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Case Report

Endometrial Stromal Sarcoma Mimicking Leiomyoma Uterus: A Rare Case Report

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ABSTRACT

Endometrial Stromal Sarcomas (ESSs) extremely very rare malignant tumors of all uterine sarcomas and occupies less than 10% of all uterine malignancies. As it mimicks uterine fibroid, it is very difficult to recognize clinically because they share common clinical features, that is why diagnosis is often made postoperatively after proper histological examination. ESSs histologically resemble stromal cells of proliferative phase of the menstrual cycle. We report a case of low-grade ESS presented clinically as a case of a uterine leiomyoma.

Keywords: Endometrial Stromal Sarcomas (ESSs), Leiomyoma uterus.

INTRODUCTION

Endometrial Stromal Sarcomas (ESSs) extremely very rare malignant tumors of all uterine sarcomas and occupies 10% but only 0.2% fall in uterine malignancies. [1] The World Health Organization (WHO) classifies endometrial stromal tumors as benign endometrial stromal nodule (ESN) and ESS. ESNs are termed benign, as they do not infiltrate myometrium. In contrast, ESSs are characterized by infiltration of myometrium. [2] Uterinesarcomas most often affect postmenopausal women. The pathogenesis of these lesions remains unknown. [1] It can be easily mistaken as leiomyoma. [1,3] As it is very difficult to recognize clinically because they share common clinical features, that is why diagnosis is often made postoperatively after proper histological examination. [3,4] Some uterine sarcomas are diagnosed during or after surgery for what is thought to be benign fibroid tumors. [4] ESSs histologically resemble stromal cells of proliferative phase of the menstrual cycle. [5,6] Characteristic pattern of this tumour is infiltration and network of branching small vessels resembling spiral arteries. [6,7] A low-grade ESS usually appears as an irregular yellowish mass involving endometrium or myometrium, possibly with focal necrosis, hemorrhage or cystic changes. [6] As these tumors have indolent growth there is chance of late recurrence and progestin therapy can reduce the risk of recurrence in adjuvant setting. [8] We report a case of low-grade ESS presented clinically as a case of a uterine leiomyoma.

CASE HISTORY

A 45 year old female attended Gynaecology outpatient department with the complaints of continuous bleeding per vagina since three months. On Ultrasonography, she was found to have a fibroid uterus, for which she was advised

surgery. Grossly, hysterectomy specimen with bilateral adnexa was overall measuring 15x 11x8.5 cm. External surface of uterus showed bossellations with areas of congestion. On cut section, uterus showed a large intramural mass which was greyish yellow in color measuring 9x7x6 cm and showed features of secondary degenerative changes. Endometrial canal was found pushed to the periphery by the mass. Cervical canal was shortened and measured 2cm in length.

On microscopy, cervix showed features of chronic cervicitis. Endometrium showed a tumor composed of uniform small round cells arranged in diffuse sheets, encircling the small blood vessels resembling spiral arterioles. Individual tumor cells were round to oval, having vesicular nucleus with scanty eosinophilic cytoplasm. Areas of hyalinization and frequent typical and atypical mitotic activity were seen. Tumor cells were seen infiltrating the adjacent myometrial smooth muscle fibers (Figure 1). Serosa was found free from tumor. Tumor emboli were seen involving both blood vessels and lymphatics. Hence, the microscopic features confirmed the diagnosis of Low Grade Endometrial stromal sarcoma.

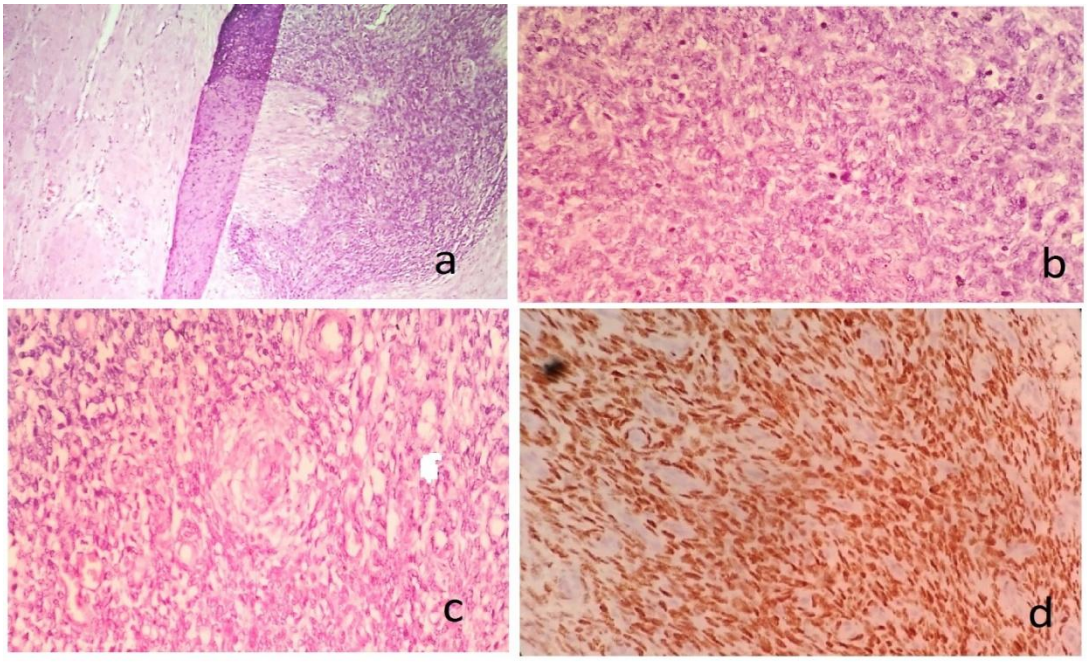

Figure 1: a) Uniform small round cells infiltrating the myometrium {H &E, X10}. b) Tumor cells with atypical mototic activity {H&E, X40}. c) Tumor cells encircling the small blood vessels resembling spiral arterioles {H&E, X40}. d) Tumor cells shows positivity for Estrogen receptor(ER) {IHC, X40}.

DISCUSSION

Endometrial Stromal Sarcomas (ESSs) are very rare malignant tumors of mesenchymal origin that constitute approximately 10-15% of all uterine sarcomas but only around 0.2% of all uterine malignancies. [1,2,9] It is second most common mesenchymal tumor of the uterus, accounting for 15%-26% of uterine sarcomas and seen commonly occurring in premenopausal women. [9]

As sign and symptoms such as abnormal bleeding, spotting, vaginal discharge, pelvic pain and mass presentations are also seen in benign leiomyoma, it is clinically difficult to diagnose LGESSs. [8,9] Also lesions such as leiomyoma, polypoid form of adenomyosis and Polyps of external cervical canal can clinically mimic LGESSs. [9] Due to its rarity and the lack of pathognomonic imaging findings, the preoperative differential diagnosis of LGESS is difficult; so many patients were preoperatively misdiagnosed as leiomyoma. [1,3,9]

Endometrial stromal nodules are well circumscribed grossly. Microscopically, Low grade ESS should have infiltrative tongues of tumor cells, low mitotic activity (<10 mitotic figures/10hpf) with mild cytological atypia and indolent behaviour. Undifferentiated endometrial sarcomas are pleomorphic with marked cytological atypia, high atypical mitotic activity (>10 mitotic figures/10hpf). [6] Management and treatment generally consists of total abdominal hysterectomy and bilateral salpingo-oophorectomy. [1,6] In the present case, clinical diagnosis was given as leiomyoma but gross finding gave us a clue of ESS with a yellow colour cut-surface which cannot be seen in case of leiomyoma.

Recent studies on receptors showed that LGESS express oestrogen (OR) and progesterone receptors (PRs), making this a potential target for treatment. Studies have shown success with the use of high-dose progestin therapy. At cytogenetic level a specific translocation t(7;17) (p15;q21) with involvement of two zinc finger genes juxtaposed with another zinc finger protein 1 and joint juxtaposed with another Zinc protein 1 was described in most of the ESS. There is a relation between chromosomal aberrations and endometrial sarcomas. Chromosomal deletion on 7p was the most common finding (55.6%) in ESS and may play a role in tumor development and progression. [9,10]

CONCLUSION

Since the prognosis of stromal sarcomas is worst when compared to benign myometrial lesions, a careful gross and microscopic examination should be done by pathologists so that the clinicians can give right mode of treatment.

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