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Case Report

Malignant presentation of uterine lymphangioleiomyomatosis

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ABSTRACT

Objective: The main aim of this case report was to present the method of diagnosis, management, and the 12-year-follow-up of a patient diagnosed with primary uterine lymphangioleiomyomatosis (LAM).
Case Report: A 47-year-old woman was admitted to the Department of Thoracosurgery due to pulmonary lesions suspected to be metastatic. The potential primary site of the neoplasm was not identified by previous imaging studies and specialist counseling. The patient had a history of total abdominal hysterectomy without ovaries due to a uterine tumor recognized as cellular leiomyoma and left salpingo-oophorectomy due to a solid ovarian tumor also recognized as leiomyoma. She had previously undergone the removal of a left kidney angiomyolipoma. After histopathological examination of the pulmonary lesions and repeated evaluation of the ovarian and uterine tumors, the diagnosis of primary uterine LAM with metastases to the ovary and the lungs was established. Although new metastatic lesions occurred, the patient remained in good condition during the 12-year-follow-up.
Conclusion: The history of our patient and review of the literature suggest that although uterine LAM presents malignant features (i.e., metastasis), the disease is long lasting and the patient can be in good condition for a number of years.

Introduction

Lymphangioleiomyomatosis (LAM) is a rare disease characterized by the proliferation of abnormal smooth muscle cells. The disease usually affects women of reproductive age. LAM may be sporadic or associated with the tuberous sclerosis complex (TSC), which is an autosomal dominant, multisystem condition characterized by the development of hamartomas and neoplasms at various body sites, especially the skin, central nervous system, and kidney. Mental retardation and epilepsy are also frequent in patients with TSC [1].

The exact etiology of LAM is currently unknown, despite recent progress in understanding its pathology [2,3]. LAM occurs mainly in the interstitium of the lungs, however, cases of extrapulmonary LAM have also been reported. LAM has been detected in several body regions, including the retroperitoneum, mesentery, ureter, biliary tract, liver, ovary, adrenal gland, and inguinal or supraclavicular lymph nodes [4–6]. We present a rare case of uterine LAM with malignant course of the disease.

Case Report

A 47-year-old woman was admitted to the Department of Thoracosurgery of Wielkopolska Center of Pulmonology and Thoracosurgery (Poznan, Poland) in January 2010 for a consultation regarding lung lesions resembling distant metastases. The lesions in the lungs were diagnosed in November 2008 during a routine chest X-ray examination. The patient had previously received counseling by a surgeon and internist. Previous imaging studies had failed to recognize the site of origin of the lung metastases. On admission to the Department of Thoracosurgery, the patient was in good condition without any clinical symptoms.

The patient had a history of total abdominal hysterectomy without ovaries because a uterine tumor was recognized as a cellular leiomyoma in 2001. In November 2008, she had left salpingo-oophorectomy due to a solid ovarian tumor. The tumor

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was surprisingly recognized as an ovarian leiomyoma during the first histopathological examination. Immunohistochemical results were positive for desmin, vimentin, and smooth muscle actin, and negative for CD1, inhibin, cytokeratin AE1/AE3, S100 protein, and CD34. The expression of CD31 was weak. The expression of Ki67 was found in 3% of the cell nucleus. Preoperative serum CA125 level was 60.3 IU/mL. In December 2009, she underwent a third laparotomy due to a left renal tumor. She had partial nephrectomy, and the final histopathological examination revealed renal angiomyolipoma.

We performed an X-ray computed tomography (CT) examination of the chest, which revealed metastatic lesions, mainly in the III and VI segments of the right lung and VI and X segments of the left lung, without fluorodeoxyglucose uptake on positron emission tomography. Several emphysematous bullae and enlargement of lymph nodes of the pulmonary hilum were also visualized. We then decided to perform video-assisted thoracoscopy with wedge tumor resection of the right lung. The diagnosis of benign metastasizing leiomyoma was made following histopathological examination with the revision of the uterine and ovarian tissue samples.

Two months later, the patient was admitted to the hospital due to dyspnea with suspected disease progression. An increased diameter of the lesion in the left lung was detected by CT scans. Thoracotomy and left lung laser wedge resection was performed in June 2010. Subsequent histopathological examination with review of the uterine, ovarian, and renal lesions revealed primary uterine LAM with metastases to the ovary and lungs. Final diagnosis was established after immunohistochemical analyses, which revealed the expression of human melanoma black protein 45 (HMB45) in the ovarian tumor and lung lesions. There was no possibility for conducting immunohistochemical analyses on the uterine tumor samples because the samples were utilized after 10 years of storage.

Experimental tamoxifen therapy was initiated due to intolerance of gestagens and disease progression. However, after 5 months of stabilization, new metastatic lesions were found in the right lung. We then stopped tamoxifen therapy, and performed right thoracotomy with wedge laser lung resection and enlarged lymph node excision. After few months of stabilization, a tumor of 68 mm × 44 mm × 52 mm in size in the retropubic area was visualized by CT examination. The tumor was excised in March

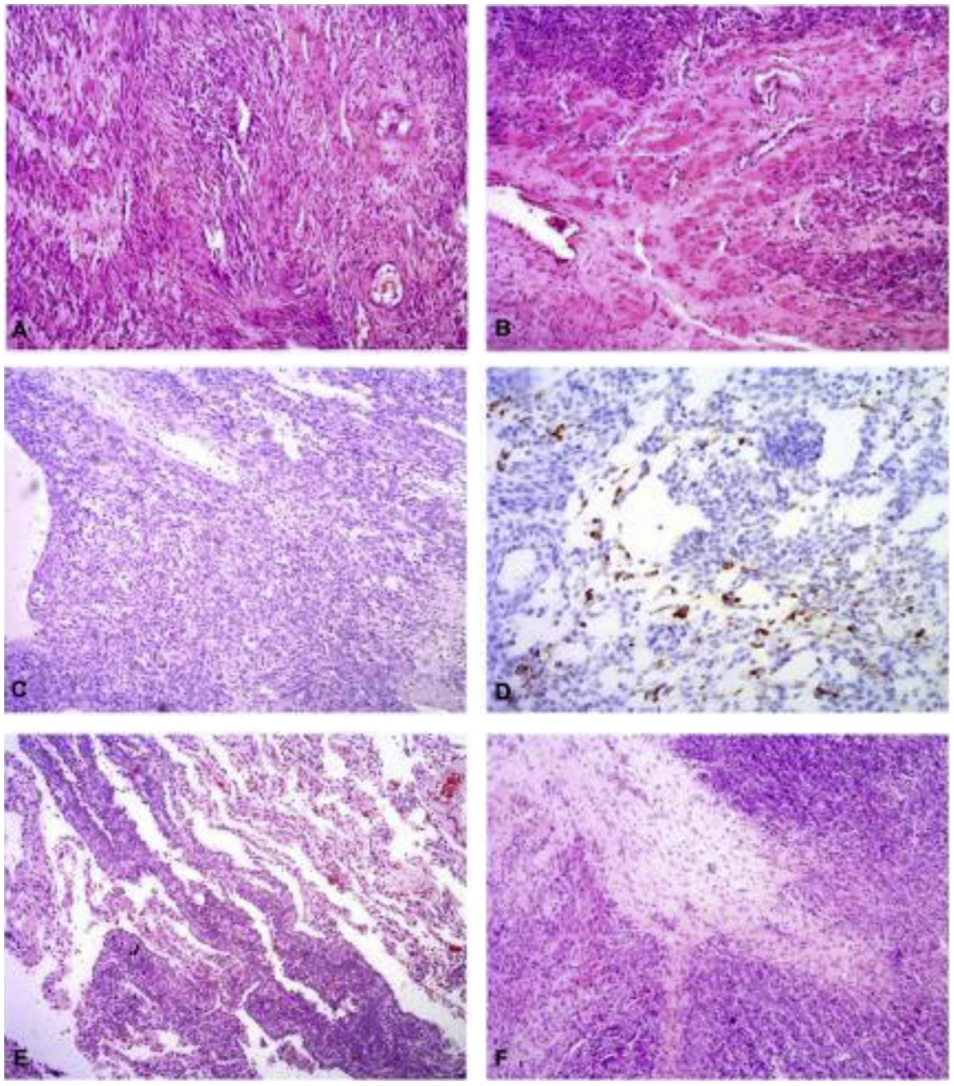

Figure 1. (A) Primary uterine lymphangioleiomyomatosis (LAM). (B) Primary uterine LAM. The photograph shows penetration of LAM cells into the myometrium. (C) LAM metastasis of the ovary. (D) LAM metastasis to the ovary with a positive reaction for human melanoma black protein 45 (HMB45). (E) LAM metastasis of the lungs obtained from the first video-assisted thoracoscopy wedge resection. The photograph presents LAM lesions within lung tissue. (F) LAM metastasis of the lungs from right thoracotomy (third resection of LAM metastasis of lungs). LAM lesions resemble uterine and ovarian tumors. All images, magnification 50×.