



Case Report

Clear cell carcinoma arising from scar endometriosis: A case report and literature review

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ABSTRACT

Scar endometriosis is uncommon and defined as the presence of ectopic endometrial glands in abdominal soft tissues after a gynecological operation. Malignant transformation has been reported but remains rare. Carcinogenesis occurs in ectopic endometrial tissue with repeated hormone stimulation during the menstrual cycle. We present a case of clear cell carcinoma directly arising from scar endometriosis after a cesarean section and review all 16 cases reported.

KEYWORDS: *Cesarean section, Clear cell carcinoma, Endometriosis*

INTRODUCTION

Endometriosis indicates functional endometrial tissues outside the uterus. It may be identified in abdominal scars after gynecological surgery and can cause pain during menstrual periods and abdominal swelling. Malignant transformation is extremely rare. Here, we present a 46-year-old patient with clear cell carcinoma (CCC) in an abdominal scar after a cesarean section.

CASE REPORT

Our patient was a gravida 2, para 1 with irregular menstrual periods and had undergone a cesarean section 18 years previously. She visited the plastic surgery department and complained about a tender mass with ulceration in the right lower abdominal wall for 2 months. Physical examination revealed 2 firm, immobile masses over the lower abdomen. The right mass was erythematous with central ulceration. Computed tomography revealed 7.0 and 9.5 cm lobulated tumors with focal cystic changes, located in the subcutaneous layer and extending into the rectus abdominis muscle. The patient underwent wide local excision.

The tumors excised from the right and left sides of the abdomen measured 7.0 cm × 5.3 cm × 4.0 cm and 6.3 cm × 3.2 cm × 2.5 cm, respectively. The overlying skin was ulcerated. On sectioning, the tumors were lobulated with an infiltrating border. Focal myxoid changes and

hemorrhage were observed. Microscopically, the tumors were composed of cysts of varying sizes lined by a single layer of flattened or cuboidal cells with atypical hyperchromatic nuclei with a clear cytoplasm [Figure 1]. Marked nuclear atypia with pleomorphism and prominent nucleoli was noted [Figure 2]. The stroma was fibrotic and hyalinized. Residual ectopic endometrial glands with stromal cells were observed [Figure 3]. Therefore, CCC arising from endometriosis was considered. After the wide excision, further operation with total hysterectomy and bilateral salpingo-oophorectomy was suggested; however, the follow-up of the patient was lost.

DISCUSSION

Scar endometriosis typically results from the implantation of normal endometrial tissues after gynecological or obstetric surgeries. The condition has an incidence of 0.03%–0.4% and may cause abdominal swelling and pain with cyclic changes during menstruation. The combination of oxidative stress from recurrent hemorrhage, inflammation, and hyperestrinism contributes to the tumorigenesis of endometriosis-related neoplasms. The overall frequency of malignant

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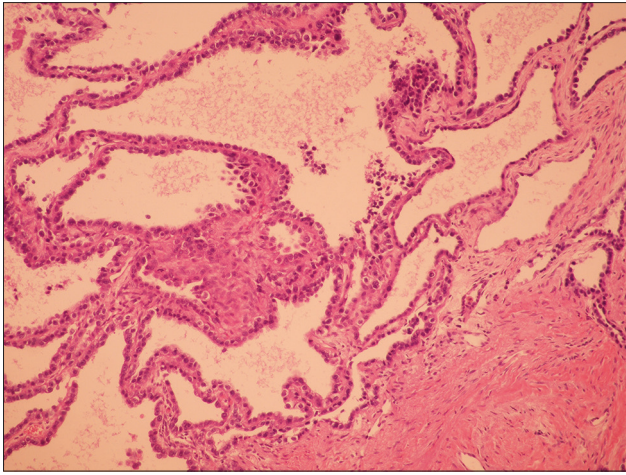


Figure 1: Multiple cysts lined by a single layer of low cuboidal epithelia are seen. The tumor cells contain atypical hyperchromatic nuclei (hobnail cells). Intraluminal secretion is also seen (hematoxylin and eosin stain, ×100)

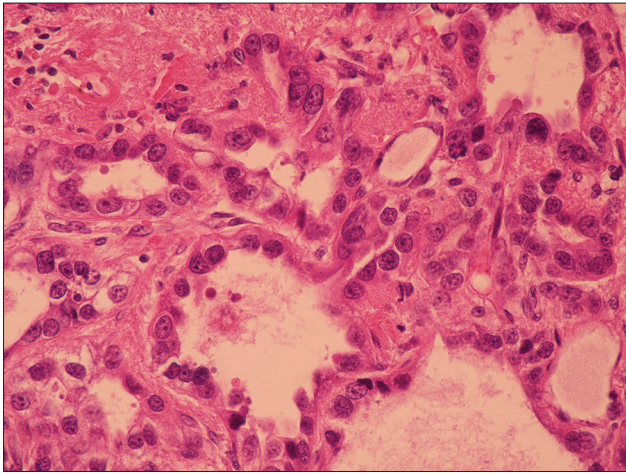


Figure 2: Tumor cells with hyperchromatic and pleomorphic nuclei and conspicuous nucleoli are seen (hematoxylin and eosin stain, ×400)

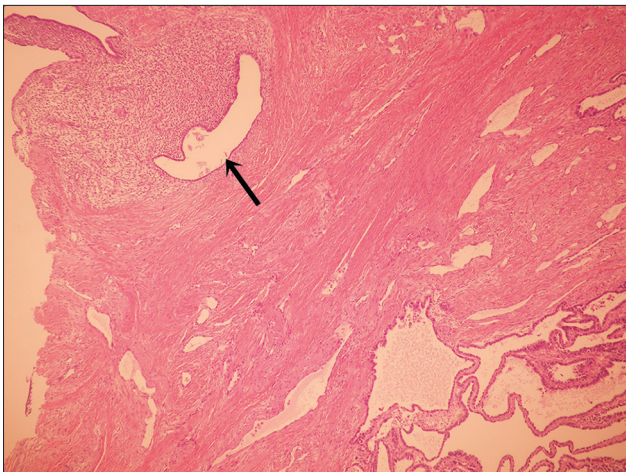


Figure 3: Benign ectopic endometrial glands can be identified at the arrow (hematoxylin and eosin stain, ×40)

transformation from endometriosis is estimated to be up to 1%. To date, only 22 cases of carcinoma arising from scar endometriosis have been reported. Among these cases, the development rate of CCC was approximately 77.3% (17/22), followed by endometrioid carcinoma (13.6%, 3/22) and serous carcinoma (0.09%, 2/22). Notably, CCC is the most common malignancy arising from extragonadal endometriosis. Therefore, we analyzed all 17 cases of CCC associated with scar endometriosis, including our case, and the details are shown in Table 1 [1-15]. Sixteen of the 17 patients (94.1%) had a surgical history of cesarean section, and the remaining one patient had undergone a laparoscopic cystectomy. Because cesarean section is the most common obstetric surgery in reproductive-aged women and may provide a long period to allow carcinogenesis, the duration from the time of surgery to CCC diagnosis varied from 9 to 30 years (mean, 17.9 years) in these cases. Most cases were originally diagnosed as a subcutaneous mass, including a simple cyst, chronic abscess and others. Only three patients had received surgery for abdominal endometriosis and local recurrence was then suspected. However, none was initially diagnosed with a malignancy.

When diagnosing a malignancy from preexisting endometriosis, the presence of benign endometrial tissue in the tumor is a key point. Histopathology revealed differentiating CCC in all cases; however, preexisting benign endometrial glands were identified in only 11 cases (64.7%, 11/17). In our case, benign endometrial tissue was observed in only a small focus from the periphery of the malignant lesion, which indicated an old hemorrhage, and was more fibrotic on gross inspection. Thus, examination of multiple sections is required and highly recommended to establish this diagnosis.

All 17 patients underwent local excision, of whom ten underwent bilateral salpingo-oophorectomy and total hysterectomy (58.8%). However, no malignant lesion was observed on uterine and ovary resection. After excising the primary tumor, adjuvant therapies with chemotherapy, radiotherapy, or a combination were administered in 16 cases. The mean follow-up duration was 14.7 months. Local recurrence in one case was attributed to a positive resection margin. Five patients died of disease (29.4%), mostly due to extensive, generalized metastasis. Particularly, three of these five cases had involvement of inguinal and pelvic lymph nodes at the initial presentation. In our case, local excision was performed; however, the basal resection margin and rectus abdominis muscle were involved by tumor cells. An additional radiotherapy session was subsequently conducted for

Table 1: The reports of 17 cases with clear cell carcinoma arising from abdominal wall endometriosis

References	Age (years)	Previous surgery	Duration (years)	Coexisting endometriosis	Surgery	Adjuvant therapy	Follow up (month)	Outcome
Shalin <i>et al</i> [1].	47	CS	Unclear	+	LE	CT + RT	7	NED
Miller <i>et al</i> [2].	38	CS	9	+	LE + ATH + BSO	RT	60	NED
Park <i>et al</i> [3].	54	CS	26	+	LE	RT	1.5	NED
Harry <i>et al</i> [4].	55	CS	30	+	LE	RT	18	NED
Bats <i>et al</i> [5].	38	CS	13	+	LE + ATH + BSO	CT	2	NED
Williams <i>et al</i> [6].	53	CS	17	-	LE + ATH + BSO	CT + RT	11	DOD
Matsuo <i>et al</i> [7].	37	LC	10	-	LE + ATH + BSO + Omen + PLD	CT	18	Recurrence
Bourdel <i>et al</i> [8].	43	CS	Unclear	Unclear	LE + ATH + BSO	CT + RT	22	DOD
Mert <i>et al</i> [9].	42	CS + RSO	Unclear	+	LE + ATH + LSO	CT	1	NED
	51	CS + ATH	Unclear	+	LE + BSO	RT	1	NED
Alberto <i>et al</i> [10].	38	CS + ATH + BSO	11	-	LE	CT + RT	3	NED
Sawazaki <i>et al</i> [11].	41	CS	18	+	LE + partial cystectomy	CT	Unclear	Unclear
Schnieber and Wagner-Kolb [12]	40	CS	15	+	LE + ATH + BSO	RT	18	DOD
Hitti <i>et al</i> [13].	46	CS	14	+	LE + ATH + BSO	-	30	NED
Ishida <i>et al</i> [14].	56	CS	24	-	LE	RT	48	DOD
Sergent <i>et al</i> [15].	45	CS	28	-	LE + BSO	CT	6	DOD
Case	46	CS	18	+	LE	RT	3	NED

CS: Cesarean section, LC: Laparoscopic cystectomy, RSO: Right salpingo-oophorectomy, ATH: Abdominal total hysterectomy, BSO: Bilateral salpingo-oophorectomy, LE: Local excision, Omen: Omentectomy, PLD: Pelvic lymph node dissection, LSO: Left salpingo-oophorectomy, CT: Chemotherapy, RT: Radiotherapy, NED: No evidence of disease, DOD: Died of disease

local control. Abdominal total hysterectomy and bilateral salpingo-oophorectomy were also suggested to determine the possibility of a coexisting malignancy. Because of the rarity of this disease, it is difficult to establish a standard treatment protocol, and no appropriate disease staging system is applicable. Most cases were treated as primary ovarian CCC with metastasis, and radical surgery with adjuvant chemo- or radiotherapy was performed.

CCC arising from scar endometriosis remains a rare phenomenon. The most affected population is women who undergone operations such as myomectomy or cesarean section that may introduce ectopic endometrial tissues into abdominal soft tissues. No definite treatment guidelines are presently available because of disease rarity. However, wide local excision and radical surgery with total hysterectomy and bilateral salpingo-oophorectomy are indicated in most cases if coexisting malignancy can be excluded. Pelvic and inguinal lymph node dissection is also recommended to predict distant metastasis and a poor outcome if any positive node is present. Adjuvant chemo- or radiotherapy may provide some benefits in local control and metastatic diseases; however, the

effect on prognosis, including the overall survival rate or progression-free survival, remains unclear.

We recommend that patients with a history of gynecological surgeries complicated by scar endometriosis undergo total wide excision with a free surgical resection margin, if possible. Furthermore, the possibility of malignant transformation should be considered and close follow-up is needed.

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Conflicts of interest

There are no conflicts of interest.

Declaration of patient consent

The authors certify that the patient have obtained appropriate consent form. 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 personal data will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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