**Suppl 1.** Summary of Previously Published Cases of ESS in Women Under 30 Years of Age

| **Author** |  **Age** | **Gravidity** | **Symptoms** | **Pre-operative diagnosis** | **Intraoperative findings**  | **Surgery performed** | **Final histo-pathologic diagnosis** | **Adjuvant treatment** | **Outcome at follow-up** |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| Bellone et al, 1990 [8] | 16 |  | Prolonged and heavy menstrual bleeding | Malignant uterine tumor | Endometrial cavity filled with hemorrhagic and necrotic polypoid masses | Total abdominal hysterectomy with bilateral oophorectomy and resection of vaginal ring | LG-ESS | Chemotherapy with doxorubicin | No recurrence at 5-year follow-up |
| Mustaphi et al, 1994 [9] | 24 |  | Heavy menstrual bleeding, dysmenorrhea, abdominal mass, dull aching pain in lower abdomen | Left-sided ovarian cyst and leiomyoma | 9 × 13.9 cm uterine mass; 15 × 15 cm retroperitoneal cystic mass with solid base adherent to the sigmoid colon anteriorly and ureter and iliac vessels posteriorly | Local resection of uterine mass and uterine reconstruction; excision of retroperitoneal mass (first surgery); total abdominal hysterectomy with bilateral salpingo-oophorectomy (second surgery)  | LG-ESS  | Pelvic radiation  | No evidence of disease at the 3-month follow-up |
| Zalameda, 2009 [10] | 24 | G0 | Heavy menstrual bleeding and hypogastric pain | To consider uterine sarcoma, left ovarian new growth with malignant features | 14 × 10 × 7 cm solid necrotic masses covering entire endometrial lining, extending up to the cervix; both ovaries converted to 10 × 10 × 8 cm hemorrhagic, necrotic, solid masses | Total abdominal hysterectomy with bilateral salpingo-oophorectomy | HG-ESS | Chemotherapy with doxorubicin | Leptomeningeal carcinomatosis was detected 1-month post-surgery before patient was lost to follow-up |
| Guzin et al, 2010 [11] | 21 | G2P1 (1011) | Prolonged and heavy menstrual bleeding, abdominal distension | Myoma | 11 × 6 × 5 cm fleshy solid mass originating from the posterior wall of the uterus | Myomectomy (first surgery); total abdominal hysterectomy bilateral salpingo-oophorectomy, pelvic lymph-node dissection, appendectomy, omentum biopsy and aspiration of abdominal fluid (second surgery)  | LG-ESS | None | Disease free for 3 years |
| Yan et al, 2010 [12] | 25 | G0 | Heavy menstrual bleeding | Myoma | 5 × 4 × 4 cm submucous mass at the posterior uterine wall | Myomectomy (first surgery); local resection and uterine reconstruction (second surgery) | HG-ESS | Hormonal therapy with mifepristone 20 mg; chemotherapy with etoposide and cisplatin | Spontaneous pregnancy 40 months after surgery and delivered a live baby at 39 weeks; no evidence of disease after delivery |
| Amant et al, 2010 [13] | 28 | G1P0, 15 weeks AOG | Pain at the right fossa | Ovarian mass  | Peritoneal spread of malignant plaques | Laparoscopy, biopsy of peritoneal lesions (first surgery); hysterotomy (second surgery)  | HG-ESS | None | Patient succumbed to the disease 6 weeks after diagnosis |
| Delaney et al, 2012 [14] | 16 | G0 | Prolonged heavy menstrual bleeding, abdominal distension | Uterine malignancy | 17 cm mass with solid and cystic components and 8 cm pedicle arising from the left fundal region | Local resection of uterine mass and uterine reconstruction | LG myxoid ESS with minimal nuclear pleo-morphism and low mitotic rate | Hormonal therapy with megestrol acetate for 8 years | Patient had spontaneous pregnancy 8 years after surgery and delivered a live baby at 34 and three-seventh weeks AOG; no evidence of disease |
| Devi et al, 2012[15] | 29 |  | Heavy menstrual bleeding, dysmenorrhea | Myoma | 6 × 6 cm lobulated mass at the anterior wall of the uterus with gray-pale yellow, soft necrotic area | Hysteroscopic myomectomy (first surgery); total abdominal hysterectomy with bilateral salpingo-oophorectomy (second surgery)  | LG-ESS | None | No evidence of disease |
| Dong et al, 2014 [16] | 25 | G0 | Heavy menstrual bleeding, shortened cycles, lower abdominal pain | Myoma | 6.1 × 5.2 × 5.9 cm submucous mass with hemorrhagic and necrotic surface at the left posterior uterine wall, 4.5 × 3.3 cm residual mass at the posterior uterine wall | Hysteroscopic myomectomy first surgery); local resection of uterine mass and reconstruction of the uterus (second surgery) | LG-ESS | Hormonal therapy with medroxy-progesterone 250 mg OD for 1 year  | Spontaneous pregnancy 6 months after medroxyprogesterone withdrawal, patient delivered a live baby at 42 weeks; no tumor recurrence |
| Dong et al, 2014 [17] | 19 | G0 | Acute hypo-gastralgia | Myoma (degeneration) | 10 × 9 cm mass with hemorrhage and necrosis in the anterior wall of the uterus | Laparoscopy, biopsy, frozen section, converted to exploratory laparotomy, local resection of uterine mass, uterine reconstruction | LG-ESS | Hormonal therapy with medroxyprogesterone acetate daily for 1 year | No signs of recurrence 33 months after surgery |
| Chhabra et al, 2017 [18] | 24 | G1P1 (1001) | Heavy menstrual bleeding, dysmenorrhea, dyspnea | Myoma | Endometrial cavity filled with multiple nodular fleshy growths with the largest nodule measuring 9 × 5 cm, 3 × 2 cm pulmonary nodule | Total abdominal hysterectomy, right posterolateral thoracotomy, excision of lung nodule | HG-ESS | Pelvic radiation | Not mentioned |
| Eamudomkarrn et al, 2018 [19] | 21 | G0 | Prolonged heavy menstrual bleeding, palpable mass at the lower abdomen | Malignant small round cell tumor suggestive of metastatic sarcoma uterine origin (biopsy obtained by endometrial curettage)  | Intracavitary polypoid mass 13 × 13 × 7 cm | Total abdominal hysterectomy, bilateral salpingo-oophorectomy, resection of enlarged pelvic nodes, omentectomy, biopsy of peritoneal nodules in the cul-de-sac | HG-ESS | Adjuvant chemotherapy with adriamycin and ifosfamide; Pelvic radiation | Rapid disease progression; succumbed 8 months post operation |
| Calin et al, 2018 [20] | 27 |  | Moderate persistent vaginal bleeding, diffuse pelvic pain | Uterine poly-fibromatosis with cervical mass | Multinodular tumor formation at the uterine fundus with largest node measuring 3 cm; entire tumor with local intramyometrial invasive character until uterine peritoneum and expansive through the cervix into the vagina | Exploratory laparotomy, interannexial total hysterectomy (first surgery); bilateral salpingo-oophorectomy, cardinal ligaments excision, partial omentectomy, lymph node sampling, re-excision of the vaginal vault (second surgery) | LG-ESS, stage Ib | None | Not mentioned |
| Chang et al, 2018 [21] | 27 | G0 | Abnormal vaginal bleeding for several days  | Uterine tumors resembling ovarian sex cord tumors (UTROSCT) and LG-ESS with adenoid, sexual cord like differentiation (previous biopsies obtained by hysteroscopy) | 3.0 × 2.3 cm fragile, soft yellow mass in the uterine cavity, 4.2 × 4.6 cm fragile dark red mass with unclear margins at the anterior wall of the uterus | Total hysterectomy with bilateral salpingectomy | LG-ESS with adenoid and sexual cord differentiation | Hormonal therapy with 250 mg medroxy-progesterone acetate and 20 mg tamoxifen daily | No evidence of disease  |
| Wong et al, 2018 [22] | 24 | G1P0,14 weeks AOG; | Painless vaginal bleeding | Spindle cell malignancy (after biopsy of cervical polyp) | 2.5 × 2.7 cm polypoid mass extending into the superior aspect of the vagina | Total abdominal hysterectomy with fetus in situ, bilateral salpingectomy, surgical staging and lymph node dissection | HG- ESS | Chemotherapy with 6 cycles of gemcitabine and taxotere | No recurrence 15 months postoperatively |
| Sohail et al, 2019 [23] | 20 | G0 | Prolonged heavy menstrual bleeding, dysmenorrhea, urinary retention | Leiomyoma and hemorrhagic cyst | 15 × 10 cm soft yellow necrotic mass in the uterine cavity extending into the vagina, retroperitoneal semisolid mass ≤ 6 cm adherent to the right pelvic wall | Abandoned total abdominal hysterectomy; local resection of the uterine mass and retroperitoneal mass | LG-ESS | Chemotherapy with doxorubicin and cyclo-phosphamide | Progression to stage IV disease (liver metastasis); recurrence of the uterine mass 4 weeks post-surgery |

ESS: endometrial stromal sarcoma; LG-ESS: low-grade ESS; HG-ESS: high-grade ESS; AOG: age of gestation; OD: once a day.