# A rare case of low-grade endometrial stromal sarcoma with myxoid differentiation and atypical bizarre cells

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**Summary**

Endometrial stromal sarcoma (ESS) is a rare mesenchymal tumor with characteristic histological appearances, consisting of diffuse infiltrate of small uniform endometrial stromal cells with a multinodular arrangement and distinct vascular pattern. Less common variants of ESS include "mixed endometrial stromal and smooth muscle tumors", "endometrial stromal tumors resembling ovarian sex cord tumors" and "endometrial stromal neoplasms with endometrial glands", and "aggressive endometriosis". Rarely do endometrial stromal tumors have a prominent fibrous or myxoid appearance which causes confusion and possible misdiagnosis as myxoid leiomyosarcoma.

In this report we present a very unusual subtype of ESS in a 32-year-old woman. The tumor revealed atypical pleomorphic bizarre cells which were stained positive only with vimentin and CD10 in an abundant myxoid matrix. A low-proliferative rate was established with MIB-1 staining. To our knowledge such appearance has not been previously reported.

*Key words:* Uterus, Endometrial Stromal Sarcoma, Myxoid change, Bizarre cells.

## Introduction

Endometrial stromal tumors account for 2-4% of uterine malignancies [1]. Histologically they are classified as endometrial stromal nodules, endometrial stromal sarcoma (ESS) and undifferentiated stromal sarcoma. ESS is the most frequent form, characterized by tumor cells which closely resemble endometrial stromal cells of the proliferative phase [2].

Less common variants of ESS include those which contain smooth muscle differentiation – "mixed endometrial stromal and smooth muscle tumors", and those associated with epithelial structures [3, 4]. The most common epithelial patterns resemble those seen in ovarian sex-cord stromal tumors – "endometrial stromal tumors resembling ovarian sex cord tumors" [5]. Endometrial stromal tumors rarely have a prominent fibrous or myxoid appearance which causes confusion and possible misdiagnosis as myxoid leiomyosarcoma and those showing osteoclast-type giant cells [6-9]. In the diagnosis of ESS, the number of mitotic fugures per 10 HPF should not exceed 10. This criterion is important in making the differential diagnosis with undifferentiated stromal sarcoma.

We present a case of ESS with abundant myxoid stroma and atypical pleomorphic bizarre cells.

## Case Report

A 32-year-old woman was admitted to the hospital with abnormal uterine bleeding of six months duration. She had been treated with progestin for the previous two months. Pelvic computed tomography (CT) revealed a well circumscribed mass, 8.5 cm in diameter, arising from the uterine corpus and resembling a fibroid (Figure 1). Dilatation and curettage was performed. Histopathological examination of the curettage material revealed the presence of atypical bizarre cells with pleomorphic, hyperchromatic nuclei, large prominent nucleoli and abundant cytoplasm, some of which had long cytoplasmic processes. These were set in a fibromyxoid matrix among mucosal fragments of proliferative endometrium (Figure 2). On histological ground alone, the possibility of a pseudosarcomatous lesion was entertained and excision of the mass was suggested to rule out malignancy. The excised fibroid-like circumscribed mass was 8.5 cm in diameter with fish-meat like softening of the cut surfaces. The same histological features were observed in the excisional biopsy. The tumor exhibited less than three to four mitotic figures per 10 HPF. There was focal invasion into the myometrium (Figure 3). Immunohistochemically tumor cells reacted for CD10 and vimentin, but there was no reaction for smooth muscle actin, cytokeratin 7 and low molecular weight cytokeratin. With these findings, our diagnosis was ESS. Since the surgical borders were focally positive with neoplastic cells, the patient underwent total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy and appendectomy. No residual tumor was seen in the hysterectomy material. The patient had no evident recurrence or metastatic disease 11 months after the operation.

## Discussion

Endometrial stromal sarcoma accounts for 2-4% of all uterine corpus malignancies [1]. The average age for ESS is 40-49 and it is generally diagnosed during the premenopausal period, but has also been reported in young women and girls [10]. In our case the patient was 32 years old. In most cases, the presenting symptom is

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Fig. 1

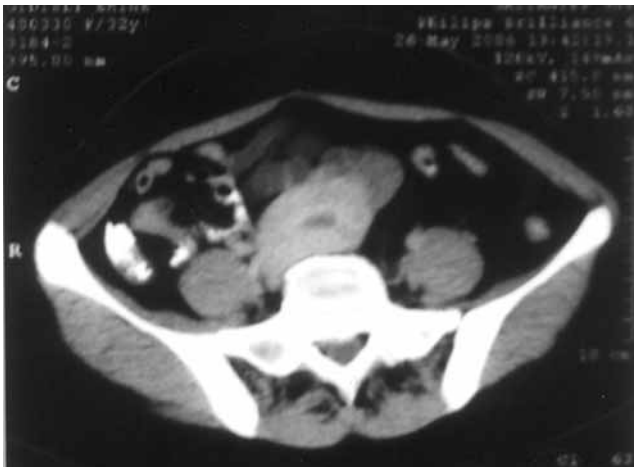

Fig. 2

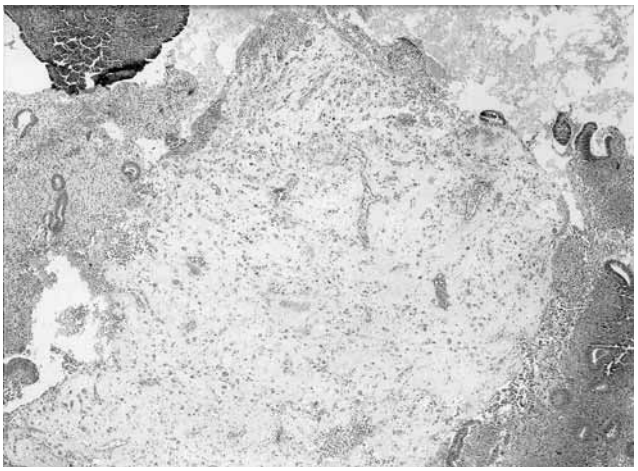

Fig. 3

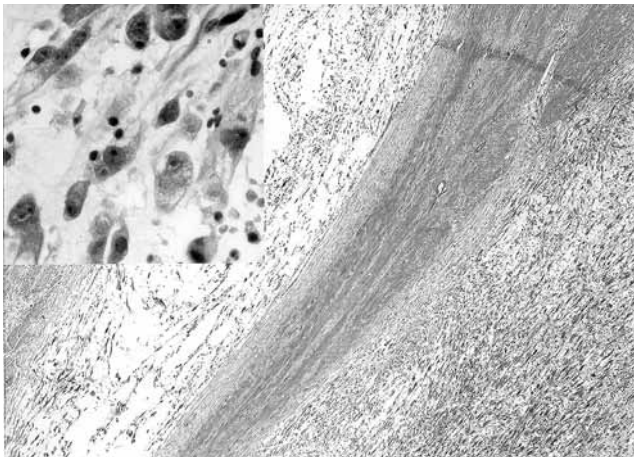

Figure 1. — Pelvic CT showed a well defined contour solid mass arising from the uterine corpus.

Figure 2. — Tumor tissue with prominent myxoid stroma and proliferative endometrial tissue fragments in curettage material (H&E x 40).

Figure 3. — Infiltration of tumor into the myometrium (H&E x 40). Inlet: Atypical bizarre cells with pleomorphic, hyperchromatic nuclei, large prominent nucleoli, and oval to spindle-shaped abundant cytoplasm in a fibromyxoid matrix (H&E x 200).

abnormal uterine bleeding, uterine enlargement or pelvic pain. The usual preoperative diagnosis is uterine leiomyoma [10]. Diffuse CD10 immunoreactivity is common in endometrial stromal tumors, as it was in our case. CD 10 positivity and smooth muscle actin negativity are important to distinguish ESS from myxoid leiomyosarcoma.

Like our case, ESS with abundant myxoid stroma has been reported but the association with large bizarre epithelial-like cells has not, to our knowledge, been described [6, 7].

We have presented an extremely rare example of low-grade ESS including both fibromyxoid stroma and atypical bizarre tumor cells. Such myxoid ESS with bizarre cells may create diagnostic difficulties, especially in curetting material. It may be mistaken as myxoid leiomyoma, myxoid leiomyosarcoma, inflammatory myofibroblastic tumor and myxoid malignant fibrous histiocytoma. Clinicopathological and immunohistochemical features may be helpful in the differential diagnosis.

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