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Case report

# Leiomyosarcoma of the uterine cervix associated with pregnancy: A case report and review of literature

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## 1. Introduction

Genital sarcomas in pregnancy are extremely rare. A review by Matsuo et al. covering the period 1955 to 2007 revealed a total of 40 cases of female genital sarcomas diagnosed during pregnancy; 37.5% uterine, 27.5% retroperitoneal, 22.5% vulvar sarcoma, and 12.5% vaginal (Matsuo et al., 2009). Mean age at diagnosis was 27.8 years and the majority of cases were diagnosed in the third trimester. The 5 year survival for all patients was quite poor at 22.2%. Leiomyosarcoma of the uterine cervix associated with pregnancy has not been previously reported. In this report we describe the case of a gravid patient with a leiomyosarcoma confined to the cervix.

### 1.1. Case

An 18 year-old female Gravida 3 Para 1011 presented to an outside clinic with several months of vaginal bleeding and abdominal pain. She was diagnosed with an intrauterine pregnancy with an estimated gestational age (EGA) of 27 weeks based on dating by ultrasound. She was also noted to have an exophytic mass protruding from the endocervical canal. Subsequent examination under anesthesia revealed a 4.2 × 3.9 cm, pedunculated mass, arising from the endocervical canal. The mass was resected and pathology was consistent with a leiomyosarcoma.

The patient was referred to our institution at 31 weeks EGA for consultation with maternal fetal medicine (MFM) and gynecologic oncology. Physical examination was consistent with a normally developed young woman, normotensive, with a BMI of 26. Ultrasound evaluation showed an appropriately grown female fetus with a normal anatomic survey. Pelvic examination showed normal external female genitalia and vaginal tube. The cervix was 50% effaced and the external cervical os was 1 cm dilated. There was a remnant of a stalk at the 9:00 position in the proximal endocervical canal.

Pathology review at our institution revealed a 4.2 cm smooth polypoid fleshy mass on a narrow stalk. Microscopic examination revealed a cellular spindle cell neoplasm composed of highly atypical and focal bizarre giant cells with a high mitotic rate (11 mitoses/10 high power fields). Marked cytologic atypia was diffuse, but tumor cell necrosis was not prominent (Figs. 1, 2). Subsequent immunostaining showed the tumor to be focally positive for smooth muscle actin and muscle specific actin. Desmin, caldesmon, AE1/AE3, S100 and HMB45 were negative; p53 stained a few scattered nuclei; p16 showed strong diffuse block staining; Ki67 showed a high proliferation rate. A diagnosis of high grade leiomyosarcoma was made which was confirmed by expert consultation. The non-ulcerated surface was covered by benign endocervical epithelium confirming a submucosal configuration (Fig. 3). The tumor appeared to involve the entire polyp. While there was a small amount of stroma at the surgical margin, a definitive assessment of the margin status was not possible.

Computerized tomography of the chest, abdomen, and pelvis demonstrated a gravid uterus with no evidence of metastatic disease. After counseling by MFM and based on the pathology showing a high grade leiomyosarcoma the decision was made for delivery of the fetus at 33 to 34 weeks. She completed corticosteroids at 31 2/7 weeks. She was admitted with preterm labor and received a rescue dose of corticosteroids at 33 1/7 weeks. Assessment of fetal lung maturity was not performed. The patient desired definitive management of the high grade leiomyosarcoma and underwent primary low transverse cesarean section followed by exploratory laparotomy, total abdominal hysterectomy, and bilateral salpingectomy at 33 2/7 weeks. She delivered a viable female infant with APGARS 6 and 8 and weight of 2180 g. Intraoperative findings included normal ovaries and fallopian tubes and no evidence of extra-uterine disease. Final pathology showed no residual leiomyosarcoma. The patient had an uncomplicated postoperative course and has no evidence of recurrent disease by clinical exam and CT scan of the chest, abdomen, and pelvis at 13 months from her diagnosis. The infant was admitted to the neonatal intensive care unit post-delivery. She was treated for 3 days with phototherapy for hyperbilirubinemia. She required supplemental oxygen therapy for 9 days for transient tachypnea of the new born. She was discharge to home on day of life 19 with no feeding or other issues. At 1 year of life her pediatric examination showed normal growth and she had met her developmental milestones.

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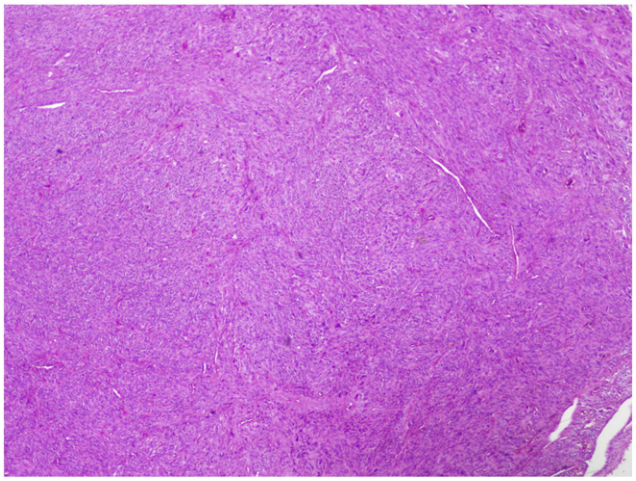

**Fig. 1.** The tumor is composed of atypical spindle cells (H&E, 20×).

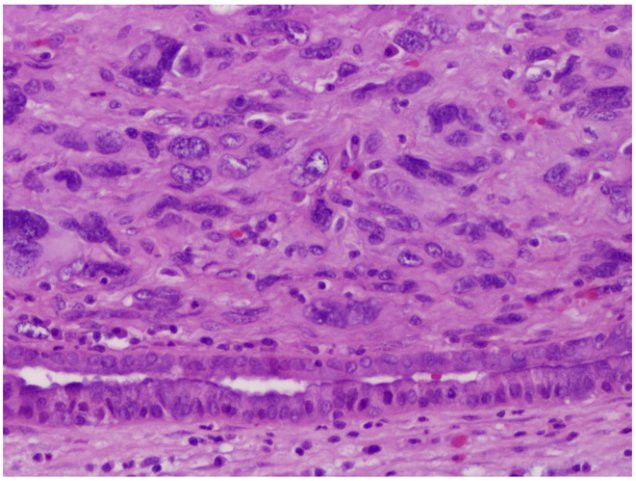

**Fig. 3.** The pedunculated tumor is lined by endocervical epithelium which is apposed to the underlying endocervical canal (H%E, 400x).

## 2. Discussion

We performed a literature search in PubMed and Medline databases without language restriction from 1960 to 2016. All articles were initially screened for title and abstract and full texts of eligible articles were subsequently selected. The search terms were "sarcoma, cervix, pregnancy, and leiomyosarcoma". Four case reports were discovered. Table 1 shows a summary of reports of cervical sarcomas in pregnancy. The 3 early case reports did not characterize the type of sarcomas using modern diagnostic criteria and nomenclature (Diaz-Bazan and Masferrer, 1960; Adinolfi and Persico, 1961; Onarir et al., 1967). The modern diagnosis of leiomyosarcoma is based on the work of Bell et al. (1994) and includes a combination of features including diffuse moderate to severe cellular atypia, a mitotic count of ≥10 MFs/10 HPFs, and coagulative tumor cell necrosis (Bell et al., 1994). Generally tumors with any 2 of these 3 features are diagnosed as clinically malignant leiomyosarcoma. The clinical circumstances of the case report by Schiavone et al. was quite similar to the current case, although the sarcoma in their case was called "high-grade" rather than leiomyosarcoma based on inconclusive immunohistochemical staining (Schiavone et al., 2011).

The diagnosis of leiomyosarcoma diagnosed during pregnancy requires expert pathological evaluation. The diagnosis of sarcoma in pregnancy can be difficult due to pregnancy induced histologic changes seen in benign leiomyomata including hemorrhage, necrosis, and degenerative changes. Muezzinoglu et al. in a review of leiomyomas in pregnancy described several cases of smooth muscle tumors with extensive hyaline and infarct necrosis (Muezzinoglu and Corakci, 1996). Coagulative cell necrosis was not observed. Cellular atypia when present was mild and focal and usually associated with hyaline type necrosis. Mitotic activity was low, less than 5/10 HPF. The diagnosis of leiomyosarcomas that meet the "three-feature" Bell criteria are considered high grade tumors (Bell et al., 1994). In the current case the tumor met the criteria proposed by Bell and was considered a high grade leiomyosarcoma.

Fertility sparing management options should be considered and discussed with patients desirous of future childbearing. In certain selected cases, surgical procedures such as myomectomy, polypectomy, and conization, during or after pregnancy, and dilation and curettage after delivery, could be performed. If no residual disease is found, surveillance rather than hysterectomy could be considered. Good outcomes and subsequent pregnancy have been reported with conservative surgery for patents with uterine smooth muscle tumor of uncertain malignant potential and low grade uterine leiomyosarcoma (Campbell et al., 2015; Salman et al., 2007). In the current case expert pathologic examination confirmed a high grade cervical leiomyosarcoma. Extrapolating from the literature on uterine leiomyosarcoma, high grade has been shown to a significant risk factor for recurrence and death and hysterectomy is considered the treatment of choice (Giuntoli et al., 2003).

In the current case CT scan was used as the imaging modality and the chest, abdomen, and pelvis were evaluated. The dose of radiation delivered to the fetus was 20 to 50 mGy which is below the dose threshold for malformations, growth retardation, mental retardation, and death; however, the risk of carcinogenesis increases approximately by a factor of 2, although it remains low in absolute term, less than one in 250 (Tremblay et al., 2012). Ultrasound and magnetic resonance imaging are not associated with risk and are the imaging modalities of choice in a pregnant patient.

The decision to deliver at 33 2/7 weeks EGA was based on several factors. The timing of delivery is based on the balance between the risk of cancer progression and the risk of delivery of a premature fetus. Leiomyosarcomas are aggressive tumors and patients whose disease appears to be initially confined to the uterus may experience a clinically aggressive disease course (Giuntoli et al., 2003). There is quite limited experience with leiomyosarcomas in pregnancy. Based on the uncertain pathologic margin status and the appearance of a residual cervical stalk on speculum exam there was a concern for residual disease. The patient was extensively counseled concerning natural history of

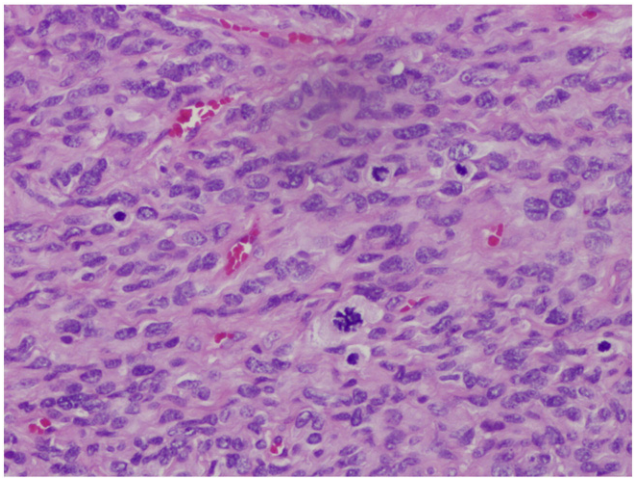

**Fig. 2.** The tumor contains numerous mitoses (H&E, 400×).