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Current Therapy of Catamenial Pneumothorax

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ABSTRACT

Catamenial pneumothorax, or monthly recurring pneumothorax associated with menstruation, has been reported with increasing frequency in recent years. A representative case illustrates the clinical syndrome, particularly the intraoperative findings. Characteristic of this disorder are a peak incidence in the late twenties or early thirties, recurrent right-sided pneumothoraces occurring at the onset of menstruation, and an association with pelvic endometriosis. Pathologically, there is a consistent pattern of intrathoracic, especially diaphragmatic, foci of ectopic endometrial tissue. There is also a strong association with diaphragmatic fenestrations, though their significance is controversial. Traditional therapy has involved treatment with estrogens, danazol, or thoracotomy with mechanical pleurodesis. These methods have proven, through a large meta-analysis, to be associated with a relatively high rate of recurrence. Subsequent advances in hormonal therapy, along with the development of minimal access surgery, have led to an evolution in management. Despite uncertainty as to the etiology of catamenial pneumothorax, diagnosis of the condition is straightforward and modern treatment is successful in preventing recurrence.

INTRODUCTION

First described by Maurer et al. in 1958, catamenial pneumothorax, once considered rare, has been reported with increasing frequency in recent years [Blanco 1998, Joseph 1996, Van Schil 1996]. It characteristically involves females in their third and fourth decades of life, and the patient frequently has a history of pelvic endometriosis. These women experience recurring chest pain and pneumothorax coinciding with the onset of menstruation. Interestingly ninety percent of these pneumothoraces occur on the right side [Joseph 1996, Slasky 1982]. Various theories have been put forward to explain the pathophysiology of recurrent pneumothoraces in these patients, yet to date none has proven conclusive. However, there does seem to be a correlation between the recurring symptoms and the presence of functional endometrial tissue in the thorax [Joseph 1996, Yamazaki 1980]. The traditional methods of treatment have involved hormonal manipulation with exogenous estrogens or danazol, or thoracotomy with a variety of methods to attempt pleural symphysis [Dotson 1993, Slasky 1982, Yamazaki 1980]. Most reports of catamenial pneumothorax are isolated, making it difficult to assess the efficacy of treatment. Meta-analysis of cases reported prior to 1996 revealed an unexpectedly high incidence of recurrence with these traditional therapies [Joseph 1996]. This information, along with technological improvements in hormonal therapy and the development of minimal access surgery, have led to an evolution of treatment strategies for managing catamenial pneumothorax [Blanco 1998, Dotson 1993, Mack 1992, Slabbnyck 1991, Van Schil 1996]. The following case highlights the salient features of this unusual malady, demonstrates dramatically the surgical findings, and illustrates some of the therapeutic advances in the treatment of catamenial pneumothorax. The current therapy of catamenial pneumothorax is reviewed, with recommended treatment algorithms based on the published data.

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CASE REPORT

A thirty-three year old female was referred with a one-year history of recurrent right-sided chest pain. Her pain always occurred on the day of onset of menstruation. Chest radiographs twice documented pneumothorax. Her past medical history was significant for endometriosis, diagnosed at laparoscopy, and recurrent urinary tract infections. In addition, along with her husband, she was participating in an infertility program in an attempt at pregnancy. She was on no medications. Physical examina-

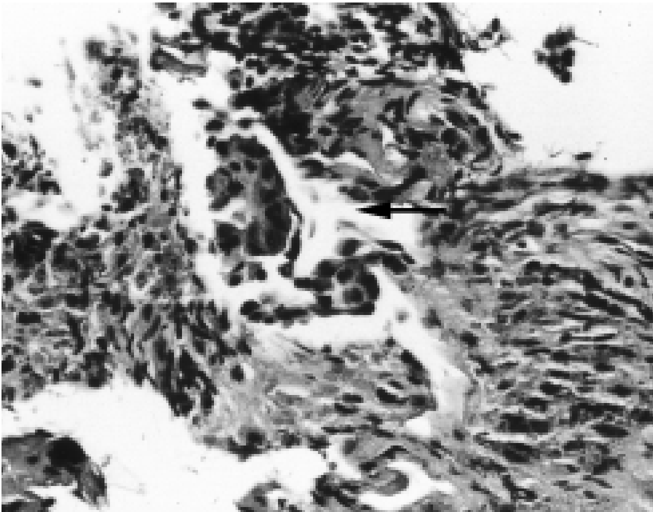

Figure 1. Photomicrograph of biopsy specimen from the diaphragmatic implants. Hemosiderin pigment is present as well as endometrial stroma forming rosettes of a glandular nature (→).

tion was unremarkable. A CT scan of the chest performed at the referring facility revealed a 3-mm contrast-enhancing lesion in the uppermost aspect of the right lobe. The CT scan was otherwise clear. Video-assisted thoracoscopic surgery (VATS) was next undertaken (see Movie ◎). At the apex of the right lung the lesion seen on CT scan was resected after the surgeons mobilized some localized adhesions and performed a limited pleurectomy. At histology this area contained only localized emphysematous changes and venous dilatation. Upon the surface of the diaphragm multiple small, discrete brown nodules were found. These were biopsied and fulgurated. Small diaphragmatic fenestrations were also noted. When examined at pathology these lesions subsequently proved to be foci of endometrial stroma along with hemosiderin deposits (see Figure 1, ◎). Mechanical pleurodesis with closed catheter drainage was then completed. After removal of the chest tube on postoperative day 2 the patient received an intramuscular injection of 3.75 mg leuprolide, a gonadotropin-releasing hormone (Gn-RH) analogue. This was to be repeated one month later for a total of two menstrual cycles. However, two weeks after her discharge from the hospital she experienced her next menstruation, most likely because the initial dose of leuprolide was given too late in her cycle. With this she experienced recurrent chest pain but no pneumothorax could be demonstrated on chest X-ray. She then received monthly injections of leuprolide for four months to maintain amenorrhea. Subsequently, she has resumed normal menstruation as well as her infertility evaluation with no further pain or pneumothoraces.

DISCUSSION

Catamenial pneumothorax has been estimated recently to account for 2.8% to 5.6% of cases of pneumothorax occurring in women [Blanco 1998]. Despite forty years of reports on this entity, controversy still exists as to the etiology of catamenial pneumothorax. Four theories have been proposed: thoracic endometriosis with pleural implants; the presence of pulmonary blebs or bullae; increased intrapulmonary concentration of prostaglandin PGF2, a potent bronchoconstrictor associated with menstruation; and the presence of diaphragmatic fenestrations allowing the passage of free intraperitoneal air, presumably released with expression of the cervical mucus plug during menstruation, into the thorax [Dotson 1993, Slasky 1982, Van Schil 1996, Yamazaki 1980]. The presence of intrathoracic, particularly diaphragmatic, endometrial implants is the most consistent finding in these women on the basis of intraoperative observations [Blanco 1998, Joseph 1996, Slasky 1982, Van Schil 1996, Yamazaki 1980]. In one large meta-analysis, of 48 cases of catamenial pneumothorax that required a surgical procedure, 42 patients had evidence of diaphragmatic fenestrations, endometrial implants, or both [Joseph 1996]. There is uncertainty even as to the origin of these intrathoracic foci of endometrial tissue and the diaphragmatic fenestrations. For example, it is unclear if the presence of functional endometrial implants on the diaphragm leads to the development of fenestrations during successive menstrual cycles, or if these communications are congenital and allow the passage of microfoci of endometrial tissue from the pelvis into the thorax [Joseph 1996, Slasky 1982]. In addition, the association of pelvic endometriosis with catamenial pneumothorax has led to theories of hematogenous spread of endometrial foci or the occurrence of coelomic metaplasia as the origin of ectopic endometrial tissue in these patients [Van Schil 1996].

Despite the uncertainties surrounding the pathophysiology of catamenial pneumothorax, the diagnosis is straightforward based on the temporal association of chest pain with the onset of menses [Blanco 1998, Slasky 1982]. The initial treatment for the primary episode follows the same guidelines as for other causes of spontaneous pneumothorax [Getz 1983, Massard 1998]. Recurrent symptoms prompt intervention based on the frequency of pneumothorax and individual patient factors. Traditional methods of therapy involved hormonal suppression of ovulation with oral contraceptives, estrogen blockade with danazol, and surgical procedures involving thoracotomy with possible bleb resection, apical resection, apical pleurectomy, and/or some form of pleurodesis [Dotson 1993, Massard 1998, Slasky 1982, Yamazaki 1980]. In the meta-analysis performed by Joseph and Sahn in 1996, follow-up data were available in 21 of 28 patients treated medically and in 20 of 42 patients treated surgically with these traditional methods [Joseph 1996]. In this series, the largest reported to date, patients who received hormonal treatment experienced significantly higher recurrence of pneumothorax from six months onwards. At six months, only 50% of medically treated patients were free of pneumothorax compared with greater than 95% of surgically treated patients ($p < 0.002$) [Joseph 1996]. At one year, less than 40% of medically treated patients were without recurrence, com-

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pared with 75% of surgical patients ($p < 0.03$). Although surgery proved superior, the rate of recurrence was significantly higher than results reported for recurrent spontaneous pneumothorax, which historically is less than 1% [Getz 1983, Massard 1998].

Since this data, however, experience with Gn-RH analogues has accumulated [Blanco 1998, Dotson 1993, Slabbnyck 1991, Van Schil 1996]. These agents have been very successful in preventing recurrent catamenial pneumothorax but require prolonged amenorrhea, up to one year, to be effective [Dotson 1993]. This may prove troublesome in some patients, as in the case presented here in which the patient desired pregnancy. The combination of a Gn-RH analogue and surgical pleurodesis, either with thoracotomy or increasingly with video-assisted thoracoscopic surgery (VATS), may shorten the duration of hormonal suppression necessary to achieve cure. This approach was used successfully in the case presented here as well as in other reports in the literature [Blanco 1998, Van Schil 1996]. To be most effective, hormonal suppression with these agents should begin at least one cycle prior to surgical intervention. At surgery any blebs or bullae should be resected and an apical resection considered. Apical pleurectomy may enhance the effectiveness of mechanical pleurodesis to achieve pleural symphysis [Getz 1983, Massard 1998].

Patients who experience recurrence after treatment with the above management plan are candidates for either a prolonged trial of menstrual suppression with Gn-RH analogues or another attempt at a combined trial of medical and surgical therapy. Thoracotomy should be strongly considered for patients requiring reoperation. If possible, a transaxillary or muscle sparing approach should be performed. The report of Slasky et al. describes resection of all or part of the central tendinous portion of the diaphragm containing the area of fenestrations, at the time of pleurodesis, in three patients with catamenial pneumothorax. Success was achieved in all three women with cure of their recurring pneumothoraces associated with menstruation [Slasky 1982]. If adopted this technique probably should be reserved for patients undergoing open thoracotomy due to the potential for hepatic injury and the need for reconstruction of the diaphragm.

Catamenial pneumothorax is therefore an increasingly recognized concern in young women. It may be diagnosed with a history of recurring pneumothoraces, notably right-sided, associated with the monthly onset of menses. Associated risk factors include a history of pelvic endometriosis. Tube thoracostomy is used to treat the initial episode if respiration is compromised or if a large pneumothorax is present. Specific treatment with Gn-RH analogues is indicated for recurrence. Video-assisted thoracoscopic exploration or mini-thoracotomy may shorten the duration of treatment needed to achieve cure. Mechanical pleurodesis, with resection of any blebs or bullae, and possibly apical pleurectomy and/or resection, should be performed at the time of the surgical procedure. If further surgery is planned for recurrence after this initial therapy, an open approach through a transaxillary or muscle sparing incision should be performed. One may consider resection of the central tendon of the diaphragm if defects in this region are noted.

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REVIEW AND COMMENTARY

1. Editorial Board member L023 writes:

The paper does not mention or discuss the possibility of spontaneous hemothorax that can also be associated with this condition.

Authors' Response by Henry F. Tripp, MD:

In this report, I attempted to limit the discussion to catamenial pneumothorax, but it is true that there are a spectrum of disorders that can be collected under the term "thoracic endometriosis" and are felt to result from the same basic pathophysiology. These include catamenial hemothorax and hemoptysis. The reference by Joseph and Sahn does a nice job of categorizing these syndromes.

Editorial Board member NL11 writes:

I would ask the authors for more details of their technique. A few more sentences will add more value to the explanation. The discussion is fine in my opinion.

Authors' Response by Henry F. Tripp, MD:

The patient was intubated with a double-lumen endotracheal tube and positioned in the left lateral decubitus position, with the table flexed in its mid-portion and placed in reverse Trendelenberg. We used a single trocar, inserted into the seventh interspace, in the mid-axillary line, to introduce a 10-mm straight videocamera. This was exchanged for a 30° angled camera to complete the exploration. Two additional incisions were made in the fourth interspace beyond the anterior and posterior to the respective axillary lines. The apical adhesions were mobilized using endoscopic scissors. These were also used, with an electrocautery attachment, for performing the apical pleurectomy. The biopsies were taken using a mediastinoscopy ball-type biotome, and the fenestrations were demonstrated with a Maryland dissector. A 35-mm endoscopic stapling device, with vascular staples, was used to perform the apical resection. The mechanical pleurodesis was accomplished with a bovie scratch pad, using specially angled thoracoscopic ringed-forceps. A single 28 Fr. Chest tube was inserted through the initial incision and positioned under video guidance and connected to underwater drainage. The patient was extubated in the operating room.

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