Case Report

Clear Cell Carcinoma Arising from Endometriosis in a C-section Scar: A Case Report with Literature Review

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Endometriosis is a frequent benign disorder. Malignancy arising in association with endometriosis mainly includes endometrioid carcinoma (70%), sarcoma (25%) and clear cell carcinoma (5%). The incidence rate of abdominal surgical scar endometriosis is between 0.03% and 1.08% of women undergoing pelvic surgery. Malignant transformation arising on the background of scar endometriosis is extremely rare.

Here, we report a case of clear cell carcinoma arising from endometriosis in a cesarean section scar in a 48-year-old woman. The patient presented to the emergency department with a palpable abdominal mass and constipation. A CT scan revealed a mass in the infraumbilical suprapubic rectus abdominis muscle region at the site of a cesarean section scar. A biopsy of this mass revealed an epithelioid neoplasm with papillary features, which is positive for AE1/AE3, PAX-8, and Napsin, suspicious for malignancy of gynecologic or upper urinary tract origin. The patient underwent an exploratory laparotomy, abdominal wall resection and reconstruction, and total abdominal hysterectomy with bilateral salpingo-oophorectomy. The resulting specimen included a 13.7 x 10.0 x 7.0 cm portion of red-yellow muscle and adipose tissue from the abdominal wall. Microscopic examination showed clear cell carcinoma arising from endometriosis in a cesarean section scar. ER, PR, and FOLR1 are all negative, compatible with the diagnosis. There was also metastasis to 6 out of 14 total lymph nodes submitted. The omentum, appendix, uterus, ovaries, fallopian tubes, and cervix were all benign, with the uterus containing a disordered and proliferative endometrium.

Although clear cell carcinoma arising from malignant transformation of endometriosis in the abdominal wall after cesarean section is extremely rare, for a female patient with a history of gynecologic or obstetric surgery, developing an abdominal wall mass, the possibility of a primary malignancy arising from endometriosis should be considered in the differential diagnosis. A tissue biopsy with histological evaluation and ancillary studies should be performed to ensure early detection of malignancy with proper next step treatment.
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Key Words: *Endometriosis, Malignant Transformation, Clear Cell Carcinoma, Abdominal Wall Scar*

INTRODUCTION

Endometriosis is a condition defined by the presence of endometrial-like tissue outside the uterine cavity, commonly involving the ovaries, peritoneum, and pelvic organs.[1] While the condition is generally benign, malignant transformation can occur, with histological subtypes such as endometrioid carcinoma, sarcoma, and CCC being least common.[2]

Abdominal wall scar endometriosis is an uncommon complication following gynecologic or obstetric surgeries, particularly C-sections.[3] Malignancy arising from scar endometriosis is extremely rare, with only a few cases documented in the literature. This report highlights a case of CCC arising from endometriosis in a C-section scar and reviews its clinical, diagnostic, and therapeutic implications.

CASE PRESENTATION

A 48-year-old female presented to the emergency department with a palpable abdominal mass and constipation. Her past medical history included diabetes, hypertensive disorder, and polycystic ovary syndrome. She had undergone one C-section, performed over 20 years ago. Her family history was significant for breast cancer in her sister, mother, and maternal grandmother, however, genomic testing was negative.

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Initial evaluation with a CT scan revealed a 13.7 x 10.0 x 7.0 cm mass in the infraumbilical suprapubic rectus abdominis muscle at the site of her C-section scar. A core biopsy of the mass revealed an epithelioid neoplasm with papillary features. Immunohistochemistry was positive for AE1/AE3, PAX-8, and Napsin, suggesting malignancy of gynecologic or upper urinary tract origin.

Based on these findings, the patient underwent an exploratory laparotomy, abdominal wall resection and reconstruction, and total abdominal hysterectomy with bilateral salpingo-oophorectomy. The abdominal wall resection yielded a 13.7 x 10.0 x 7.0 cm portion of red-yellow muscle and adipose tissue. Serial sectioning of the mass revealed cut surfaces ranging

from tan-pink and papillary to tan-white and fibrous with diffuse hemorrhagic cystic areas.

Histological evaluation confirmed CCC arising from endometriosis within the C-section scar (**Figure 1**). Immunohistochemical staining was negative for ER, PR, and FOLR1, consistent with the diagnosis. Lymph node analysis identified metastatic carcinoma in 6 out of 14 lymph nodes received (**Figure 2**). The remaining specimens, including the omentum, appendix, uterus, ovaries, fallopian tubes, and cervix, were grossly and microscopically unremarkable. The uterus showed a disordered and proliferative endometrium without evidence of malignancy.

Postoperatively, the patient completed six cycles of chemotherapy with paclitaxel and carboplatin. A subsequent CT scan unfortunately revealed new and progressive retroperitoneal lymphadenopathy, including worsening left common iliac lymphadenopathy.

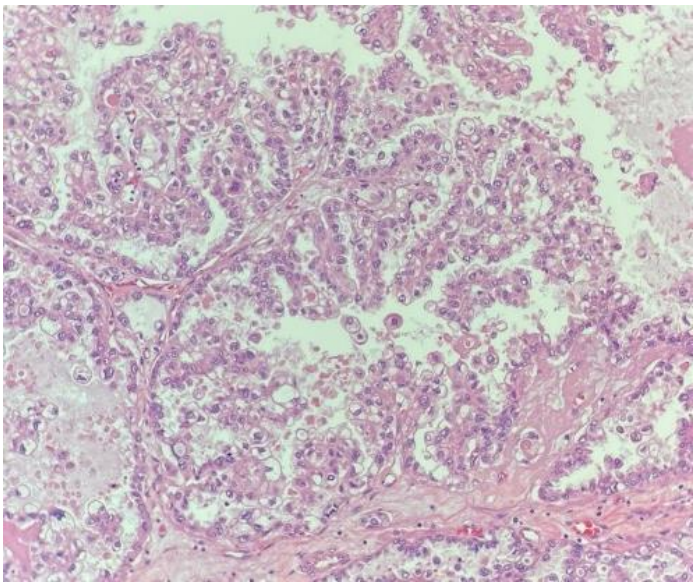

Figure 1. Hematoxylin and eosin stain (400x magnification) of the abdominal wall mass showing a papillary architecture of cells with a clear cytoplasm, prominent nucleoli, and pleomorphism, consistent with clear cell carcinoma.

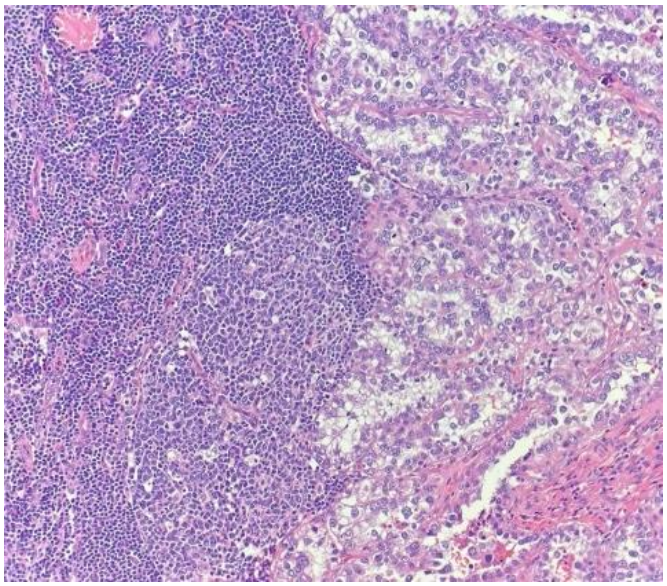

Figure 2. Hematoxylin and eosin stain (200x magnification) of a left pelvic lymph node showing metastatic clear cell carcinoma.

DISCUSSION

The development of CCC in association with endometriosis is rare, particularly in abdominal wall scars. While the exact mechanism underlying malignant transformation is not fully understood, there are multiple proposed theories. Chronic inflammation associated with ectopic endometrial tissue is thought to create a pro-tumorigenic environment due to repeated cycles of tissue injury and repair potentially driving genetic instability.[4] Hormonal influences, particularly estrogen and progesterone, may stimulate the proliferation of ectopic endometrial cells.[5] Also, mutations in genes involved in cell growth and survival, such as ARID1A and PIK3CA, have been implicated in the pathogenesis of CCC.[5] These genetic alterations are frequently observed in CCC arising in other settings and may play a role in the transformation of endometriosis into malignancy.[5]

Scar endometriosis is believed to occur from the direct implantation of endometrial cells into surgical wounds during pelvic procedures.[3] These implanted cells may persist and proliferate under the influence of ovarian hormones, forming endometriotic lesions in the scar tissue.[5] While benign scar endometriosis typically presents within a few years of the surgical procedure, malignant transformation can occur years later, as seen in this case.

The clinical presentation of abdominal wall masses associated with scar endometriosis often mimics conditions such as hernias, appendicitis, ectopic pregnancy, or desmoid tumors.[6] Patients may present with nonspecific symptoms such as localized pain, swelling, or mass effect symptoms, such as constipation, as seen in this patient.[6] Imaging studies, particularly CT or MRI, can provide more information regarding the lesion and help characterize features. However, histological evaluation remains the definitive diagnostic method as many benign and malignant conditions have overlapping radiographic appearances. As seen in this case, histopathology and immunohistochemistry provided crucial information to distinguish CCC from other neoplasms.

Due to the rarity of this malignancy, there is no standard treatment plan. The most widely accepted treatment in the literature is surgical excision with adjuvant chemotherapy, most commonly a combination of paclitaxel and carboplatin, as was given to the patient in this case.[7] Radiation therapy is particularly considered in cases of lymph node involvement or recurrent disease. Any defect in the abdominal wall was also repaired, and some patients underwent endometrial curettage or hysterectomy, bilateral salpingo-oophorectomy, omentectomy, and lymph node resection.[7] The patient in this case received all of these treatments due to the severity and spread of her disease. Unfortunately, this patient experienced disease progression despite chemotherapy and was evaluated for radiation therapy.

The prognosis of CCC arising from an abdominal wall C-section scar is generally poor due to its aggressive nature and the tendency for late diagnosis.[8] This rare malignancy often presents decades after the initial surgical procedure by which time it may already be advanced and include lymph node involvement or metastasis. Surgical resection with negative margins offers the best chance for local control, but recurrence is common and lymph node metastasis significantly worsens outcomes.[8]

A review of the literature reveals limited cases of CCC arising from scar endometriosis, emphasizing its rarity. Most reported cases emphasize the importance of early detection, as delayed diagnosis often results in advanced disease with a poorer prognosis. The importance of surgical management and the multidisciplinary collaboration among surgeons, oncologists, and radiologists are also emphasized to optimize outcomes for patients with this rare condition.

CONCLUSION

CCC arising from scar endometriosis is an exceedingly rare but important differential diagnosis for abdominal wall masses in women with a history of gynecologic surgery. This case highlights the need for heightened clinical suspicion, particularly in patients presenting with abdominal wall masses and a history of C-section.

Definitive diagnosis requires histopathological evaluation and immunohistochemical studies. Early recognition and surgical intervention are critical for optimal patient outcomes. Further research is needed to better understand the pathogenesis, establish treatment guidelines, and improve outcomes for this rare condition.

CONFLICTS OF INTEREST
None.

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