Endometrial stromal sarcoma presented as endometrial polyp: a rare case

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Abstract

Endometrial stromal sarcomas are rare malignant mesenchymal tumors, occurring in the age group of 45–55 years. We report a case of low-grade endometrial stromal sarcoma in a 39-year-old woman, whose ultrasound examination suggested a large endometrial polyp. Polypectomy followed by laparoscopic total hysterectomy with bilateral salpingo-oophorectomy was performed. The final histological examination showed no residual disease, implying that the tumor was totally resected during hysteroscopy. This case report highlights that endometrial stromal sarcoma is to be included in the differential diagnosis of endometrial polyp though rare.

Keywords: Endometrial stromal sarcoma, Hysteroscopy, Endometrial polyp, Uterine malignancy, Uterine sarcoma, Mesenchymal neoplasm

Introduction

Endometrial stromal tumors (EST) are rare tumors and according to the latest 2014 WHO classification are divided into four categories: endometrial stromal nodule (ESN), low-grade endometrial stromal sarcoma (LG-ESS), high-grade endometrial stromal sarcoma (HG-ESS), and undifferentiated uterine sarcoma (UUS) [1]. ESS accounts for < 1% of all uterine tumors with an annual incidence of 1–2 per million women [2, 3].

Although the early diagnosis and treatment is essential for the prognosis, it is often delayed due to the non-specific signs and symptoms [4].

In our article, we report a case of low-grade endometrial stromal sarcoma, presented as an endometrial polyp in a young patient, with a brief review of its incidence, diagnosis, management, and prognosis.

Case report

Endometrial stromal sarcoma (ESS) is a rare clinical entity, particularly among young women. Herein, we report a case of a 39-year-old woman, gravida 2, para 2, with no history of exposure to tamoxifen neither to unopposed estrogens, who was referred to our center in March 2019 due to abnormal vaginal bleeding. The transvaginal ultrasound examination showed a large endometrial polyp and its hysteroscopic removal was scheduled. Grossly, the tumor measured 3 × 2 × 2 cm, and was a relatively circumscribed, lobulated oval mass. Although the hysteroscopic impression was that of an endometrial polyp the histological examination showed LG (low grade)-ESS (Fig. 2). The tumor cells were spindle-shaped with mild atypia, and few mitoses (3–4 per 10 high power fields), and formed fascicles that infiltrated between the muscle bundles of the myometrium. Immunohistochemically, the tumor cells were positive to CD10 and negative to h-caldesmon, desmin, and a-smooth muscle actin. Preoperatively, CT (computed tomography) scan was reported as with no suspicious findings and total hysterectomy with bilateral salpingo-oophorectomy was then decided.

Total hysterectomy with bilateral salpingo-oophorectomy was completed laparoscopically with no intraoperative or postoperative complications. The final histological examination showed no residual disease,
implying that the tumor was totally resected during hysteroscopy. In conclusion, our patient with stage Ia LG-ESS was assessed by the oncology council and was recommended observation alone with no need of adjuvant treatment complying with the National Comprehensive Cancer Network (NCCN) consensus guidelines [5].

Discussion

The ESS are rare tumors and the mean age of the affected women ranges between 45 and 55 years and the main symptoms and findings are abnormal uterine bleeding, pelvic pain, uterine mass, or endometrial polyp. The only way to definitively rule out the presence of sarcomatous tissue in such cases is the removal of the myometrous or/and endometrial lesion [6]. Our patient presented at 39 years due to abnormal vaginal bleeding, which is a rarity in itself. Due to their non-specific signs and symptoms LG ESS tumors are difficult to diagnose in the early stage or preoperatively [4].

The definitive treatment is total abdominal hysterectomy, bilateral adnexectomy, and excision of all grossly detectable tumor. Adjuvant radiotherapy (RT) may be used in order to achieve a high-locoregional control rate and should be determined on a case-by-case basis. In cases of LG-ESS stage 3–4 and in cases of recurrent disease, endocrine therapy with medroxyprogesterone, tamoxifen, gonadotropin-releasing hormone (GnRH) analogues and aromatase inhibitors is possibly indicated. With regards to the role of lymphadenectomy, it is not indicated except for cases with suspicious lymphadenopathy noted on preoperative imaging, since the risk of lymph node metastases is around 10% [7–10].

In our case, the 39-year-old patient with stage Ia LG-ESS was assessed by the oncology council and was recommended observation alone with no need of adjuvant treatment.

Regarding prognosis, tumor stage and patient’s age are the most important prognostic factors. The 5-year survival rate is over 90% for patients with tumor stage I–II, while it is around 50% for those with stages III–IV. The risk of recurrence in LG-ESS is estimated to be 10–20%. Recurrences may occur after more than 10–30 years and may appear locally in the vagina, pelvis, or as distant metastases. There is no association between distant metastases and recurrences with positive lymph nodes. In cases with negative lymph node status, the recurrence rate is reported to be up to 30% within 2 years [11].

Conclusions

Taking into account the fact that ESS is a rarely encountered malignancy of uterus, an accumulation of reports of this clinical entity is, therefore, essential to better understand its natural course and to improve treatment options. Furthermore, this case report highlights that ESS is to be included in the differential diagnosis of endometrial polyp though rare.

Abbreviations

CT: Computerized tomography; ESN: Endometrial stromal nodule; ESS: Endometrial stromal sarcoma; EST: Endometrial stromal tumors; GnRH: Gonadotropin-releasing hormone; HG: High grade; LG: Low grade;
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