Case report

Massive pleural endometriosis

S.D. Moffatt, John D. Mitchell*
Department of Cardiothoracic Surgery, Stanford University, Stanford, CA, USA

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Abstract

We present a patient who was referred to our thoracic surgical service with a massive, loculated right pleural effusion accompanied by significant ascites. Thoracotomy and decortication were required and pleural biopsy led to a diagnosis of endometriosis. Aggressive medical therapy was subsequently initiated but hysterectomy with bilateral oophorectomy was required due to poor symptom control and the inability to rule out a neoplastic process. There are less than 15 reported cases of endometriosis presenting with both pleural effusion and ascites. Thoracic surgeons presented with such a scenario should be cognizant of this pathological entity.

Keywords: Endometriosis; Pleural biopsy; Ascites; Pleural effusion

1. Case report

A 37 year-old African–American woman was referred to our service with a massive pleural effusion and ascites after presentation to the emergency department with complaints of dyspnea and abdominal pain. The patient had a prior medical history significant for endometriosis, initially diagnosed at the age of 23 when she presented with abdominal pain and infertility. Treatment at that time included medical therapy for her endometriosis, and an exploratory laparotomy performed at the age of 26 for persistent pelvic pain. At the same age, she underwent right thoracotomy for recurrent pleural effusions; nodular implants on the visceral and parietal pleural surfaces were encountered which yielded endometrial tissue, and she underwent talc pleurodesis. She was treated with oral contraceptive pills for 5 years thereafter, and then discontinued her hormonal therapy without consultation of a physician. She remained asymptomatic until 6 months prior to presentation.

Physical examination on admission to the hospital revealed diminished breath sounds to the right lung field and a distended abdomen with ascites. Cytological examination of the chocolate-colored abdominal and chest fluid was negative for malignancy and endometrial cells. Her CA-125 level was normal. Her hemoglobin was 11.0 g/dl; white blood cell count, electrolytes, creatinine and liver function tests were normal.

A chest X-ray and computed tomography (CT) of the chest (Fig. 1) and abdomen were performed. The X-ray demonstrated a significant right pleural effusion with loss of lung volume. The CT scan revealed multiple loculated fluid collections compromising the right lung and displacing the heart into the left hemithorax, ascites, a complex left adnexal cyst, an abnormal soft tissue mass in the right adnexa and a mass superior to the uterus.

Operative intervention was recommended for diagnosis and to relieve the patient’s dyspnea given the multi-loculated nature of her pleural effusion and prior history of pleurodesis. Preoperative fiberoptic bronchoscopy was unremarkable other than for moderate extrabronchial compression at the subsegment level in the right lung. Thoracoscopic examination revealed chocolate brown fluid and dense adhesions necessitating open thoracotomy and decortication. A dense, cream-colored nodular peel was noted on the visceral and parietal pleural surfaces, with the visceral component causing considerable restriction of the underlying lung. Pathological examination of the pleura and fibrous peel revealed small foci of glandular tissue resembling endometrial glands and stroma, consistent with endometriosis. Postoperatively, the patients’ lung reexpanded and she was able to resume normal activities. She was prescribed Leuprolide (11.25 mg intramuscularly every 3 months) and Premarin (0.625 mg postoperatively daily) following consultation with the Gynecology service.

The patient was readmitted 4 weeks after discharge with recurring abdominal pain to the Gynecology service. Repeat abdominal CT scanning demonstrated an increase in the amount of intra-abdominal ascites and the continued
presence of pelvic pathology as previously described. Due to her ongoing symptoms and the inability to rule out a neoplastic process, laparotomy was recommended. Extensive pelvic endometriosis was noted; hysterectomy, bilateral salpingo-oophorectomy with lysis of adhesions and bilateral ureterolysis were performed. Final pathology revealed endometriosis involving the uterus, left ovary, pelvic sidewall and small bowel. Her postoperative recovery was unremarkable.

Nine months after her initial presentation, she is well. She is pain free and denies dyspnea or other respiratory symptoms. She has been maintained on Leuprolide for control of her residual endometrial disease.

2. Discussion

Although endometriosis is usually limited to pelvic organ involvement, extrapelvic endometriosis is well recognized. Its epidemiology, etiology and natural history, however, remain obscure. In fact, it remains unclear if lesions distant from the pelvis even represent the same disease process [1].

Thoracic endometriosis typically affects multiparous women in their mid-30s, with preferential involvement of the right hemithorax in greater than 90% of cases. Most patients will present with catemenial pneumothorax, hemothorax, or both. The differential diagnosis of pleural endometriosis includes metastatic adenocarcinoma as well as a variety of mesothelial proliferative disorders [2]. After the diagnosis is established and the presenting symptoms have been addressed (often with tube thoracostomy), treatment is usually directed at hormonal suppression to prevent recurrence. Pleurodesis, either with talc or by mechanical abrasion, has been used in recalcitrant cases [3].

Massive bloody ascites is rarely secondary to endometriosis, and is more likely encountered in the setting of hepatic tumors, carcinomatous peritonitis or cirrhosis of the liver. In the absence of liver disease, ascites due to endometriosis is commonly mistaken for ascites caused by ovarian neoplasms and therefore this entity is seldom recognized before surgical exploration of the abdomen. The tumor marker for ovarian neoplasms, CA-125, can be elevated in endometriosis as described in previous case reports [4].

Massive ascites associated with a clinically significant pleural effusion caused by intra-abdominal endometriosis is an even rarer phenomenon, with fewer than 15 cases described in the literature since the first report by Brews in 1954 [5,6]. As in Meig’s syndrome, ascitic fluid can reach the pleural cavity by transdiaphragmatic lymphatics. We suggest that in this particular case, endometrial implants on the parietal pleura itself may have been responsible for the associated pleural effusion.

Due to the rarity of cases, treatment of disseminated endometriosis (abdominal and intrathoracic) is anecdotal. Hormonal treatment is often tried as initial therapy, but surgical intervention is often mandated in order to exclude malignancy. In addition, it is unlikely the loculated effusion in the present case would have responded to medical therapy alone. Of the 14 reported previous cases, only two were managed medically whereas the remaining required medical and surgical management [5]. Given her residual endometrial disease, our patient has been maintained on hormonal suppressive therapy to prevent recurrence of effusion.

3. Conclusion

In summary, endometriosis presenting with massive
pleural effusion is rare. Thoracic surgeons presented with such a scenario should be cognizant of this pathological entity. Hormonal therapy should be incorporated into the treatment regimen to minimize recurrence. If associated with ascites, the similarity to neoplastic pelvic pathology in a young population mandates increased awareness and aggressive diagnostic intervention.

References