A Rare Case of Isolated Primary Endometrial Carcinoma of the Abdominal Wall

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Abstract:

Endometriosis is a common disease in women of reproductive age that is characterized by the occurrence of endometrial tissues outside of the uterus. In some rare cases, sites of endometriosis can also potentially develop into carcinoma. We present a case of a patient found to have a rare form of primary endometrial carcinoma of endometrial cells in the abdominal wall that were suspected to be seeded from a prior abdominal surgery (cesarean section) performed nearly 25 years ago. There is not much known in terms of favorable treatment plans and the prognosis is overall generally poor.

Introduction:

In most developed countries, carcinoma of the uterus is the most frequently diagnosed gynecological malignancy, with a peak incidence between 65-74 years of age, usually in postmenopausal women [1]. In the US, it affects 1-2% of women, with roughly 20-30 new cases per 100,000 women per year. Furthermore, it is the fourth most common cancer in women (after breast, lung, and colorectal cancer). Endometrial cancers can generally be divided into two histological types, the majority of which are those of endometrial origin (Type I) that tend to affect women who are approaching menopause. On the other hand, Type II endometrial cancers (which make up roughly 10-20%) are cancers that originate mostly from other cell types, such as serous, clear cell, mucinous, and even undifferentiated cells.

Several risk factors are associated with the development of endometrial carcinoma, the most important factor being long-term exposure to increased estrogen levels. This would include things such as nulliparity, early menarche and late menopause, PCOS, obesity, and unopposed estrogen replacement therapy to name a few. Additionally, some genetic mutations, such as in the PTEN tumor suppressor gene, are also associated with Type I endometrial cancers. Conversely, protective factors would include low estrogen states or high progestin/progesterone levels; this would include things such as multiparity, combination OCPs, and even regular physical exercise. Type II cancers are usually estrogen-independent and are more associated with endometrial atrophy, and therefore post-menopausal women.

Diagnosis is commonly achieved via endometrial sampling, commonly performed as a part of pelvic exams. Alternatively, a biopsy can be obtained via hysteroscopy or dilation & curettage. Pathologically, positive results would reveal endometrial hyperplasia either with or without atypia. Furthermore, results could also reveal a pronounced proliferation of disorganized glandular tissue, which is characteristic of endometrial adenocarcinoma. Additionally, imaging (i.e. transvaginal/abdominal ultrasonography, CT, MRI, etc.) can also play a role in identifying abnormalities in structures and assessing for any metastatic spread commonly found in the lungs and pelvis. There are no routine screening tests for endometrial cancer as there are for cervical and breast.

Endometriosis is a common and chronic disease in women of reproductive age that is characterized by the occurrence of endometrial tissues outside of the uterus [2]. In some extremely rare cases, sites of endometriosis can also potentially develop into sites of carcinoma, which is what this case study hopes to explore.

Clinical Course:

A 45-year-old female, with no significant past medical history, presented to the emergency department with complaints of an abdominal lump in her left lower quadrant that she had noticed two days prior. The patient also noted some drainage of red-tinged fluid as well. She was alert, and comfortable and did not present with any acute distress. At that time, the patient denied any abdominal pain, nausea, vomiting, fevers, chills, changes in bowel habits, or any other gastrointestinal symptoms. She also denied any gynecological symptoms and noted that she was still menstruating. She did admit to having a previous cesarean section nearly 25 years ago. The patient also mentioned an intentional 80 lb weight loss in the previous few months while on a Weight Watchers diet. Vital signs were unremarkable except for tachycardia of 120 beats per minute (BPM). A complete review of systems was wholly negative. Physical examination revealed a soft, non-distended, and non-tender abdomen with cystic fluid collection in the left lower quadrant that was inferior to the lateral edge of a prior C-section scar. A slightly mobile 3 cm mass in the subcutaneous space could be felt in the left lower quadrant below the incision site. Additionally, deep to the incision site was an additional 4-5 cm mass that seemed adherent to the surrounding tissue with an overlying large fluid collection; no obvious cellulitis or inflammatory changes on the skin surface could be appreciated. Compression of the area expressed a large amount of thin hemorrhagic fluid with air bubbles. General surgery was consulted; the wound was incised, drained, and repacked with ribbon gauze. The remainder of the abdomen, as well as the physical exam, was unremarkable. Lab work was significant for low Hgb levels of 9.9 g/dL and an elevated alkaline phosphatase level of 146 units/L. Blood cultures were negative. The patient was started on clindamycin.

Initial CT imaging revealed a large complex soft tissue mass with fluid and multiple pockets of air in the left ventral abdominal wall of the lower abdomen involving the rectus abdominis muscle measuring 10x11x12cm. Mass was concerning for abscess and possible malignancy. Additional CT findings included enlarged left inguinal and left iliac chain lymph nodes. Follow-up transvaginal ultrasound imaging of the abdomen revealed a normal appearance of the uterus and right ovary, with non-visualization of the left ovary; there was redemonstration of a cystic mass, similar to what was found on abdominal CT. Repeat CT abdomen imaging revealed an unchanged solid cystic mass and lobulated lesions of the left hemipelvis.

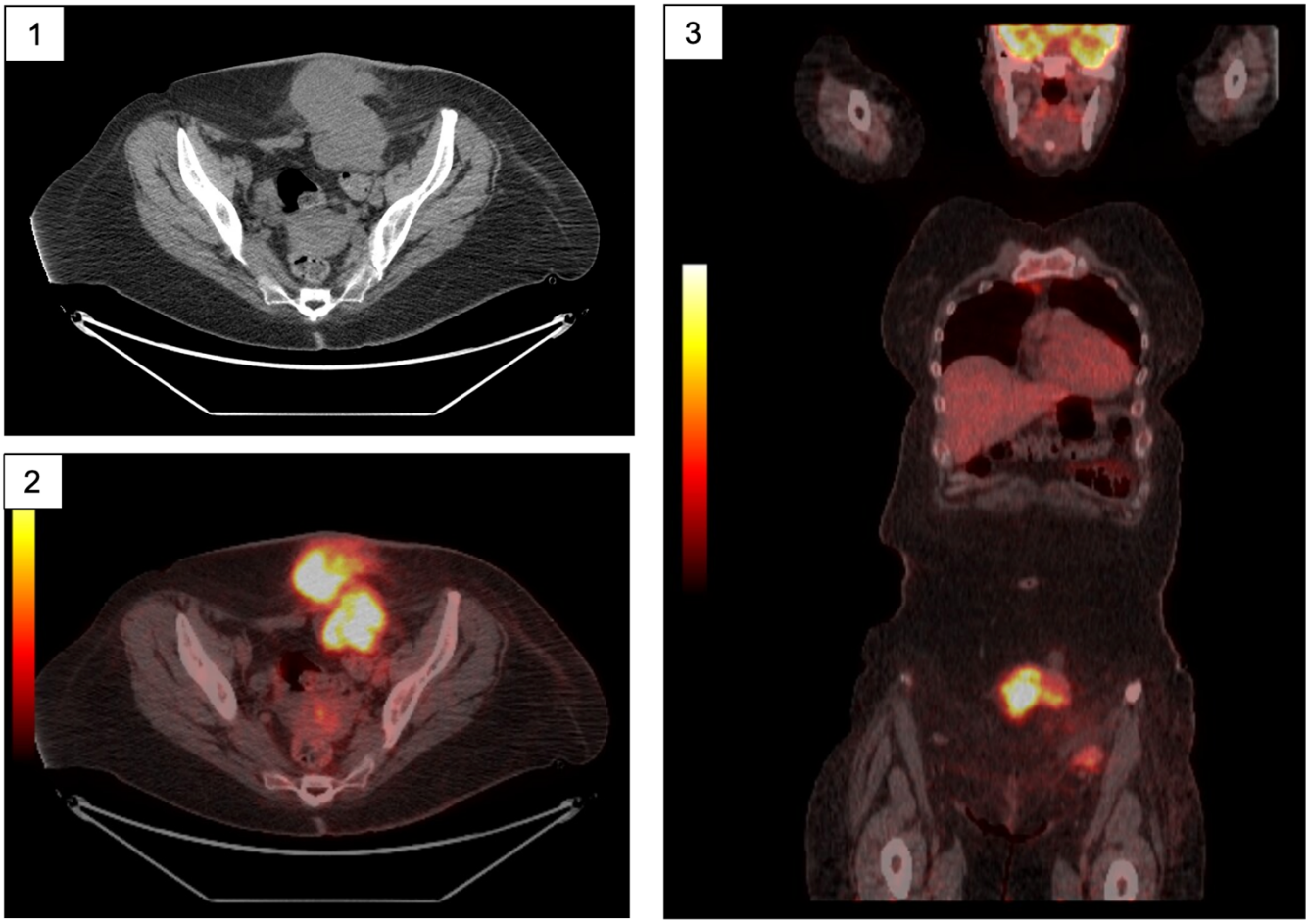

Figure 1: CT imaging revealing complex soft tissue mass in the left ventral abdominal wall.
Figure 2: PET CT of hypermetabolic soft tissue mass from Figure 1, extending through the abdominal wall into the subcutaneous tissue.
Figure 3: PET CT skull-base to mid-thigh revealing the hypermetabolic soft tissue mass of the left ventral abdominal wall.

The patient underwent an exploration and evacuation of the abdominal wall hematoma. Surgical examination suggested that the mass was arising from the abdominal wall itself, within and on the surface of the rectus sheath, which was consistent with CT findings. An abdominal wall incisional biopsy of the mass revealed soft tissue that was consistent with endometrial carcinoma with positive immunostains for CA-19-9, CA125, and increased P53 expression. The next step was to determine if the mass was primary, arising in endometriosis or metastasis from an ovarian or uterine primary cancer. A follow-up PET scan revealed a large lobulated and markedly hypermetabolic soft tissue mass within the left anterior pelvis that extended through the abdominal wall to the subcutaneous soft tissues, consistent with a primary malignancy. There was also a soft tissue mass at the apex of the right adrenal gland with hypermetabolic activity and a Standardized Uptake Value (SUV) of 12.03 concerning for adrenal metastatic disease. Final pathology reports confirmed a diagnosis of aggressive clear cell carcinoma. Furthermore, cytology from cervical swab specimens revealed atypical squamous cells of undetermined significance.

The patient followed up with Gynecology/Oncology and was agreeable to 3-6 cycles of chemotherapy (carboplatin + taxol) with consideration for surgery thereafter.

Discussion:

One of the rarest forms of endometriosis is that of the abdominal wall; this includes cesarean scar endometriosis. It remains overall a challenging condition, and the increasing number of cesarean sections and laparotomies will likely increase the rate of abdominal wall endometriosis. Even further, a synchronous development of endometrioid-type endometrial carcinoma originating from the foci of an abdominal wall endometriosis is an extremely rare phenomenon, with only ~50 cases of endometriosis-associated abdominal wall cancers recorded as of 2021. A study by Mihailovici et al. analyzed data from 48 cases with endometriosis-associated abdominal wall cancers [3]. All patients had undergone uterine surgery of some form, mostly C-sections. The data showed that the average time between the initial surgery and the diagnosis of cancer was roughly 19 years with a confidence interval of about 8 years. The patient in our case study had undergone her C-section roughly 25 years ago, which is within the average range of the study above. Similarly, the study also revealed that while the surgery occurred nearly 2 decades ago on average prior to diagnosis of carcinoma, symptoms only began within 6 months of diagnosis, which was also the case in our case study as our patient had only begun to experience her symptoms within a week. An important thing to point out is that only this, and at least one other study, report the development of endometrioid-type endometrial cancer that originates from the foci in scar tissue of the abdomen likely secondary to seeding of tissue from prior uterine surgery as what was documented. Malignant transformation of an endometriosis associated with surgical scars is extremely rare, with an estimated incidence of less than 0.3% [4]. Given the rare nature of this event, with only very few numbers of cases recorded in literature, there is no standardized treatment. Adjuvant therapy can be beneficial, which is what our patient had begun. Further reports show that in addition to adjuvant chemotherapy and radiotherapy, done in 74% and 30% of cases respectively, surgical treatment is common [5]. Some studies suggest that the recorded prognosis of this condition is poor, however, it is unclear if this is in regards to more common conditions where there was a metastasis from primary uterine carcinoma to the abdominal wall.

Conclusion:

Given the rare nature of this condition, more literature and investigation of recorded treatment approaches and outcomes are needed to specify a favorable treatment plan. The exact mechanism of abdominal wall endometrial carcinoma can usually be explained by hematogenous dissemination from the site of trauma, with seeding of neoplastic cells after direct contact between the tumor and the wound. However, in this case study, the primary tumor is believed to be the site of the abdominal wall itself, with negative findings of cancer in the uterus itself. Further studies with long-term results are needed to determine an optimal approach to this rare condition.

References:

1. Luz R, Leal R, Simões J, Gonçalves M, Matos I. Isolated abdominal wall metastasis of endometrial carcinoma. Case Rep Obstet Gynecol. 2014;2014:505403. doi:10.1155/2014/505403
2. Carsote M, Terzea DC, Valea A, Gheorghisan-Galateanu AA. Abdominal wall endometriosis (a narrative review). Int J Med Sci. 2020;17(4):536-542. Published 2020 Feb 10. doi:10.7150/ijms.38679
3. Mihailovici A, Rottenstreich M, Kovel S, Wassermann I, Smorgick N, Vaknin Z. Endometriosis-associated malignant transformation in abdominal surgical scar: A PRISMA-compliant systematic review. Medicine (Baltimore). 2017;96(49):e9136. doi:10.1097/MD.0000000000009136
4. Paulino E, de Melo AC, da Silva VF. Endometrioid Carcinoma Arising from an Endometriosis-Associated Abdominal Wall Scar. Am J Case Rep. 2020;21:e922973. Published 2020 Jun 1. doi:10.12659/AJCR.922973
5. Şahin O, Tokgözoğlu N, Taşcı T. Localized endometrioid cancer in the abdominal wall with synchronous early-stage endometrial cancer. Journal of Turkish Society of Obstetric and Gynecology. 2021;18(3):264-266. doi:10.4274/tjod.galenos.2021.85530