Recurrent spindle cell sarcoma in pregnancy: a case report

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Abstract

Malignant Fibrous Histiocytoma (MFH) is a soft tissue sarcoma usually involving limbs and retroperitoneum. MFH of the anterior abdominal wall is rare with a high recurrence rate. Though wide surgical excision is the mainstay of treatment, it creates large abdominal wall defects that need reconstruction. The addition of chemotherapy and radiotherapy optimize the chances of survival and cure. We report a case of twice recurrent pleomorphic spindle cell sarcoma of the anterior abdominal wall in pregnancy, with adverse maternal and fetal outcome.

Key words: Pregnancy, Spindle cell sarcoma, Anterior abdominal wall, Adverse maternal and fetal outcome, Suboptimal management

Introduction

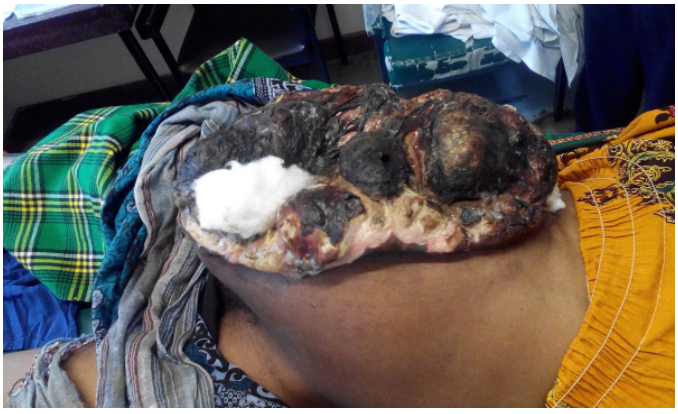
Malignant Fibrous Histiocytoma (MFH) refers to a group of aggressive soft tissue tumours characterized by a cartwheel growth pattern arising from fibroblasts and hystiocytes (1). Known risk factors include radiation exposure, surgical incision or burn scars (2-4). MFH is histologically classified into storiform-pleomorphic, myxoid, giant cell, inflammatory and angiomatoid types. The first two types are high grade, whereas the others are low grade. Of these, the storiform-pleomorphic is the most common type (70%). It comprises spindle-shaped cells arranged in short fascicles in a cartwheel or storiform pattern, along with plump histiocytic cells showing mitotic features (5).

The most common clinical presentation is an enlarging painless soft-tissue mass in the thigh (6). We present a patient who had twice recurrent anterior abdominal wall pleomorphic malignant spindle cell sarcoma that resulted in adverse maternal and fetal outcome.

Case Report

A 40 year old para 3+0 gravida 4 of unknown gestation presented at the Kenyatta National Hospital in active labour with a huge fungating, purulent necrotic anterior abdominal mass as shown in Figure 1. Six weeks prior to presentation, she developed a painful rapidly growing ulcerating mass on her abdomen at the site of a tumour that had been excised twice prior to her pregnancy. Three weeks later, she experienced abnormal abdominal movements, which prompted a visit to a nearby local facility where pregnancy was diagnosed by a positive urine beta human chorionic gonadotropin test.

Figure 1: Left superolateral view of the anterior abdominal wall mass



Her past medical history revealed that she had had surgical excision of a 3 by 2 by 2 cm anterior abdominal wall mass three years before presentation. It was histologically diagnosed as a malignant pleomorphic spindle cell sarcoma of the anterior abdominal wall that had invaded the subcutaneous tissue. Two years later, the mass recurred, and it measured 20 by 16 by 10 cm. Distant metastases were ruled out by an abdominal computerized tomography scan and a chest radiograph. The mass was excised and the anterior abdominal wall defect was repaired with a mesh. Histology revealed a high grade pleomorphic fibrosarcoma that was staged at T2b, NX M0. There was no history of chemotherapy or radiotherapy. Following this excision, she was on three monthly depomedroxyprogesterone acetate injections for nine months, but was amenorrheic for about six months after she stopped receiving them, which is when she likely conceived the current pregnancy.

On examination at admission, she had normal vital signs. The anterior abdominal wall mass measured 40 by 30 cm, extending from the epigastric to suprapubic regions as shown in Figure 1. It was exquisitely tender, purulent, malodorous, fungating and necrotic with irregular margins, thereby precluding performance of Leopold's manoeuvres or fetal monitoring. Vaginal examination revealed active labour in breech presentation. Four hours later, she expelled a grossly normal two kilogram macerated male fetus with a grossly normal placenta and umbilical cord. The fourth stage of labour was uneventful.

Investigations done during labour included a haemogram, urea/electrolytes and creatinine and liver function tests. They revealed a mild anaemia of 8g/dl, leukocytosis with neutrophilia (29.30 x 10^9/L and 85.8% respectively). Glomerular and tubular functions were deranged (Urea at 13.9 mmol/L and Creatinine at 139 umol/l) and liver function was normal. Postnatally, she received three units of whole blood, intravenous hydration and antibiotics. Two weeks after delivery, the oncology team prescribed Etoposide 1000mg/m^2, Ifosfomide 1800mg/m^2 and Mesna 1750 mg/m^2 (Day 1 – 5) in 21 – 25 day cycles. However, before the chemotherapy could begin, the patient developed a septic enterocutaneous fistula and died before surgical intervention.

Discussion

Although malignancies may occur in pregnancy in up to 0.1% of cases, sarcomas are rare (7). MFH often occurs as a mass in the lower and upper extremities (68%), with only 16% occurring in the abdominal cavity. The peak incidence is 61-70 years (6). Large tumour size, high histological grade, presence of distant metastases and myxoid subtypes are adverse prognostic indicators (6,8). The current case presented with a large recurrent high-grade pleomorphic tumour with no previously demonstrated metastases in pregnancy.

Smaller, superficial tumours metastasize less frequently than the larger, more deeply located tumours (4), with up to 90% of distant metastases occurring to the lungs (4,6). This patient, despite having a huge recurrent tumour, did not display any clinical stigmata of pulmonary metastasis. However, since there were no fresh radiological investigations undertaken during this admission, pulmonary metastases could not be conclusively ruled out.

Though surgery is the mainstay of treatment (9), it was not possible to assess the adequacy of the initial and second excisions, as the histological reports did not indicate whether or not there were negative margins. Although chemotherapy and radiotherapy have been shown to enhance the probability of cure (9,10), there was no indication that this patient received either prior to her current presentation. The chemotherapy regimen prescribed during her terminal admission was Etoposide and Ifosfamide. However, she died before receiving the first course due to a high output enterocutaneous fistula complicated by sepsis, in agreement with the findings of Campos *et al* (11). She would likely have benefited from chemotherapy and radiotherapy after surgical excision following the first recurrence.

On the adverse fetal outcome, it is difficult to determine whether there was a separate factor that could have accounted for the fetal demise apart from the malignancy, as the fetus, cord and placenta were grossly normal. However, the presence of the necrotic anterior abdominal wall mass would likely have precluded surgical access to expedite delivery, even if it had been possible to diagnose a non-reassuring fetal status prior to fetal demise.

Conclusion

Malignancies complicate pregnancy and pregnancy complicates malignancies (7). Wide excision is recommended, with chemotherapy and radiotherapy being shown to optimize survival (9). Ultimately, this patient received suboptimal treatment during her initial recurrence, which may ultimately have been responsible for her adverse fetal and maternal outcomes.

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