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Case Report

Laparoscopic en bloc resection of a para-cervical cancer with OHVIRA syndrome

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ABSTRACT

Objective: There are some reports of cervical cancer with uterus didelphys but a case of clear cell carcinoma (CCCC) with Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA) syndrome is extremely rare. The aim of this paper was to report a case of CCCC with OHVIRA syndrome and the difficulty in making a preoperative diagnosis.
Case report: A 65 years old woman presented with postmenopausal bleeding and pelvic examination showed right paracervical mass. Preoperative confirmation of cervical carcinoma was difficult due to the location of the mass, which was inaccessible by cervical punch biopsy. Pelvic examination revealed a large mass in pelvic cavity without parametrial invasion and ultrasound showed approximately 70 mm cervical tumor. Laparoscopic surgery revealed clear cell carcinoma of the para-endocervix with OHVIRA syndrome.
Conclusion: In the case of cervical carcinoma with OHVIRA syndrome, laparoscopic surgery is preferable for the diagnosis and management.

Introduction

The prevalence of clear cell cervical carcinoma is 3–10% of adenocarcinomas. Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA) syndrome is a congenital anomaly of the female genital tract characterized by obstructed hemivagina, uterus didelphys, and ipsilateral renal anomaly [1]. It is a Müllerian duct anomaly due to abnormal embryologic development of the urogenital system. There are many different type of mullerian anomalies [2–4] there are some reports of cervical cancer with uterus didelphys but a case of CCCC with OHVIRA syndrome is extremely rare. The aim of this paper was to report a case of CCCC with OHVIRA syndrome and the difficulty in making a preoperative diagnosis.

Case report

A 65-year-old female, gravida 3, para 2, was transferred to our institution with genital bleeding for a month, 10 years after menopause. During her deliveries, no anomalies were noted. The patient could be had no in-utero DES exposure by limited record of past history. On speculum examination, there was no mass on the cervix. On pelvic examination, there was an approximately 7 × 7 cm solid mass in the right paracervical area. A single cervix was seen on colposcopy, which was unsatisfactory, as the squamo-columnar junction was not visualized. Cytology as well as biopsy of the cervix was normal. Magnetic resonance imaging (MRI) revealed uterine didelphys, right renal agenesis and large mass that measured 70 mm located right side of endocervix (Fig. 1A). A staging computed tomography (CT) scan of the chest and abdomen did not reveal evidence of lymph node metastasis or distant metastasis. Preoperative diagnosis of a possible malignancy was difficult due to the location of the paracervical mass being inaccessible through the vagina. Total laparoscopic hysterectomy with bilateral salpingo-oophorectomy was performed. After insertion of the trocars, inspection of abdominoplevic organs was done. Right paracervical mass was identified. The procedure was started by opening the peritoneum lateral to the right psoas muscle from the level of the right common iliac artery to the insertion of the right round ligament. The right paravesical and pararectal spaces were developed and opened. The right infundibulopelvic ligament was isolated,

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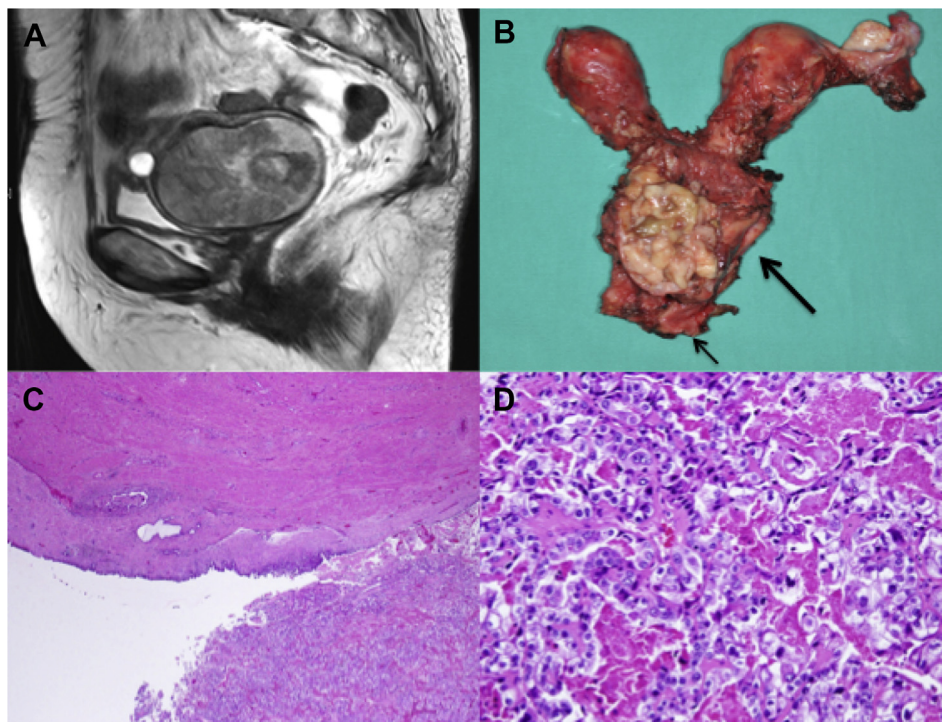
Fig. 1. A) MRI shows cervical tumor that measures 70 mm with uterus didelphys. B) Specimen showed para-cervical tumor with uterus didelphys. Big arrow indicated left side of uterine cervix whereas small arrow indicated right side of uterine cervix. C), D) Histology shows clear cell carcinoma with a stromal invasion of 1 mm in depth and negative of lymphovascular invasion.

coagulated then cut using Ligasure™ (Covidien). Since there was right renal and ureteral agenesis, dissection of the right parametrium was easy. Right uterine artery was coagulated and ligated near its origin from the hypogastric artery. The right parametrium was dissected and cut lateral to the right paracervical mass. Similar steps were done on the left but care was done during dissection of the left parametrium avoiding the left ureter. The vagina was circumferentially incised below the cervix. The specimen was then placed into a retrieval bag and removed vaginally. During hysterectomy, the paracervical mass was resected en bloc with the right parametrium and the uterus with no significant tumor free margin, due to the size of the mass. Frozen section showed right side cervical tumor (Fig. 1B). When the frozen section revealed high-grade adenocarcinoma of the cervix, the surgeons proceeded with pelvic lymphadenectomy. Postoperative histopathology confirmed clear cell adenocarcinoma, with 1 mm depth of invasion, negative of lymphovascular invasion and surgical margin, pT1b2N0M0 (Fig. 1C and D). Following the surgery, the patient received concurrent chemoradiation therapy with no evidence of disease 12 months after completion of therapy.

Discussion

The authors reported a very rare case of CCCC with OHVIRA syndrome. There are only two case reports of cervical cancer with OHVIRA syndrome [5]. Watanabe et al. reported a case of endometrioid adenocarcinoma of the semiobstructed side of the uterine cervix with OHVIRA syndrome [6]. Although cytology of the visible part of the uterine cervix was normal, adenocarcinoma was diagnosed by biopsy of the semiobstructed vaginal septum. Kaba et al. reported a case of endometrioid adenocarcinoma of the non-blind side of cervix with OHVIRA syndrome [7]. Cervical cytology and endocervical curettage were normal, in spite of a 70 mm mass seen on MRI. However, endometrial biopsy showed endometrioid

adenocarcinoma. In both published cases, they reported that preoperative investigations, such as cytology and endocervical curettage, failed to detect cervical malignancy, and these tests have the risk of false negative results. Compared to the two cases, this patient presented with postmenopausal bleeding and pelvic examination showed right paracervical mass. The ectocervix was devoid of cervical mass. Preoperative confirmation of cervical carcinoma was difficult due to the location of the mass, which was inaccessible by cervical punch biopsy. This is the first reported case of CCCC with OHVIRA syndrome in the literature, to the best of our knowledge.

Laparoscopic surgery was chosen for the diagnosis and management of this patient, because it is a less invasive approach compared to open surgery. Aside from less incidence of complications and blood loss, magnification of deep pelvic structures is better during laparoscopy than laparotomy, making the procedure less difficult [8–12]. Furthermore, en bloc resection with paracervical tumor was possible without causing ureteral injury, due to agenesis of the ipsilateral ureter. However, attention has to be paid for the spillage of tumor cell while removing the uterus. Placing the specimen into a retrieval bag before removing it is recommended for suspicious cases of malignancy. Treatment of CCCC is based on that of a squamous cell carcinoma due to the rarity of type of tumor [13,14]. Thomas et al. conducted a retrospective multi-institutional review and analyzed 34 patients of CCCC [15]. The author reported stage I and IIA CCCC patients with excellent 3-year overall survival rate (91%) indicating that clear cell histology by itself does not appear to portend a poor prognosis. Tumor size and grade had no impact on progression free survival or overall survival, but the presence of positive lymph nodes is a strong predictor of adverse prognosis. This patient had negative lymph node and surgical margins as well as shallow cervical stromal invasion of 1 mm in spite of the large size of tumor, indicating better prognosis. Nevertheless, long-term follow up is necessary. Carcinogenesis of CCCC with OHVIRA syndrome is totally different from CCCC without

Müllerian anomalies. There is a report about the association of CCCC and vaginal and Müllerian anomalies [15]. Vaginal adenosis is induced in association with Müllerian duct anomalies, which may provide a basis for carcinogenesis [16]. This patient had cervical tumor located near the endocervix (paraendocervix) with only microinvasion, in spite of large mass. It seemed that it degenerated from persistence of Müllerian epithelium connected to cervical mesenchyme. Carcinogenesis of CCCC from Müllerian anomaly is considered, that clear cell carcinoma may have arisen from the paracervix of Müllerian-derived columnar epithelium that have persisted for a long time and was subjected to genetic and hormonal changes [17].

Conflict of interest

The authors declare that there are no conflicts of interest.

Acknowledgement

References

[1] Kaminski PF, Maier RC. Clear cell adenocarcinoma of the cervix unrelated to diethylstilbestrol exposure. Obstet Gynecol 1983;62:720–7.

[2] Li YC, Lee CL, Cheng MH, Kuo HH. Laparoscopic creation of neovagina and neocervix, followed by their reconstruction with polytetrafluoroethylene graft/buccal mucosa and pudendal artery perforator flap. Gynecol Minim Invasive Ther 2015;4(4):160.

[3] Lee JM, Huang CY, Wu KY, Yen CF, Chern B, Lee CL. Novel technique of neovagina creation with uterine serosa in the treatment of vaginal agenesis associated with mullerian agenesis. Gynecol Minim Invasive Ther 2014;3:50–3.

[4] Kobayashi A, Fukui A, Funamizu A, Ito A, Fukuhara R, Mizunuma H. Laparoscopically assisted cervical canalization and neovaginoplasty in a woman with cervical atresia and vaginal aplasia. Gynecol Minim Invasive Ther 2017;6(1):31–3.

[5] Smith NA, Laufer MR. Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome: management and follow-up. Fertil Steril 2007;87:918–22.

[6] Watanabe Y, Etoh T, Nakai H. Adenocarcinoma of the lower female genital tract in patients with Herlyn-Werner-Wunderlich syndrome. Am J Obstet Gynecol 2012;207:e5–6.

[7] Kaba M, Gungor T, Baser E, Ozdal B, Sirvan L. Cervical cancer in a patient with uterus didelphys and obstructive hemivagina, ipsilateral renal anomaly (OHVIRA) syndrome. Arch Gynecol Obstet 2013;288:229–30.

[8] Yoshiki N. Single-incision laparoscopic myomectomy: a review of the literature and available evidence. Gynecol Minim Invasive Ther 2016;5(2):54–63.

[9] Ulm MA, Ginn DN, ElNaggar AC, Tillmanns TD, Reed KM, Wan JY, et al. A Comparison of outcomes following robotic-assisted staging and laparotomy in patients with early stage endometrioid adenocarcinoma of the uterus with uterine weight under 480G. Gynecol Minim Invasive Ther 2016;5(1):25–9.

[10] Terao Y, Kitade Mari, Kusunoki S, Fujino K, Ujihira T, Kimura M, et al. Surgical and oncological outcome of laparoscopic surgery, compared to laparotomy, for Japanese patients with endometrial cancer. Gynecol Minim Invasive Ther 2016;5(2):64–8.

[11] Lee CL, Huang KG, Wu PJ, Lee PS, Yen CF. Long-term survival outcome of laparoscopic staging surgery for endometrial cancer in Taiwanese experience. Taiwan J Obstet Gynecol 2014;53:57–61.

[12] Wu KY, Lertvikool S, Huang KG, Su H, Yen CF, Lee CL. Laparoscopic hysterectomies for large uteri. Taiwan J Obstet Gynecol 2011;50:411–4.

[13] Manvelyan V, Khemarangsan V, Huang KG, Adlan AS, Lee CL. Port-site metastasis in laparoscopic gynecological oncology surgery: an overview. Gynecol Minim Invasive Ther 2016;5(1):1–6.

[14] Korhonen MO. Adenocarcinoma of the uterine cervix. Prognosis and prognostic significance of histology. Cancer 1984;53:1760–3.

[15] Robboy SJ, Kaufman RH, Prat J, Welch WR, Gaffey T, Scully RE, et al. Pathologic findings in young women enrolled in the National Cooperative Diethylstilbestrol Adenosis (DESAD) project. Obstet Gynecol 1979;53:309–17.

[16] Barnes L, Eveson JW, Reichart P, Sidransky D. World Health Organization classification of tumours: pathology and genetics of head and neck tumours. Lyon: IARC; 2005. p. 168–75.

[17] Waggoner SE, Anderson SM, Van Eyck S, Fuller J, Herbst AL. Human papillomavirus detection and p53 expression in clear-cell adenocarcinoma of the vagina and cervix. Obstet Gynecol 1994;84:404–8.