

Uterine Sarcoma; An Unusual Presentation of Endometrial Stroma Carcinoma

Khalid Ali, Fauzi Alhakmi, Ecem Mimoglu, Shagufta Rafiq, Samia Azad, Ibrahim Abbasher, Nageen Naseer and Raouf Sallam

BACKGROUND

Uterine sarcomas are a rare heterogeneous group of tumours accounting for approximately 1% of all female genital tract malignancies, with endometrial stromal sarcomas (ESS) accounting for only 0.2% of these [1]. The rarity of these tumours presents challenges for diagnosis, with cases often misdiagnosed as benign leiomyoma, consequently delaying necessary treatment.

CASE REPORT

A 49-year-old woman presents initially with menorrhagia. Her EUA, hysteroscopy, and D&C revealed no significant abnormalities. The endometrium appeared unremarkable and a Mirena was inserted. Her symptoms persisted and a pelvic USS revealed a 5cm fibroid. Ten months later, the patient presented again with heavy PV bleeding. An urgent pelvic US revealed a significant increase in the fibroid size, now measuring 10.3 x 11.3 x 10 cm. No other pelvic or abdominal abnormalities were noted. The patient agreed to start LHRHa injections for symptomatic management, however was ineffective and she re-presented three months later, still in significant pain. Her abdomen was distended with a large pelvi-abdominal mass reaching the level of the umbilicus. She was considered for an urgent laparotomy, total abdominal hysterectomy and bilateral salpingo-oophorectomy. Laparotomy revealed an enlarged uterus reaching the umbilical level, with multiple metastatic lesions covering the uterine surface, bowel, and omentum. The planned surgical intervention was abandoned, and biopsies were taken. CT-TAP revealed distant widespread metastases. Palliative treatment was decided following discussion with the MDT, patient, and immediate family.

DISCUSSION

This case highlights the importance of considering uterine sarcomas as a differential for a presentation of vaginal bleeding with a uterine mass. ESSs are very rare; only comprising 0.2% of all uterine malignancies and less than 10% of all epithelial uterine sarcomas [2]. Clinical factors tend to be poor differentiators between ESS and benign leiomyoma – the age of onset is fairly similar in both diseases and symptoms largely mimic one another [3,4]. Although the rarity of ESS makes it an unlikely differential, a recent literature review has estimated that 90% of women with ESS present initially with uterine bleeding, and 70% present with uterine enlargement [3]. Although only 25% of ESS cases are asymptomatic, roughly 30-50% present with extra-uterine spread [5]. The presence of large and rapidly growing leiomyomas should therefore prompt investigation for ESS, as it has notably similar symptoms [6].

The patient's symptoms were investigated with a pelvic ultrasound on this occasion. Ultrasound diagnosis of ESS is often unreliable as it may suggest an incorrect diagnosis of adenomyosis or leiomyoma, with MRI also producing similar results [3,6]. Thus, there can be substantial difficulty in distinguishing between ESS and leiomyoma pre-operatively, as it can mimic both the physical appearance and imaging characteristics of benign leiomyoma. It is unsurprising that this has led to 75% of women with ESS receiving an incorrect leiomyoma diagnosis [7].

Peri-operative diagnosis of ESS is also very difficult due to an appearance that can often mimic highly cellular leiomyomas, although the presence of large, thick-walled muscular vessels would make ESS the more likely differential [6]. In this patient's case, extensive metastases and haemorrhagic lesions were clear signs of malignancy. Although the patient had undergone D and C on initial presentation, this unfortunately also has poor diagnostic reliability due to the similarity between ESS and normal endometrium [7]. ESS is also likely to grow through the intramural sections of the uterus, making histopathological diagnosis even more challenging [6].

It appears that immunohistochemistry is a better approach to accurately diagnose ESS. There are several disease-specific markers, including CD10, inhibin, and oestrogen/progesterone receptor positivity [5,6]. These immunological markers can also be useful to differentiate between ESS and leiomyosarcoma. Immunohistochemistry of our patient was negative for all markers except Cyclin-D1, which is an unusual presentation for ESS.

CONCLUSION

ESS is a rare sub-type of uterine sarcoma, with poor consensus in literature on its treatment and prognosis. Data from large, randomised control trials is needed to enable better care for patients suffering from this disease. Although it is widely agreed that surgery is the best initial treatment, the role of adjuvant therapy is not well-described beyond some evidence for the use of hormonal therapies. The rarity of these tumours presents challenges for diagnosis, with cases often being misdiagnosed as benign leiomyoma and consequently delaying necessary treatment. Clinicians should have a low threshold for suspecting malignant disease and should be wary of the false confidence that can arise from a leiomyoma diagnosis.

REFERENCES

