

Low grade endometrial stromal sarcoma presenting as a cervical polyp in a young female: A rare case report

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ABSTRACT

Endometrial stromal sarcomas ESS are rare malignant mesenchymal tumors that usually develop in the uterus in perimenopausal women. The report describes a rare extra uterine low grade ESS presenting as a cervical polyp in a young female for which local tumor resection was performed. Histological examination revealed a spindle cell tumor. The diagnostic challenge was to differentiate ESS from cellular leiomyoma and other spindle cell tumors for which immunohistochemistry was carried out. The case report highlights that ESS is to be included in the differential diagnosis of spindle cell tumor in cervix even though rare. We also discuss the differential diagnosis of cervical polypoidal lesions.

Key words: Cervix, endometrial stromal sarcoma, extra uterine, young age

INTRODUCTION

Endometrial stromal sarcomas (ESS) are very rare tumors constituting to about 0.2% of all uterine malignancies.^[1] ESS most often occurs in perimenopausal women of age ranging between 42 and 55 years.^[1,2] ESS occurring in an extra uterine sites are rare, with primary low grade ESS occurring in cervix reported very rarely.^[3,4] To the best of our knowledge, our case is the third to be reported with ESS arising in cervix in a young female. The aim of the report is to highlight a case of low grade ESS with unusual location in an unusual age group, hence keeping in mind the ESS as a part of differential diagnosis, while reporting a case of spindle cell tumor of the cervix.

CASE REPORT

A 20-year-old unmarried girl patient presented with excessive bleeding per vagina of 1½ months duration.

Her menstrual cycle was regular. Pervaginal examination revealed a reddish brown polypoidal growth measuring 5 cm × 3 cm arising out of the cervical rim with an irregular surface and surface bleeding. A clinical diagnosis of infected leiomyomatous polyp was considered. Ultrasound examination revealed no abnormalities in the uterus. Polypectomy carried out and the specimen sent for histopathological examination. The girl gave history of similar polyp in the cervix 1½ year ago which was removed. However, the histopathology slides were not available for review. The patient is on follow-up to look for recurrence.

Gross appearance

Gross examination revealed a single irregular polypoidal mass measuring 4 cm × 2.5 cm × 2 cm with a stalk measuring 1.5 cm in length. The entire tissue was routinely processed, embedded in the paraffin blocks and sections stained with hematoxylin and eosin stain and immunohistochemistry subsequently.

Microscopic examination

Sections showed a well-circumscribed lesion composed of plump spindle shaped cells [Figure 1a] with mild atypia, elongated nuclei with eosinophilic cytoplasm and interspersed thin walled blood vessels [Figure 1b]. Two to three mitotic figures were noted per 10 high power fields. Focal ulceration, necrosis and granulation tissue were also seen. No glandular elements identified. Based on the

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histological findings the diagnosis of spindle cell tumor with possibility of smooth muscle tumor of unknown malignant potential or endometrial stromal tumor was considered. Immunohistochemistry was sent for further evaluation.

Immunohistochemistry showed CD10 to be strongly positive, [Figure 2] WT1 weakly positive, focal positivity of h-caldesmon, β -catenin, SMA and scattered desmin positivity. Cytokeratin (CK) and epithelial membrane antigen (EMA) were negative. A final diagnosis of low grade ESS was given.

DISCUSSION

Endometrial stromal tumors are rare uterine malignancies and low grade ESS accounts for 0.2% of all uterine malignancies.^[1] The tumor frequently occurs in women in the age group 40-55 years though there are few cases showing occurrence of ESS in a young women.^[1,2,5] The exact etiology of the tumor is not known but a few associations such as ovarian polycystic disease, estrogen use and tomosifen therapy have been shown.^[2] Most patients with ESS present with abnormal vaginal bleeding, pelvic pain and dysmenorrhea but about 25% of the patient are asymptomatic and are detected incidentally. About one-third of patients with ESS show recurrent local

disease.^[1,2] ESS is usually located in the uterus but has been reported in extra uterine sites such as ovary, fallopian tubes, omentum, retroperitoneum, vagina, vulva, sigmoid colon and round ligament.^[6] There are very few reported cases in the literature of ESS in the cervix. Boardman *et al.*, in their study have reported a case of low grade ESS occurring in ectocervix in a patient with carcinoma breast who was on hormonal therapy.^[3] Another low grade ESS case that is reported in the cervix was in 44-year-old female patient presenting with a hemorrhagic mass in the endocervix arising in a foci of endometriosis.^[4] All the reported cases of ESS in cervix have been in the perimenopausal age group and the present case occurring in a young girl is very unusual.

Cervical polyp, leiomyomatous polyp, endometrial polyp, sarcoma botryoides, mixed mullerian adenosarcoma and rarely ESS are some of the lesions which present as polypoidal lesions in cervix. Cervical polyps show endocervical glands and thick walled blood vessels while an endometrial polyp will show endometrial glands and thick walled vessels. Mixed mullerian tumors are typically seen in older age group and show glands. Our case had spindle cell morphology and did not show any glandular elements as confirmed by CK and EMA negativity. Some of the tumors of cervix with spindle cell component include leiomyoma, rhabdomyosarcoma and ESS. Rhabdomyosarcoma occurs in younger age group and the absence of Cambium layer, rhabdomyoblasts and myxoid stroma ruled out sarcoma botryoides. The closest differential diagnosis of ESS is cellular leiomyoma, however strong positivity of CD10 favors ESS.^[7,8] Diffuse positivity of WT1 positivity is characteristic in ESS^[9] and is positive in our case, but may be weakly positive in leiomyoma also.^[10] Though h-caldesmon is characteristic of smooth muscle tumors, the focal positivity in our case could be due to smooth muscle differentiation.^[8] This is probably why desmin, SMA and β -catenin, which are smooth muscle markers were also weakly positive.^[8] Recently, a fusion of two zinc finger genes JAZF1 and JAZ1 has been reported to be useful in differential diagnosis of leiomyoma and ESS.^[6]

Endometrial stromal tumors are classified into endometrial stromal nodule, low grade ESS and undifferentiated endometrial carcinoma/uterine sarcoma by World Health Organization 2003. Differentiation of endometrial stromal nodule and ESS is done by establishing the well-circumscribed nature of the lesion in endometrial stromal nodule in contrast to irregular, infiltrating margins in ESS. However, this can only be done in a hysterectomy specimen and was not possible in our case as the patient was young and hysterectomy was not carried out. As the patient gave history of recurrence and ESS can present rarely as a polypoidal lesion,^[9] the diagnosis of low grade ESS was favored.

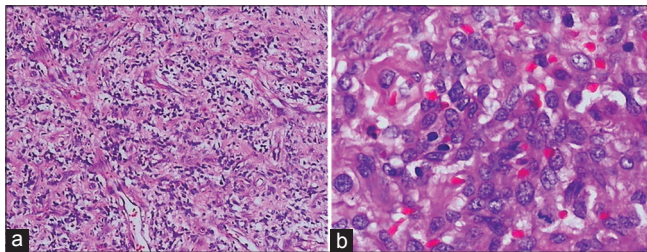


Figure 1: (a) Thin walled blood vessels with plump spindle shaped tumor cells (H and E, $\times 100$) and (b) plump spindle shaped tumor cells and a mitotic figure noted (H and E, $\times 400$)

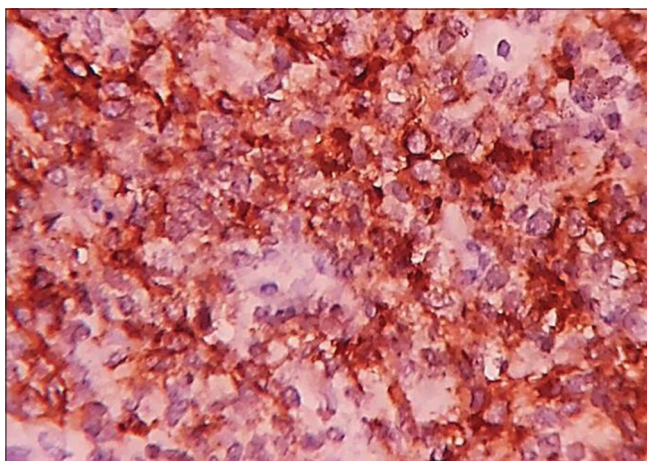


Figure 2: Strong membranous positivity of spindle shaped tumor cells

CONCLUSION

We report a case of ESS which occurred in an unusual age group and an unusual location and suggest that ESS be kept in mind while reporting a spindle cell tumor in cervix, even though rare.

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