

Case Report

Uterine sarcoma in a 14 year-old girl presenting with uterine rupture

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Introduction

Uterine sarcomas are rare tumors with poor prognosis and even more rare in adolescents. They predominantly occur after the fourth decade and represent less than 3% of all genital tract malignancies (Seddon and Davda, 2011). The symptoms of uterine sarcomas are almost identical to leiomyomas such as abnormal uterine bleeding (60%), abdominal pain (50%), and gastrointestinal or genitourinary symptoms (30%). Uterine rupture and hemoperitoneum are rare presentations (Philip et al., 2011).

The uterine sarcomas consist of *pure mesenchymal tumors* (leiomyosarcoma (LMS), endometrial stromal sarcoma (ESS), undifferentiated endometrial stromal sarcoma (UES)), and *mixed epithelial and mesenchymal tumors* (adenosarcoma (AS) and adenosarcoma with sarcomatoid overgrowth (ASSO)). Carcinosarcoma (CS) is reclassified as the metaplastic form of endometrial cancer but the aggressiveness resembles uterine sarcomas. STUMP is a smooth muscle tumor with uncertain malignant potential (D'Angelo and Prat, 2010). Characteristic pathologic features include high mitotic index, hypercellularity, nuclear atypia, tumor cell necrosis, tumor size and intratumoral hemorrhage. Pathologic diagnosis may be problematic because several variants have confusing, subtle tumor characteristics and overlapping morphologic features that make distinguishing among the entities challenging (Tiirumani et al., 2013; Shah et al., 2012).

In this case report, we describe a large, rapidly growing tumor with subsequent uterine rupture and hemoperitoneum in a 14 year old African girl.

Case

A 14 year old, thin, African female, gravida 0, was admitted to our clinic with abdominal discomfort and increased abdominal circumference. She had no subsequent menstrual abnormalities or previous hormone use. Her past medical history was unremarkable and she had no surgeries before. There was no family history of uterine myomas or malignancy. Abdominal palpation showed a mobile, nontender mass extending towards the umbilicus. Pelvic ultrasonography revealed solid heterogenous uterine masses of 97×70 mm and 46×43 mm protruding from left uterine wall. MRI showed the invasion of all uterine layers. Tumor markers revealed normal, except CA 19-9 was 43.1 U/ml. For definitive treatment, laparotomy with frozen section pathology was planned. Exploration during laparotomy showed a 3×3 cm mass on the posterior wall of the uterus. Another mass of 5×6 cm invaded the endometrial cavity through the vagina with polypoid protrusions. All lesions were excised. Frozen section pathology revealed leiomyomata uteri with no malignant transformations and myomectomy alone was performed. She recovered uneventfully and was discharged on the second postoperative day. The final pathology report revealed a benign leiomyomata uteri. The result seemed irrelevant with the macroscopic appearance and we asked for a second opinion. The consultation, as well, showed benign tumor with spindle cells suggesting leiomyoma. Immunohistochemical staining was not performed since there were no heterologous elements in the slides.

Eight months after the surgery, she, for the second time, attended to our clinic with abdominal discomfort, fatigue, nausea and vomiting. She looked pale and dyspneic. Abdominal palpation revealed a 20 week gestation sized, nonmobile, tender mass. Hb level was 9.3 g/dl. USG and abdominal MRI revealed a $10 \times 10 \times 24$ cm heterogenous mass with predominantly solid components suggesting uterine sarcoma. CA -125 (2005.5 U/ml) and Ca 19-9 (92.6 U/ml) were elevated. She was hospitalized and during the preoperative diagnostic evaluation, the hemoglobin levels started to decrease towards 7.1 g/dl. Reevaluation of the patient revealed massive intraabdominal bleeding and she was emergently referred for laparotomy. On gross examination during laparotomy, bulky nodular masses were scattered around the peritoneum, intestines and the liver. These studded masses measured from 0.5 to 15 cm in diameter. Cut surfaces of solid masses appeared grayish white and myxoid with cystic changes. The anatomic contours of the

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pelvic structures were hard to differentiate because the rupture of the uterus distorted the adjacent structures. The left uterine artery was ripped off and was ligated immediately. The supportive tissues of the uterus, a 20 cm, irregular solid mass originating from the uterine wall and the ovaries were excised through the presumed anatomic cleavages. The scattered masses were all removed. She was discharged from the hospital uneventfully two weeks later.

The pathology revealed “adenosarcoma with stromal overgrowth”, a high grade sarcoma and she was referred to the oncology department for chemotherapy.

Discussion

This was an unusual case of an aggressively growing uterine sarcoma occurring in a fourteen year old girl.

Sarcomas, occurring under the age of 20 are quite rare. Predisposing factors are unclear but pelvic radiation therapy and tamoxifen, a selective estrogen receptor blocker is associated with high grade uterine sarcomas (D’Angelo and Prat, 2010). Hormonal stimulus like pregnancy, oral contraceptives or ovarian tumor may potentiate the mesothelium and submesothelial mesenchyme to differentiate into sarcomas. Clinical presentation is generally vaginal bleeding, pain, abdominal mass or severe uremia due to pressure. In our case, the rapidly growing tumor caused the rupture of the uterus and the patient was referred for emergency laparotomy due to massive intraabdominal bleeding.

There are a few cases of sarcomas in the literature with devastating presentations like uterine rupture and hypovolemic shock. De Roy and Wiegerinck (1986), described a large rapidly growing tumor with hemoperitoneum in a 15 year old girl. They concluded that the hemorrhage was due to the ruptured vascular connection rather than the uterine rupture and they preserved the uterus with the resection of the tumor solely. A case of a uterine tumor at an older age, was reported by Kim (2013), recently. The complaints, preoperative diagnostic features and the intraoperative macroscopic appearance of the mass, resembled those of our patient. The microscopic findings revealed spindle cells and hemorrhagic necrosis but immunohistochemical staining revealed rhabdomyosarcoma, atypical for that age. In another case by Stadsvold et al. (2005), a low grade myxoid endometrial sarcoma in a nulliparous 16 year old girl was managed by local resection and uterine reconstruction. Another remarkable point with our case was the dissemination of the solid and cystic masses over the serosal surfaces. There is a case report by Arneja (2011) about disseminated peritoneal leiomyomatosis in an 18 year old girl. Benign dissemination was explained by the smooth muscle metaplasia of subperitoneal mesenchymal stem cells of the secondary müllerian system. She was treated with hysterectomy and bilateral salpingoopherectomy and showed complete regression. In our case, it is probable that the pathology of the initial surgery might have been misdiagnosed and if it was a sarcoma initially then the dissemination was not due to the metaplasia of the müllerian system but rather the dissemination of the malignant cells during the first operation. Another point of view implies that morcellation of a uterine leiomyoma in a laparoscopic myomectomy may rarely cause dissemination. In our case, the patient had a myomectomy as a predisposing factor but it was an enucleation not a morcellation of the mass. In most cases firm diagnosis can be problematic. If a mass is noted in the uterine wall, the differential diagnosis will include a benign leiomyoma versus a uterine sarcoma. Endometrial biopsies are inadequate and often confounded by sampling error and bias. The role of imaging in the assessment of uterine sarcoma has not been well described given the rarity of the disease. Generally USG and MRI are used for pretreatment assessment of local uterine spread. CT is generally confined to extrauterine spread and metastasis (Shah et al., 2012). Tumor size, mitotic index, grade, age, race, tumor cell necrosis, adnexial involvement, sarcomatous overgrowth, and myometrial invasion are the factors that influence the prognosis of sarcomas. Standard primary surgical treatment for uterine sarcomas includes total

hysterectomy with or without salpingoopherectomy considering fertility preservation. Surgical staging of pelvic and paraaortic nodes may be helpful for carcinosarcoma and has less value for other uterine sarcoma types. Aggressive tumor debulking may be valuable for extrauterine spread. Chemotherapy may be beneficial in advanced stage uterine sarcomas. Although most uterine sarcomas are radiosensitive, the overall survival does not change with radiotherapy. Hormonal therapy may be useful for metastatic or recurrent sarcomas with estrogen and progesterone receptors. We, in this case, planned TAH + BSO preoperatively but intraoperatively, the pelvic structures were found to be unrecognizable due to uterine rupture. We performed tumor debulking, bilateral salpingoopherectomy and removed all solid and cystic masses (Fig. 2). The specimens from the second surgery revealed necrosis, atypia, sarcomatous components accompanied with myoid and chondroid elements (Fig. 1A,B) and focal glandular components. The major component of the tumor was the sarcomatous spindle cells and immunohistochemical staining showed positive for vimentin and negative for SMA, eliminating leiomyosarcoma.

According to WHO classification it was “adenosarcoma with stromal overgrowth”; a subgroup of mixed epithelial and mesenchymal tumors. The survival of our patient is unpredictable due to advanced stage uterine sarcoma; she is still on her fourth chemotherapy line with ifosfomide and etoposide.

Since a certain number of sarcomas will not be diagnosed until after hysterectomy, the patient will be best managed by those

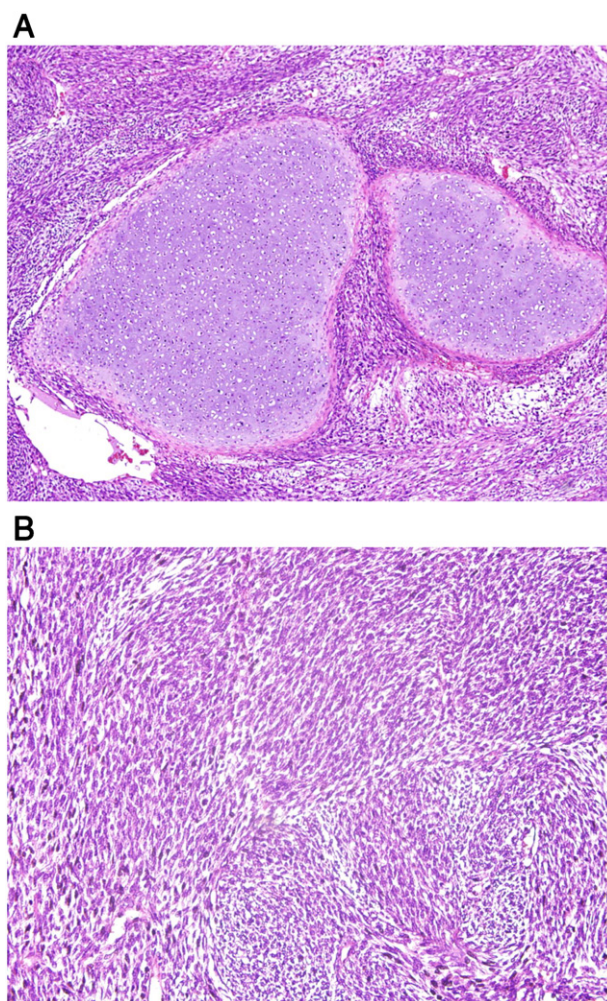


Fig. 1. (A) Chondroid differentiation in tumoral tissue (hematoxylin and eosin, original magnification $\times 40$). (B) Mesenchymal tumor with crossing branches of spindle cells (hematoxylin and eosin, original magnification $\times 100$).

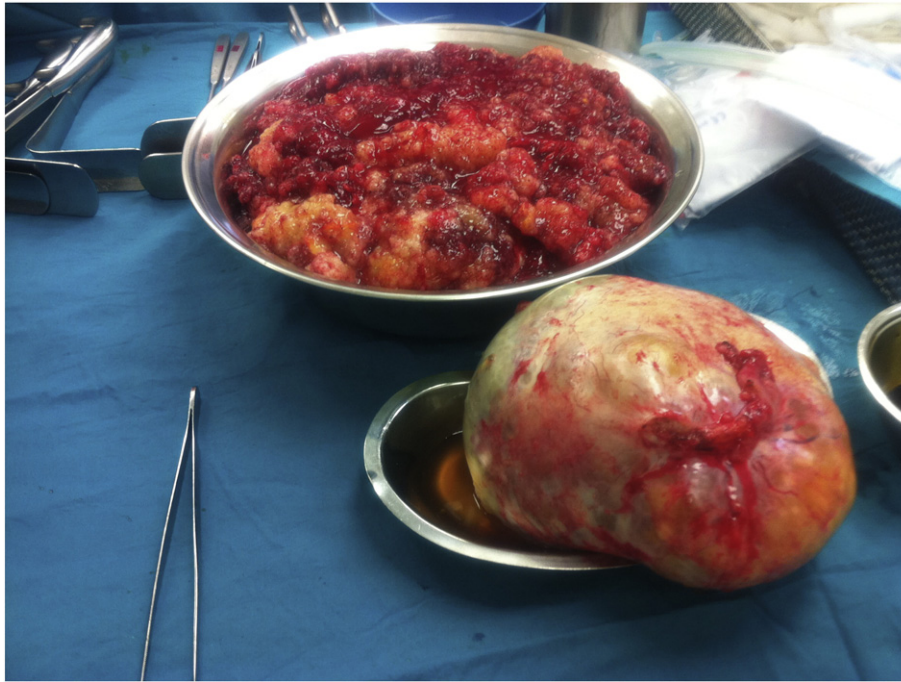


Fig. 2. Mesenchymal tumor of uterus and bulky masses.

with specialized expertise such as oncologic surgeons and gynecopathologists when a uterine sarcoma is suspected (Karlan et al., 2014).

Conclusion

Rapidly growing tumors may suggest uterine sarcomas which are quite rare in adolescents. Uterine rupture or hypovolemic shock are infrequent presentations. Preoperative diagnosis may be controversial since there are no clear criteria for malignancy. The experience of the gynecologic surgeons and gynecopathologists constitutes the hallmark of definitive diagnosis and treatment.

Conflict of interest statement

The authors declare that there are no conflicts of interest.

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