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Case report

Epithelioid angiosarcoma arising in a uterine leiomyoma with associated elevated CA-125: A case report

Sarah V. Strickland[a,*], Mark R. Kilgore[a], Elise J. Simons[b], Mara H. Rendi[a]

[a] Department of Pathology, University of Washington, Seattle, WA, United States
[b] Women's Cancer Care of Seattle, Section of Obstetrics and Gynecology, Northwest Hospital & Medical Center, University of Washington Medicine, Seattle, WA, United States

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ABSTRACT

We describe the case of a 67 year old female with longstanding uterine leiomyomas who presented with fatigue, weight loss, elevated CA-125 and an enlarging mass arising from the posterior uterine fundus. Histologic sections of the mass contained a leiomyoma with interspersed foci of malignant epithelioid cells forming anastomosing vascular channels. The neoplastic cells were diffusely positive for CD31 and FLI1, supporting the morphologic impression of epithelioid angiosarcoma. Few cases of epithelioid angiosarcoma arising within a leiomyoma have been described. In this report we discuss this association and describe its relation with elevated CA-125.

1. Introduction

Angiosarcomas are aggressive neoplasms of endothelial origin which typically arise in the skin or superficial soft tissues of elderly patients and account for approximately 4% of sarcomas (Toro et al., 2006). Although rare, within the gynecologic tract, angiosarcoma has been described to arise from the cervix, ovary, vulva, vagina and uterus (Kruse et al., 2014). We present the case of a 67 year old female who developed epithelioid angiosarcoma within a pre-existing uterine leiomyoma. To the best of our knowledge, this is only the third time this association has been reported and the association with a significantly elevated CA-125 has not previously been described.

2. Case report

A 67 year old G0 woman with a history of uterine leiomyomas diagnosed in adolescence presented to her primary care practitioner with a chief complaint of fatigue and unintentional weight loss. She reported scant bloody vaginal discharge but denied abdominal distension, pain, or changes in her bowel and bladder habits. The patient had been in menopause since age 53 and, at that time, was told her uterus was enlarged due to leiomyomas to that of 3–4 months' gestation. She had no other relevant medical or surgical history. Upon examination the patient had a BMI of 16.5 kg/m^2 and examination of her abdomen revealed a firm mass arising from the pelvis and extending above the umbilicus, approximately 5–6 cm below the xiphoid process. A pelvic exam was performed, which was significant for a small amount of blood in the vaginal vault, but she was unable to tolerate further exam. Endometrial sampling was precluded due to severe distortion of anatomy by the large mass. Due to her symptoms and physical examination findings the patient underwent laboratory evaluation and computed tomography (CT) scans of the abdomen and pelvis. Laboratory studies were significant for anemia (hemoglobin 8.2 g/dL, hematocrit 26.2%) as well as an elevated CA-125 (237.5 U/mL) and CT of the abdomen and pelvis (Fig. 1) revealed a 21 × 18 × 15 cm heterogeneous and septated mixed density pelvic mass. It was unclear whether the origin of the mass was uterine or ovarian, however, the mass contained a large area of calcification suggestive of a calcified leiomyoma. Subsequent abdominal and pelvic magnetic resonance imagining (MRI) studies again showed a large heterogeneous pelvic mass measuring 24 × 20 × 12 cm with calcification noted in the right lateral aspect. The uterus and ovaries could not be visualized and no gross evidence of extra-uterine disease was identified. A CT of her thorax revealed three pulmonary nodules measuring up to 0.7 cm in maximal dimension however these were felt to be non-specific. Evaluation by a gynecologic oncologist was suspicious for possible uterine sarcoma, and surgical resection was recommended for definitive diagnosis and management. The patient therefore underwent subsequent exploratory laparotomy, total abdominal hysterectomy and bilateral salpingo-oophorectomy. Intraoperative findings were significant for a large mass confirmed to be arising from the posterior fundus of the uterus with a smooth external contour. The bilateral ovaries appeared atrophic without adnexal masses and there was no evidence of extrauterine disease or adenopathy. The enlarged uterus and adnexa

* Corresponding author at: Department of Pathology, University of Washington, 1959 NE Pacific St, Box 356100, Seattle, WA 98195, United States.
E-mail address: sstri012@uw.edu (S.V. Strickland).

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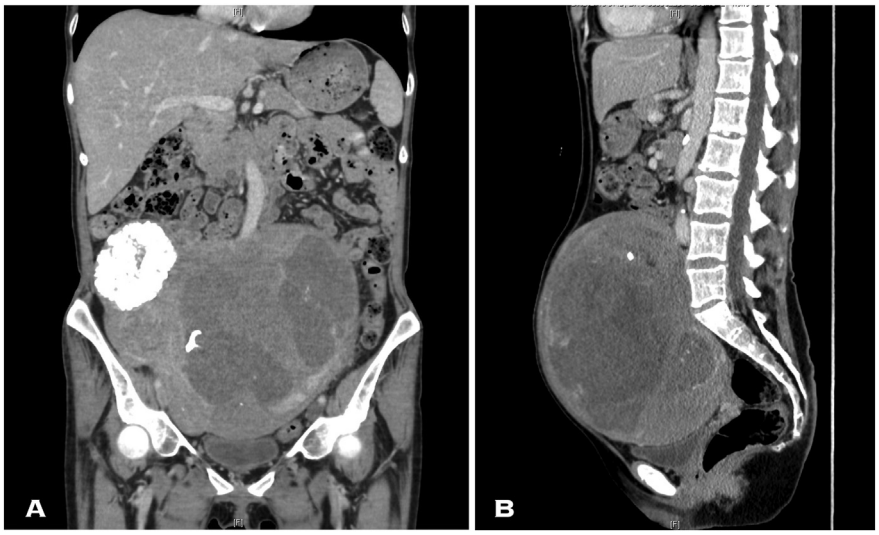

Fig. 1. Abdominal and pelvis computed tomography (CT) images. A) Coronal and B) sagittal views demonstrating a heterogeneous and septated mixed density pelvic mass with an area of calcification in the right lateral aspect.

were removed on bloc without morcellation of the specimen. Intraoperative frozen section was consistent with a hemorrhagic degenerating leiomyoma without evidence of malignancy.

3. Pathologic findings

The uterus measured 25 cm in maximal dimension and weighted 3901 g. The myometrium was distended by three well-circumscribed intramural masses, the largest of which measured 17 cm and contained areas of hemorrhage, necrosis and calcification. The intervening myometrium, endometrium, bilateral ovaries and fallopian tubes were grossly unremarkable.

Histologic sections (Fig. 2) of the largest intramural mass were composed predominantly of spindled cells arranged in a fascicular pattern with areas of hyalinization and calcification, consistent with a longstanding leiomyoma. Interspersed throughout the leiomyoma and associated vessel lumens there was a multifocal and distinct population of pleomorphic and epithelioid cells which formed sheets and atypical anastomosing channels containing red blood cells. Abundant mitotic activity and necrotic debris were also present. The epithelioid neoplastic cells were not present within the endometrium or myometrium which was sampled outside of the leiomyoma. Immunohistochemical

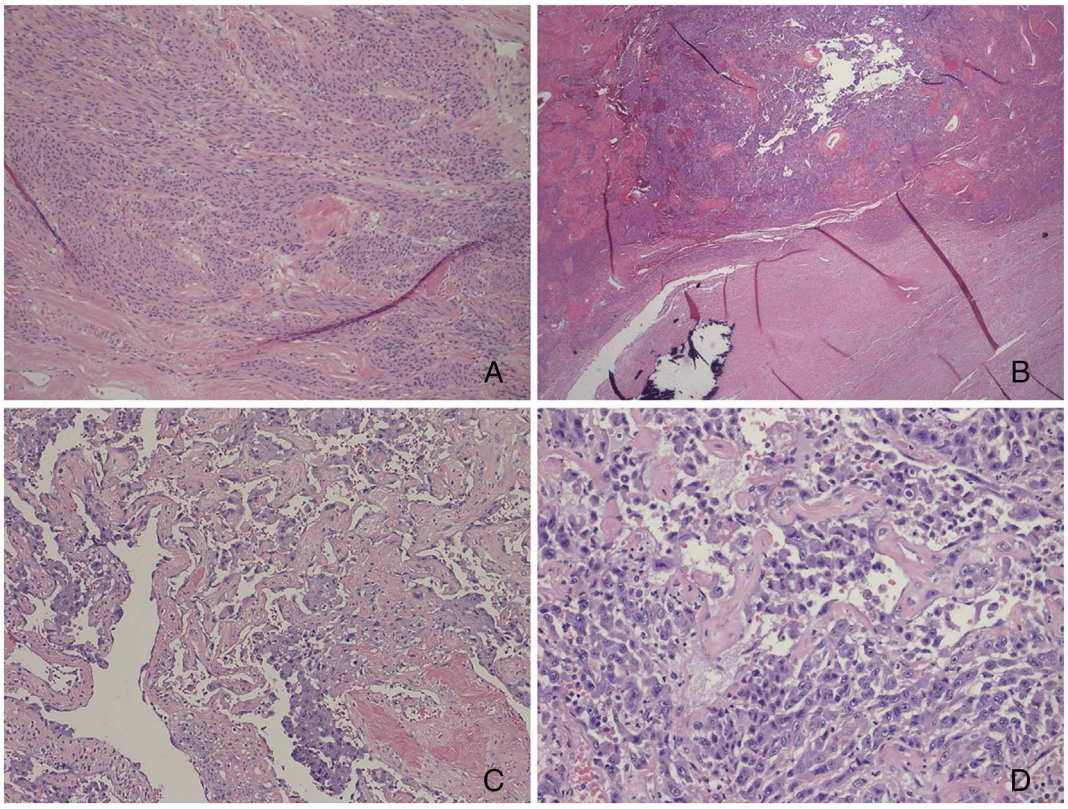

Fig. 2. Histologic features of largest intramural uterine mass. A) Bundles of smooth muscle with conventional appearance of leiomyoma, B) low-powered view (40 ×) of neoplastic cells interspersed amongst bundles of smooth muscle. C–D) Anastomosing vascular channels lined by enlarged and atypical epithelioid cells.