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Cytologic-Pathologic Correlation

Primary gastric extra-uterine endometrial stromal sarcoma

Ming Jin, MD, PhD [a], Jordan P. Reynolds, MD [b], Shelley I. Odronic, MD [b], Paul E. Wakely Jr., MD [a,*]

[a] Department of Pathology, The Ohio State University Wexner Medical Center, Columbus, OH
[b] Robert J Tomsich Pathology and Laboratory Medicine Institute, Cleveland Clinic, Cleveland, OH

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ABSTRACT

Endometrial stromal sarcoma (ESS) is an uncommon uterine neoplasm, but its occurrence as an extra-uterine primary (EESS) is exceedingly unusual, and the fine-needle aspiration (FNA) cytopathology of EESS is rarely described. We hereby present 2 women with primary gastric EESS whereby the FNA cytopathology of this rare entity showed a population of cytologically monotonous oval-spindle shaped cells. This cytopathology is correlated with the subsequent histopathology. EESS is another, albeit rare, diagnostic consideration along with gastrointestinal stromal tumor, schwannoma, glomus tumor, and leiomyoma of cytologically bland neoplasms of the stomach that can be encountered using endoscopic ultrasound-guided FNA biopsy.

1. Introduction

Endometrial stromal sarcoma (ESS), an uncommon/rare mesenchymal neoplasm of the uterus, is commonly quoted as representing only 0.2% of all uterine malignancies [1]. Its occurrence at extra-uterine sites is even more exceptional. Primary extra-uterine endometrial stromal sarcoma (EESS), also referred to as "*endometrioid* stromal sarcoma" is diagnostically formidable for a variety of reasons including its extra-uterine location, atypical clinical presentation, variable histology, and lack of a specific immunohistochemical (IHC) profile. Principal extra-uterine sites of EESS include the ovary, bowel wall, abdomen, peritoneum, pelvis, and vagina [2-4]. The cytopathology of EESS is rarely described in the literature. We hereby present the cytologic-pathologic correlation of two examples of primary gastric EESS. No evidence of a primary uterine ESS was identified in either patient. To our knowledge, only one prior instance of EESS arising in the stomach is reported in the English literature [5].

2. Case report

2.1. Case 1

A 37-year-old woman presented for workup of an inguinal hernia. Computed tomographic scan demonstrated an asymptomatic mass in the lesser curvature of the stomach. Esophagogastroduodenoscopy and endoscopic ultrasound (EUS) confirmed a 4 cm. submucosal mass at the gastric antrum. EUS-guided fine needle aspiration (FNA) biopsy performed at an outside hospital issued a diagnosis of a low-grade neuroendocrine tumor despite negative staining of tumor cells for cytokeratin AE1/3, synaptophysin, and chromogranin on a cell-block. After transfer to our medical center a review of the smears and additional stains led to a diagnosis of "low-grade mesenchymal tumor" from the cytologic specimen. Subsequent laparoscopic gastric wedge resection was performed.

It was subsequently discovered that the patient underwent vaginal hysterectomy 6 years earlier for repeatedly abnormal pap smears. Slides from that hysterectomy (obtained from an outside hospital) were reviewed and showed no evidence of a uterine lesion.

2.2. Case 2

A 54-year-old woman presented with bloating, gas, and increased frequency of stool that worsened with stress and anxiety that was attributed to irritable bowel syndrome. A gastric mass was identified with esophagogastroduodenoscopy. Gastric and duodenal biopsies were negative. EUS found a 1.7-cm nodule arising in the gastric fundus that led to an EUS-guided FNA biopsy. A cytologic diagnosis of "spindle cell neoplasm, favor gastrointestinal stromal tumor" was made. Subsequent gastric wedge resection was performed.

Upon follow-up, a 2-cm mass in the left uterine wall was diagnosed clinically as a leiomyoma. Gynecologic organs were otherwise unremarkable. Endometrial biopsy showed only disordered proliferative endometrium.

3. Pathology

3.1. Case 1

Cytologic smears were only modestly cellular and arranged primarily in loose clusters. Uniformly small, bland cells had rounded-

* Corresponding author. Department of Pathology, Wexner Medical Center @, The Ohio State University College of Medicine, 405 Doan Hall, 410 West 10th Avenue, Columbus, Ohio 43210. Tel.: +1 614 293 9232; fax: +1 614 293 7626.
E-mail address: paul.wakely@osumc.edu (P.E. Wakely).

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ovoid nuclei, evenly dispersed chromatin, no or inconspicuous nucleoli and scant to moderate cytoplasm with indistinct cell borders (Fig. 1 A and B). Some cell clusters were accompanied by short capillary segments. Many areas had a slightly myxoid background. Additional IHC staining of the cell-block by us demonstrated positivity with vimentin and CD56, and negative staining with smooth muscle myosin heavy chain, desmin, CD34, CD117, DOG-1, S-100, smooth muscle actin, muscle specific actin HHF-35, and h-caldesmon.

A $3.4 \times 2.7 \times 1.7$ cm irregular, red-brown, subserosal gastric mass was found upon resection. The mass extended from the muscularis propria into the submucosa and displayed a densely cellular proliferation of monotonous small cells with ovoid to spindle nuclei and scant cytoplasm having a smooth "pushing" as well as a slightly jagged infiltrative border. Variable architectural patterns included solid, pseudoangiomatous, reticular, myxoid, and epithelioid features such as sex-cord like foci. The latter were in cords, trabeculae, and nests (Fig. 1C-F). Multiple foci of lymphovascular invasion were present. There was no evidence of endometriosis. IHC performed on the resected specimen showed diffuse expression of vimentin, CD10, and β-catenin (nuclear), while stains for cytokeratin AE1/3, CAM 5.2, epithelial membrane antigen (EMA), estrogen receptor (ER), progesterone receptor (PR), inhibin, smooth muscle actin (SMA), desmin, h-caldesmon, calponin, CD34, CD117, DOG-1, S-100, ERG, synaptophysin, and chromogranin were negative. Flourescence in situ hybridization analysis showed no rearrangement of the *JAZF1*, *PHF1*, or *YWHAE* gene regions. The locus and probes (break apart probe) used were 7p15 (3′JAZF1, 5′JAZF1), 6p21.32 (5′PHF1, 3′PHF1), and 17p13.3 (3′YWAE, 5′YWAE) correspondingly. A final diagnosis of EESS was made.

3.2. Case 2

Smears were similar to those of case 1 except that syncytial cell clusters were larger, more numerous, and had a second population of dispersed single cells. Similar to case #1, cells were uniformly-sized with rounded, ovoid to spindled nuclei, inconspicuous to absent nucleoli, occasional nuclear grooves and slight irregularity. Cells had a minimal amount of non-vacuolated cytoplasm with ill-defined cell borders (Fig. 2A-C). No extracellular matrix material was present. Small lymphocytes were interspersed throughout. IHC performed on the cell-block was negative for cytokeratin AE1/3, CAM 5.2, synaptophysin, chromogranin, and CD117.

Resection showed a 1.5 cm well circumscribed gastric mass with a tan, partially necrotic cut surface. Overlying mucosa and serosa were not fixed to the mass. A fascicular, and storiform pattern of proliferating ovoid-spindled cells punctuated by small spiral arterioles with about one mitoses per 10 high power field was seen. Foci of

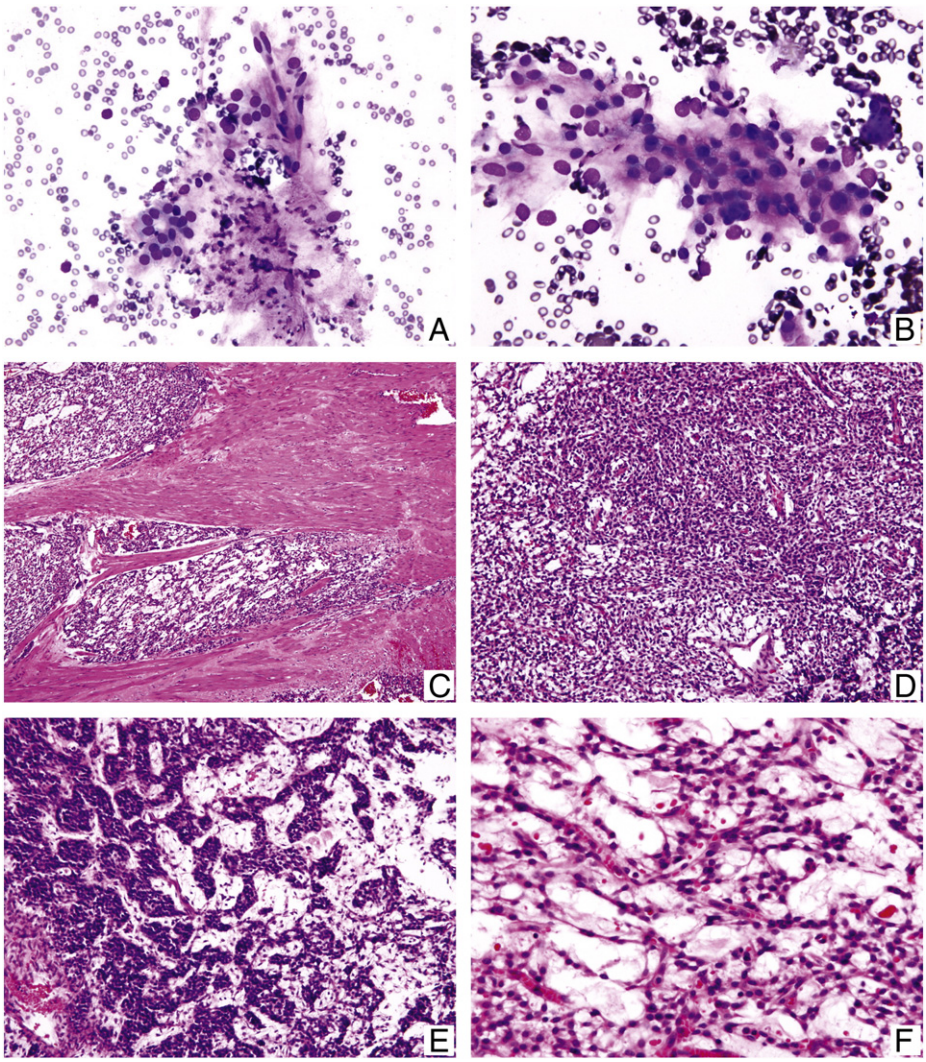

Fig. 1. Case 1. (A and B) Small syncytial clusters of loosely aggregated isomorphic cells are set in a sparse fibromyxoid stroma with a "clean" smear background. Romanowsky stain. Resection specimen shows large nests of tumor cells infiltrating the adjacent gastric muscularis propria (C) with a variety of patterns including solid (D), sex-cord like (E), and pseudoangiomatous (F). Hematoxylin and eosin stain.