Gynecologic Oncology Reports 45 (2023) 101139

Contents lists available at ScienceDirect

Gynecologic Oncology Reports

journal homepage: www.elsevier.com/locate/gynor

Case report

Carcinosarcoma of the uterus, derived from subserous cystic adenomyosis, presenting as an acute abdomen: A case report and review of the literature

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ARTICLE INFO

Keywords:
Acute abdomen
Carcinosarcoma
Cystic adenomyosis
Malignant transformation
Subserous

ABSTRACT

When a woman presents with an acute abdomen with cystic lesions in the abdominal cavity, the differential diagnosis includes torsion or rupture of an ovarian tumor. We report our experience with a 54-year-old nulliparous woman who underwent emergency surgery for a suspected ruptured ovarian tumor. Intraoperative examination revealed disruption of a cystic tumor that had developed externally from the fundus of the uterus. The patient, who was taking aspirin because of a history of medullary infarction, reported lower abdominal discomfort for several days. When she sought care, she was referred to the gynecology department where transvaginal ultrasonography and contrast-enhanced computed tomography showed a poorly toned mass with a maximum diameter of 20 cm posterior to the uterus. She also had a large amount of ascites reaching around the liver and the spleen.

She underwent an emergency laparotomy for a presumed diagnosis of acute abdomen caused by a ruptured ovarian tumor with intra-abdominal bleeding. Intraoperative examination revealed normal adnexae bilaterally, but there was a cystic tumor in the pouch of Douglas that was strongly adherent to the surrounding intestines. This mass was connected to the posterior uterus by a stalk and appeared to be continuous with the uterine tissue. The postoperative pathological diagnosis was carcinosarcoma derived from subserous cystic adenomyosis. This is the first case report of carcinosarcoma developing from subserous cystic adenomyosis in the English literature as far as we know.

1. Introduction

Uterine adenomyosis is typically diffuse, but it can occur as a focal lesion. (Siegler and Camilien, 1994) Adenomyosis consists of heterotopic endometrial glands and stroma, and 11.7% of the cases are located in the subserous space. (Sakamoto, 1991) Cystic degeneration is often seen in leiomyomas but rarely reported in adenomyomas. (Calagna et al., 2015) Cystic adenomyosis is a rare type of adenomyosis that most often occurs in women of reproductive age. Because there are few case reports of cystic adenomyosis, its clinical characteristics have not been definitively established.

Noncystic adenomyosis is a common gynecological condition; the heterotopic tissue can serve as a nidus for endometrial cancer (Habiba et al., 2018) or clear cell carcinoma. (Baba et al., 2016) Here, we report our experience with a patient who underwent emergency surgery for a suspected rupture of an ovarian tumor but who ended up having a very rare condition: disrupted cystic adenomyosis that developed externally from the fundus of the uterus and that had a component of carcinosarcoma.

2. Case presentation

A 54-year-old nulliparous Japanese woman had a medical history of medullary infarction, vertebral artery dissection, hyperlipidemia, and rheumatoid arthritis. Her medications included aspirin, folic acid, lansoprazole, a statin, and methotrexate. She had been aware of lower abdominal discomfort for several days; when she sought care, she was referred to the gynecology department. Her vital signs at the initial visit were stable, with a blood pressure of 118/76 mm Hg, a pulse of 77, and a temperature of 37.6 ℃. There was marked tenderness and rebound pain over her entire abdomen. Laboratory analysis showed evidence of anemia, with a hemoglobin of 7.8 g/dL and a hematocrit of 25.8%. Her

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https://doi.org/10.1016/j.gore.2023.101139
Received 24 October 2022; Received in revised form 23 December 2022; Accepted 17 January 2023
Available online 21 January 2023

cancer antigen-125 (CA-125) level was elevated to 264.8 U/mL. Transvaginal ultrasonography revealed multiple uterine fibroids and a mass > 15 cm in size on the dorsal side of the uterus. Intraperitoneal fluid accumulation was observed in the abdominal cavity, suspected to be intra-tumoral fluid or hemorrhage. Contrast-enhanced computed tomography (CT) showed a poorly toned mass posterior to the uterus with a maximum diameter of 20 cm (Fig. 1A) and a large amount of ascites reaching around the liver and the spleen (Fig. 1B). We made the decision to perform an emergency laparotomy for a presumed diagnosis of acute abdomen caused by a ruptured ovarian tumor and intra-abdominal bleeding.

At laparotomy, a large amount of dark red ascites was seen. The uterus had multiple subserous and intramuscular myomas, and both ovaries were grossly normal. A cystic tumor was present in the pouch of Douglas; this mass was tightly adherent to the surrounding intestines and dorsal uterus, and it appeared to be continuous with the posterior uterine surface, emerging from that surface on a stalk. The tumor was ruptured and actively bleeding; hence, we ligated the suspensory ligament of the ovary and the uterine artery to cut off blood flow to the upper uterus. We then dissected the tumor and surrounding adhesions and removed it en bloc with the upper uterus and both adnexae. Intraoperative blood loss, including ascites, totaled about 4500 mL, and the patient received 8 units of packed red blood cells during surgery.

Postoperative pathological examination revealed multiple uterine myomas, but there were no histological abnormalities of the ovaries or fallopian tubes. A stalked cystic mass emerged from the posterior wall of the uterus (Fig. 1C, 1D), and the cyst wall showed evidence of endometriosis. The cyst contained a mixture of epithelial carcinoma components, with adenoductal and papillary structures, and mesenchymal sarcoma components that were spindle-shaped and intricately arranged (Fig. 2A, 2B). On immunohistochemical staining, the area of carcinoma was strongly positive for epithelial membrane antigen (Fig. 2C), and the area of sarcoma was strongly positive for CD10 (Fig. 2D) and vimentin. The final pathological diagnosis was carcinosarcoma. The ascitic fluid cytology was negative for tumor cells.

Three months after surgery, the patient underwent a second procedure: laparotomy to remove the residual uterine cervix, together with a pelvic lymph node biopsy and partial omentectomy. There were no residual lesions in the abdominal cavity and no malignant findings in the excised specimens. The patient received six courses of adjuvant chemotherapy with paclitaxel and carboplatin, and no evidence of recurrence has been seen since the initial emergency surgery approximately one year ago.

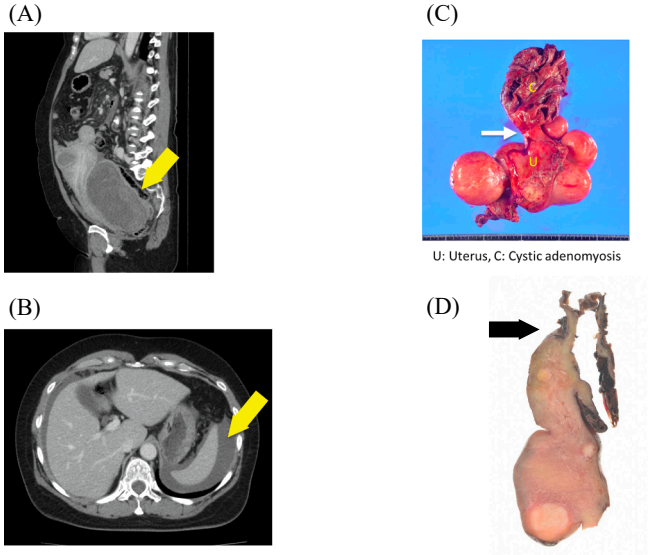

Fig. 1. Preoperative imaging and surgical specimen. A. Sagittal section on contrast-enhanced computed tomography (CT) shows a poorly toned mass on the dorsal surface of the uterus with a maximum diameter of approximately 20 cm (arrow); B. cross-sectional view on contrast-enhanced CT shows massive ascites around the spleen (arrow) and the liver; C. multiple uterine fibroids are noted in the amputated uterus; the cyst stems from the posterior wall of the uterus (arrow); D. the transition zone from the myometrium to the cystic tissue can be seen on the cut surface of the excised specimen (arrow).

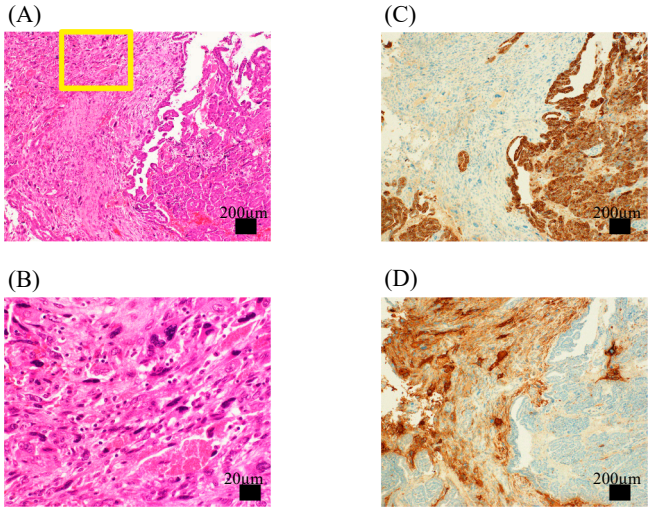

Fig. 2. Microscopic findings. A. A weakly magnified image of the cyst wall shows carcinoma components with adenoductal and papillary structures on the right and mesenchymal sarcoma components on the left; B an enlarged image of the yellow frame in part A shows spindle-shaped tumor cells that are intricately arranged; C. the cancerous part of the tumor stains positive for epithelial membrane antigen on immunohistochemistry; D. CD10 immunostaining is strongly positive only in the area of the sarcoma. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

3. Discussion

Cystic adenomyosis is rare; according to a review by Xu et al., there are even fewer cases of subserous cystic adenomyosis. (Xu et al., 2022) Cystic adenomyosis can be broadly classified into 2 groups based on the age of onset: in the juvenile-onset group, symptoms develop early after menarche; in the late-onset group, symptoms develop when patients are in their 30 s or older. (Cucinella et al., 2013) The mechanism is thought to be congenital dysplasia of the Müllerian ducts in the juvenile-onset group and an acquired cause in the late-onset group. (Cucinella et al., 2013) Our patient's age of onset and her lack of dysmenorrhea suggest a late-onset mechanism.

Although malignancy occurring in the background of uterine adenomyosis is a known entity, only 3 patients have been reported so far in whom malignant transformation was observed against a background of cystic adenomyosis (Table 1). (Ohta et al., 2008; Mori et al., 2015; Gomez et al., 2021) Two patients developed clear cell carcinoma, (Ohta et al., 2008; Gomez et al., 2021) and one developed endometrioid carcinoma. (Mori et al., 2015) Clear cell carcinoma and endometrioid carcinoma are the major histological types of malignancy that typically arise in the context of endometriosis (Koike et al., 2013); ours is the first report to describe the presence of a sarcoma component, together with carcinoma arising from subserous cystic adenomyosis.

The exact mechanism of how adenomyosis develops is still unknown, although several theories have been proposed. (Koike et al., 2013) Subserous adenomyosis may develop as a variant of pelvic endometriosis. (Sakamoto, 1991) The foremost hypothesis involving deep endometriosis is stromal invagination of the endometriosis tissue into the inner layer of the myometrium, with subsequent gland invasion.

Table 1
Review of the literature on malignant transformation of the subserosal cystic adenomyosis.

References	Age	Clinical presentation	Surgical approach	Histology	Adjubant therapy	Final outcome
Ohta et al. (Ohta et al., 2008)	54	Hypermenorrhea	TAH + BSO + OMT	Clear cell carcinoma	Chemotherapy	metastasis to the liver
Mori et al. (Mori et al., 2015)	67	Asymptomatic	TAH + BSO	Endometrioid carcinoma	Chemotherapy	NED
Gomez et al. (Gomez et al., 2021)	65	Constipation and urinary frequency	TAH + BSO	Clear cell carcinoma	None	NED
Present case	54	Acute abdomen	Supravaginal amputation + BSO	Carcinosarcoma	Surgery + Chemotherapy	NED

TAH: total abdominal hysterectomy; BSO: bilateral salpingo-oophorectomy; OMT: omentectomy; NED: no evidence of disease

(Calagna et al., 2015) Similar to the mechanism of development, there is no consensus on the definition of an adenomyotic lesion. However, several terms have been used to define these large cysts, including adenomyotic cysts, cystic adenomyoma, and cystic adenomyosis. (Cucinella et al., 2013)

The symptoms of cystic adenomyosis are nonspecific and include dysmenorrhea, chronic pelvic pain, and abnormal uterine bleeding. Cystic adenomyosis can also be asymptomatic and may not be detected until it increases in severity. The differential diagnosis of cystic adenomyosis includes subserous uterine fibroid degeneration, ovarian endometriosis, and uterine malformation. Ultrasonography and pelvic magnetic resonance imaging (MRI) are reportedly useful for differentiating between these conditions. (Ho et al., 2008) However, because of atypical clinical manifestations and rare conditions—such as in our patient—women with symptomatic cystic adenomyosis are often misdiagnosed. (Xu et al., 2022) Our patient had an acute abdomen and anemia resulting from intra-abdominal bleeding, necessitating an emergency laparotomy. Therefore, detailed preoperative imaging, including pelvic MRI, could not be performed. Even if we had been able to obtain an MRI, this imaging modality often attributes the characteristics of cystic adenomyosis to endometriotic cysts derived from the ovary. The diagnosis in this situation may be challenging, but surgeons performing emergency surgery for patients with acute abdomens need to include lesions other than ovarian tumors in the differential diagnosis, and patients should be prepared and consented for a hysterectomy.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

CRediT authorship contribution statement

Kaoru Hashizume: Data curation. **Masafumi Toyoshima:** Conceptualization, Data curation, Investigation, Writing – original draft, Writing – review & editing. **Tatsunori Shiraishi:** Conceptualization, Data curation, Formal analysis. **Yuta Ueno:** Investigation. **Akihito Yamamoto:** Investigation. **Rieko Kawase:** Investigation. **Yoshimitsu Kuwabara:** Supervision. **Takashi Sakatani:** Data curation, Investigation. **Shunji Suzuk:** Supervision.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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