Catamenial pneumothorax: lessons learned and literature review

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Kardiochirurgia i Torakochirurgia Polska 2009; 6 (1): 73–76

Abstract

Catamenial pneumothorax is a rare clinical entity. The aetiology of this syndrome is unknown and lack of consistent findings have led to different aetiological theories. We present a 32-year old woman with catamenial pneumothorax and its recurrence after surgical management. A detailed review of the literature since the first description is also presented.
Key words: catamenial pneumothorax, spontaneous pneumothorax, endometriosis.

Introduction

Recurrent pneumothorax in women during menstruation was first described by Maurer et al. in 1958 [1] and Lillington et al. coined the term catamenial pneumothorax [2]. Catamenial pneumothorax (CP) is defined as recurrent spontaneous pneumothorax occurring within 72 hours before or after the onset of menstruation [3]. This clinical entity is considered to be rare but the true incidence could be higher than suggested. More cases have been reported in the last decade (102) than in the previous decade (42). Although over 247 such cases have been described in the English literature the precise pathophysiology is still uncertain. The hypotheses available could not explain all the findings described in the literature. We describe a young lady with catamenial pneumothorax, lessons learned and a literature review.

Report

A married woman 32 years old presented with recurrent right-sided pleuritic chest pain, right shoulder pain, dyspnoea and dry cough. The episode started two days after the onset of menstruation. Right pneumothorax was detected and she was treated with intercostal chest drainage. Pneumothorax recurred in two months time during the week of menstruation. Her past medical history includes pelvic endometriosis for which she required bowel resection 8 years ago. She was treated with danazol for 6 months and discontinued to have children. After completing her family she had an etonogestrel contraceptive implant. This progesterone implant was removed 5 months before the first episode of pneumothorax.

Following her second attack she was referred for surgical management. Clinical examination was unremarkable except for right-sided pneumothorax. At the time of admission chest X-ray showed 10% pneumothorax on the right side. CT scan of chest did not reveal any additional abnormality.

Right video-assisted thoracoscopy was performed which showed adhesion of the apex of the right upper lobe to the chest wall. The entire lung surface was normal and there were no bullae or nodules. The central portion of the right dome of the diaphragm appeared to have multiple fenestrations; the largest one was 1 cm in diameter (Fig. 1). The right pleural cavity was freely communicating with the peritoneal cavity and the right lobe of the liver was visible through these holes. The surface of the diaphragm in other areas appeared very thin and overlying vessels looked numerous and dilated.

Muscle sparing right thoracotomy was performed and the chest was entered through the 8th intercostal space. The membranous part of the right diaphragm was excised and replaced with bovine pericardium using 3/0 continuous prolene suture. No pleurodesis was performed.

Microscopic examination of the resected specimen showed focal active chronic inflammation, mesothelial reaction and some aggregates of macrophages containing haemosiderin (Fig. 2). There was no evidence of endometrial stroma or glands. The patient recovered uneventfully and continued to do well until 2 months later, when she had recurrence of a small basal pneumothorax. The pneumothorax resolved spontaneously. She was put on gonado-

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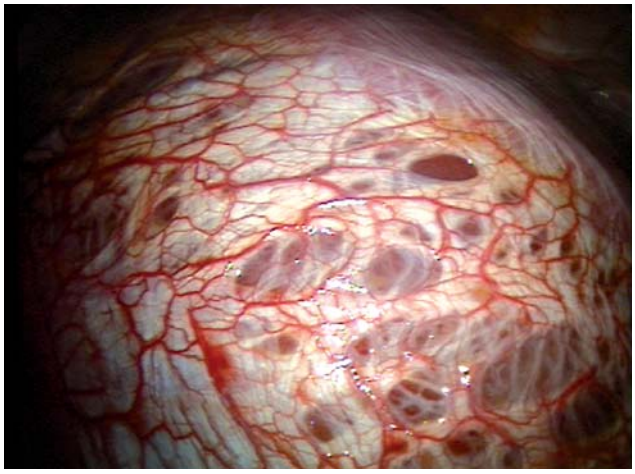

Fig. 1. Thoracoscopic picture of the central tendon of the right hemi-diaphragm showing multiple fenestrations. In some parts the tissue is thinned out. Note prominent blood vessels

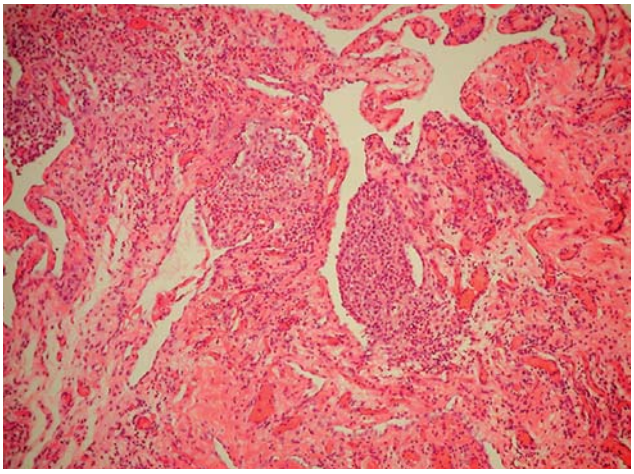

Fig. 2. Histology of resected central part of the diaphragm. Infiltration of chronic inflammatory cells and congested vessels. There was an aggregate of macrophage containing haemosiderin

tropin-releasing hormone agonist following recurrence of pneumothorax. Since starting the hormonal therapy she has remained symptom free.

Discussion

In recent years more cases of catamenial pneumothorax have been described. In the past the prevalence of this peculiar condition has been quoted as 1-5.6% in women suffering from spontaneous pneumothorax [3-5]. Recent studies indicate that the true incidence is higher than previously thought [6-8]. This apparent increase is partly due to the awareness of this condition and partly because of use of thoracoscopy in the management of spontaneous pneumothorax, which greatly facilitates thorough examination of the lung surface and specially the diaphragm [6].

Catamenial pneumothorax occurs in the third and fourth decade of life. Age range is 15-54 years [3, 7, 9]. Chest pain, cough and dyspnoea associated with menstruation are the commonest symptoms. Chest pain may radiate to the right shoulder. Pneumothorax occurs predominantly on the right side [9-13]. Most women experience recurrent episodes of pneumothorax before the definitive diagnosis and treatment is made.

Despite a clear association between the occurrence of pneumothorax and menstruation there is no clear explanation of its pathogenesis. Various hypotheses have been published in the literature but a unifying concept is lacking. According to one hypothesis it was postulated that during menstruation the dissolving cervical mucosa allows passage of air through the fallopian tube into the peritoneal cavity. Air reaches the pleural cavity through congenital fenestration of the diaphragm [3]. In young females spontaneous pneumoperitoneum has been reported following physical effort, sexual intercourse, postpartum knee-chest exercise and pelvic examination [14, 15]. But the fact that there is evidence of recurrence of pneumothorax in patients who had hysterectomy [16, 17] and tubal ligation [18] challenges this theory. Since spontaneous pneumoperitoneum can occur without menstruation, the question also remains why CP always occurs during menstruation.

Diaphragmatic defect plays an important role in the pathogenesis but it is not the primary cause. The simple fact that CP has a temporal relationship with menstruation and its clinical manifestations can be controlled by hormonal manipulation indicates that endometriosis is the key to the pathogenesis of CP. Endometriosis in women is common and occurs in 15% of menstruating women. The incidence of pelvic endometriosis in this group of patients was reported to range from 18% to 70% [4-9, 12]. But the true incidence could be higher because pelvic endometriosis may remain asymptomatic and the diagnosis was not actively looked for in many cases. Moreover the age distribution of CP differs from that of primary spontaneous pneumothorax but is similar to that of endometriosis. In fact thoracic endometriosis was diagnosed in over 50% of cases that underwent surgical exploration [7]. Maurer et al. were the first to associate CP with Sampson's theory of endometriosis [1] and subsequent literature also supported the idea [7-10, 16]. According to Sampson's theory, endometrial tissue refluxes into the peritoneal cavity and flows with the peritoneal fluid in a clockwise current and eventually resides in the sub-diaphragmatic space. It enters the chest through pre-existing diaphragmatic microchannels, congenital perforations or by tissue invasiveness. Pleuroperitoneal communications through the right side of the diaphragm have been implicated in the pathogenesis of many clinical entities including hepatic hydrothorax, Meigs syndrome, pancreatic ascites and chylous ascites [19]. Once in the chest cavity the endometrial tissue can be implanted on the diaphragm or visceral pleura. Implanted endometrial tissue undergoes cyclic changes with the menstruation and can cause diaphragmatic perforation, haemothorax and pulmonary air leak. In CP diaphragmatic defects are found in 15-60% of cases [6-10, 20, 21]. But

diaphragmatic defects are not always discernible; they range from tiny pinhole size to a centimetre or more in diameter [19, 22]. Diaphragmatic defects are common on the right side. The passage of endometrial tissue through the diaphragm is favoured by the thoracoabdominal pressure gradient. In addition to this the relatively fixed solid liver along with the contracting right dome of the diaphragm cause a 'piston' action allowing air to be sucked into the pleural cavity from the peritoneal space through the diaphragmatic defects [19]. Menstruation allows an open connection between the abdominal cavity and the ambient air. In some cases air probably leaks from the lung due to sloughing of retrogradely implanted endometrial tissue on the surface. Pleural endometriosis was not a common feature in the published literature [9]. This explains the predominance of CP on the right side. Endometrial tissue has been diagnosed histopathologically on the diaphragm with or without holes. Most cases of thoracic endometriosis involve the right side and this finding strongly supports the direct extension of endometriosis from the abdomen through the diaphragm into the right pleural space. This theory is supported by the fact that many authors have claimed excellent results following repair of diaphragmatic defects [6, 8, 10, 23]. These reports have a mean follow-up of 6.6-48 months. It is interesting to note that the recent publications have found a higher rate of diaphragmatic defects than the previous ones [5-10, 20, 21]. In the past the defects were probably overlooked in some patients. It is possible that the defects were too small to identify even with video magnifications. Recurrence after diaphragmatic repair, tubal ligation or hysterectomy may happen due to the presence of implanted endometrial tissue on the lung surface.

Metaplastic transformation of cells lining the pleural cavity has also been implicated in the development of thoracic endometriosis and CP [24]. Pleura develop from coelomic epithelium, the same as for the peritoneal cavity and endometrium, and it might have the potential to develop into endometriosis. Degenerating endometrium liberates metaplasia-inducing substances which pass through the diaphragmatic microchannels and induce metaplastic changes of the pleura. This theory fails to explain the right sided predominance and lacks additional basic and experimental data.

Haematogenous spread of endometrial tissue and its homing in lung has been observed in an animal experiment [25]. Pulmonary endometriosis causing catamenial haemoptysis has been well documented [9, 26]. Metastatic spread of endometrial tissue on the surface of the lung can proliferate and slough in synchrony with the menstrual cycle, giving rise to spontaneous pneumothorax. In humans haematogenous spread of endometrial tissue is more likely to occur with uterine manipulation, trauma, pregnancy, labour and surgery. Haematogenous spread can give rise to thoracic endometriosis but fails to explain the preponderance of pneumothorax on the right side as in the hormonal theory suggested by Rossi and Goplerud. Haematogenous spread is probably responsible for bronchopulmonary endometriosis [19, 26].

Korom et al. collected surgical findings of 140 cases of catamenial pneumothorax. They found evidence of thoracic endometriosis in 52.1% of cases. In their review diaphragmatic perforation with or without presence of endometrial tissue was the commonest finding at surgical exploration. This finding was supported by others [5-12, 23]. Other findings including endometriosis of the visceral pleura, pulmonary blebs and apical scarring were noted [4-10]. A small proportion of cases had no pathological finding [7].

Due to lack of understanding of pathogenesis of this syndrome no consensus has been reached on the correct treatment of this problem. The uncertainty has been compounded because of the rarity of this disease and lack of long-term follow-up in treated cases. Since recurrence is common pleurodesis is performed by most authors [3-10]. Because of the young age of the victims mechanical pleurodesis is preferred over talc pleurodesis. Unfortunately pleurodesis alone has a high rate of recurrence [7-9].

A suspected endometrial lesion on the pleura and diaphragm needs excision. The localised area of diaphragmatic defects is repaired by suture or an endoscopic stapler [6, 23]. A large area of diaphragm with multiple holes needs excision and patch repair with prosthetic material [5]. In an animal model prosthetic repair of a larger diaphragmatic defect with synthetic material is preferred to a simple suture [28]. Repair of diaphragmatic defect when present produces significantly improved outcome [7, 8, 10]. However, diaphragmatic defects are not always discernible. Failure to address diaphragmatic defects (macroscopic or microscopic) may lead to recurrence, which is common at the base. Mechanical pleurodesis does not address the diaphragmatic surface, which is the location of pathology and site for recurrence. Recent reports have recommended the use of polyglactin mesh to close any occult defect [8, 9]. Polyglactin mesh over the diaphragm apparently augments basal pleurodesis [8]. Thoracoscopy remains the best diagnostic approach. Better visualisation of the entire thoracic cavity with added magnification is an important advantage [5, 7, 23]. Thoracoscopy will allow repair of diaphragmatic defects with limited excision of diaphragm and mechanical pleurodesis. But a larger diaphragmatic defect or excision should be performed by open thoracotomy to prevent failure of repair and diaphragmatic hernia [5, 6]. Resection of the diaphragm using an endoscopic stapler may fail. There is no doubt that surgical treatment offers a better option to prevent recurrence of pneumothorax [5, 10] but the patient may still experience catamenial chest pain [8].

Hormonal therapy has been used to control endometrial implants. Gonadotropin-releasing hormone (GnRH) agonist, danazol, continuous oral contraceptive pill and progestational drugs are used to treat CP. These drugs suppress endometrial tissue, but do not cause complete regression. Hormonal therapy alone is inadequate to control CP. GnRH agonist appears to be more effective than other hormonal therapies, allowing menstruation [7, 8, 13]. GnRH agonist is

suggested for a limited period due to its side effects. But pneumothorax may recur after stopping it. GnRH significantly prevents recurrence when used in combination with surgery and several authors have recommended its use [5-8, 10, 13, 23].

In the presented case, the occurrence of the basal pneumothorax two months postoperatively may have been multifactorial. First, electing not to do pleurodesis was one factor. The other factor stems from using bovine pericardium (the only tissue available during the procedure), being less inhibiting to endometriosis when compared to Gore-Tex replacement. The third factor that may have contributed to this early recurrence could be that hormonal therapy was not started early in the postoperative period. Early start of hormonal treatment is recognised to mature the pleurodesis effect.

Conclusion

Incidence of CP is higher than suggested and it should be suspected in menstruating women with recurrent spontaneous pneumothorax especially on the right side. Video-assisted assessment is important to diagnose this problem. Although retrograde implantation of endometrial tissue is favoured for the pathogenesis of catamenial pneumothorax, there may also be other aetiological mechanisms involved in this clinical entity. Perioperative GnRH agonist should be offered to patients to allow maturation of pleurodesis and prevent recurrence.

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