Case report

Thoracic endometriosis as cause of recurrent pneumothorax

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Case report

We present a 44-year-old woman with no personal history of relevance but for a heterozygous mutation G1691A of the gene of Leiden factor V. Her history starts 2 years ago with two consecutive, 1 month apart, right pneumothoraces that were diagnosed as primary spontaneous pneumothorax (PSP). In both cases she was treated with endopleural aspiration and respiratory physical therapy, with full recovery.

The patient remained asymptomatic for a year, but she suffered another right spontaneous pneumothorax (Figure 1). She was treated with endopleural aspiration. Videothoracoscopy was performed and revealed a bullae in the inferior lobe, a fibroid plaque on the posterior thoracic wall and a hypervascularized lesion with multiple fenestrations localized at the diaphragmatic pleura. A biopsy was taken from the hypervascularized lesion and showed pleural with fibrosis with a hyperplasic mesothelial reaction. Resection of the bullae and pleural abrasion were performed.

The next year the patient suffered her fourth episode of spontaneous pneumothorax and was treated again with endopleural aspiration and respiratory physical therapy. After 1 month, she presented her fifth pneumothorax that did not respond to endopleural aspiration and required a right post-lateral videothoracoscopy. Multiple pleural diaphragmatic fenestrations were observed and were biopsied. Finally, the anatomopathology revealed pleural endometriosis, which was considered to be the cause of the recurrent pneumothorax. On review of her history, the patient explained that her pneumothorax coincided with the beginning or the end of menstruation. With all these clinical features and the outcome, the patient was diagnosed with catamenial pneumothorax.

Following the last episode, medical hormonal treatment for endometriosis was considered, the oral contraceptives were rejected due to her trombophilic state, so the patient was started on gonadotrophin-releasing hormone agonists, trimestral intramuscular triptorelin. At the year of follow-up, the patient had no further episodes of pneumothorax.

Discussion

Pneumothorax is defined by the presence of air in the pleural cavity. It was first described by Itard and then Laennec in 1803 and 1819, respectively. Pneumothorax is classified in PSP, occurring in healthy people, or as secondary pneumothorax (SSP), when it is associated with an underlying lung disease. A type of SSP is the catamenial pneumothorax (CP) caused by endometriosis and it was first described by Maurer et al. in 1958.

Endometriosis is a condition characterized by the presence of endometrial tissue out of the uterine cavity. It has been estimated that it affects 10% of the women in reproductive age, and the incidence of extrapelvic endometriosis in these women is approximately 12%. CP is the most frequent presentation of thoracic endometriosis and it is defined as recurrent pneumothorax in women during their reproductive age. CP occurs within 72 h from the
onset of menstruation, even though there have been cases reported outside of this period.\(^6\) CP, catamenial haemothorax, catamenial haemoptysis and lung nodules have been termed as the thoracic endometriosis syndrome.\(^2\) Typically, the CP is unilateral and right sided, but left-sided or bilateral pneumothoraces have occasionally been reported.\(^7\)

Three theories have been proposed to explain the CP: (i) the coelomic metaplasia based on the common embryologic origin of both mesothelium and endometrium and that under appropriate pathogenic stimuli differentiation of precursor cells in the serosae towards endometrial cells could occur, (ii) lymphatic or haematogenous embolization from uterus or pelvis and (iii) retrograde menstruation with subsequent transperitoneal–transdiaphragmatic migration of endometrial tissue. None of these theories, however, can explain all the clinicopathological presentations of CP, which suggest that the disease has probably a multifactorial cause.\(^7\)

Cancer Antigen (CA) 125 has been known to be associated with endometriosis. An elevated serum CA125 level is associated with any process that irritates the mesothelial cells, for example endometrium, endocervix, pleura, peritoneum or pericardium. Increased levels of CA125 have also been associated with endometriosis-related CP; for this reason, the assessment of serum CA125 levels may be useful in the early diagnosis of thoracic endometriosis in females of reproductive age presenting recurrent PSP.\(^7,8\)

The management of CP strategies include surgery and hormonal treatment. The aim of medical treatment is to block the hormonal support to the existing endometrial implants and to prevent further seeding; the different options available are oral contraceptives, prostegstives, danazol and gonadotrophin-releasing hormone agonists.\(^9\) The recurrence rate after hormone therapy seems high, at \(~50\%).\(^7\) Thoracic surgical techniques have been varied and include diaphragmatic resection or placation of the fenestrations seen at thoracoscopy, the insertion of a mesh or patch over these fenestrations, electrocoagulation of the endometriosis deposits and pleurodesis. The recurrence rates after surgical management of CP seem less frequent than with hormone therapy, with documented recurrence rates of 30%.\(^10\) Combined therapy, surgery and hormonal treatment, offer a lower recurrence.\(^2,7,11\)

In conclusion, CP is a condition that must be suspected in women at reproductive age with recurrent pneumothorax. The coincidence of the pneumothorax with menstruation must put us on alert for pneumothorax related with thoracic endometriosis. CA125 levels may be of aid if CP is suspected. The association of surgical and hormonal treatment seems to be the best treatment option, which will require collaboration between thoracic surgeons, gynaecologists and family physicians.

Conflict of interest: None declared.

References

