Case report

Endometrial stromal sarcoma presenting with metastatic deposits in the retroperitoneum and mesocolon

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Introduction

Endometrial stromal sarcoma is a rare, low grade tumour of the uterus. It has the propensity for local recurrence and distant metastases many years after hysterectomy for the original tumour. Endometrial stromal sarcomas pose a diagnostic challenge when present in an extrauterine site. This is case of a patient presenting with metastatic deposits of an endometrial stromal sarcoma occurring in the retroperitoneum and mesocolon 16 years after the diagnosis of the primary tumour.

Case History

A 62 year old lady presented with acute onset left loin pain and was diagnosed to have a urinary tract infection. An ultra sound scan of the abdomen revealed a left flank mass in line with the mid ureter causing hydronephrosis and hydroureter. Computerised Tomography (CT scan) of the abdomen revealed the presence of two contrast enhancing masses: one a hyperdense mass with central hypodensity in the

midline anteriorly and the other a large mass posterolaterally situated to the left of the aorta and inferior to the left kidney (Fig.1).

Fig.1. CT scan of the abdomen: Two contrast enhancing masses: a hyperdense mass with central hypodensity in the midline anteriorly (A) and a large posterolateral mass situated inferior to the left kidney (B).

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A vascular retroperitoneal tumour and a tumour attached to the mesocolon were found at laparotomy. Both masses were resected.

Macroscopically both tumours had a similar appearance. The outer surface was smooth and bosselated. The cut surface had a solid tan colored, homogenous appearance with a vague nodularity. Histological examination revealed the presence of a cellular tumour with a haemangiopericytomatous vascular pattern

(Fig 2a). The tumour was composed of uniform predominantly oval small cells arranged concentrically around blood vessels. The tumour cells showed cytoplasmic positivity for CD10 (Fig 2b) and strong nuclear positivity for oestrogen receptors (Fig 2c), progesterone receptors and bcl-2. The tumour cells were negative for CD34 (Fig 2d), CD 99 and cytokeratin. A diagnosis of a low grade endometrial stromal sarcoma was made.

Fig.2. 2a) Histological appearance of the tumour: A cellular tumour with a haemangiopericytomatous vascular pattern (H&E x400) 2b) Cytoplasmic positivity for CD10 (CD 10 x400) 2c) Strong nuclear positivity for estrogen receptors (ER x400), 2d) Negativity for CD34 (CD 34x400)

Discussion

Endometrial stromal sarcoma is a rare tumor of the uterus accounting for only 0.2% of all genital tract malignant neoplasms (1). It is composed of proliferating uniform oval to spindle shaped cells of endometrial stromal type that are supported by numerous thin walled arteriolar type vessels. Low grade endometrial stromal sarcoma is characterized by slow clinical progression, repeated local recurrences and occasional metastases (1).

Sites of metastases include the lung, intestinal wall, retroperitoneum, deep soft tissue of the extremities, liver, bone and brain (2-4). This patient presented with metastatic deposits in the retroperitoneum and mesocolon.

Endometrial stromal sarcoma may pose a diagnostic challenge when present in an extrauterine site. Given the clinical and radiological features in this patient our first differential diagnosis was of a primary retroperitoneal tumour with metastatic deposits in the mesocolon. Microscopy revealed a low grade neoplasm with a haemangiopericytomatous vascular pattern. Therefore, our differential diagnosis included a haemangiopericytoma, monophasic synovial sarcoma and solitary fibrous tumour.

This patient had been diagnosed as having a low grade stromal sarcoma of the uterus following hysterectomy performed for menorrhagia 16 years earlier at the age of 46 years. This vital piece of clinical information directed us towards the diagnosis of endometrial stromal sarcoma.

Endometrial stromal sarcoma presents with abnormal uterine bleeding in women in the perimenopausal age group (5). Late pelvic and abdominal recurrences and metastases can occur many years after the diagnosis of the primary tumour (1). In some patients the primary tumour may even have been misdiagnosed as a smooth muscle tumour and a high degree of suspicion is needed to come to a diagnosis.

Histological features that favor a diagnosis of endometrial stromal sarcoma include multinodularity seen on low power, the presence of spiral arteriole like vessels, and the deposition of coarse collagen fibers among the tumour cells (6). The majority of endometrial stromal sarcoma stain positive for CD10, oestrogen receptor, progesterone receptor and bcl-2 and are negative for CD34 as was demonstrated in this case. However, bcl-2 is not useful to distinguish endometrial stromal sarcoma from its mimics (7). Haemangiopericytoma, solitary fibrous tumor and synovial sarcoma do not express oestrogen receptors but may show at least focal nuclear staining with progesterone receptors and most cases of haemangiopericytoma and solitary fibrous tumor express CD34 and some demonstrate a degree of CD10 positivity (7). A small antibody panel of CD10, anti estrogen receptor and anti CD 34 has been recommended to distinguish endometrial stromal sarcoma from tumours with a predominant haemangiopericytomatous growth pattern (7).

Conclusion

Endometrial stromal sarcomas pose a diagnostic challenge when present in

an extrauterine site. The presence of a haemangiopericytomatous vascular pattern may result in diagnostic confusion with soft tissue tumours including haemangiopericytoma, synovial sarcoma and solitary fibrous tumour. A high degree of clinical suspicion, subtle histopathological features and judicious use of immunohistochemistry will aid in the diagnosis.

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