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#### Annals of Diagnostic Pathology

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Cytological-Pathological Correlation

## Low-grade endometrial stromal sarcoma presenting as multiple pulmonary nodules: A potential pitfall in fine needle aspiration and core biopsy specimens - A Cytological - Pathological Correlation



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#### ARTICLE INFO

# Keywords: Low-grade endometrial stromal sarcoma Solitary fibrous tumor Synovial sarcoma Neuroendocrine tumor Leiomyosarcoma Fine needle aspiration

#### ABSTRACT

Low-grade endometrial stromal sarcoma (LGESS) is the second most common malignant mesenchymal tumor of the uterus. The most common location is the uterine corpus, but it can also primarily arise in a variety of extrauterine locations such as pelvis, ovary, abdominal cavity, vagina, and vulva. We are reporting a case of a 47-year-old female with no significant medical history who presented with multiple pulmonary nodules. Fine needle aspiration (FNA) specimen revealed spindle cell neoplasm consistent with the diagnosis of LGESS. The differential diagnosis included neuroendocrine tumor, synovial sarcoma, solitary fibrous tumor, smooth muscle tumors, and peripheral nerve sheath tumors. The clinical, cytological, and histopathologic details of this case, as well as a discussion of the potential pitfalls and differential diagnosis of spindle cell lesions of the lung are described.

#### 1. Introduction

Low-grade endometrial stromal sarcoma (LGESS) is the second most common malignant mesenchymal tumor of the uterus with majority occurring in the perimenopausal period [1]. The differential diagnosis would have to include a number of monomorphic spindle cell neoplasms. These may include, depending on the site of FNA, endometriosis, ovarian stromal tumors, smooth muscle tumors, peripheral nerve sheath tumors, synovial sarcoma, and solitary fibrous tumor, among others.

#### 2. Case report

A 47-year-old female who never smoked, presented with abdominal pain at an outside institution. A CT scan revealed multiple bilateral lung nodules and therefore a CT-guided fine needle aspiration (FNA) and a core biopsy of the largest 1.0 cm solid lung nodule was performed at the outside institution (OI #1) in June 2015. The diagnosis of "Atypical cells present, consistent with carcinoid" was made. The patient was then referred to New York-Presbyterian Hospital/Weill Cornell Medicine for further evaluation. The pathology slides were reviewed within the Weill Cornell Medical College (WCMC) Department of

Pathology and Laboratory Medicine.

The FNA smears revealed singly scattered and loosely cohesive groups of relatively uniform spindled tumor cells with scant to moderate delicate cytoplasm (Fig. 1A-C). The neoplastic cells exhibited round to oval nuclei with fine chromatin and inconspicuous nucleoli. Naked nuclei of tumor cells were noted and no mitoses were observed. Clusters of tumor cells attached to delicate blood vessels were also seen. The background was clean and no significant stromal matrix component was noted. The concomitant core biopsy specimen showed proliferation of bland spindle cells with mild cytologic atypia. (Fig. 2A) Immunohistochemistry stains performed on the core biopsy specimen, by the outside institution, showed the lesional cells to be positive for CD56 (Fig. 2B) and negative for chromogranin, synaptophysin, AE1/ AE3, CK20, CK7, CD117, CAM5.2, CD45 Leukocyte Common Antigen (LCA), and TTF-1. At WCMC, we performed additional immunostains on de-stained immunoslides. The lesional cells were positive for CD10 (Fig. 2C) and negative for PAX8. Ki-67 showed a proliferative index of 1-2%. Based on the immunohistochemical profile, the diagnosis of a neuroendocrine tumor (NET) was unlikely and we were favoring a diagnosis of metastatic endometrial stromal sarcoma. However, a precise diagnosis could not be rendered on the limited tissue material, and additional tissue sampling for further characterization of the lesion was

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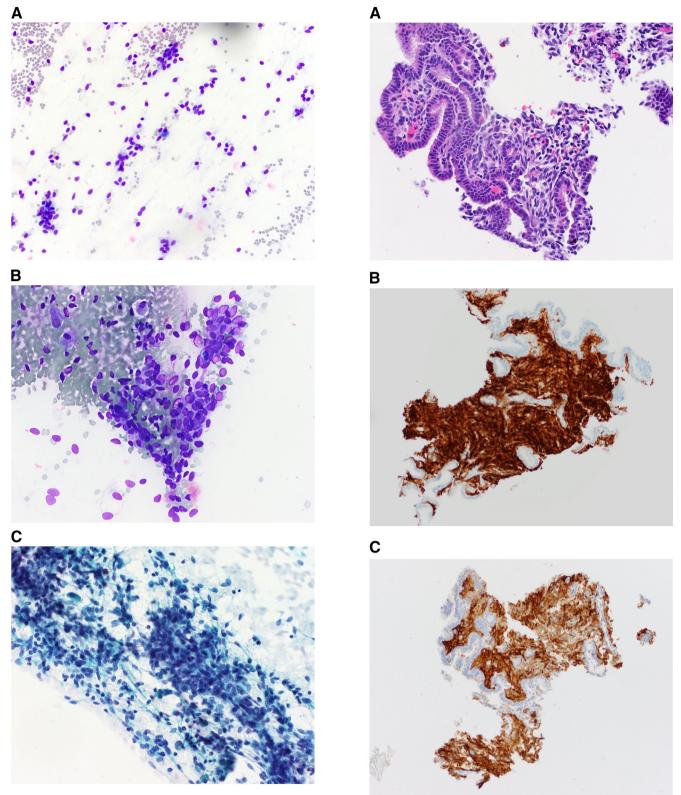


Fig. 1. FNA showing singly scattered and loosely cohesive groups of relatively uniform spindled tumor cells with scant to moderate delicate cytoplasm. The neoplastic cells exhibited round to oval nuclei with fine chromatin and inconspicuous nucleoli. Occasional "comet cells" and naked nuclei of tumor cells were noted. Clusters of tumor cells attached to delicate blood vessels were also seen. (A–B) DQ stain and (C) PAP stain; A  $\times$ 10, B–C  $\times$ 40.

**Fig. 2.** The core biopsy specimen showed proliferation of bland spindle cells with mild cytologic atypia. (A) H&E stain (B) Immunostain for CD56 is positive in the lesional cells. (C) Immunohistochemistry CD10 performed on the destained slides is positive in the lesional cells.  $A-C \times 40$ .

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Repeat CT chest showed 10 nodules in each lung, some of which increased in size while others decreased in size. The previously

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