CASE REPORT

Synovial sarcoma in an HIV-positive pregnant woman and review of literature

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SUMMARY

We present a first case of synovial sarcoma in an HIV-positive pregnant woman. This 28-year-old woman was diagnosed with synovial sarcoma, a high-grade malignant soft tissue sarcoma, involving her left thigh during the first trimester of her pregnancy. She underwent surgical treatment in the form of hip disarticulation at 30 weeks' gestation. She was subsequently delivered by emergency caesarean section (CS) at 34 weeks' gestation when she presented with wound sepsis and a scan revealed static growth in a small for gestational age fetus. Prompt diagnosis and treatment of this aggressive tumour is important and should involve a multidisciplinary approach, with a balanced consideration of the maternal and fetal outcomes.

BACKGROUND

Sarcomas are rare cancers which can affect almost any part of the body and can develop in the muscle, bone, nerves, cartilage, tendons, blood vessels and the fatty and fibrous tissues.¹ There are about a hundred different subtypes of sarcomas, but the main types include: bone and soft tissue sarcomas.¹ Synovial sarcomas are high-grade malignant soft tissue sarcomas; they account for 5%–10% of soft tissue sarcomas and arise most frequently in the extremities of young adults.² Overall, the diagnosis of cancer occurs in one per 1000 pregnancies; the most common being cancers of the breast, uterus and cervix, lymphomas and melanomas.³ The diagnosis of synovial sarcoma in pregnancy is even more rare. Our literature search revealed that only 12 cases of synovial sarcomas during pregnancy have been reported since 2006. We present a first case of synovial sarcoma in an HIV-positive pregnant woman.

CASE PRESENTATION

We present the case of a 28-year-old Caucasian woman with a background of two previous caesarean sections (CS) and a diagnosis of HIV infection since the age of 21. During her routine HIV clinic appointment, she was found to be pregnant. A mass was also noted on her left thigh (figure 1), so she was referred for urgent ultrasound scan (USS) and consultant-led antenatal care. Unfortunately, she defaulted from most of her care, apart from her left thigh USS and her dating and anomaly obstetric scans. This led to a significant delay in diagnosis and initiating her treatment plan. The USS of the

left thigh mass raised suspicion of a soft tissue sarcoma. The subsequent MRI scan demonstrated a large heterogeneous soft tissue mass within the left medial thigh. The lesion, which had replaced most of the distal fibres of the left adductor longus muscle belly, was encasing the femoral vessels and measured at least 14 cm (craniocaudal) by 10 cm (anteroposterior) by 8 cm (mediolateral). Infiltration of the left sartorius muscle was also noted in that region with no signs of osseous infiltration (figure 2).

She was referred to the regional sarcoma service where a biopsy confirmed a histological diagnosis of synovial sarcoma (figure 3). A CT scan showed no evidence of distant metastases. At this point, she was 27 weeks' pregnant and the initial stage of the cancer was T2b, N0, M0, G2. Two operative choices were discussed with her. The first being leg sparing surgery with en bloc tumour resection and reconstruction of the femoral vessels, free flap to cover the defect and postoperative radiotherapy (60Gy). The second option was hip disarticulation surgery which would be more curative but involve extensive postoperative rehabilitation. She opted for hip disarticulation surgery.

Initially, her HIV viral load was detectable at 130 copies/mL and her CD4 T-cell count was 895 cells/ μ L. She had a long history of poor compliance with her antiretroviral therapy which included Truvada, Atazanavir and Ritonavir. Raltegravir was added at 25 weeks of pregnancy to intensify her regimen. This led to a reduction of her viral load to <40 copies/mL by 28 weeks of pregnancy.

OUTCOME AND FOLLOW-UP

In view of the high grade and aggressive nature of the tumour, she had her initial surgery at 30 weeks of pregnancy under a multidisciplinary team involving the obstetric, orthopaedic, HIV, oncology and sarcoma specialists. Prophylactic steroids were administered because of the risk of premature delivery. Postoperatively, the cancer staging was pT2b, N0, M0, G2.

She later presented at 34 weeks of pregnancy with sepsis arising from an infected leg stump wound which required surgical management. An obstetric USS showed a small for gestational age (SGA) fetus with an estimated fetal weight under the 10th centile and static growth. An emergency delivery by CS was therefore planned in addition to the wound exploration and debridement. A healthy female neonate weighing 1756 g was delivered. She required two further wound debridement

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Figure 1 Mass on patient's left upper thigh.

procedures and subsequent long-term intravenous antibiotics. She was discharged home 4 weeks postoperatively with a peripherally inserted central catheter for long-term administration of intravenous antibiotics in the community which she required for a total of 11 weeks. She also had a plan for intensive rehabilitation and surveillance in the form of clinical review and chest X-rays every 3 months for 2 years and biannually until year 5 and then annually until year 10.

DISCUSSION

Sarcomas are rare cancers which can develop in the muscle, bone, nerves, cartilage, tendons, blood vessels and the fatty and fibrous tissues.¹ There are about a hundred different subtypes of sarcomas, but the main types include: bone and soft tissue sarcomas.¹ Synovial sarcomas are high-grade malignant soft tissue sarcomas; they account for 5%–10% of soft tissue

Figure 2 MRI image of the lesion, which had replaced most of the distal fibres of the left adductor longus muscle belly, very close to the femoral vessels. Infiltration of the left sartorius muscle was also noted in that region with no signs of osseous infiltration.

Figure 3 Monophasic fibrous synovial sarcoma. Microphotograph showing monomorphic, small spindle cells arranged in fascicles (H&E stain; 10× magnification).

sarcomas and arise most frequently in the extremities of young adults.² The poor prognosis associated with synovial sarcomas is related to a high rate of local recurrence and late metastases.² As with our patient, diagnosis is often delayed due to the slow tumour growth and benign nature of the presenting symptoms, leading to delayed presentation or referral.

Overall, the diagnosis of cancer occurs in one per 1000 pregnancies; the most common being cancers of the breast, uterus and cervix, lymphomas and melanomas.³ The diagnosis of synovial sarcoma in pregnancy is even more rare. Our literature search revealed that only 12 cases of synovial sarcomas during pregnancy have been reported since 2006. Of the 12 cases of synovial sarcomas during pregnancy that have been reported since 2006, five were pulmonary,^{3–7} three were renal,^{8–10} two involved the head and neck,^{11 12} one involved the abdominal wall,¹³ and the last, like our patient, involved the upper left thigh.¹⁴ The three patients who passed away during the course of follow-up all had the diagnosis of pulmonary synovial sarcomas. Patients were aged between 19 and 38 years and presented at various gestations between 8 and 34 weeks, with signs and symptoms related to the primary site of the tumour.

The recommended treatment for synovial sarcoma is surgery followed by radiotherapy; chemotherapy is reserved for patients with advanced or metastatic disease,¹⁵ although experts still debate the role of chemotherapy in preventing metastases and improving survival. Indeed, the management plan for all 12 patients involved surgical resection of the tumour, however the choice of additional therapy was not as consistent. Two of the patients went on to have adjuvant chemotherapy and radiotherapy. Five patients only had chemotherapy either before or after surgery, while one patient had only radiotherapy with fetal shielding following surgery. Two patients declined any adjuvant therapy, and for the last two patients there was no mention of any other treatment. Our patient had ablative rather than limb-sparing surgery and the excision margins were very wide. For these reasons, the consensus at the multidisciplinary team meeting was that postoperative radiotherapy was not indicated. It is worth noting that her HIV status and the fact she was post-partum had no impact on this decision. If she had attended her follow-up appointment, the role of chemotherapy would have been discussed as this could still be used alongside surgery for this tumour type.

With regard to fetal outcomes, three patients opted to terminate their pregnancy. Of the nine patients who progressed with their pregnancy, only one delivered at term and this was by CS. Four of the five patients who delivered preterm were also delivered by CS. It was not always obvious why a CS was performed. Mode of delivery in pregnant patients with cancer is based on a multidisciplinary approach and in conjunction with the obstetric recommendation.¹⁶ There was no record of how or when the remaining three patients delivered. Our patient was delivered by CS in view of her previous two CS, the SGA fetus at 34 weeks, and the fact that a combined operation with the orthopaedic team was also required. Her baby girl was born in a very good condition, but was taken to the special care baby unit for 15 days because she was born prematurely weighing less than the 10th centile, and to provide our patient with the extra support during her postoperative recovery. As of when she was discharged from community care 15 weeks postnatally, her baby remained well and was meeting all developmental milestones.

People who have a weakened immune system, such as those with HIV, have an increased risk of sarcoma. However, the most common type of sarcoma associated with HIV infected patients is Kaposi's sarcoma. Kaposi's sarcoma is caused by human herpes virus 8 and is considered an AIDS-defining illness. There can be involvement of the skin, mouth, gastrointestinal and respiratory tract. Since 1971, there has been 14 reported cases of Kaposi's sarcoma in pregnancy in both seropositive and seronegative patients.^{15 17–19}

To the best of our knowledge, this is the first reported case of synovial sarcoma in an HIV-positive pregnant woman. The only other case of synovial sarcoma in an HIV-positive patient in the literature was a case of pulmonary synovial sarcoma in an HIV-positive man.²⁰ Our case was particularly interesting because of the multidisciplinary input required from the obstetric, HIV, orthopaedic, sarcoma, oncology, neonatology, microbiology, tissue viability and physiotherapy teams.

As with most cases of cancer during pregnancy, this case presented challenges surrounding the need to investigate and treat the woman optimally, while minimising harm to the fetus. From a diagnostic point of view, oncology cases in pregnancy can be investigated using different imaging modalities, such as CT and MRI, depending on gestation and locality of the tumour. Imaging should not be withheld simply because of the potential toxicity to the fetus without considering the risk of compromising maternal outcomes. It is important to realise that if the benefit of a test is indisputable, then almost all imaging tests expose the fetus to such low levels of radiation that they are not a cause for concern.²¹

From a therapeutic and ethical point of view, consideration had to be given to the timing of the hip disarticulation. In view of the aggressive nature of the tumour, it was important not to delay surgery, yet allow the patient sufficient time to fully consider the implications of having life changing limb amputation during pregnancy and the extensive rehabilitation that would follow her surgery. As with the other patient with synovial sarcoma involving the thigh,¹⁴ because of the tumour location, size and proximity to the main femoral vessels, it was deemed that the optimal curative surgery for our patient was a hip disarticulation. Whereas the patient from the literature review declined the operation and rather opted for limb-sparing surgery and adjuvant chemotherapy and radiotherapy, with positive results seen on her follow-up MRI, our patient chose to have hip disarticulation as she did not wish to take the risk of recurrence. Hip disarticulation is an amputation through the hip joint capsule, removing the entire lower limb, with closure of the remaining

musculature over the exposed acetabulum.²² Early and intensive physical and psychological rehabilitation before and after surgery can result in most patients being completely independent and ambulatory at the end of 1 year.²²

The use of radiotherapy in pregnancy is well documented depending on the tumour location and type. Radiation to the pelvis is best avoided because of high fetal exposure. When administered in the first trimester, exposure to radiation has been linked to fetal anomalies, miscarriage and intrauterine growth restriction (IUGR).²³ There is a theoretical risk of childhood cancers following maternal exposure to radiation in pregnancy.²³

Chemotherapy has been used in different oncology cases in pregnancy too and is considered safe in the second and third trimester. In the first trimester it has been associated with malformations, miscarriage and IUGR.²³ There is better maternal survival when chemotherapy is administered during the second trimester when compared with delaying treatment until the post-natal period.²³

Overall, prompt diagnosis and treatment of this aggressive tumour are important and should involve a multidisciplinary approach, with a balanced consideration of the maternal and fetal outcomes.

Patient's perspective

I was very happy for all the care I received even after having my baby. I am very pleased that you are writing up my case so that everyone can learn about this rare condition in pregnancy.

Learning points

- ▶ To the best of our knowledge, we present the first reported case of synovial sarcoma in an HIV-positive pregnant woman. This is significant because of the diagnostic, therapeutic and ethical challenges faced when managing the case.
- ▶ As with most cases of cancer during pregnancy, it was important to investigate and treat the woman optimally, while minimising harm to the fetus.
- ▶ Particular to this case was the need to not delay surgery, in view of the aggressive nature of the tumour, yet allow the patient sufficient time to fully consider the implications of having life changing limb amputation during pregnancy and the extensive rehabilitation that would follow her surgery. Such complexities required multidisciplinary input from the obstetric, HIV, orthopaedic, sarcoma, oncology, neonatology, microbiology, tissue viability and physiotherapy teams.

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