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Clinical Features and Treatment Outcomes of Pulmonary Metastases in Low-Grade Endometrial Stromal Sarcoma

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Introduction

Low-grade Endometrial Stromal Sarcoma (LGESS) is a rare and indolent tumor that originates from the stromal tissue of the uterus. It is part of a broader group of uterine sarcomas that includes various malignant tumors of mesenchymal origin. LGESS is distinguished by its relatively slow growth, less aggressive nature, and propensity for late recurrence. The presence of pulmonary metastases in patients with LGESS is uncommon but can significantly alter the clinical course and prognosis of the disease. Pulmonary metastases are an important consideration in the management of these patients due to their potential impact on survival, quality of life, and response to treatment. Through an in-depth exploration of the clinical characteristics and treatment options, the article aims to provide a comprehensive understanding of how these metastases affect the management of LGESS and the implications for patient care [1].

Description

Endometrial stromal sarcoma is a rare neoplasm originating from the endometrial stroma, the supportive tissue in the uterine lining. It accounts for only 0.2-0.5% of all uterine cancers. Histologically, LGESS is characterized by a proliferation of endometrial stromal cells, often resembling the normal stroma of the uterus but with the presence of cellular atypia and increased mitotic activity. These tumors are typically classified as low-grade due to their indolent behavior, though they can occasionally exhibit more aggressive features. Patients with LGESS are often diagnosed in their reproductive years, though it can occur at any age. The clinical presentation can vary, but most patients report symptoms related to abnormal uterine bleeding, pelvic pain, or a palpable mass. LGESS tends to be estrogen receptor-positive, which may provide insight into its pathophysiology and therapeutic strategies. Despite being generally less aggressive than other types of uterine sarcomas, LGESS can spread to distant organs, with the lungs being a common site of metastasis. Pulmonary metastasis in LGESS is a rare but serious complication. Lung metastases typically develop years after the initial diagnosis and are often seen in patients with recurrent or advanced disease. The mechanisms underlying pulmonary metastasis in LGESS are not entirely understood but are believed to involve hematogenous spread. The pulmonary vasculature is a common site for the dissemination of tumor cells due to the high blood flow through the lungs, making them a frequent target for metastasis in various malignancies, including uterine sarcomas [2].

Clinical manifestations of pulmonary metastases in LGESS may include persistent cough, hemoptysis, pleuritic chest pain, or dyspnea. However, many patients remain asymptomatic, and the metastases are often detected incidentally during follow-up imaging for suspected recurrence or progression of the primary uterine tumor. Radiological findings include multiple nodular lesions, which may be diffuse or localized to one lung. These findings are suggestive of metastatic disease and warrant further investigation. The diagnosis of pulmonary metastasis in LGESS is primarily based on imaging studies, particularly chest X-rays, Computed Tomography (CT) scans, and occasionally magnetic resonance imaging (MRI). Histologically, pulmonary metastases from LGESS retain the characteristic features of the primary tumor, such as the presence of endometrial stromal cells with a similar architectural pattern. Immunohistochemical staining for estrogen and progesterone receptors may also aid in confirming the diagnosis, as these markers are frequently expressed in LGESS [3].

Surgical resection of pulmonary metastases is considered the primary treatment option when the metastases are limited in number and are resectable. Pulmonary metastasectomy can offer a potential curative approach, especially in patients with isolated, well-defined lesions. The role of surgery is most beneficial in patients with a good performance status who have no evidence of further systemic disease. The success of surgery depends on the size and location of the lesions, as well as the patient's ability to tolerate the procedure. In a study of patients with pulmonary metastases from uterine sarcomas, including LGESS, surgical resection of metastases was associated with prolonged survival in a subset of patients. However, long-term outcomes can vary, and recurrence of pulmonary metastases after surgical resection is not uncommon. Patients undergoing surgery should be closely monitored for signs of recurrence through regular imaging. Systemic therapy for pulmonary metastases in LGESS typically involves chemotherapy or hormone therapy, as these tumors often express estrogen and progesterone receptors. Chemotherapy has limited efficacy in treating LGESS, but it may be considered in cases of widespread metastasis or when surgery is not an option. Doxorubicin-based regimens are commonly used for uterine sarcomas, but response rates remain modest. These therapies aim to inhibit estrogen-driven tumor growth and can be an option for patients with hormone-sensitive pulmonary metastases. The efficacy of hormone therapy may vary based on the receptor status of the tumor and the individual patient's response [4,5].

Conclusion

Pulmonary metastases in low-grade endometrial stromal sarcoma represent a significant challenge in the management of this rare malignancy. Although the clinical features of LGESS are often indolent and patients may remain asymptomatic for years, the development of pulmonary metastases can alter the prognosis and therapeutic approach. Surgical resection offers the best chance for long-term survival in patients with limited metastatic disease, while systemic therapies, including chemotherapy and hormone therapy, remain the mainstay of treatment for advanced cases. Early detection, close monitoring, and a multimodal treatment approach tailored to the individual patient remain critical for managing pulmonary metastases in LGESS. Despite the rarity of the disease, continued investigation and clinical trials are essential to optimize treatment strategies and improve the prognosis for patients with pulmonary metastases from low-grade endometrial stromal sarcoma.

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Conflict of Interest

None.

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