had recurrences while taking these medications. In one patient, this was after a total abdominal hysterectomy and bilateral salpingooophorectomy. This patient was on hormonal replacement for the management of hormonal withdrawal symptoms. Three patients received a GnRH agonist post-operatively. One patient stopped this therapy after 1 month due to side effects and subsequently developed recurrent CP. No patients had a recurrence while taking a GnRH agonist either pre-operatively or post-operatively.

3.2. Intra-operative findings/surgical management

Of eight patients treated for catamenial pneumothorax, thoracoscopic evaluation confirmed diaphragmatic endometrial implants in five (Fig. 1). One patient had an endometrial implant visible on their visceral pleural surface (Fig. 2). Diaphragmatic fenestrations were observed in four of the five patients with diaphragmatic endometrial implants. Two patients had apical blebs in addition to one or more of the previously described findings. Two patients did not have any identifiable intra-thoracic pathology at the time of initial thoracoscopy. One of these had parenchymal implants observed at a repeat procedure for recurrent CP and failed pleurodesis (Table 1).

Four patients with fenestrations had them closed by excision with mechanical staplers. One patient had an endometrial diaphragmatic implant excised. Pleurodesis was initially accomplished by mechanical abrasion in seven and parietal pleurectomy in one. The latter patient, who was later in the series, also had talc insufflation directed at the diaphragm at the time of parietal pleurectomy. This procedure was chosen as personal preference of the surgeon to prevent a diaphragmatic recurrence.

Three patients recurred post-operatively. One had a loculated basilar pneumothorax and was successfully managed with a catheter and doxycycline pleurodesis. A GnRH agonist was recommended to the second patient, but she chose a hysterectomy with bilateral salpingooophorectomy.
She was placed on hormonal therapy to control hot flashes and had recurrent CP with this. She subsequently underwent repeat thoracoscopy with pleurectomy. At the repeat procedure, no adhesions were observed. Visceral parenchymal endometrial implants were identified and resected. This patient subsequently recurred again with a loculated pneumothorax along the diaphragm. A small catheter was placed and the basilar space was pleurodesed with doxycycline. The third patient had a small pneumothorax and was managed conservatively. Other than recurrence, there were no post-operative complications. Three of eight patients currently have chronic catamenial chest pain without radiographic abnormalities.

Follow-up ranged from 27 to 63 months with a mean of 48 months (Table 1). Asymptomatic patients were followed yearly and patients with a recurrence were evaluated at the onset of symptoms. In addition, a telephone interview was obtained from all patients at the time of data collection.

4. Discussion

Catamenial pneumothorax is a subclass of secondary spontaneous pneumothoraces occurring in women usually in their third or fourth decade of life. Concurrence of pneumothorax with initiation of menstruation is classic and a previous diagnosis of endometriosis is frequent. Although the majority of patients develop symptoms within 72 h, occasionally it can be seen within 96 h of the onset of menses.

Historically, CP has been considered a rare cause of spontaneous pneumothorax. The reported incidence of catamenial pneumothorax is only 2.8–5.6% in women suffering from spontaneous pneumothoraces [3]. Our series and a recent prospective study of catamenial pneumothorax would suggest that the incidence is actually much higher [4]. We found CP to be the cause of pneumothorax in eight of 24 (33%). This may reflect increased awareness of this pathology with more directed history taking and careful visual inspection at the time of surgery.

Despite the fact that CP has been known to exist for almost half a century, the etiology has not been clearly defined. Endometriosis has often been blamed for the initiation of the sequence of events in this condition, though the existence of associated pelvic endometriosis has been accounted for in only 20–70% of patients with catamenial pneumothorax [5,6]. A pre-operative history of pelvic endometriosis was identified in 50% of our patients with CP. As of yet, a history of pelvic endometriosis has not been shown to play a specific role in the development or prevention of CP, so we do not recommend additional interventions looking for this pathology in asymptomatic patients with the diagnosis of CP.

Although no single theory can explain all of the findings in either endometriosis or CP, there are three extant theories of the pathogenesis of CP that may be relevant to CP:

(1) Sampson’s theory of retrograde menstruation and implantation [7,8].
(2) Entry of endometrial cells into the venous system and ‘metastatic’ spread of endometrial tissue.
(3) Coelomic metaplasia [9–11].
In our series, diaphragmatic implants were observed in five of eight patients. Frank diaphragmatic fenestrations were observed in 50% of the patients in our series; all patients with fenestrations also had diaphragmatic endometrial implants. This, and the fact that endometrial tissue has been observed at the borders of the defects [12], would suggest that endometrial implants are the cause for the fenestrations.

Catamenial pneumothorax has a predominant right sided occurrence (100% in our series in the vicinity of 90% in others) [1,4,6]. The preference for the right hemi-diaphragm is attributed by some to the well-described ‘piston effect’ exerted by the liver [13]. Visceral pleural implants were observed in 1/8 of our patients initially, and in a second patient at a repeat procedure. The presence of intra-parenchymal endometrial implants observed by others in CP, as well as catamenial hemoptysis, is best supported by the ‘metastatic’ hypothesis. This also explains endometrial tissue reported in distant regions such as the eye, knee, and brain [5,9].

There is a wide variation in the prevalence of diaphragmatic fenestrations with endometrial implants in patients with CP, 23-88% [1,4,15]. As previously mentioned, in our series, every patient with diaphragmatic fenestrations also had diaphragmatic endometrial implants, strongly suggesting that the latter causes the former.

It has been hypothesized that elevated levels of prosta- glandins in the peri-menstrual period might sensitize pulmonary blebs and make them more prone to rupture [16]. In our study, apical blebs were identified and resected in two patients (25%). However, at the time of surgery, none of the blebs appeared to have ruptured, and both of these patients also had parenchymal or diaphragmatic implants (Table 1). Further, in spite of undergoing complete excision of their bullous disease, one patient had recurrent CP following surgical treatment.

Endometrial parenchymal and/or diaphragmatic implants were observed in 6/8 patients at the initial operation and one additional patient with parenchymal implants identified at a second operation (Table 1). Also, 6/8 patients had multiple sites of pathology identified. The proportion of findings do not appear to be related to the severity of disease or risk for recurrence as two of the three patients who recurred had no pathology identified at the time of initial thoracoscopic. As 7/8 patients (87%) had either diaphragmatic or visceral pleural implants identified, our data strongly supports Sampson’s theory as the predominant etiology of CP.

Our data further demonstrate that hormonal therapies that allow for menses do not prevent CP, even in patients who have had a hysterectomy with bilateral salpingoophorectomy. There were no CP observed in patients receiving the GnRH agonist, Lupron. This drug appears to effectively suppress CP and may be of use in the peri-operative management of these patients. Pre-operative patients, who are not going to surgery with-in their current cycle, or post-operative patients, to allow for maturation of adhesions before challenging the pleurodesis, may benefit from this therapy. We do not recommend long term management of CP with this drug because it is poorly tolerated and the long term effects of chronic hormonal ablation in pre-menopausal women is unknown.

Finally, regarding surgical technique, some have reported that pleural abrasion alone is necessary for the management of CP. We would argue on the contrary, that as much of the pathology be addressed as possible through a minimally invasive approach. Apical blebs and parenchymal implants should be resected. Diaphragmatic fenestrations should be excised or closed. Additionally, because of the high recurrence rate observed, we would agree with others that an additional procedure to specifically address the risk of basilar recurrence should be performed [10,17]. Polygalactin mesh placed on the diaphragm appears to augment pleurodesis in this difficult area [10]. Another option is the selective use of talc along the diaphragmatic surface, as one surgeon used in this series. However, it is unclear if the potential risks of intrapleural talc outweigh the benefits in this young population. Certainly, the standard mechanical pleurodesis that most surgeons perform does not sufficiently address the diaphragmatic surface, which we and others find to be the location of the majority of the pathology in CP and the site of many recurrences.

5. Conclusion

In summary, the etiology of catamenial pneumothorax appears by our data to be most often due to retrograde menstruation with seeding of the diaphragm and/or visceral pleural surfaces. These endometrial implants subsequently necrose in response to the normal hormonal cycle and allow the passage of air. The incidence of CP appears to be far greater than historically thought so we must be vigilant in collecting the data through directed questioning and careful thorascopic inspection. The operative management for women with recurrent spontaneous pneumothorax should include careful inspection of the diaphragmatic surface. For those with CP, we believe that all intrathoracic pathology be addressed and an additional procedure to augment diaphragmatic pleurodesis should be performed. Hormonal therapy that allows for menses is ineffective in suppressing CP. One should consider a short course (<6 months) of a GnRH agonist in pre-operative patients not having surgery within their cycle and early post-operative patients to allow for a maturation of the pleurodesis.

References


