Case Report

Laparoscopic Posterior Pelvic Exenteration with Radical Vulvectomy for Intestinal-type Vulvar Adenocarcinoma

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Abstract

Vulvar intestinal adenocarcinoma is a rare malignancy. The most significant predictor of advanced vulvar cancer is achieving complete resection, although determining the optimal treatment for this rare histologic type remains uncertain. We report the case of a 63-year-old woman with a primary vulvar tumor suspected of having rectal invasion and inguinal lymph node metastases based on preoperative magnetic resonance imaging and computed tomography scans. To achieve complete resection of stage IIIC intestinal-type vulvar adenocarcinoma, we performed a laparoscopic posterior pelvic exenteration (PPE) and radical vulvectomy, along with bilateral inguinal lymph node dissection. This case report highlights the use of a novel hybrid procedure that combines laparoscopic PPE with radical vulvectomy and bilateral inguinal lymph node dissection for vulvar adenocarcinoma of the intestinal type. Laparoscopic PPE can be considered a minimally invasive approach for vulvar tumor when complete resection is achievable with an appropriate safety margin.

Keywords: Hybrid surgery, laparoscopic posterior pelvic exenteration with radical vulvectomy, vulvar adenocarcinoma of intestinal type

Introduction

Vulvar cancer is an uncommon disease comprising only 0.65% of female cancer cases.[1] As of October 2023, only 35 cases of intestinal-type adenocarcinomas have been published in the literature.[2,3] It is still unclear where intestinal-type vulva adenocarcinomas originate from according to the fifth edition of the WHO classification,[4] although it is speculated to arise from a cloacal remnant.[5] Due to its extremely low incidence, the prognosis and optimal treatment are not well established. Here, we present a case of primary intestinal-type adenocarcinoma of the vulva in a patient who underwent laparoscopic posterior pelvic exenteration (PPE) and radical vulvectomy.

Case Report

A 63-year-old woman presented to our hospital with discomfort caused by a vulvar mass. On examination, a tumor measuring approximately 3 cm and presenting with an ulcer was detected on the left labia minora [Figure 1a]. These findings were further confirmed by magnetic resonance imaging, which revealed a 3 cm tumor in the left vulva, close to the rectum [Figure 1b]. The tumor was suspected to have invaded the rectum. Computed tomography indicated possible metastases in the inguinal lymph nodes but showed no distant metastases. Tumor biopsy confirmed the diagnosis of intestinal-type adenocarcinoma. Preoperative upper and lower endoscopic examinations revealed no abnormalities. Therefore, the patient was scheduled to undergo radical surgery to achieve complete resection.

The surgical procedure involved laparoscopic PPE with radical vulvectomy and bilateral inguinal lymph node dissection. Trocars were inserted, including an umbilical

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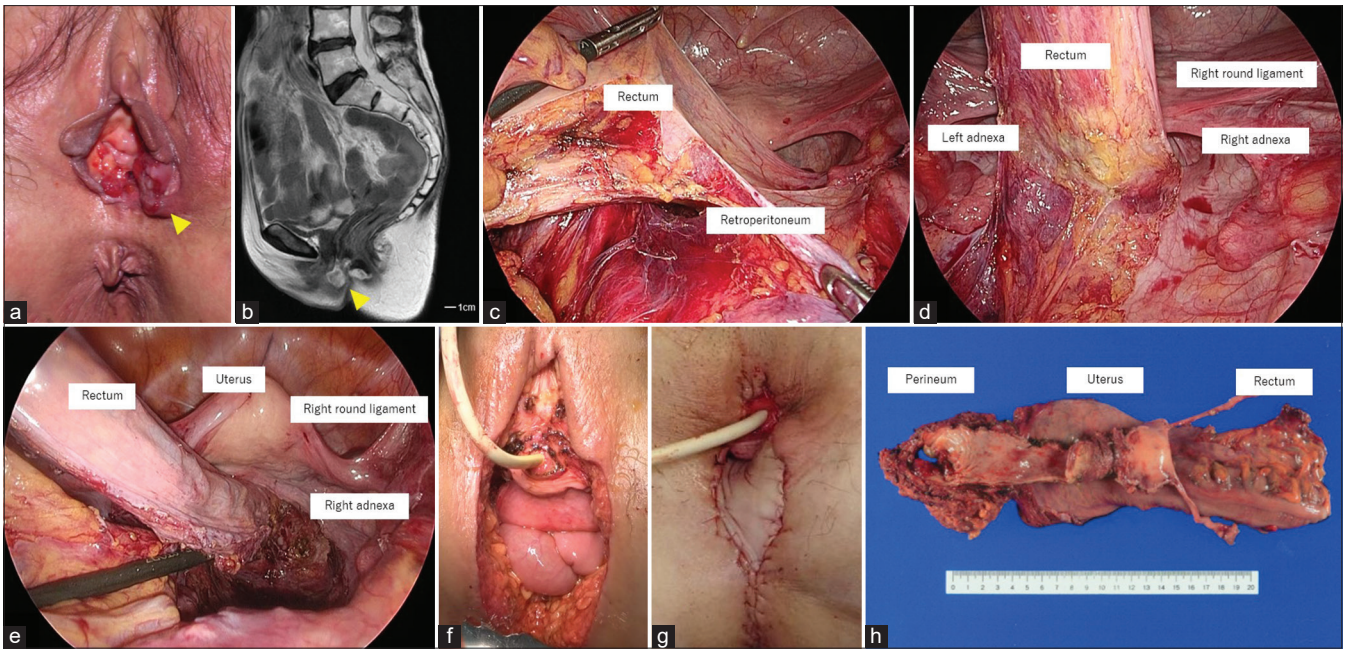

Figure 1: (a) Perineal ulcer approximately 3 cm in diameter on the right side of the vagina, (b) T2-weighted magnetic resonance imaging (sagittal) shows the tumor (yellow arrowhead) (Scale bar = 1 cm), (c-e) laparoscopic intraoperative procedure, (f and g) Reconstruction of the perineum, (h) The postoperative specimen

trocar (12 mm), right and left lateral and left lower abdominal trocars (5 mm), and a right lower abdominal trocar (12 mm). The surgery began with rectal resection, where the inferior mesenteric artery and vein were severed, and the retroperitoneum was incised [Figure 1c]. The retroperitoneum was incised along the fascia propria of the rectum, sparing the inferior abdominal nerve [Figure 1d]. Mobilization of the rectum was performed until the anorectal muscles were exposed [Figure 1e]. The colon was resected at the junction of the sigmoidal and descending colon. Following colon resection, a laparoscopic total hysterectomy was performed, involving dissection of the bilateral fallopian tubes, ligamentum propria of the ovaries, and parametrium. The peritoneum of the vesicouterine pouch was dissected to preserve the bladder and urethra. In addition, the anterior wall of the vagina was incised transabdominally. Subsequently, the vulvar approach was initiated after laparoscopic PPE. A resection line was drawn, including a 2 cm margin from the tumor located on the left labia minora, encompassing the clitoris and anus. The resection line was connected to the incision in the vaginal wall through laparoscopy, and the tumor was removed along with the perineum, rectum, and uterus [Figure 1f]. Due to the extensive defect resulting from radical vulvectomy, perineal reconstruction, including the posterior wall of the vagina, was performed using a left gracilis musculocutaneous flap [Figure 1g]. The postoperative specimens included the perineum, uterus with bilateral adnexa, and rectum with the anus [Figure 1h]. To optimize the operation time, bilateral inguinal lymph node dissection and colostomy were performed simultaneously by two teams comprising gynecologists and colorectal surgeons. The total operation time was 9 h and 2 min, with a blood loss of 398 mL, and no blood transfusion was required. No perioperative complications occurred, and the patient was discharged 16 days after surgery.

The postoperative diagnosis confirmed vulvar adenocarcinoma of the intestinal type, classified as FIGO Stage IIIC (pT2N2bM0). Histopathological findings are shown in Figure 2. The vulvar tumor exhibited deep stromal invasion [Figure 2a], although the rectum and anus remained unaffected. The tumor showed a combination of well-to-moderately differentiated tubular adenocarcinoma and mucinous carcinoma components [Figure 2b], including a notable presence of goblet cells with intestinal differentiation [Figure 2c]. Immunohistochemical analysis revealed that the tumor cells were negative for p16 and PAX8 but diffusely positive for SATB2 [Figure 2d-f], confirming the diagnosis of intestinal-type adenocarcinoma.

Bilateral inguinal lymph node metastases were identified in seven of the 11 lymph nodes that were larger than 5 mm in size. Surgical margins were negative, and the patient received adjuvant radiotherapy at a dose of 50 Gy for the inguinal lesion with lymph node metastasis. After irradiation, the patient experienced pain in the inguinal lesion due to cellulitis, which was effectively managed with antibiotic treatment. There was no evidence of recurrence 6 months after treatment.

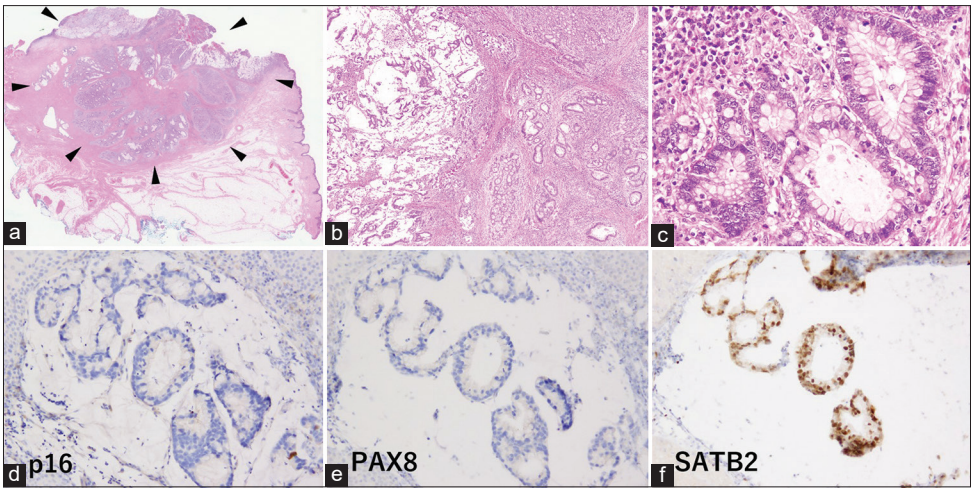

Figure 2: (a) Deep stromal invasion (black arrowheads), (b) Well-to-moderately differentiated tubular adenocarcinoma and mucinous carcinoma (H and E, ×40), (c) Intestinal differentiation with goblet cells (H and E, ×400). Immunohistochemical test results were negative for p16, (d) and PAX8, (e) Positive for SATB2, (f) D-F, ×200. PAX8: paired box gene 8, SATB2: Stabilin-2

Discussion

This is a rare case of intestinal-type adenocarcinoma of the vulva in which laparoscopic PPE with radical vulvectomy and bilateral inguinal lymph node dissection was performed.

The significance of achieving complete resection is well established as the most important indicator of prognosis for Stage III/IV vulvar cancer.[6] Pelvic exenteration is considered a therapeutic option for advanced vulvar cancers. However, to date, only four out of 32 cases in the limited literature data[7-9] have undergone radical vulvectomy and bilateral inguinal lymph node dissection for vulvar adenocarcinoma of the intestinal type. In certain cases, radical local excision of the invasive lesion could be considered feasible[10,11] to preserve the functionality of adjacent organs. However, in the present case, the preoperative evaluation revealed suspected rectal invasion by the vulvar tumor, which posed a challenge in preserving the rectum and anus. Therefore, the patient opted for radical vulvectomy combined with rectal and anal resections.

Due to the rarity of this tumor type, there is no established management for inguinal lymph node metastases. Negative prognostic factors, as indicated by a systematic review, include large lesion size (>2 cm) and lymph node involvement.[12] Computed tomography scans in this case revealed possible metastases in the inguinal lymph nodes. A retrospective study focused on squamous cell carcinoma of the vulva found that the method of surgical dissection of the groin (bulky positive groin nodes-nodal debulking versus full groin dissection) before radiation therapy did not significantly impact survival as an independent factor.[13] However, there is a lack of data specifically available for intestinal-type adenocarcinoma.

Given these uncertainties, we decided to proceed with a complete bilateral inguinal lymph node dissection. There is still disagreement on the function of adjuvant therapy in females with groin node metastases. In three reported cases,[14-16] no adjuvant therapy was administered, and no relapses were observed after 12, 16, or 38 months. GOG37 suggested that radiation following radical vulvectomy and inguinal lymphadenectomy significantly decreased cancer-related deaths and reduced local relapses.[17] Taking this into consideration, we opted for adjuvant radiotherapy in line with the GOG37 recommendation, despite the fact that intestinal-type adenocarcinoma may not be highly sensitive to this treatment.

There are considerable benefits to pelvic exenteration from minimally invasive surgery (MIS) performed laparoscopically with a higher magnification view. MIS is used in approximately 3% of pelvic exenteration procedures for gynecological malignancies.[18] In this case, laparoscopic PPE was performed for intestinal-type adenocarcinoma of the vulva, ensuring an adequate safety margin. Laparoscopic rectal resection was feasible due to the absence of intra-abdominal disease and no history of pelvic surgery. In addition, a laparoscopic hysterectomy was performed to prevent pyometra due to vaginal stenosis. In cases where uterine preservation was performed, postoperative complications, such as infection and partial detachment of the skin flap, resulting from wound-closing tension have been reported.[19] It was hypothesized that by performing a concurrent hysterectomy, wound-closing tension would be relieved during the closure of the vagina and skin flap. The mobility of the remaining vagina increased after hysterectomy, making the closure relatively easier, and no postoperative complications were observed. However, further accumulation of such cases is necessary to determine the appropriateness of performing hysterectomy in cases without

uterine lesions. Previous studies have shown that pelvic exenteration performed using MIS resulted in significantly less intraoperative blood loss than laparotomy (900 vs. 1550 mL, $P < 0.01$).[20] This translates to a reduced need for blood transfusion, despite a longer mean operative time (640 vs. 432 min, $P < 0.01$). Similarly, in our case, the patient did not require blood transfusion despite the longer operative time required for bilateral inguinal lymph node dissection and perineal reconstruction. This case report of a hybrid procedure is valuable because we combined laparoscopic PPE with radical vulvectomy, bilateral inguinal lymph node dissection, perineal reconstruction using a gracilis musculocutaneous flap, and colostomy for the management of intestinal-type vulvar adenocarcinoma.

Laparoscopic PPE can be considered a minimally invasive approach for vulvar tumor when complete resection is achievable with an appropriate safety margin.

Ethics approval

This study was approved by the appropriate research ethics committee (Approval number: 2018-120).

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Author contributions

T.N. and M.K-Kato designed the study. T.N. wrote the initial draft of the manuscript. M.K-Kato and H.Y. assisted with manuscript preparation. All other authors contributed to the critical manuscript review. All authors have approved the final version of the manuscript and agreed to be accountable for all aspects of the study. We ensured that questions related to the accuracy or integrity of any part of the study were appropriately investigated and resolved.

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Data availability statement

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

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Conflicts of interest

There are no conflicts of interest.

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