Spontaneous Pneumothoraces due to Metastatic Endometrial Stromal Sarcoma in a Woman Infected with HIV

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Figure 1. Chest radiograph in posteroanterior view (A) demonstrates bilateral pneumothoraces as well as several pulmonary nodules and masses. Computed tomography images of the chest (displayed in lung windows) in axial (B and C) and coronal (D) planes demonstrate solid and cavitary/cystic predominantly subpleural pulmonary nodules and masses. Bilateral pneumothoraces appear to be secondary to subpleural lesions, which were ultimately proven to be pulmonary metastases of endometrial stromal sarcoma.

A 41-year-old woman with HIV/AIDS (CD4 439 cells/mm³ on antiretroviral therapy) presented with cough, dyspnea, and pleuritic chest pain. Chest imaging showed bilateral pneumothoraces as well as numerous solid and cystic subpleural pulmonary nodules and masses (Figure 1). Concurrent abdominopelvic imaging revealed uterine and bilateral adnexal enlargement. CA-125 was 41.5 U/ml (normal, <20 U/ml). Pneumothoraces were managed with bilateral tube thoracostomy. Surgical lung biopsy yielded a diagnosis of metastatic endometrial stromal sarcoma (ESS) (Figure 2A). Two years earlier, a uterine mass had been diagnosed as leiomyoma by biopsy.

In advanced HIV infection (CD4 <200 cells/mm³), pneumothoraces are typically associated with opportunistic infections. Our patient's reconstituted immunity, previous uterine pathology, and elevated CA-125 prompted consideration of alternative etiologies such as pulmonary metastases or thoracic endometriosis.

Obtaining a definitive diagnosis of ESS is challenging. Common obstacles include inadequate sampling and confusion with other spindle cell entities due to overlapping cytopathologic profiles. In one retrospective series, 40% of uterine ESS cases were initially misdiagnosed as benign lesions, resulting in a mean diagnostic delay of 143 months (1). In our case, positive CD10 staining of lung biopsy tissue helped secure the diagnosis of metastatic ESS (2). Early detection is critical, because 5-year survival for stage III/IV disease is only 66% (3, 4). Our patient was initially managed with hysterectomy and megestrol acetate (5). Unfortunately, her disease progressed, and she was switched to exemestane, an aromatase inhibitor. ■

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Some of the contents of this case report have been previously reported in the form of an abstract/poster presentation (5).

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Figure 2. After nondiagnostic percutaneous and transbronchial lung biopsies, video-assisted thorascopic biopsy revealed a diagnosis of stage IV endometrial stromal sarcoma (ESS) with lung metastases. (A) Microphotograph (CD10, 10×) showing a spindle cell lung nodule with diffusely positive CD10 staining. CD10 antigen is normally expressed in benign and malignant uterine stroma and useful in distinguishing ESS from other spindle cell neoplasms. (B) Microphotograph (hematoxylin and eosin, 10×) of the uterus showing an intramyometrial vascular channel with conspicuous lymphovascular invasion by ESS.

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