SELECTED REPORTS

Catamenial Pneumothorax in Sisters*

James M. Hinson, Jr., M.D.; Kenneth L. Brigham, M.D.; and James Daniel, M.D.

Two sisters had documented pelvic endometriosis and catamenial pneumothoraxes. Both were typical of the 54 reported cases of catamenial pneumothorax in that pneumothoraxes were always on the right and occurred only at menses with onset in the fourth decade. One patient was asymptomatic during 11 months of menstrual suppression with hormones but subsequently required surgery where diaphragmatic perforations were found. The other patient was treated with isoxazole ethisterone with no pneumothoraxes while under suppression. These patients are unlike any of the previously reported cases of familial pneumothoraces. We have not found a previous report of familial catamenial pneumothorax.

Maurer et al. first reported, in 1958, pneumothorax associated with menses as a distinct entity. Now there are at least 54 cases in the literature. Although a few cases of familial spontaneous pneumothorax exist, we can find no report of familial catamenial pneumothorax.

Case Reports

Case 1

After a long history of dysmenorrhea, patient 1, a black 35-year-old woman, gravida 2, para 2, abortus 0, had the diagnosis of endometriosis made by laparoscopy and by biopsy in September 1974. In May 1975, she suffered a right-sided pneumothorax coinciding with menses. This was treated with closed chest tube thoracotomy. At the onset of her next menses, she had a second right-sided pneumothorax but was not treated. At the onset of her menses in late June 1975, she developed a third right-sided pneumothorax which enlarged up to day three of her period when a chest tube was inserted. After three days, the chest tube was removed and the patient was placed on suppressive doses of norethynodrel with mestranol (Eoonid E). She was followed-up as an outpatient for 11 months on this regimen and required up to 20 mg per day to prevent breakthrough bleeding. She had no pulmonary symptoms during that time and repeated chest x-ray films showed no pneumothorax.

In June 1977, the patient was placed on a cyclic regimen of norethynodrel with mestranol and on the first day of her first period, she presented with a 15 percent right-sided pneumothorax which enlarged over the next three days. At that time, she had a right thoracotomy and parietal pleurectomy. The right hemidiaphragm had multiple small perforations and several darkly-pigmented spots were found to contain fibrous granulation tissue and hemosiderin. Following surgery, she was asymptomatic until October 1977 when further dysmenorrhea led to therapy with isoxazole ethisterone (Danazol), with resolution of the symptoms.

Case 2

This woman, a 34-year-old gravida 2, para 2, abortus 0, younger sibling of patient 1, presented in June 1977 on the first day of her menses with shortness of breath, pleuritic chest pain, and a 20 percent right-sided pneumothorax seen on chest x-ray film. She gave a history of dysmenorrhea at the onset of each menstrual period for the previous nine months but had not sought medical attention. The pneumothorax resolved without therapy over five days. No symptoms of pelvic endometriosis were elicited, but she had palpable uterosacral nodules on pelvic exam.

She then had irregular spotted menses until October 1977 when she became pregnant. She underwent therapeutic abortion in early December 1977 and was without chest pain until late December when she presented on the first day of her next menses with chest pain and a 20 percent right-sided pneumothorax.

In January 1978, before her next menses, this patient underwent laparoscopy and was found to have left uterosacral and left ovarian implants typical of endometriosis. There were no lesions on the abdominal surface of the diaphragm. A regimen of isoxazole ethisterone, 800 mg per day, was started and she remained asymptomatic until August 1978. Then, one month after discontinuation of the isoxazole ethisterone, the patient had her typical syndrome and again had a 20 percent pneumothorax. Pleurodesis or castration was recommended but refused. The patient had a small apical pneumothorax at her next menses in September 1978. Surgery was again refused. The patient recovered uneventfully from both episodes. Her next menses was due in October 1978. However, the patient became pregnant and was asymptomatic through her normal delivery. Although she has failed to report for medical follow-up, her sister reports that she continues to have monthly chest pain.

These patients have 11 other siblings, seven males and four females. None of the other siblings has had a documented pneumothorax. One sister (40 years old, gravida 2, para 2, abortus 0) has had chest pain unassociated with menses, but she underwent a total abdominal hysterectomy at age 38 for fibroid tumors. Two sisters, ages 28 and 28, have had dysmenorrhea without chest pain or dyspnea, and random chest x-ray films during their menses have shown no pneumothoraces.

Discussion

The two sisters described both are typical of reported cases of catamenial pneumothorax. The characteristics were summarized by Lillington et al. as follows: (1) involvement of the right hemithorax (there are recent exceptions); (2) close temporal relationship with menses; (3) no pneumothorax except with menses; (4) onset in the third or fourth decade (similar to the onset of symptoms of pelvic endometriosis); and, with a few exceptions, (5) prevention while on suppressive hormones.

Both of our patients had pelvic endometriosis and the one who came to thoracotomy (case 1) also had...
diaphragmatic perforations. Perforations of the diaphragm have been demonstrated in six previous cases.8,9,10,11 and diaphragmatic implants in only eleven.4,9,10,12 At least 13 of the reported 54 cases had no thoracotomy or pleuroscopy.4,5,13

The pathogenesis of catamenial pneumothorax is not clear. Maurer et al8 and others8,14 thought that the pleural air came through the uterus and Fallopian tubes to the peritoneum and entered the chest through diaphragmatic perforations. The recent findings of pleural defects at pleuroscopy seem to support this.8 However, most subsequent authors have implicated thoracic endometrial implants.2,3,5,9,12,13 In reviewing autopsy data, Kovaric and Toll15 found that patients with pulmonary endometriosis usually had bilateral parenchymal lesions, but pleural and diaphragmatic lesions were limited to the right side. They suggested mesenchymal metaplasia or embolization through lymphatics or blood vessels secondary to pregnancy, abortion, or trauma as the source of parenchymal implants and migration through diaphragmatic defects (which are more common on the right) as the source of the pleural lesions. This is supported by the fact that five of seven reported patients with holes in the diaphragm (including our patient 1) also had pleural endometriosis.3,8,10

Rossi and Gopleur13 postulated that parenchymal lesions might release prostaglandin F2α, a smooth muscle constrictor (and bronchoconstrictor) known to occur in the endometrium,16 and this could cause local air trapping and lung rupture. Blood prostaglandin levels are low in anovulatory patients,17 and suppressing ovulation usually prevents catamenial pneumothorax; however, this mechanism does not account for the right-sided nature of the pneumothorax.

A third theory, by Lillington et al,9 holds that the swelling of intraparenchymal endometrial tissue at menses causes a check-valve airway obstruction resulting in hyperinflation and rupture. Although similar to the PGF2α theory, this would not require ovulatory cycles, and thus, could explain the one case of the syndrome occurring without ovulation.4 This patient had bilateral involvement suggesting intrapulmonary endometriosis, and in fact, no pleural implants were found.

Although catamenial pneumothorax has usually been treated by pleural ablation, this has not always prevented recurrence.9,13 There has been general success with cyclic or suppressive hormonal therapy,4,5,9,13 but failures are also reported.4 Isoxazole ethisterone (Dana-zol), a steroid derivative which inhibits gonadotropin release, has been reported to be 87.5 percent effective in relieving symptoms of pelvic endometriosis.18 Our patient is the second we know of to receive this drug for control of presumed intrathoracic endometriosis, and she responded well while taking the drug but had recurrent pneumothorax when the drug was stopped.

We can find no previous report of catamenial pneumothorax in siblings. Familial pneumothorax has been reported both as idiopathic and with Marfan’s syndrome and alpha1-antitrypsin deficiency. These are usually in tall, thin males although one group was in third decade females.1 In the latter cases, the pneumothoraces were bilateral in two sisters and on the left in a third. None of the patients had pelvic endometriosis, and two had extensive pulmonary bullae. There is a report of one family in which twin sisters had left-sided pneumothoraces and three brothers had the same problem on the right side.19 These, too, were thought secondary to blebs. Neither of our patients was above average height or had bullae or emphysema on chest x-ray film. Neither of our cases fits the described pattern of other familial pneumothoraxes.

We have described the occurrence of the typical syndrome of catamenial pneumothorax in two sisters. We do not know whether this represents chance occurrence or a familial predisposition to intrathoracic endometriosis.

REFERENCES

1 Maurer ER, Schaal JA, Mendez FL. Chronic recurring spontaneous pneumothorax due to endometriosis of the diaphragm. JAMA, 1958; 168:2013-14
2 Davies R. Recurring spontaneous pneumothorax concomitant with menstruation. Thorax 1968; 23:3790-93
15 Kovanicck JL, Tol'l GD. Thoracic endometriosis with recurrent spontaneous pneumothorax. JAMA 1966; 196:595
17 Pickles VR. The menstrual stimulus in puberty. J Physiol 1986; 183:968-701

CHEST, 80: 5, NOVEMBER, 1981

CATAMENIAL PNEUMOTHORAX IN SISTERS 835

Downloaded From: http://journal.publications.chestnet.org/pdaccess.ashx?url=/data/journals/chest/21256/ on 04/18/2017