

Myxoid Liposarcoma at Thigh during
Pregnancy: A Case Report and Literature
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Abstract

Background: Liposarcomas in pregnancy are peculiar clinical scenarios that are continuously being reported in the English literature for their rarity, challenges in management and speculations of association between pregnancy and disease kinetics. Most cases reported are with liposarcomas in the retroperitoneum. We here add another case whose lesion is at left thigh with optimal tumor excision and split-thickness skin graft reconstruction to augment both prognosis and aesthetics. A literature review for cases reported after 2000 will be presented.

Method: Case report and literature review.

Results: A 33-year-old female with left thigh tumor noted 1 year prior to her second pregnancy found the lesion growing in size at 18 weeks of gestation. Characteristics of the tumor included soft, elastic, fixed, painless, without discharge or discoloration, and sized about 5x5cm. Myxoid liposarcoma was diagnosed under histological examination after tumor biopsy under local anesthesia. She soon received complete tumor resection with free margins confirmed, followed by split-thickness skin graft reconstruction of the wound to meet cosmetic concerns. She currently continues with her pregnancy uneventfully and no tumor recurrence is observed. A literature search on Pubmed using keywords with sarcoma and pregnancy found 6 entries prior to the current study after year 2000. Patient characteristics, diagnosis modality, pathology histology subtypes, maternal and fetal outcomes are organized and presented.

Conclusion: Liposarcoma in pregnancy is rare but deserves practitioners' attention, for early detection and proper surgical management could greatly improve disease outcomes. Life-long surveillance after surgery is also important for local recurrence rate is high. Further studies for the interplay of growth hormones and sex hormones during pregnancy and their effects on soft tissue sarcomas are called for, as they might be of valuable treatment or prevention bases.

Introduction

Sarcomas account for less than 1% of all cancer, among which Soft Tissue Sarcomas (STS) are the most common [1]. Liposarcomas are one of the most common subtypes of STS with predilections for males, age 40-70 years and at retroperitoneum or lower extremities [2]. The World Health Organization classified liposarcomas into well-differentiated, myxoid, round-cell, pleomorphic and dedifferentiated subtypes; the first two subtypes belong to the low-grade lesions with better prognosis and higher incidence rates, which largely depends on the histologic subtypes, resection margin and age of the patients [2,3]. With free surgical margin, the 5-year overall survival rate can be 68-80%, although the local recurrence rate 75% [3,4]. Liposarcoma in pregnancy are peculiar clinical scenarios that are continuously being reported in the English literature for their rarity, challenges in management and speculations of association between pregnancy and disease kinetics. Most cases reported are with liposarcoma in the retro peritoneum, yet we here add another case whose lesion is at left thigh with optimal tumor excision and split-thickness skin graft reconstruction to augment both prognosis and aesthetics.

Case report

This 33-year-old female without known underlying conditions is gravida 2, para 1, with singleton and received her regular antepartum care without significant events. Her first child was born prematurely but healthy at the gestation age of 36+2 weeks under the stress of acute gastroenteritis, 2 years prior to the current pregnancy. Her body mass index was measured to be 24.9 and an increase of body weight due to pregnancy was noted. A painless, soft and elastic tumor on the anteromedial aspects of her left thigh sized about 4x4cm was noticed one year prior to her second pregnancy. Upon inspection, there was no discoloration, discharge or itchiness. She denied history of blunt trauma, contact dermatitis or insect bites. Tumor biopsy for pathology study was suggested then

but postponed by the patient because she was still breastfeeding her first child. The procedure was further postponed after she conceived her second child. However, the tumor started to grow in size at a noticeable speed around 18 weeks of gestation age, reaching the size about 5x5cm.

Tumor excision was immediately arranged for her, of which histology report revealed myxoidliposarcoma, histologic grade 2, mitotic rate of 2 per 10 high-power fields, and with positive margins. Microscopic examination of the lesion showed a mixture of relatively monotonous or oval shaped nuclei and small lipoblasts in a prominent myxoid stroma (Figure 1). Some arborizing and chicken-wire capillary vasculature are noted. Focal high-grade primitive rod cells with high nuclear-cytoplasm ratio are also seen. Tumor markers are focally positive for S100 and CD 117. Further tumor survey with magnetic resonance imaging could only identified increased infiltrations in the subcutaneous space surrounding the operation site without definitive evidence of residual tumor or suspected bony lesions. Wide resection of the tumor under epidural anesthesia was soon arranged for her (Figure 2), when intraoperative frozen pathology and repeated tumor excisions were done until all margins were free. The final operation wound sized about 15cmx 18cm with the whole thickness of the tissue. To repair the wound, split thickness skin grafts were fixed and she had recovered well without complications. Up to the time manuscript submission, she is 29+2 weeks into her pregnancy with estimated fetal body weight of 1511gm, which is appropriated for the fetal gestational age. No recurrence of liposarcoma was identified and regular tumor surveillance is arranged for her.

Results and discussion

Liposarcomas in pregnancy, although rare, are not unheard of in the English literatures. As early as 1937, Smith has reported 4 cases of pregnancy coincide with sarcomas treated by surgery or radiotherapy and resulted in promising maternal and fetal outcomes [5]. Jeng et al. also compiled a list of cases reported up to year 2000 with a total number of 52 cases [6]. Another 6 entries, this current report included, are reported since then and are listed in Table 1

[2-4,6-8]. When examining the 2 sets of data, it could be observed a trend of improved maternal and fetal outcome in recent years. This is postulated to be because of better diagnostic tools, anesthesia choices, surgical techniques and neonatal care quality comparing to those before year 2000. It should also be noted that prompt tumor excision upon diagnosis to limit tumor growth, adjacent organ invasion and metastasis are helpful for maternal survival, as reported by Hou [2], Lopes [3], Oh [7] and this current study. Pregnancy continuation is plausible and feasible with continuous fetal monitoring and current anesthetic methodology. Newborn outcomes are promising with corticosteroids use for premature fetal lung maturation and intensive neonatal cares. In contrast, patients postponed tumor excision until time of Cesarean section or postpartum seem to have poor outcome, as described by Jeng [6] and Tebes [4].

The principal aim of surgery is to remove tumor with free margins, as this is one of the strongest prognostic factors. If margin is left positive, adjuvant chemotherapy and radiotherapy should be considered, although supportive evidence is yet to be obtained. In this current case with tumor at thigh, margin-free lesion excision might not be as challenging as those in the retro peritoneum with multiple organs, major vessels and nerves involvement, especially with intrauterine pregnancy to take consideration. However, the aesthetics of the large operation wound becomes of concern, especially for young women like our patient. Meticulous operation planning with split-thickness skin graft wound reconstruction by specialized plastic surgeon adds to the surgical outcomes. Other prognostic factors include the tumor histology subtypes, grading and patient age; which are of less or no roles for the practitioners. Cases listed in Table 1 are agreeable to these assertions, for myxoid subtypes are commonly diagnosed and with better prognosis; while de-differentiated subtypes are of poor maternal outcomes or require adjuvant management.

The association between pregnancy and liposarcoma tumor kinetics has been speculated. Cantin and McNeer suggested that pregnancy does not promote tumor growth, the estrogen-progesterone environment during pregnancy could have a favorable impact on the sarcoma growth, while hormone therapy might play a role in managing metastatic disease [9]. Similar views are presented by a recent Swedish population-based case-control study from 1988 to 2009 by Wagner et al. [1]. The authors found that lower BMI is associated with increased risk for soft tissue sarcomas; while shorter

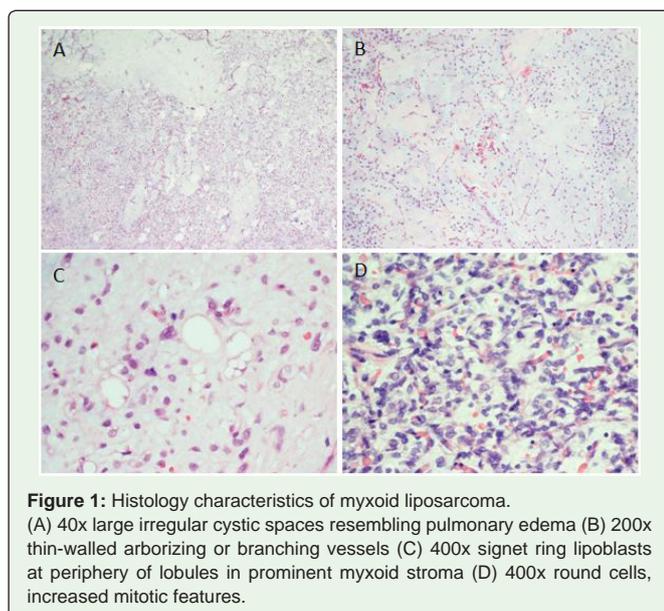


Table 1: Overview of liposarcoma in pregnancy cases reported in English literature.

Case	Age	Gestational age at diagnosis/operation	Diagnosis	Pathology	Pregnancy outcome	Prognosis
Ker, 2017	33	18 weeks/18 weeks	Anteromedial aspect of left thigh; 5x5cm by inspection	Margin free Myxoid liposarcoma; Mitotic index 2/10 HPF (+) S100,CD 117	Ongoing pregnancy	No recurrence at the time of publication
Huo, 2015 [2]	27	16 weeks/20 weeks	Retroperitoneal tumor 12x8cm by ultrasonography; 16x9cm by MRI	Margin free Low-grade myxoid liposarcoma; Mitotic index 5/50 HPF (+) Vimentin	GA 37 weeks by Cesarean section	No recurrence 6 months post-operation
Oh, 2014 [7]	39	28 ⁺² weeks/29 weeks	Retroperitoneal tumor 35x26x17cm by ultrasonography, MRI and CT	De-differentiated liposarcoma; (+) MDM2, CDK4, Ki-67	GA 29 weeks by Cesarean section	No recurrence 3 months post-operation and adjuvant radio-chemotherapy*
Lopes, 2009 [3]	33	13 weeks/13 weeks	Retroperitoneal tumor 22x20x20cm by CT	Well-differentiated liposarcoma	GA 37 weeks by Cesarean section	No recurrence 1 year post-operation
Jeng, 2005 [6]	33	12 weeks [#] /36 weeks	Recurrent retroperitoneal tumor 9.3x6.0cm, 6.7x6.3cm by ultrasonography at diagnosis and 25x20cm at tumor excision	Myxoidliposarcoma Metastasis to peritoneum para-aortic lymph nodes and omentum	GA 36 weeks by Cesarean section	Recurrence at 4 months post-operation and expired
Tebes, 2001 [4]	22	13 weeks/ 3 weeks postpartum	Right adnexal tumor 6cm by ultrasonography at diagnosis, 8x9x10cm by MRI at 26 weeks, 20 cm at tumor excision	Poorly differentiated liposarcoma	GA 29 weeks of twin males by vaginal delivery	Adjuvant chemotherapy/ Persistent disease and expired 6 weeks post-operatively
Matsuda, 2000 [8]	-	-/ 5-7 weeks postpartum	2 cases	Myxoid liposarcoma	Healthy	No recurrence

* 30 sessions of radiotherapy in addition to doxorubicin 12.5mg/m2 and ifosfamide 2,000 mg/m2 for 3 months

Incomplete tumor resection without adjuvant therapy 2 years prior to presentation

! MESNA, doxorubicin, ifosfamide for 1 month

stature at puberty, increased parity and contraceptives use especially at young age have protective effects for soft tissue sarcomas. They further explained the low body weight might be as a result of tumor aggression, but this clue can be masked in weight-gaining pregnant women, making disease detection even more difficult. On the other hand, Growth Hormones (GH) and Insulin-Like Growth Factor 1 (IGF1) increase target cell malignant transformations, while Sex Hormones (SH) promote tissue cell differentiation and reduce tissue responsiveness to GH/IGF1. This explained why short stature at puberty, pregnancy, menopause and continuous contraceptives exposure seemingly exert protective roles to STS, as these phenomena are surrogates to low GH/IGF1 and high SH levels. Although conclusive evidence is yet to be obtained, they offer potential treatment and prevention targets.

In conclusion, liposarcoma in pregnancy is rare but deserves practitioners' attention, for early detection and proper surgical management could greatly improve disease outcomes. Life-long surveillance after surgery is also important for local recurrence rate is high. Further studies for the interplay of growth hormones and sex hormones during pregnancy and their effects on soft tissue sarcomas are called for, as they might be of valuable treatment or prevention bases.

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