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CASE REPORT

Aggressive Clinical Behavior of a Rare Uterine Sarcoma

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Heterologous sarcomas of the uterus are rare neoplasms. A case of a pure leiomyosarcoma with a poorly differentiated liposarcomatous component is described. No carcinomatous elements were identified, thus excluding the possibility of a mixed mullerian tumor. This is the third report of such an entity in the medical literature. The disease in this patient was characterized by pulmonary metastases and recurrent disease in the abdomen developing within 5 months of optimal surgical debulking. It is speculated that totipotent primitive cells may have the ability to differentiate into an aggressive heterologous tumor. © 1998 Academic Press

INTRODUCTION

Sarcomas of the uterus are uncommon, accounting for only 3–5% of all malignant tumors of the uterine corpus [1]. Treatment is primarily surgical with or without preoperative or postoperative pelvic irradiation. Although nearly one-half of patients are diagnosed with early-stage disease, the majority of women experience both local and distant recurrences. The prognosis for these patients is very poor. Ongoing clinical trials are being conducted by the Gynecologic Oncology Group to determine whether a role exists for systemic therapy in the treatment of this malignancy.

Leiomyosarcoma is the second most common of the uterine sarcomas, after the malignant mixed mullerian tumor (MMT). In contrast to the MMT which contains both epithelial and mesenchymal elements, the leiomyosarcoma occurs almost always as a pure homologous tumor. Rarely, heterologous leiomyosarcomas such as rhabdomyosarcoma, osteogenic sarcoma, and chondrosarcoma of the uterus have been described [1].

We describe an aggressive case of a heterologous uterine sarcoma composed of both a leiomyosarcomatous component and a liposarcomatous component and suggest a hypothesis that may account for the origin of such an unusual neoplasm.

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CASE REPORT

E.B. is a 50-year-old Caucasian woman, gravida 2, para 2, with a 6-year history of uterine fibroids. In June 1996, she presented with increased vaginal bleeding, severe cramping pain, and sensations of pelvic pressure. She was found to have a large multilobulated abdominopelvic mass, which by history, had been rapidly increasing in size. An endometrial biopsy was nondiagnostic secondary to insufficient tissue. The patient underwent magnetic resonance imaging of the abdomen and pelvis and the findings were noteworthy for a very large complex mass involving the anterior aspect of the uterus measuring 16 cm in maximal diameter (Fig. 1). This was believed to represent an intramural fibroid.

In July 1996, the patient underwent an exploratory laparotomy. Intraoperative findings were significant for a 25 × 25-cm uterine mass, normal-appearing adnexa, and a normal upper abdominal survey. The mass had an irregular appearance and focal “fish-flesh” tissue was present at the surface. Other regions were covered with soft yellow adipose tissue. Frozen section analysis was that of an atypical leiomyoma versus a leiomyosarcoma.

There was no gross evidence of disease outside of the uterus and the patient underwent a full staging procedure, including total abdominal hysterectomy, bilateral salpingo-oophorectomy, pelvic lymph node sampling, and omentectomy. The final pathology demonstrated a high-grade leiomyosarcoma with areas of pleomorphic liposarcomatous differentiation, totally confined to the uterine corpus and therefore consistent with a surgical Stage I disease.

The patient refused adjunctive therapy postoperatively and developed an isolated pulmonary metastasis 5 months after surgery. She underwent thoracotomy with a left lung upper lobe wedge resection. Pathology was consistent with metastatic leiomyosarcoma. Recurrent disease was identified in the pelvis 1 month later. She has declined surgical and/or experimental



FIG. 1. Magnetic resonance image depicting a complex 16-cm mass involving the uterus.

salvage therapy and at the time of this writing has entered into hospice care with end-stage disease.

PATHOLOGIC DESCRIPTION

The final pathologic description of the uterine corpus was remarkable for a high-grade leiomyosarcoma with areas of poorly differentiated pleomorphic liposarcoma. The endometrium was atrophic. The uterine cervix, both ovaries, both fallopian tubes, omentum, and 11 pelvic lymph nodes were without evidence of malignant disease.

Staining Technique

Immunoperoxidase stains for keratin, desmin, and muscle-specific actin antibodies and a special stain for lipid vacuoles (oil red O) were used.

Leiomyosarcomatous Component

The uterine corpus specimen measured $16.2 \times 13.5 \times 10.5$ cm and weighed 1365 g (Fig. 2). It demonstrated areas of fish-flesh appearance with softening and areas of yellow necrosis. There were large regions of significant nuclear pleomorphism and increased mitotic figures (an average of 23 mitotic figures per HPF). Large areas of the specimens were

covered with a thin capsule, but there were regions which had tumor cells on the surface. Both the muscle-specific actin and desmin stained positive. Despite abundant sampling and review of multiple sections, no carcinomatous elements were identified (i.e., the keratin stain was negative), thus excluding the possibility of an MMT. The results were consistent with a leiomyosarcoma (Fig. 3, left).

Liposarcomatous Component

Multifocal large areas within the leiomyosarcoma revealed poorly differentiated pleomorphic liposarcomatous differentiation (Fig. 3, right). Both the keratin stain for epithelial components and the smooth muscle marker stains were negative in these areas. However, this tissue stained positive with the oil red O fat stain (Fig. 4). There was no evidence of a benign uterine lipoma.

Pulmonary Metastasis

The lung nodule measured $2.4 \times 2.3 \times 1.8$ cm and was consistent with a high-grade leiomyosarcoma without a liposarcomatous component.

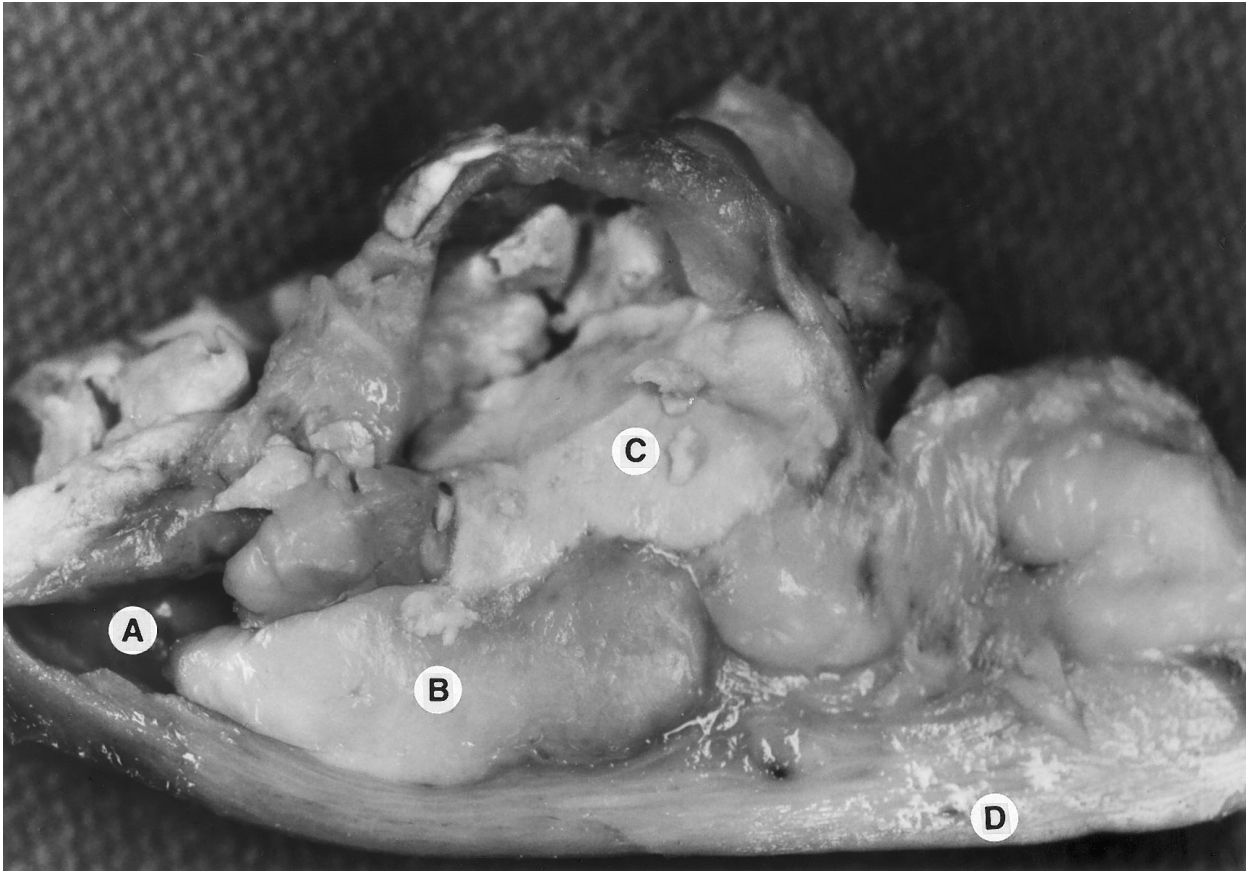


FIG. 2. Macroscopic uterine specimen containing the neoplasm, measuring $12 \times 4.5 \times 4.5$ cm and weighing 1365 g. A, cervix; B, myometrium; C, adipose tissue; D, serosal surface.

DISCUSSION

The histogenesis of uterine lipomas and uterine liposarcomas has been the subject of much speculation. Adipose tissue does not normally occur in the myometrium. Some of the theories that have been proposed to account for the presence of fat cells in the uterine myometrium include misplaced embryonic fat cells, metaplasia of muscle or connective tissue, perivascular extension of fat along uterine vessels, and derivation from pluripotential cells and embryonic multipotential components [3]. A liposarcoma of the uterus may be the result of the neoplastic transformation of a uterine lipoma.

Liposarcomas of the uterus are uncommon. In 1923, Peterson noted 2 cases of liposarcoma in an analysis of 31 cases of uterine lipomas [4]. A pure liposarcoma of the cervix in a leiomyomatous uterus was described by Veliath and colleagues in 1978 [5]. Gleeson and colleagues reported a 45-year-old patient with endometrial adenocarcinoma who also had a well-differentiated sclerosing liposarcoma involving the right ovary [6]. Schmidt and Doroszewski described a liposarcoma of the uterus in a 78-year-old patient [7].

Liposarcoma of the uterus in association with leiomyosarcoma has only been reported two other times. The first case

was described by Vakiani and colleagues in 1982 as one of a series of three heterologous sarcomas [8]. They described a 45-year-old woman who presented with metrorrhagia and an enlarged uterus. She underwent total abdominal hysterectomy and was found to have a leiomyosarcoma. In addition, there were areas of yellowish tissue with neoplastic polyhedral cells and vacuolated cytoplasm. These sections stained positive for lipids by the oil red O method. There was no epithelial component to the tumor. The patient subsequently experienced two recurrences involving the vertebral column.

In 1989, Bapat and Brustein reported the second case of a liposarcoma in combination with a leiomyosarcoma [9]. Their patient was a 55-year-old woman who presented with postmenopausal bleeding and a weight loss of 2.3 kg. A total abdominal hysterectomy and limited staging procedure was performed. Sections from a soft yellow area on the uterine specimen revealed a pleomorphic sarcoma containing many bizarre giant cells with lobulated nuclei and many cytoplasmic vacuoles which were positive by oil red O stain. Other sections demonstrated well-differentiated smooth muscle cells with abundant mitoses (more than 10 per 10 HPF), consistent with a leiomyosarcoma. This patient received two courses of chemotherapy and presented with recurrent disease in the pelvis 1 year later.

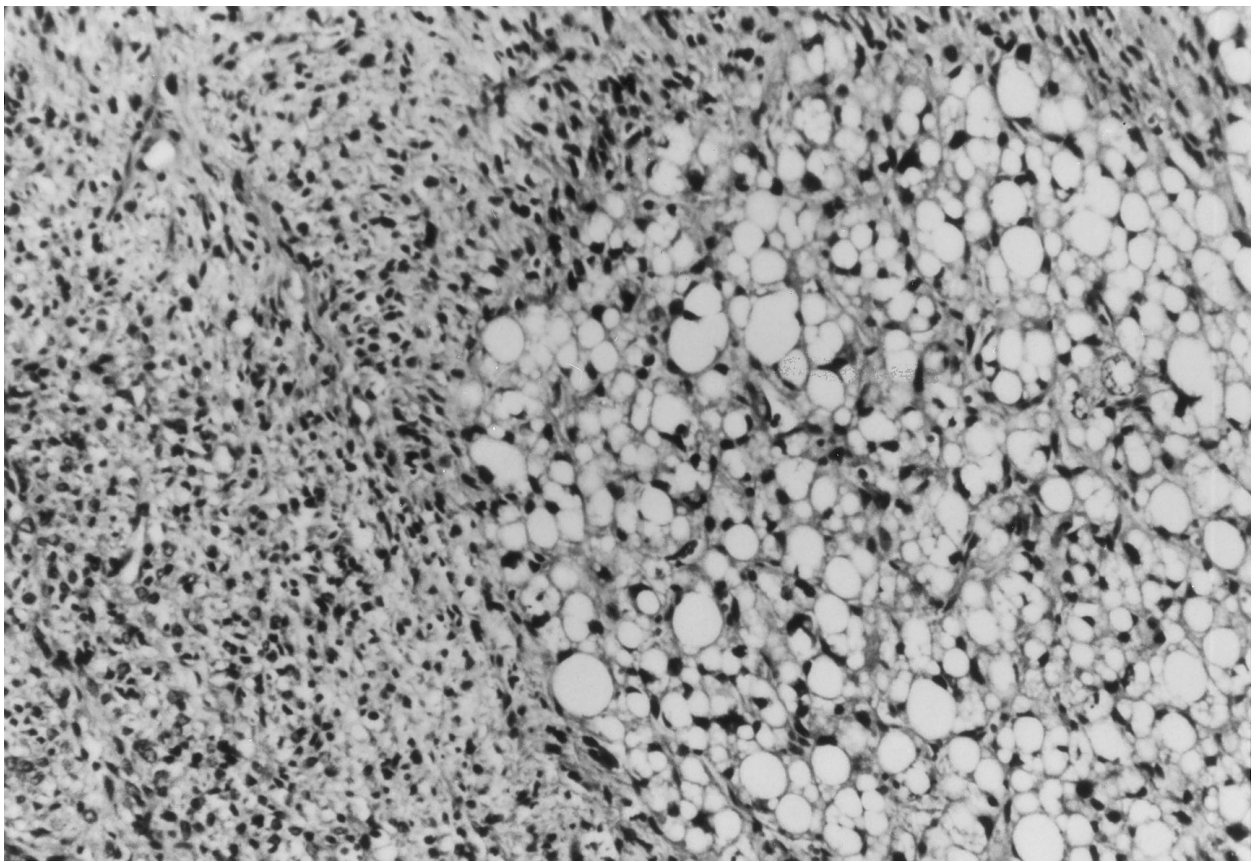


FIG. 3. Left: Positive desmin and muscle-specific actin staining in spindle cell high-grade leiomyosarcomatous component of the tumor confirmed the smooth muscle origin of this portion of the neoplasm. Right: Sections of tumor showing bizarre lipoblasts with multiple lipid vacuoles and hyperchromatic enlarged atypical nuclei consistent with a high-grade pleomorphic liposarcoma.

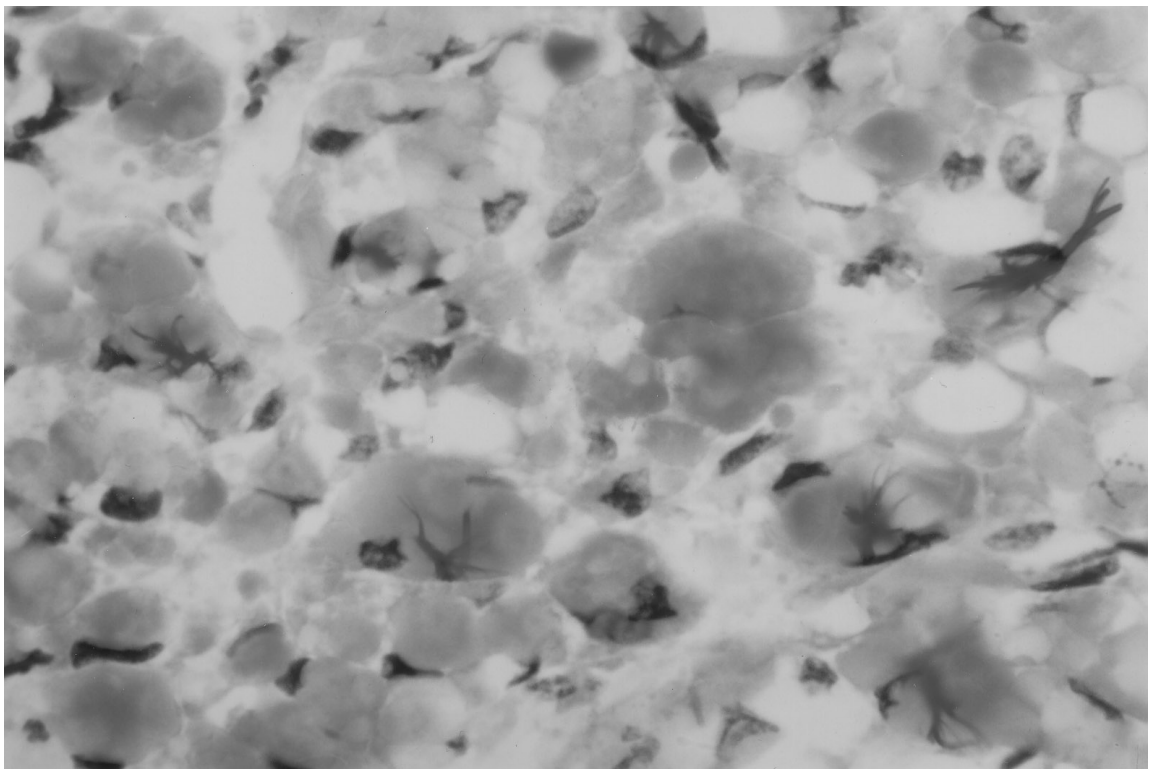


FIG. 4. Lipid stained red with oil red O stain, confirming the diagnosis of liposarcoma.

Recurrent disease is often part of the clinical course of patients who have been diagnosed with a sarcoma of the uterus. Indeed, both of the patients previously reported in the literature to have had a liposarcoma in combination with a leiomyosarcoma recurred either in the pelvis or at a distant site. Our patient recurred locally and at a distant site as well.

The finding of pleomorphic liposarcomatous components within a leiomyosarcoma could be a reflection of the capability of the mesenchyme primordium of the myometrium to differentiate into an aggressive tumor foreign to the uterus. The oncogenic stimulus which can promote such differentiation remains unknown.

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