

Diffuse leiomyomatosis of the cervix: a rare tumor in a rare location. A first report and literature analysis

Zaigui Wu^{1,*}, Fei Ruan^{1,*}, Xiaocen Niu¹, Jianhong Zhou¹

¹Zhejiang University, School of Medicine, Womens Hospital, Department of Obstetrics and Gynecology, Hangzhou (China)

Summary

Objective: Diffuse uterine leiomyomatosis (DUL) is rare and almost occurs in the corpus uteri without involving the cervix. Here the authors report a patient with DUL only involving the cervix. **Case Report:** A 42-year-old female was admitted to this hospital for prolonged menstruation and shortened menstrual cycle in September 2016. Repeated gynecologic and imaging examinations only confirmed an obvious enlargement of the cervix. After careful preoperative assessment, the patient underwent a well-prepared cervical resection procedure and the pathological report confirmed diffuse leiomyomatosis. Her menstrual cycle resumed normally after one month postoperatively. **Conclusion:** DUL only involving the cervix is extremely rare in clinic and doctors should assess symptoms and prescribe supplementary examinations individually in order to adopt the most appropriate treatment.

Key words: Diffuse uterine leiomyomatosis (DUL); Cervix; Cervical resection procedure; Diagnosis; Appropriate treatment.

Introduction

Diffuse uterine leiomyomatosis (DUL) has been reported to be an extreme type of multiple uterine leiomyomata with diffuse dissemination of numerous small (usually ≤ 3 cm) and ill-defined benign leiomyomata throughout the entire myometrium [1]. Since Lapan and Solomon first described this disease in 1979 [2], less than 60 cases of DUL are found in the English literature and all of them were reported to occur in the corpus uteri while the cervix appears to be spared [3]. Here the authors report a unique case of diffuse leiomyomatosis occurring only in the cervix not involving the corpus uteri.

Case Report

The patient, a 42-year-old woman, gravida 1 para 1 presented to this gynecology outpatient in 2016 due to shortened menstrual cycle (23 days) and prolonged menstruation (15 days) for the past two years. Gynecological examination only showed a smooth and hypertrophic cervix but hard as a stone. Repeated transvaginal US revealed the cervical tissue to be thickened diffusely with plenty of blood flow and no obvious limited mass was seen in the cervix. MRI examination further confirmed the same result (Figure 1). The initial diagnosis was cervical neoplasm and a cold knife resection of the total cervix were carried out. Diffuse hyperplasia of the entire cervical stroma around the tube and local nodules less than 1 cm and ill-defined were seen intraoperatively. Tissue was prepared with routine histological procedures and proved to be diffuse leiomyomatosis (Figure 2). Normal menstrual cycle resumed normally after one month postoperatively. Written informed consent was obtained from the patient.

Discussion

Although leiomyomas are the most common benign uterine tumor, DUL are extremely rare in clinical and this is the first report for cervical diffuse leiomyomatosis to the present authors' knowledge. The difference between diffuse



Figure 1. — The complete cervix is enlarged diffusely with spontaneous hypointensity on T1-weighted and hypersignal intensity on T2-weighted MRI.

*Co-first author



Figure 2. — High power view (HE $\times 40$) shows nodules composed of uniform, spindled smooth muscle cells.

leiomyomatosis and ordinary leiomyomas is that the former constitutes innumerable, ill-defined, and small smooth-muscle nodules. Genetic analysis shows non-random X-chromosome inactivation and different nodules having different alleles inactivated in diffuse leiomyomatosis [1], while the exact pathogenesis is still unclear due to the limited case numbers.

As ordinary leiomyomas, the clinical manifestation has no specificity [3]. Patients have mild or even no symptoms in the early and later have menorrhagia, menstrual changes, etc. The only abnormality of the present patient was shortened menstrual cycle and prolonged menstruation. The present authors hypothesize that it was associated with the enlarged cervix tube and impaired cervical smooth muscle contraction which can be proved by the recovery of menstrual cycle postoperatively.

Ultrasound scan is still the first line imaging for its diagnose [4]. The main appearance is diffuse symmetrical uterine enlargement and countless inseparable hypoechoic masses without evidence of acoustic shadowing for pseudocapsule because of absent normal muscular tissue among smooth-muscle nodules. The size of each nodules varies from less than 1 mm to less than 1 cm, occasionally larger but no more than 3 cm [1]. In the present patient the US did not detect any mass in the cervix. The role of MRI can not be underestimated as its appearance parallels the gross pathological appearance just like a “pebble-filled purse” [5]. The MRI in the present patient revealed obvious hypertrophy within the entire cervical submucosa which is more obvious after enhancement. The largest difference of the present case is that the nodules were extremely little so no mass was detected by US and MRI.

Treatment is also similar to ordinary leiomyomas, but because of too many myoma nodules, myomectomy would not be the best choice. Total hysterectomy is mainly suited

for patients with severe symptoms and no fertility requirements. The conservative treatments include drugs like gonadotropin-releasing hormone analogue (GnRHa), hysteroscopic myomectomy, uterine artery embolization (UAE), and high-intensity focused ultrasound ablation (HIFU). Literature has reported patients conceived spontaneously with successful delivery after GnRHa treatment or hysteroscopic myomectomy [6]. A case report also demonstrated the safety and efficacy of HIFU [7]. UAE was a highly effective treatment for DUL but its potential damage to the function of uterine and ovarian have limited its use in cases with fertility requirements [8]. Case in this report had a long history of one year with benign clinical course and morphology according to the unchanged symptom and cervical size and texture, hence the authors suggested a diagnostic cervix resection.

In short, DUL is very rare in clinics and the present authors have reported a diffuse leiomyomatosis of the cervix for the first time. For these rare cases doctors should analyze symptoms and supplementary examinations individually and adopt the most appropriate treatment.

References

- [1] Baschinsky D.Y., Isa A., Niemann T.H., Prior T.W., Lucas J.G., Frankel W.L.: “Diffuse leiomyomatosis of the uterus: a case report with clonality analysis”. *Hum. Pathol.*, 2000, 31, 1429.
- [2] Lapan B., Solomon L.: “Diffuse leiomyomatosis of the uterus precluding myomectomy”. *Obstet. Gynecol.*, 1979, 53, 82S.
- [3] Ip P.P., Tse K.Y., Tam K.F.: “Uterine smooth muscle tumors other than the ordinary leiomyomas and leiomyosarcomas: a review of selected variants with emphasis on recent advances and unusual morphology that may cause concern for malignancy”. *Adv. Anat. Pathol.*, 2010, 17, 91.
- [4] Shimizu Y., Yomo H., Kita N., Takahashi K.: “Successful pregnancy after gonadotropin-releasing hormone analogue and hysteroscopic myomectomy in a woman with diffuse uterine leiomyomatosis”. *Arch. Gynecol. Obstet.*, 2009, 280, 145.
- [5] Pai D., Coletti M.C., Elkins M., Ladino-Torres M., Caoili E.: “Diffuse uterine leiomyomatosis in a child”. *Pediatr. Radiol.*, 2012, 42, 124.
- [6] Shimizu Y., Yomo H., Kita N., Takahashi K.: “Successful pregnancy after gonadotropin-releasing hormone analogue and hysteroscopic myomectomy in a woman with diffuse uterine leiomyomatosis”. *Arch. Gynecol. Obstet.*, 2009, 280, 145.
- [7] Chen L., Xiao X., Wang Q., Wu C., Zou M., Xiong Y.: “High-intensity focused ultrasound ablation for diffuse uterine leiomyomatosis: A case report”. *Ultrason. Sonochem.*, 2015, 27, 717.
- [8] Koh J., Kim M.D., Jung D.C., Lee M., Lee M.S., Won J.Y., et al.: “Uterine artery embolization (UAE) for diffuse leiomyomatosis of the uterus: clinical and imaging results”. *Eur. J. Radiol.*, 2012, 81, 2726.

Corresponding Author:

JIANHONG ZHOU, M.D.

Zhejiang University, School of Medicine
Womens Hospital

Department of Obstetrics and Gynecology
Hangzhou 310006 (China)

e-mail: zhoujh1117@zju.edu.cn