Case Reports

Epithelioid leiomyosarcoma of the uterus: computed tomography findings

M.C. Hsieh[1], C.Y. Lin[2], J.C.W. Chien[1], C.J. Cheng[3], W.P. Chan[1, 2]

[1]Department of Radiology, Taipei Medical University-Wan Fang Hospital, [2]Department of Radiology, [3]Department of Pathology, School of Medicine, Taipei Medical University (Taiwan)

Summary

Uterine epithelioid leiomyosarcoma is a rare neoplasm. There have been no previous reports describing computed tomography (CT) findings for this tumor. A 31-year-old woman presented with a heterogeneous enhancing mass, with internal septa, in the uterus, which was shown on CT images. Histological diagnosis was compatible with epithelioid leiomyosarcoma.

Key words: Computed tomography (CT), Leiomyosarcoma, Sarcoma, Uterus.

Introduction

Epithelioid leiomyosarcoma arising in the uterus is extremely rare [1-5]. No computed tomography (CT) images of this tumor have been reported before. Since CT findings for this tumor may resemble those of other uterine malignancy, we report a case with histologically confirmed epithelioid leiomyosarcoma in the uterus, with characteristic findings on CT imaging.

Case Report

A 31-year-old woman suffered from a palpable growing mass over the right lower quadrant of her abdomen for two months. She was also bothered by increased menstrual flow. Physical examination revealed a palpable tumor in her lower abdomen. Blood analysis showed anemia (hemoglobin, 6.8 g/dl) and normal CA 125 (12.9 U/ml). Ultrasound (US) revealed a huge cystic mass with septa measuring 14.8 × 10.1 × 14.9 cm at the uterus with endometrial hyperplasia. Doppler US indicated hypervascularity of the tumor. CT scan showed a huge heterogeneous enhancing cystic mass with internal septa and thickened walls (Figure 1). No detectable enlarged lymph nodes in the obturator or internal iliac chain were noted. Our initial diagnosis was mucinous or serous cyst adenocarcinoma of the uterus.

The patient underwent total hysterectomy and left salpingo-oophorectomy. At surgery, multiple myoma nodules and adenomyosis of the uterus with surrounding bloody ascites were seen. Grossly, the uterus had a poorly defined margin with tan-colored and marked cystic degeneration filled with serous and bloody fluid. On serial sections, focal myxoid change was seen. Microscopy showed an epithelioid and spindle cell tumor of the uterine corpus with multifocal cysts and myxoid degeneration. These epithelioid tumor cells had eccentric cytoplasm with eosinophilic and inclusion-like cytoplasm. The nuclei were vesicular and had prominent nucleoli. In some areas, spotty eosinophilic changes with shrinkage of nuclei were also seen. Marked nuclear atypia, characterized by pleomorphic and hyperchromatic nuclei with some abnormal mitosis features was noted. No frank coagulation necrosis was identified. In the more cellular proliferative area, at least ten mitosis features (some abnormal) per 10 high-power field were noted. The diagnosis was compatible with epithelioid leiomyosaroma. The tumor was confined to the uterine corpus, free of the serosal surface; the endometrium was not involved by the tumor.

The patient did not receive adjuvant therapy after surgery. Regular follow-up US and serum CA 125 revealed no evidence of recurrent disease over two years.

Discussion

Leiomyosarcomas are the most common type of uterine sarcoma, but variants of these tumors, such as epithelioid leiomyosarcomas, are extremely rare. Only a few cases of these tumors arising from the uterus have been reported before [1-5]. They can easily mimic leiomyomas of the uterus on clinical diagnosis [6].

Kato *et al*. [7] reported two cases of epithelioid sarcoma in the thighs and calf, respectively, with obviously elevated serum CA 125 levels. However, as in our case, epithelioid leiomyosarcoma in the uterus revealed a normal CA 125 level.

Epithelioid leiomyosarcoma occurs in the third to seventh decades, and ranges in size from 2.7 cm to 30 cm [1-5]. Patients often complain of a palpable growing mass in the lower abdomen, with lower abdominal discomfort, menorrhea and bloody (or non-bloody) vaginal discharge. Our patient had a similar clinical manifestation.

Epithelioid leiomyosarcoma is a subtype of leiomyosarcoma and is defined by histological findings of rounded to polygonal cells with abundant eosinophilic cytoplasm in more than 50% of the tumor [1, 8-10]. On microscopic examination, the epithelioid cells have round nuclei and are eosinophilic in approximately 75% of cases and, less

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Fig. 1A

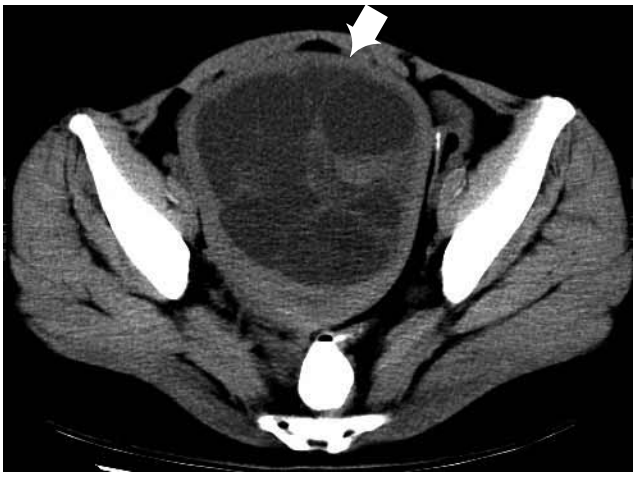

Fig. 1B

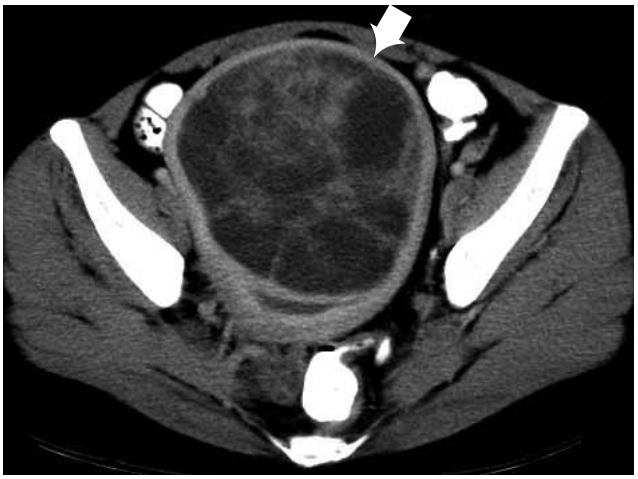

Fig. 1C

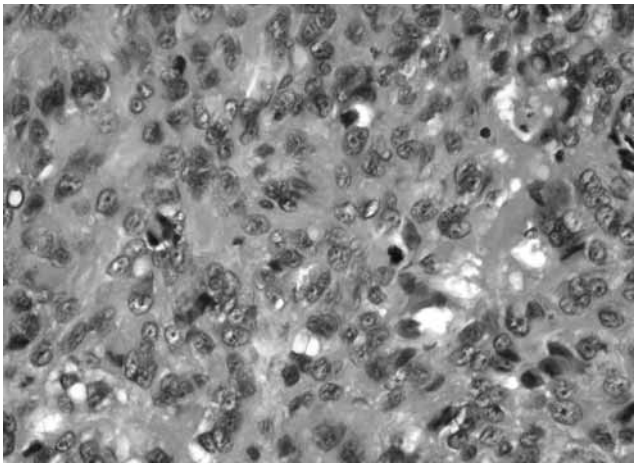

Fig. 1D

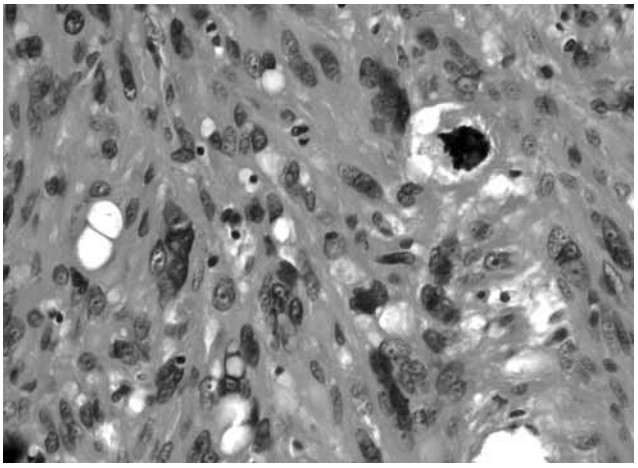

Figure 1. — Palpable mass in the lower quadrant of the patient's abdomen with increased menstrual flow for two months.
(A) CT scan without contrast enhancement showing a heterogeneous cystic tumor (arrow) in the uterus.
(B) CT scan after contrast medium administration showing a heterogeneous cystic mass (arrow) with a thickened, obviously enhanced wall and internal septa in the uterus.
(C) Photomicrograph of the specimen obtained from the uterine mass shows epithelioid and spindle cells with eccentric and eosinophilic cytoplasm. Mitosis features can be easily identified (H&E × 200).
(D) The tumor cells show hyperchromatic or vesicular nuclei with prominent nucleoli. Atypical mitosis feature is also seen (H&E × 400).

frequently, vacuolated or with clear cytoplasm [1, 8-10]. Nuclear atypia, 5 or more mitoses per 10 high-power fields), tumor cell necrosis, and tumor size together constitute the best criteria to predict malignancy [2, 9, 11]. In our case, the high mitotic index, nuclear atypia, and large tumor size all indicated malignancy.

To the best of our knowledge, there have been no previous reports describing radiological findings of epithelioid leiomyosarcoma in the uterus. In our case, US disclosed an echogenic mass with multiple cystic components in the uterus. CT scan showed a uterine tumor with solid and cystic components, consistent with cystic and myxoid degeneration of the tumor on histology. On CT imaging, the tumor also showed internal septa and heterogeneous enhancement, without hemorrhage or necrosis. Histology also showed no calcification or necrosis.

CT findings of a uterine tumor with cystic and solid components may include epithelioid trophoblastic tumor and uterine adenocarcinoma. Coulson *et al*. [11] reported a 9.8 cm cystic midline mass contiguous with the uterus, with an elevated serum beta-human chorionic gonadotropin (β-hCG) level, later proved to be an epithelioid trophoblastic tumor. Nalaboff *et al*. [12] showed one case with a heterogeneous enhancing mass with irregular endometrial thickening of the uterus, resembling our case, on CT images. However, uterine epithelioid leiomyosarcoma lacks the abnormal serum β-hCG level and presents with internal septa on CT images, as in our case.

Uterine epithelioid leiomyosarcoma is commonly treated by total abdominal hysterectomy with bilateral salpingo-oophorectomy [2]. Chemotherapy and hormonal therapy have been used in a few patients with

tumor recurrence or metastasis [1, 2]. Distinct metastasis is not common [2, 13]. Miyajima *et al*. [10] reported that the size and staging of the tumor were the most important predictors of a patient's prognosis. Other important prognostic factors included the mitosis rate and coagulative tumor cell necrosis in uterine sarcomas [10, 14].

In summary, we have reported the CT findings of an epithelioid leiomyosarcoma in the uterus in a 31-year-old woman. The diagnosis of this rare tumor should be considered in the case of a heterogeneous enhancing mass with internal septa on CT imaging.

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Address reprint requests to:
W.P. CHAN, M.D.
Department of Radiology
Taipei Medical University-Wan Fang Hospital
111 Hsing-Long Road, Section 3
Taipei 116 (Taiwan Republic of China)
e-mail: wingchan@tmu.edu.tw