

<u>TITLE</u>	A Rare Case of Stromal Sarcoma of the Endometrium with Sex-Cord-Like Differentiation
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<u>KEYWORDS</u>	Uterus, endometrial stromal sarcoma, sex-cord-like elements, glandular elements
<u>DIAGNOSIS</u>	High-Grade Endometrial Stromal Sarcoma With Sex-Cord-Like Differentiation
<u>SUMMARY</u>	<p>A grey firm uterine tumor of 8 cm in diameter was detected in a 35 y old patient. Histology revealed a biphasic growth pattern with glandular or epitheloid and sex-cord-like elements as well as a spindle cell component. The spindle cells resemble to a certain degree stromal cells of the endometrium. More than 10 mitoses/HPF were counted. IH disclosed a strong expression of vimentin and only focal positivity for CD99 and smooth muscle actin, no cytokeratin.</p> <p>In summary, the diagnosis of a rare case of a high-grade endometrial stromal sarcoma with sex-cord-like differentiation was stated.</p>
<u>CASE REPORT</u>	<p><i>Clinical Record</i> 35 years old female with spontaneous vaginal bleeding from the uterus. Manual examination revealed a deformation of the uterus due to a big firm tumor. Sonography raised the suspicion of tumorous growth beyond the organ. No other clinical symptoms. No elevated tumor associated antigens.</p> <p><i>Surgeon´s Report</i> The tumorous mass was confined to the uterus. No spreading in the abdominal cavity, no visible metastases in the liver, peritoneum or other organs. No lymph node enlargement. The lesion could be removed in toto.</p>

Macroscopy

During operation an uterus mass was found, 8 cm in diameter with subtotal infiltration of the myometrium ([Fig. 1](#)). The uterine surface was not infiltrated. In the cavum there were rests of coagulated blood. The sectioned surface showed a predominantly solid lesion with hemorrhagic necrosis. The color of the solid tissue varied from grey to white. The consistency was mainly firm.

Histology

The tumor showed two different growth pattern ([Fig. 2](#)), a spindle cell and a more glandular or sex cord-like differentiation. Both were directly adjacent to each other.

The spindle cell component consisted of small partly elongated, partly ovoid or round tumor cells resembling endometrial stromal cells. The cytoplasm appeared inconspicuous, sometimes relatively pale ([Fig. 3](#)) while in other parts the cells were smaller with a more intensely stained cytoplasm (Fig. 3 insert). The nuclei differed from small and round to large hyperchromatic and polymorphic.

The second component showed an epithloid appearance ([Fig. 4](#)) with a major part of sex-cord-like differentiation ([Fig. 5](#)) and a minor part of pseudo-glandular growth pattern ([Fig. 6](#)). The round or polygonal cells were arranged in tubules and cords roughly simulating granulosa cell elements.

The mitoses in both components were found to be between 5 up to 15 per high power field.

Immunohistochemistry (IH)

In IH different growth pattern appeared even more characteristic when vimentin is disclosed ([Fig. 7, 8 and 9](#)). A focal positivity of the smooth muscle antigen was found ([Fig. 10](#)). MIB1 was detectable in up to 50% of the tumor cells indicating a high proliferative activity ([Fig. 11](#)).The IH results are summarized in the following table.

CD 99 +	(focally)	CK 8, 18, 19	-
actin +	(focally)	CA 125	-
Inhibin -		CK 7	-
Vimentin +		CEA	-
S 100 -		EMA	-

MIB 1: positive in 25 - 50% of the tumor cells

DISCUSSION

The most relevant differential diagnosis was that between a low grade vs. a high grade endometrial stromal sarcoma (ESS). The criteria [1] to distinguish one from the other are:

Imperative Criteria:

- 10 or more than 10 mitotic figures per 10 HPF are indicative for a high grade sarcoma even if otherwise the histological features are identical.
- Tumor cells must resemble endometrial stromal cells.

Optional Criteria:

- Higher degree of cellular atypia in high grade sarcoma, however, no correlation with clinical outcome was observed [2].
- Invasive periphery, in particular angio-invasive.
- Nuclear pleomorphism and hyperchromasia.
- Vascular channels, irregularly distributed.
- Huge cells with multiple nuclei.
- Only foci of myogenic differentiation.
- Heterologous elements (chondro, rhabdo etc.) may be present.
- Necrosis.

Following these criteria a high grade ESS was presumed although in high-grade ESS almost never foci of sex-cord-like or glandular differentiation are seen. In a study of Silverberg and Kurman [2] only in 2 of 21 high grade ESS such elements were observed.

Further differential diagnoses were considered:

- other types of sarcomas, e.g. smooth muscle sarcoma, fibrosarcoma, angiosarcoma, neurogenic sarcoma etc. – all excluded by ICH
- endometrial stromal nodule – too many mitoses, polymorphism too strong
- carcinosarcoma – no cytokeratin expression
- metastatic carcinoma – no cytokeratin expression

After exclusion of the other possibilities and taking together the morphologic and immunophenotypic features the final diagnosis of a rare case of an high grade endometrial stromal sarcoma with sex-cord-like differentiation was set.

REFERENCES

1. Norris HJ, Taylor HP (1966) Mesenchymal tumors of the uterus: A clinical and pathological study of 53 endometrial stromal tumors. *Cancer* 755-766
2. Silverberg SG, Kurman RJ (1991) Tumors of the uterine corpus and gestational trophoblastic disease. *Atlas of Tumor Pathology, AFIP, 3rd Series, fascicle 3*

Fig. 1: Uterine Stromal Sarcoma



Fig. 2: Malignant Tumor of the Endometrium

35 y.
old
female

HE
x 200

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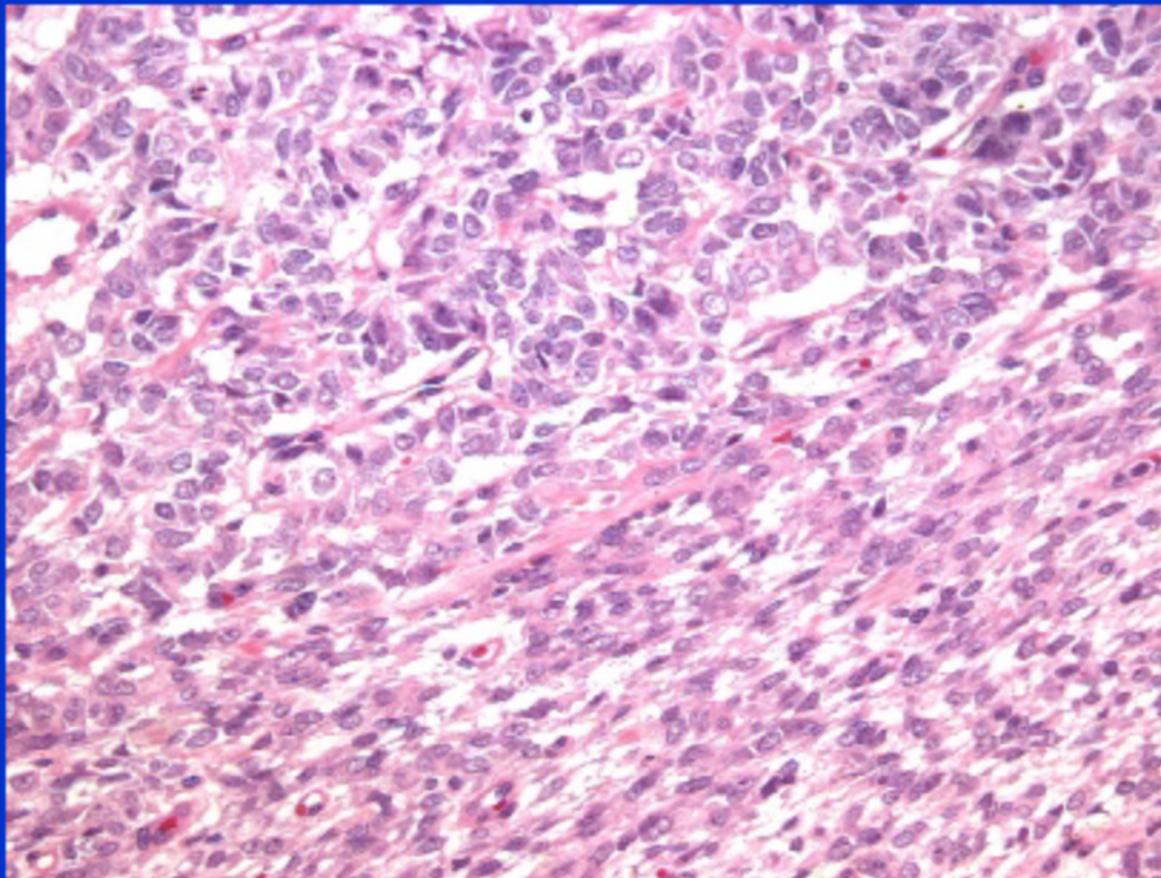


Fig 3: Malignant Tumor of the Endometrium

35 y.
old
female

HE
x 200

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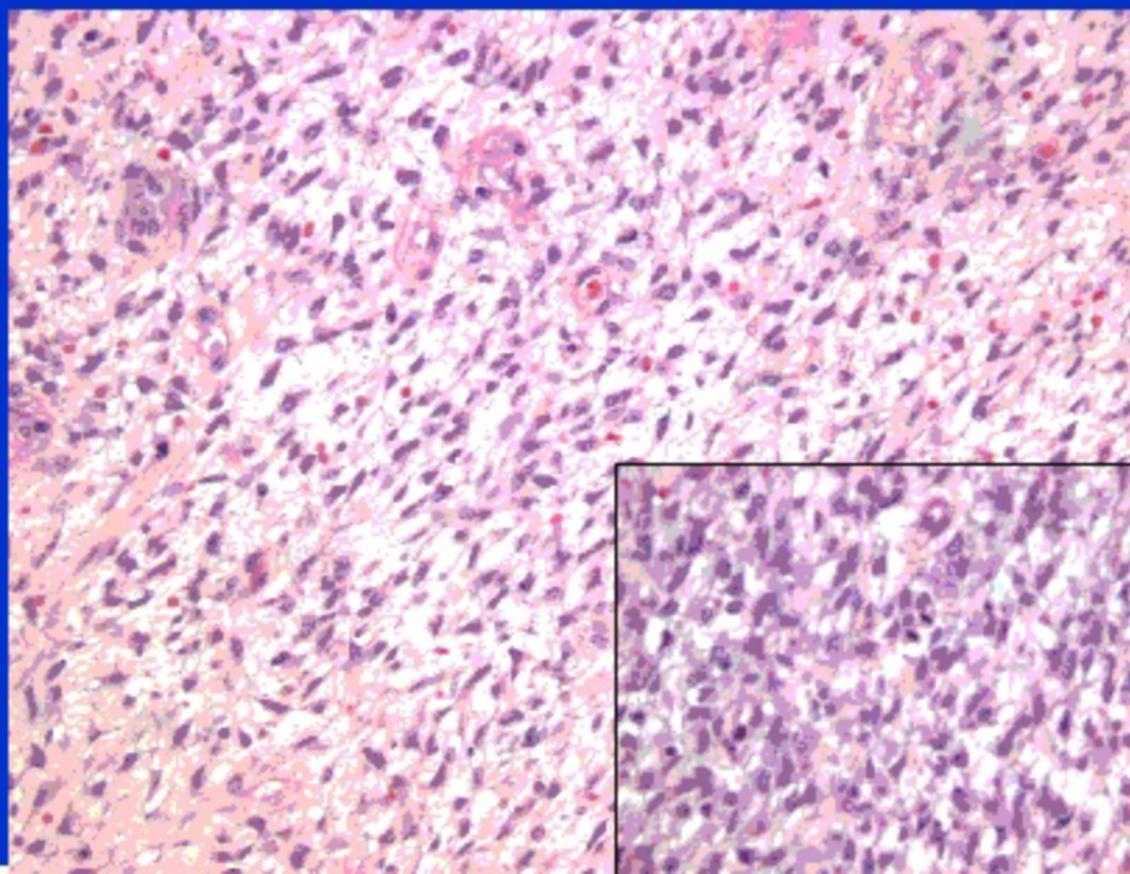


Fig. 4: Malignant Tumor of the Endometrium

35 y. old
female

H&E
x 200

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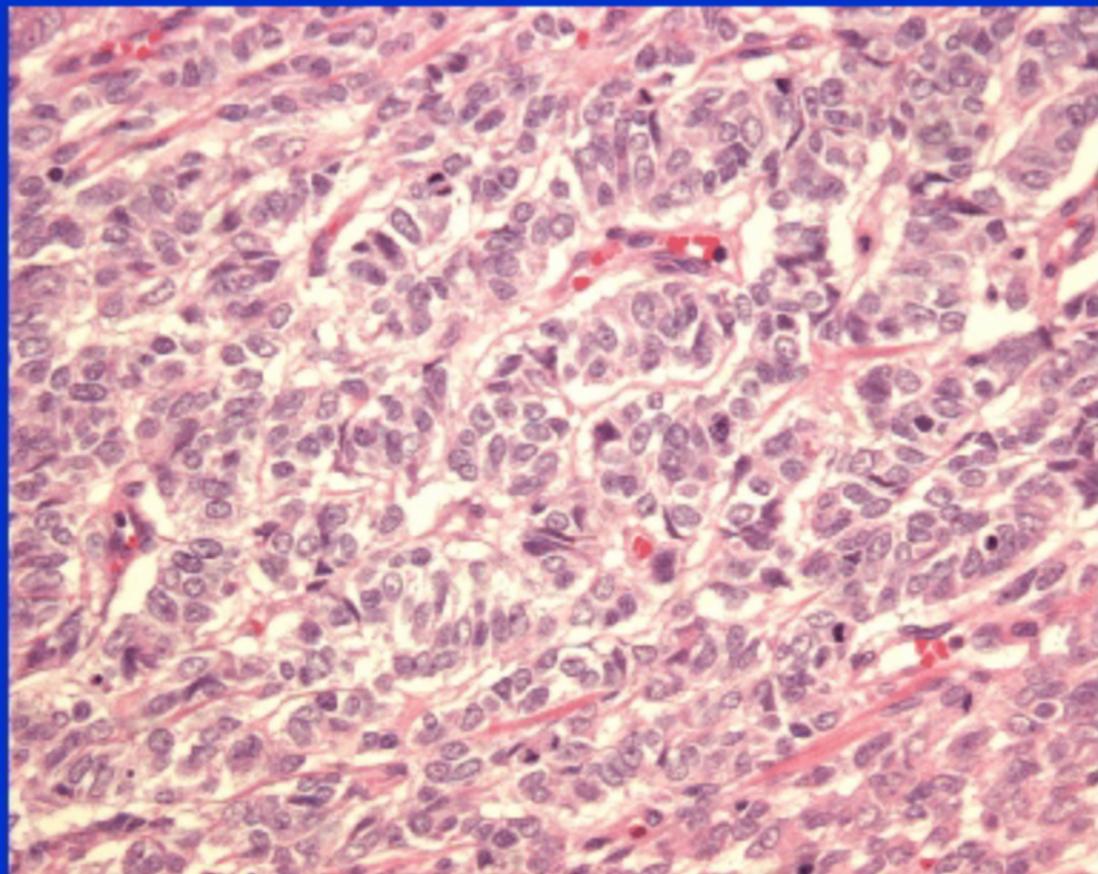


Fig. 5: Malignant Tumor of the Endometrium

35 y. old
female

HE
x 200

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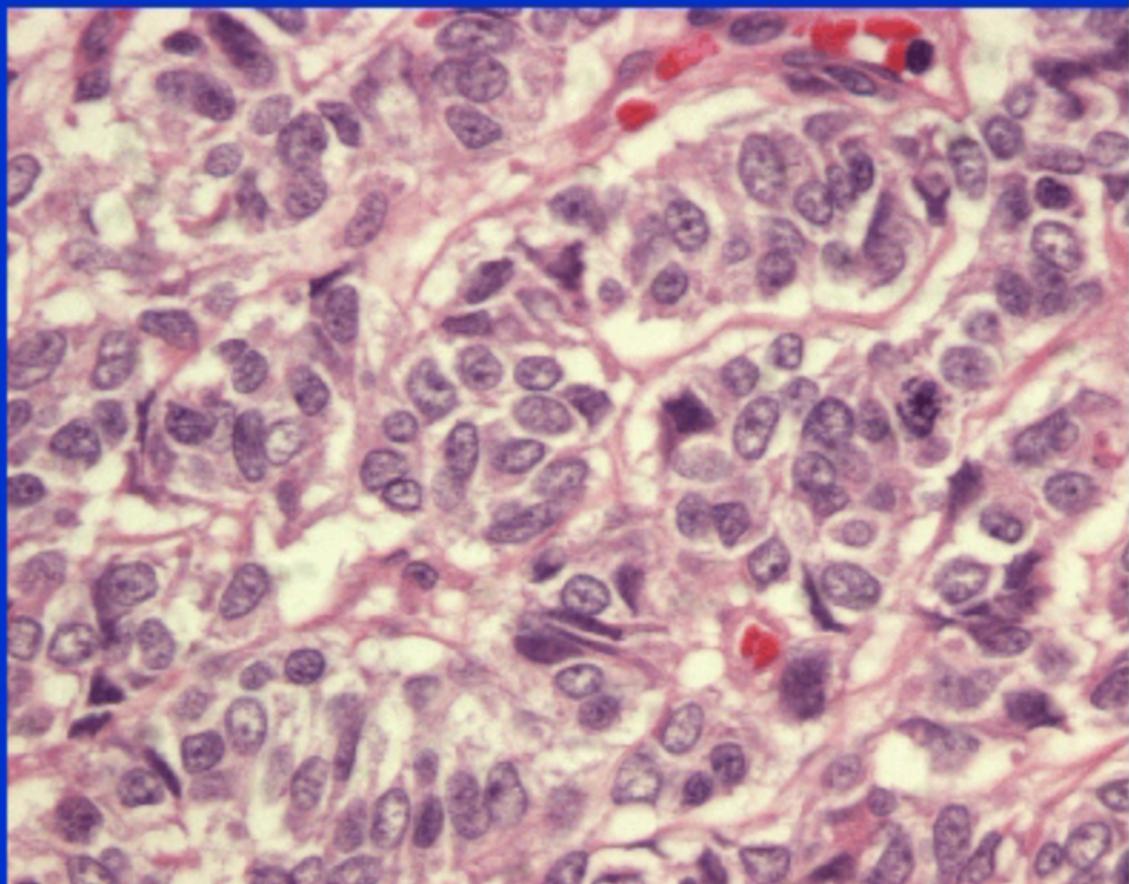


Fig. 6: Malignant Tumor of the Endometrium

35 y. old
female

H&E
x 400

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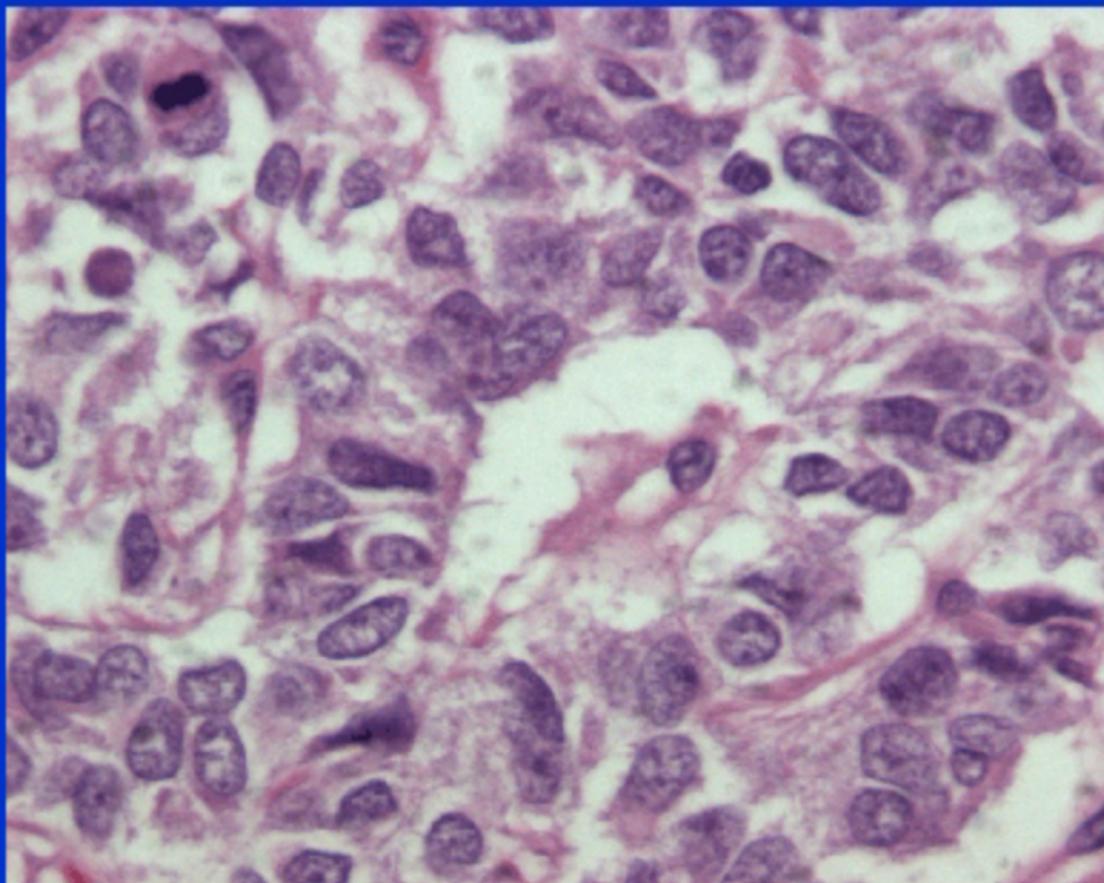


Fig. 7: Malignant Tumor of the Endometrium

35 y.
old
female

vim
x 200

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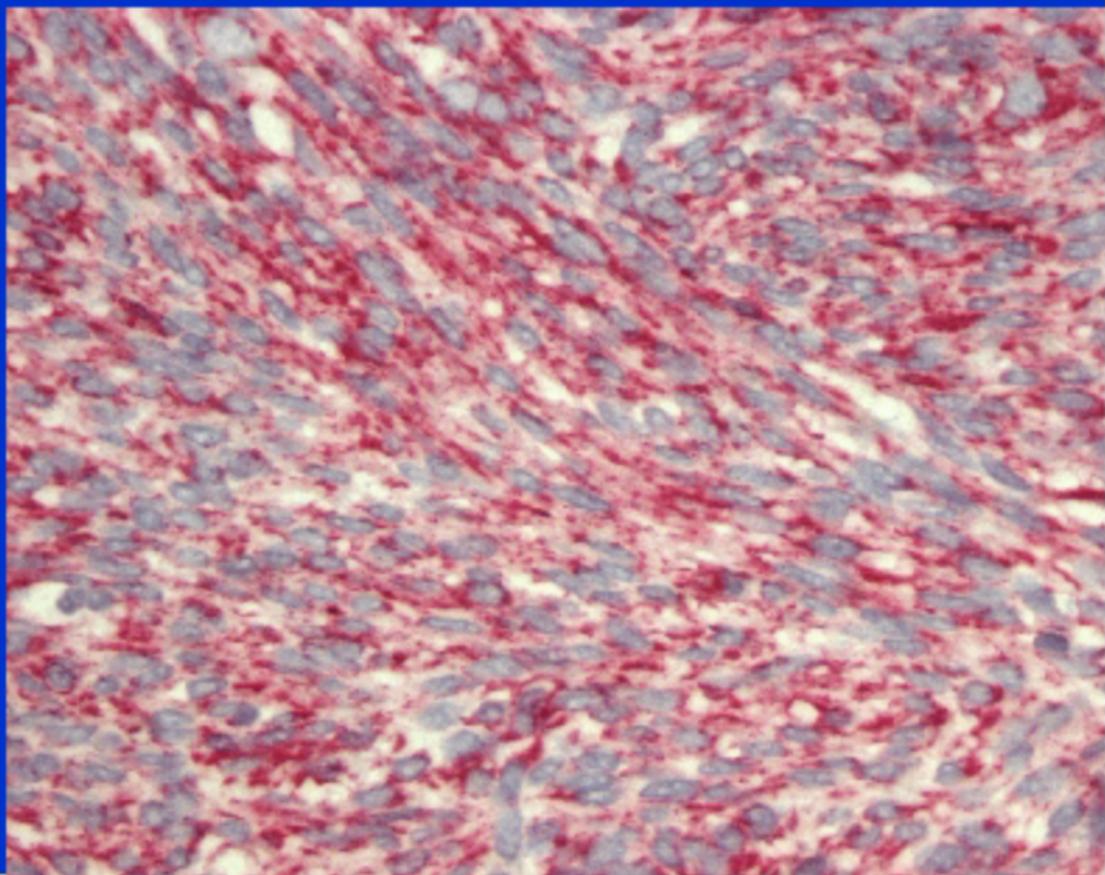


Fig 8: Malignant Tumor of the Endometrium

35 y.
old
female

vim
x 200

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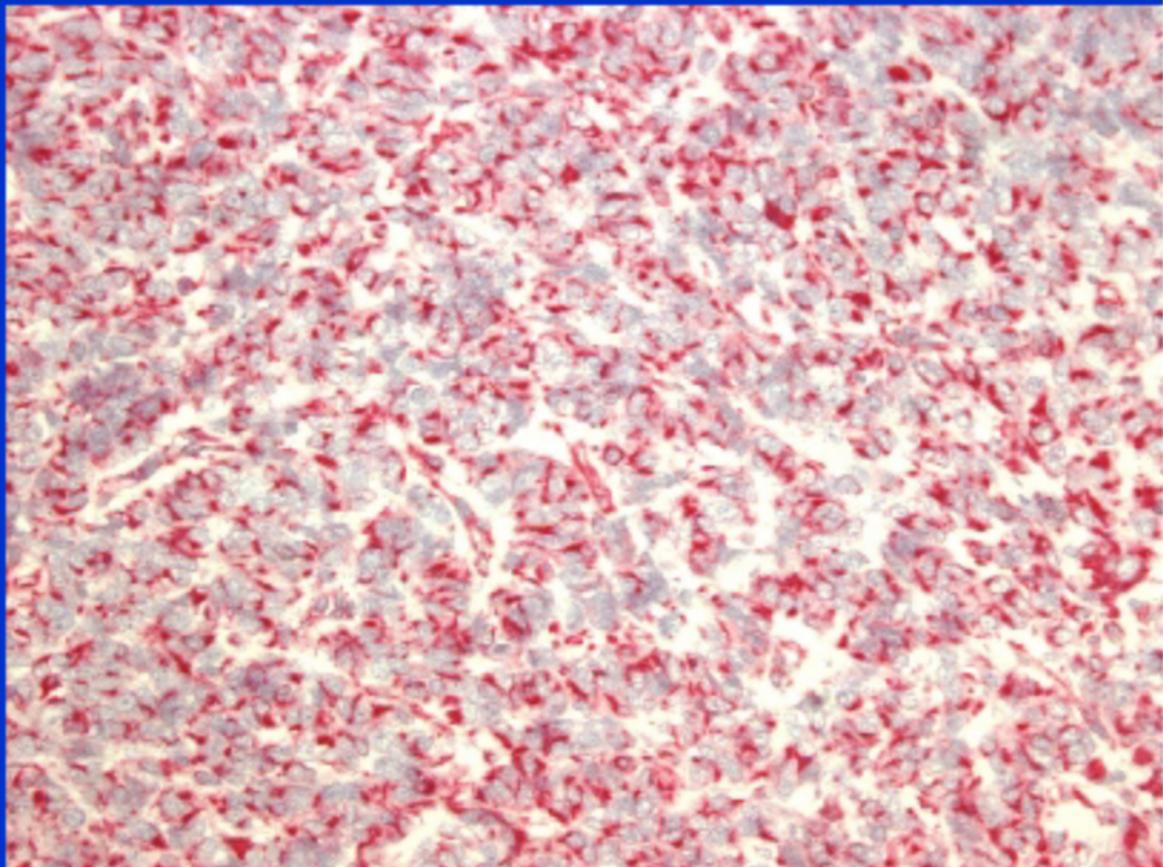


Fig. 9: Malignant Tumor of the Endometrium

35 y.
old
female

vim

x 200

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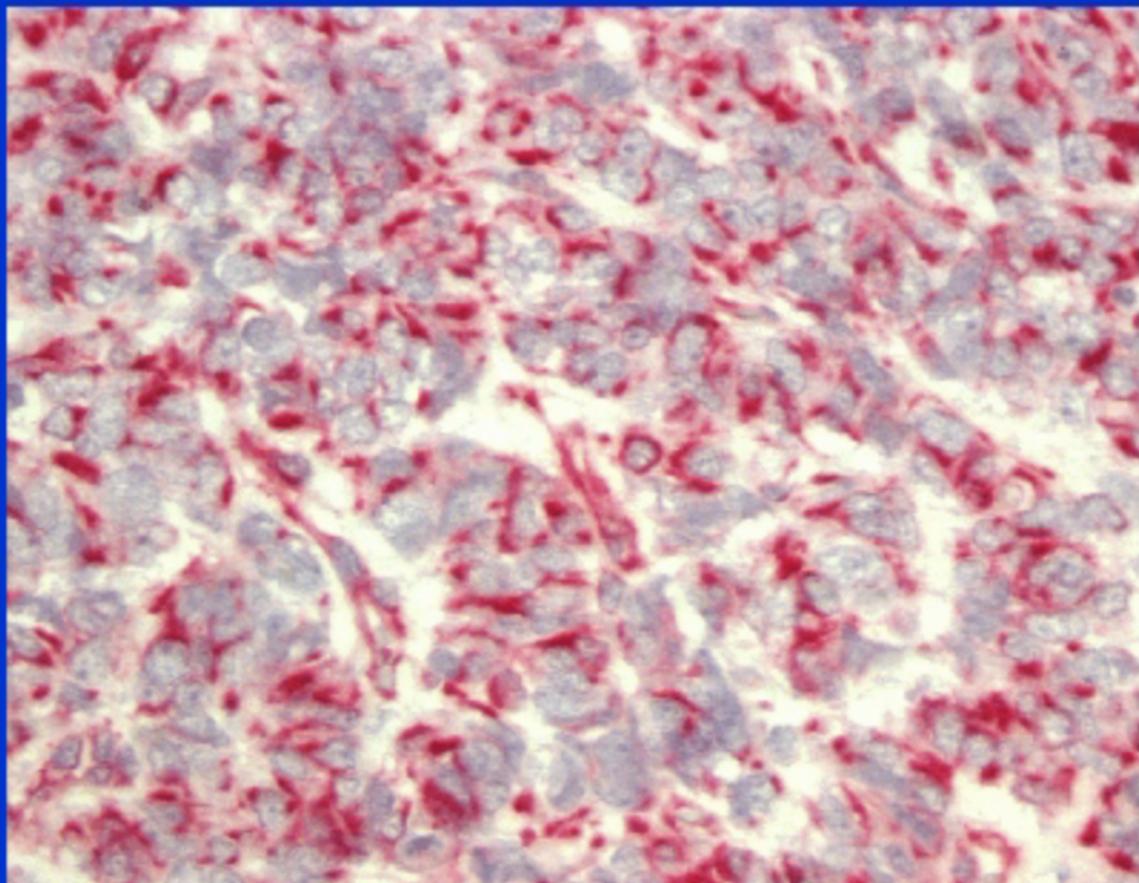
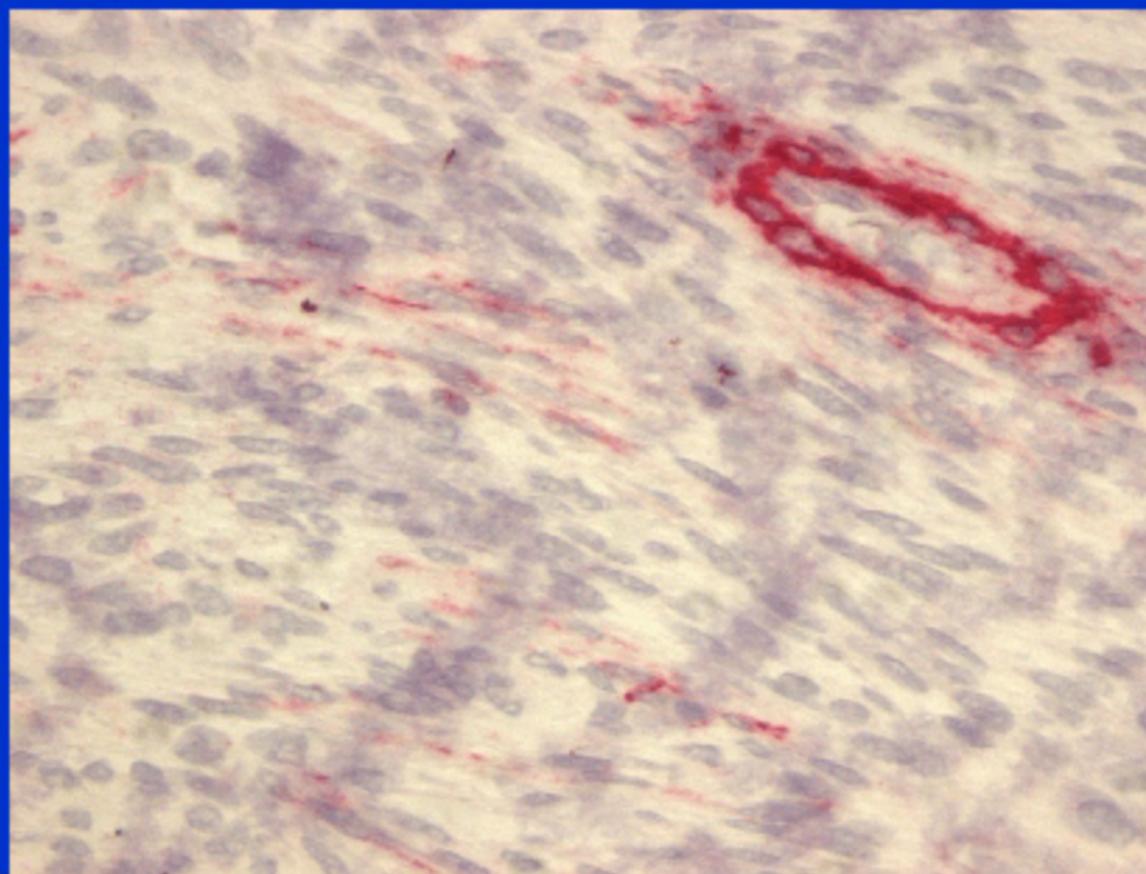


Fig. 10: Malignant Tumor of the Endometrium



35 y. old
female

smooth
muscle

x 200

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Fig 11: Malignant Tumor of the Endometrium

35 y. old
female

MIB1

x 200

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