

Endometrial stromal sarcomas related to endometriosis

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【Introduction】 It is estimated that about 1% of women with endometriosis develop malignant neoplasms and the ovary is the primary involved transformation site. Most malignant tumors, which arise from endometriosis are usually endometrial adenocarcinomas. The development of endometrial stromal sarcoma (ESS) is rare. Moreover, the extra-uterine ESS is very rare and only about 80 cases of which have been reported in literature. Half of those cases were associated with endometriosis. The behavior of extra-uterine ESS is reported to have poor prognosis than uterine ESS even though low-grade.

【Patients and Methods】 We present the study of 9 patients with endometrial stromal sarcoma in our hospital from 2003 to 2016. We reviewed the treatment, prognosis and whether they had any association with endometriosis or not.

【Results】 The 6 of 9 cases are still alive with no relapse after surgery. The other 3 cases have died. In the no relapse 6 cases, 5 cases were uterine ESS and had no association with endometriosis. They were diagnosed after surgery which had done with suspect of uterine tumor. They received only surgery (hysterectomy and both salpingo-oophorectomy) and have no relapse. One of those 6 cases was extra-uterine ESS with low grade located in the abdomen and had a focus of endometriosis which was contiguous with the tumor, confirming its origin and immunostaining showed estrogen-receptor-positive[ER+] and progesterone-receptor-positive[PR+]. This patient had surgical removal and received a postoperative progestin therapy for 3 years. After stopping hormonal therapy, she is still alive with no relapse. The 3 deceased cases were extra-uterine ESS with metastasis to pelvic tissue, peritoneum, liver lymph nodes, and so on. They had no past medical history of endometriosis. 2 of 3 had surgery, postoperative chemotherapy and radiation. 1 case received chemotherapy and progestin therapy without surgery because of its high stage.

【Conclusion】 The common treatment of ESS is complete surgical removal of ESS. In the case of uterine ESS, hysterectomy and both salpingo-oophorectomy is a standard surgery and have good prognosis. Postoperative chemotherapy and radiation said to be little efficacy. In the case of low grade ESS with ER and PR positive, hormonal therapy like progestin, gonadotropin-releasing hormone agonist(GnRHa), aromatase inhibitor is said to be effective. On the other hand, in the case of extra-uterine ESS, the standard treatment is the same with that of uterine ESS, but we cannot expect good prognosis. However, in the case of extra-uterine ESS associated with endometriosis, as one of the case we reported, adjuvant progestin therapy may be effective.

Keywords : endometriosis, endometrial stromal sarcoma, progestin therapy

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