A CASE OF PEUTZ-JEGHERS SYNDROME WITH ENDOMETRIAL ADENOMYOMATOUS POLYP

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Peutz-Jeghers syndrome, which has autosomal dominant inheritance, shows pigmentation in lip and skin. It also has features of hamartomatous polyp over the gastrointestinal tract, while sometimes developing tumor in genital tract. Known tumors in female genital tract include cervical adenocarcinoma, sex cord tumor, etc. Adenomyomatous polyp in uterus is also one of the rare diseases, which seldom develop in the absence of hormone treatment such as Tamoxifen. Currently, there is one case of Peutz-Jeghers syndrome patient with endometrial adenomyomatous polyp. Authors found mole-like lesions on the transvaginal ultrasonogram in a typical Peutz-Jeghers syndrome patient without pregnancy history and previous chemotherapy. After confirming its diagnosis of adenomyomatous polyp on resectoscopic biopsy, we report our findings along with brief literature review.

Keywords: Peutz-Jeghers syndrome, Adenomyomatous polyp, Resectoscopy

Case Report

A twenty-eight-year-old female visited our hospital with vaginal spotting for about a month. She did not experience pregnancy and had characteristic dark-colored pigmentation in skin and lip. She has a history of bowel resection seventeen and twelve years ago. She did not remember the reasons of bowel resection. As for Family history, the patient’s mother died from cervical cancer. She had no history of previous hormonal therapy, such as Tamoxifen. In a transvaginal ultrasonogram, mole-like appearance of vesicle pattern on the transvaginal ultrasonogram.

Peutz-Jeghers syndrome (PJS) is known to have autosomal dominant inheritance and is related to the STK11 gene. The symptoms include dark-colored pigmentation from the lip and buccal mucosa to fingers and to the toes, along with hamartomatous polyp over the entire gastrointestinal tract. Moreover, in addition to gastrointestinal tract, it can cause tumor in breast, genital tract, lung, etc. Known tumors in female genital tract include cervical adenocarcinoma, ovarian sex cord tumor, granulosa cell tumor, etc. adenomyomatous polyp in uterus has stromal component consisting of smooth muscle cells and it occasionally develops among women who received hormonal therapy such as Tamoxifen. It has a feature of mole-like appearance of vesicle pattern on the transvaginal ultrasonogram.

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like lesion. Two months after performing primary resectoscopy, the secondary resectoscopic polypectomy was performed under general anesthesia. The endometrium was still filled with Swiss cheese like lesion and the diagnosis of adenomyomatous endometrial polyp was confirmed.

The submitted endometrium from the primary curettage and the secondary resectoscopic biopsy showed the similar histologic finding each other. The endometrium revealed several pieces of irregular sized polyps measuring 1.5×1.5 cm of the largest one and aggregating volume was about 20 mL. The polyps were composed of irregularly distributed endometrial glands and prominent bundles of smooth muscle cells in the stroma as well as combined plasma cells. Theses bundles of smooth muscle cells were variable in proportion. The endometrial glands revealed cellular proliferation and focal glandular crowding. Also, relatively usual endometrial polyps were present around adenomyomatous polyps (Fig. 1). More than 8 polyps were found at the duodenum and resected polyps showed hyperplastic and dilated glands with intervening smooth muscle bundles which were compatible with hamartomatous polyp (Fig. 2). Two years after surgery, there was no recurrence of symptoms and no thickened uterine endometrium on regular ultrasonographic examination but amenorrhea continued after resectoscopy without menopausal symptoms. Because the patient wants to preserve uterus, even though there are the risk of endometrial cancer, the prophylactic hysterectomy is not performed.

**Discussion**

This case is considered to be the second endometrial adenomyomatous polyp in patient with PJS on literature review. PJS can cause tumor in genital tract in addition to gastrointestinal tract, which is often precancerous. It is known that the occurrence of such cancer is fifteen times more likely compared to the general population. Even though gynecologic neoplasms related to PJS include cervical adenocarcinoma, endometrial cancer, epithelial ovarian tumor, sex cord stromal ovarian tumor, etc., specific occurrence rate is unknown [1-3].

Endometrial polyp is frequent lesion in uterus, which shows histologically focal hyperplasia of basalis with a localized overgrowth of gland, stroma, and blood vessels. Adenomyomatous polyp has