Uterine Intravenous Leiomyomatosis with Pulmonary Spread.

Farah Al-Naimy, Ruṣaida Alwaqfī, Ismail Matalka, Hussam Telfah, Shadi AL-Hamouri, Isam Lataifah, Department of pathology and laboratory medicine, King Abdullah University Hospital.

To report a case of Uterine intravenous leiomyomatosis (IVL) with pulmonary extension.

A 47 year old Jordanian female patient presented with shortness of breath and cough of two month duration. She was found to have bilateral multiple lung nodules seen on chest CT scan. The patient had undergone hysterectomy in 2008 and was diagnosed with uterine intravenous leiomyomatosis. Wedge resection of one of the lung nodules was performed and sent for histopathology examination. A diagnosis of pulmonary extension from her previous uterine IVL was made. Follow up of the patient shows improvement on hormonal treatment.

Uterine intravenous leiomyomatosis is a very rare disease characterized by intravascular proliferation of a histologically benign-looking smooth muscle cell tumor mass, but not invading the tissue. Less than 300 cases have been reported with distant extension. The neoplastic smooth muscles of IVL are histologically and cytogenetically similar to benign leiomyomata but might behave in a “malignant” fashion, with not only involvement of pelvic veins, IVC, renal veins, pulmonary artery and right cardiac chamber, but also distant metastasis, such as lung, brain, and lymph nodes. The radiological presentation is usually multiple nodules. Making the diagnosis of distant spread IVL is very difficult with strict pathological diagnostic criteria; it is based on histological features, immunohistochemical staining and the history of hysterectomy for IVL. Surgery combined with hormone therapy is believed to be the best choice of treatment for distant spread of IVL.

Pulmonary spread of IVL is a very rare condition and should be considered in women of reproductive age with a history of uterine leiomyoma who present with solitary or multiple pulmonary nodules of the lung. Long-term follow up is required for patients with IVL for early detection of recurrence or distant metastases.