Maternal Metastatic Synovial Sarcoma in Pregnancy

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Abstract

Pregnancy with known metastatic cancer is rare. A case of a woman with known metastatic synovial sarcoma in pregnancy is presented in this study. Her disease progressed with an enlarging and new pulmonary metastasis requiring bilateral pulmonary metastasectomies, both during pregnancy and after delivery. Her pregnancy was uncomplicated otherwise, and she had a successful at term vaginal delivery of a healthy female baby. During her second pregnancy, her disease aggressively recurred at eight weeks gestation without therapeutic options. Thus, palliative care was initiated and she died shortly thereafter. The implications of sarcoma growth and the potential thromboembolic risk with active malignancy in pregnancy is discussed in the study.

Résumé

La grossesse accompagnée d’un cancer métastatique connu est rare. Cette étude présente le cas d’une femme enceinte atteinte d’un sarcome synovial métastatique connu. Sa maladie a progressé sous forme d’une nouvelle métastase pulmonaire en croissance nécessitant une métastasectomie pulmonaire bilatérale pratiquée pendant la grossesse et après l’accouchement. Sa grossesse s’est déroulée sans autre complication et elle a accouché à terme par voie vaginale d’une fille en bonne santé. Au cours de sa deuxième grossesse, son cancer a récidivé de manière agressive à huit semaines de grossesse sans qu’aucune option thérapeutique ne soit possible. Par conséquent, des soins palliatifs ont été mis en place et elle est décédée peu de temps après. Cette étude examine les conséquences de la croissance du sarcome et le risque thromboembolique potentiel lié aux tumeurs malignes actives durant la grossesse.

Keywords: neoplasm, pregnancy, sarcoma, surgery, synovial
Introduction

While roughly 9% of cancer diagnoses occur in women of reproductive age, only 0.02–0.1% of these occur in pregnancy.1 Entering a pregnancy with a known metastatic cancer is even rarer. A case of pregnancy in a woman with known metastatic synovial sarcoma (SS) is presented in this study. SS is commonly diagnosed in adolescents and young adults with a predilection for extremities as the location of primary disease.2 The 5-year survival is roughly 76% with SS, however 38% develop metastatic disease with grade III tumors size; a tumor greater than 5 cm in size is associated with the worst survival rate.3

Case

A 26-year-old gravida 2 para 0 abortus 1 patient with a history of metastatic SS was referred for prenatal care. She was otherwise healthy with an 11-pack-a-year smoking history. In the year preceding her pregnancy, she underwent a surgical resection of a 7.5 cm synovial sarcoma of the left knee joint and reconstruction using megaprosthesis. The pre-operative staging computed tomography (CT) scan was negative, apart from two small right lung lesions thought to be granulomas. The post-operative CT showed metastatic disease with three small right lung lesions. The patient was treated with doxorubicin at 75 mg/m², for four cycles with a good response. She was then given a break with plans for surgical resection if the lesions grew back. The patient was aware that the outcome was guarded. At follow-up with Medical Oncology, the patient happily disclosed her pregnancy. Referrals were sent to Obstetrics and Thoracic Surgery.

The patient was seen by Obstetrics at 10 weeks 2 days. The risks of the pregnancy with metastatic sarcoma, including disease progression were reviewed, and she remained committed to continuing the pregnancy. The prenatal lab work was normal, and aneuploidy screen was found to be low risk. The maternal echocardiogram showed normal cardiac structure and function.

A CT scan at 13 weeks showed the progression of the pulmonary metastases in both lungs. Thoracic surgery recommended an excision as this was the only identified location of metastatic disease. At 16 weeks, she underwent a right thoracotomy, and a resection of three right-sided metastases. She then underwent a left VATS wedge resection of one of the metastases at 19 weeks. Given the active malignancy in pregnancy, a prophylactic anticoagulation with low molecular weight (LMWH) was initiated for the remainder of pregnancy.

Serial fetal ultrasound surveillance showed normal anatomy and growth. A CT scan at 31 weeks showed a new 6×7 mm nodule in the apical segment of the right lower lobe and a 9 mm nodule in the left upper lobe. MRI of the left hip/thigh at 35 weeks gestation showed no evidence of local disease recurrence. Given the isolated recurrence, further surgical resection was recommended. Induction of labour occurred at 37 weeks with a cervical foley catheter and oxytocin. A live female baby was born vaginally with a birthweight of 2590 g. Postnatal course was unremarkable. Prophylactic anticoagulation with LMWH was given for 6 weeks postpartum.

Two months postpartum, she underwent another left VATS wedge resection of a new left-sided metastases. One month later underwent a right VATS wedge resection of a new right-sided metastatic deposit. A follow-up CT scan, 3 weeks post-operative, demonstrated no evidence of residual or new metastatic disease.

Three months later, she returned to the clinic with a second planned pregnancy at 8 weeks 4 days. A routine follow up thoracic CT scan performed earlier that week demonstrated an extensive bulky mediastinal and left hilar nodal disease with encasement and obstruction of the left pulmonary artery and left mainstem bronchus. In clinic, she was visibly dyspenic and endorsed new onset hemoptysis. A consultation was obtained with Thoracic Surgery and Medical Oncology. The disease was inoperable, and the patient did not wish to pursue further systemic treatment. Given the rapid progression of the disease and her symptoms, she was transitioned to palliative care. The pregnancy, given early gestation, was left in-situ. She died from her disease 5 days later.

Discussion

Synovial sarcoma account for 5–10% of all soft tissue sarcomas.3,4 Commonly, these tumors arise in the extremities with a propensity to metastasize to the lungs.3,4 There are no known risk factors for synovial sarcoma.4–6 The standard treatment is wide surgical resection with/without radiotherapy. A systemic chemotherapy is typically reserved for metastatic disease; the role of adjuvant chemotherapy remains controversial.7

The above described case represents the first reported case of a woman starting pregnancy with a known metastatic SS. Of the 15 published case reports of SS in pregnancy, all...
but one woman was diagnosed with the disease in pregnancy. The location of primary SS in these reports is varied, with five being pulmonary, four renal, two head/neck, one abdominal wall, two involving lower extremities, and one vulvar. The vulvar SS is the only published case of pre-pregnancy diagnosis, where the woman was diagnosed and treated with surgery, brachytherapy, and external beam pelvic radiation, 2 years prior to conception. She was disease-free at the time of conception, and her pregnancy was uncomplicated with no evidence of recurrence.

Given the limited published data of cases similar to our patient, it was challenging to counsel appropriately regarding the impact pregnancy would have on her prognosis. The SS tumor cells can have estrogen receptors (ER) and/or progesterone (PR) receptors. The high circulating levels of both these hormones in pregnancy could stimulate or promote tumor growth. In our case, the pre-existing lesions grew, and new pulmonary metastases developed through the course of the pregnancy. Unfortunately, we do not have the ER/PR receptor status of her tumor cells.

This mechanism for possible tumor growth in pregnancy was our rationale to resect the growing metastases in the second trimester. It was felt that if these tumors were left untreated until the end of pregnancy, this could worsen her overall survival. The ideal time for surgery in pregnancy is the second trimester; avoiding the higher miscarriage rate of the first trimester, and theoretical teratogenic risks of anesthetic drugs. The safety of thoracic surgical intervention including VATS in pregnancy has been established. Harris et al. published a case of primary pulmonary SS in pregnancy using ifosfamide rather than surgery in pregnancy. In our case, as these were isolated metastasis, surgical resection rather than chemotherapy in pregnancy was the chosen treatment. When our patient presented with the second pregnancy, she had been experiencing progressive dyspnea over the preceding month. While this may have partially been attributable to dyspnea of pregnancy, it is unknown whether the hormonal effects of her last pregnancy propagated the rapid tumor growth in her lungs. Tragically, her symptom burden and tumor spread were such that no life-extending therapy was an option. There is limited literature surrounding the use of maternal palliative care in pregnancy. Similar principles to palliation in non-pregnant women were applied to this case. Given her early gestational age and limited life expectancy, we elected not to organize a termination of pregnancy and rather focus on supporting a comfortable death.

The other interesting clinical challenge this case presented was the venous thromboembolism (VTE) risk for patients with active malignancy in pregnancy. Independently both active malignancy and pregnancy are risk factors; however, neither in isolation provides sufficient criteria for anticoagulation. The national guidelines for VTE prophylaxis in pregnancy have not addressed this clinical situation. Typically, anticoagulation is recommended in pregnancy if the VTE risk is greater than 1%. Based on expert opinion, we elected to place her on VTE prophylaxis for the remainder of the pregnancy and for 6 weeks postpartum. A recent cohort study from Denmark would support this recommendation. They demonstrated a VTE rate of 75.2 per 10,000 pregnancies with active malignancy compared to 10.7 per 10,000 without malignancy. The present case demonstrated that LMWH VTE prophylaxis can safely be used throughout a pregnancy complicated by active thoracic malignancy and recurrent surgeries without sustaining additional complications related to anticoagulation.

Conflict of Interests
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Informed Consent
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Ethics Approval
None required for a case report.

Contributions
CN/MO wrote the first draft. All authors reviewed, edited the manuscript, and approved the final version.

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References
Maternal metastatic synovial sarcoma


