

## Two Young Women with Left-sided Pneumothorax Due to Thoracic Endometriosis

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### Abstract

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Pneumothorax associated with thoracic endometriosis (TE) generally occurs in women around 30 years old and it usually affects the right pleural cavity. We herein report two cases of TE associated with left-sided pneumothorax in young women. The prevalence of TE in younger patients may be underestimated if these cases are treated as spontaneous pneumothorax. Pneumothorax occurring in younger patients has not been reported to show laterality. TE-related or catamenial pneumothorax in young women must therefore represent a different clinical entity from the condition seen in older patients.

**Key words:** catamenial pneumothorax, thoracic endometriosis

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### Introduction

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Thoracic endometriosis (TE) syndrome includes three clinical entities: catamenial pneumothorax (CP), catamenial hemoptysis, and endometrial lung nodules (1). Pneumothorax is the most frequent presentation of TE syndrome, accounting for about 70% of cases (2). This syndrome typically occurs in women of child-bearing age and affects the right pleural cavity. We herein present two rare cases of TE causing left-sided pneumothorax in younger patients.

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### Case Reports

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#### Case 1

A 15-year-old girl attended her primary-care physician complaining of chest pain and dyspnea that had occurred on the first day of her menses. Chest radiography revealed left-sided pneumothorax, and the patient was referred to our institution. She presented with tension pneumothorax and un-

derwent emergency thoracic drainage. Chest computed tomography (CT) revealed apical blebs on the upper left lobe.

Conservative treatment was provided for the first episode of pneumothorax, but the air leakage persisted despite continuous low-pressure suction. She therefore underwent video-assisted thoracic surgery (VATS) 9 days after admission, revealing multiple yellow nodules measuring 2 mm in diameter on the diaphragm (Figure A) and white nodules measuring 2 mm in diameter on the visceral pleura (Figure B). A water seal test showed air leakage only from the apical bleb, and wedge resection of the lung parenchyma containing the apical blebs was therefore performed.

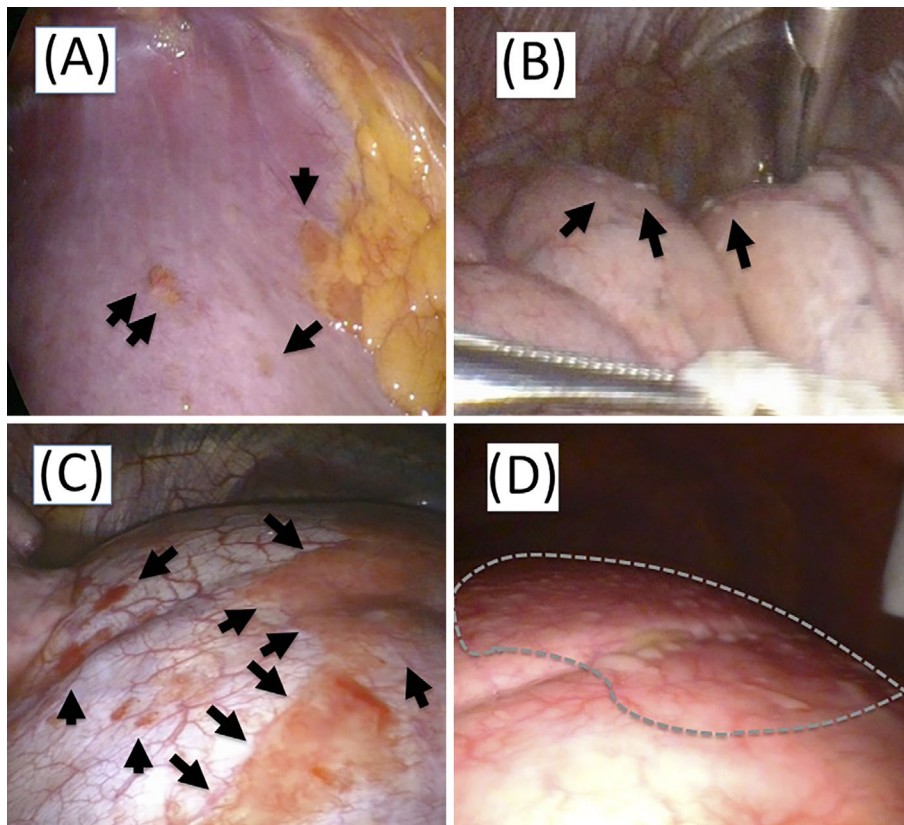
A histopathological examination of the wall of a bleb revealed endometrial stroma with positive results for expression of the progesterone receptor (PgR). As a result, the patient was diagnosed with TE. Although no pelvic lesions were seen, a gynecologist nevertheless recommended gonadotropin-releasing hormone (GnRH) analog therapy, which was declined by the patient and her parents. The left-sided pneumothorax recurred 22 days after the VATS procedure. Drainage and pleurodesis using a streptococcal prepa-

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**Figure.** Operative findings. (A, B) Case 1: Small yellow nodules on the left diaphragm (A) and white nodules on the visceral pleura (B). (C, D) Case 2: Diaphragmatic lesion (C) and nodules on the apical part of the left upper lobe (D).

ration (OK-432) was performed. The patient has remained free of recurrence over a one-year follow-up period.

### Case 2

A 20-year-old woman was admitted to our institution for left-sided pneumothorax with persistent air leakage lasting for 10 days, which had been drained by her primary physician. The patient was on the first day of her menses when she was referred to our institution. Chest CT revealed apical blebs on the left upper lobe.

The patient underwent VATS 16 days after the onset of the pneumothorax. Multiple diffuse red nodules were seen on the diaphragm (Figure C), and white nodules were seen on the visceral pleura (Figure D). A water seal test showed air leakage from the apical bleb. Wedge resection of the region of the upper lobe containing the blebs and biopsy of a diaphragmatic lesion were performed.

A histopathological examination of the wall of a bleb and of the biopsy specimen revealed endometrial stroma that was positive for PgR, estrogen receptor, and CD10. TE was diagnosed, and the patient was referred to a gynecologist, who ruled out pelvic lesions. GnRH analog treatment was initiated, and in the six months of follow-up thus far, she has not shown any recurrence.

### Discussion

Reports of endometriosis-related pneumothorax in young women are rare, and the prevalence among women of all ages remains unclear. Although the frequency of TE is unknown, endometriosis is not a rare condition, affecting 15% of all menstruating women (3). Spontaneous pneumothorax is the most common clinical manifestation of TE, occurring in 72-73% of patients (2). CP refers to spontaneous pneumothorax that occurs within 24 hours before to 72 hours after the onset of menses in women of child-bearing age (4). However, TE-associated pneumothorax can also occur outside of that time frame (5). Pneumothorax due to TE can thus be difficult to diagnose without surgical exploration.

The histopathological diagnosis of TE requires the presence of both endometrial stroma and glands, or stroma-only positive expression of estrogen or progesterone receptors (6). Approximately 40% and 30% of patients with CP have diaphragmatic abnormalities and visceral pleural endometriosis, respectively (1). The number of cases of CP- or TE-related pneumothorax in young women is probably underestimated. In general, the mean age at presentation of CP is 34-35 years, and CP affects the right hemithorax in 85-90% of cases (1). In pathologically confirmed TE, Fukuoka et al. found that only 1 of 150 cases (0.7%) showed left-sided pneumothorax (7). Lesions occurring in juveniles and in-

volving the left hemithorax are considered rare. Inoue et al. (8) recently reported that CP in juveniles manifests different characteristics from CP in adults. CP can occur in teenage girls in either hemithorax, whereas right-sided lesions are the most common in older patients.

The proposed mechanisms for TE remain controversial. Maurer et al. theorized (9) that diaphragmatic lesions result from transtubal regurgitation and transperitoneal dissemination or translocation of endometrial tissue. Endometrial tissue is said to circulate clockwise, along with the flow of peritoneal fluid in the abdominal cavity, until arriving at the peritoneal surface of the right diaphragm (10). Endometrial tissue traverses the diaphragm through microchannels, hereditary perforations, or by invasion (11). Such a process would account for the finding that CP usually affects the right lung. However, this hypothetical mechanism is only sufficient for explaining CP in mature women and is inadequate for juvenile cases. Kovaric et al. reported autopsy findings showing that pleural and diaphragmatic lesions were always right-sided, whereas pulmonary parenchymal endometriosis usually manifested as bilateral lesions (10).

Circulation of microemboli of endometrial tissue through the pelvic veins is another proposed mechanism for the peritoneal-pleural translocation of endometrial tissue (1). A universal hypothesis, the coelomic metaplasia theory, attempts to explain the occurrence of endometriosis at unusual sites and instances (10). That theory accounts for the finding that endometriosis can occur in teenagers before menarche as well as in postmenopausal woman and castrated men treated with estrogen. We therefore think that the terms "thoracic endometriosis" and "catamenial pneumothorax" do not refer to a single disease entity but rather to a broader category of entities.

In young women with left-sided or non-catamenial pneumothorax, clinicians may be excluding TE-related pneumothorax from the differential diagnosis. In addition, we may avoid surgical treatment for pneumothorax, because a young age is considered a risk factor for recurrence after VATS procedures for spontaneous pneumothorax (12). Avoiding VATS may in turn have resulted in underestimation of the prevalence of TE in younger patients. We believe that performing VATS during the early stages of pneumothorax will help ensure the timely diagnosis of TE-related pneumothorax and the initiation of an appropriate treatment or an appropriate clinical decision to observe the patient and apply conservative management.

In conclusion, the prevalence of TE-related pneumothorax

in teenage girls and young women may be underestimated if the case is treated as spontaneous pneumothorax. Although the mechanisms underlying the dissemination to or the implantation of endometrial tissue in the thoracic cavity in these age groups are still not well understood, they likely differ from the mechanisms causing TE that affects patients in other age groups.

**The authors state that they have no Conflict of Interest (COI).**

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