A case of B-cell lymphoma in a pregnant woman presenting with superior vena cava obstruction

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Summary: We present a case of non-Hodgkin's lymphoma in a pregnant woman who presented at a very young age, which is unusual, and also the management dilemmas of a mediastinal mass in pregnancy. Maternal malignancies are very rare during pregnancy; this report gives a brief overview of the management and the complications that might be encountered.

Keywords: B-cell lymphoma, pregnancy, superior vena cava obstruction

We present the case of a 20-year-old Caucasian woman who, in her first pregnancy, presented to her local hospital at 30 weeks of gestation with a two-week history of haemoptysis. On examination, there were significant findings which were confined to the respiratory system and signs of superior vena cava obstruction.

A chest X-ray revealed a right-sided mediastinal mass extending from the sternoclavicular junction to the diaphragm, confirmed by chest computed tomography (CT) scan. Mediastinal biopsy and immunohistochemistry confirmed the diagnosis of a high-grade diffuse large B-cell non-Hodgkin's lymphoma (NHL) (Stage II A B-cell lymphoma). A bone marrow biopsy was negative for malignancy and ultrasound of her abdomen and transthoracic echocardiogram were normal. Likewise, a fetal ultrasound scan was appropriate for gestation.

She was then referred to our tertiary unit and the case was reviewed by the multidisciplinary team (MDT), comprising an obstetrician, a haematologist, an oncologist and anaesthetists. A plan to commence chemotherapy was made as maternal respiratory effort was compromised due to the size and location of the mass. Also betamethasone was administered to aid fetal lung maturity as preterm delivery was contemplated.

At 32 weeks of gestation, her first cycle of chemotherapy (Rituximab–Cyclophosphamide Doxorubicin – R-CHOP 14 without vincristine) was administered. Two weeks later her blood results were within the normal range (no neutropenia) and at 34 + 5 weeks of gestation an elective caesarean section was performed under spinal anaesthesia. The procedure was uneventful and a healthy male infant was delivered (birth weight 2320 g). Two days post-delivery she received her second course of chemotherapy (R-CHOP 14 with vincristine). This was well tolerated and she was discharged home to complete her treatment as an outpatient.

DISCUSSION

Maternal malignancy is unusual during pregnancy (0.1%), and mediastinal tumours are particularly rare.¹ Lymphoma is characterized by an uncontrolled proliferation of lymphocytes and is the fourth most common malignancy during pregnancy.

The World Health Organization Classification² groups lymphomas by cell type: B-cell, T-cell and natural killer cell tumours. Hodgkin's lymphoma predominantly affects women of childbearing age, average age 25.5 years. The incidence has been reported to be one in 1000 to 6000 pregnancies.³ The diagnosis is based upon the presence of clonal malignant Hodgkin cells or multinucleated Reed Sternberg cells. In comparison, the incidence of NHL is approximately 0.8/100,000⁴ and the peak incidence occurs at a mean age of 42 years. In contrast to this, our patient was 20 years old. NHL is differentiated from Hodgkin's lymphoma by the absence of Reed Sternberg cells.

This neoplasm begins in lymphoreticular tissue and spreads throughout the lymphatic system. The usual clinical signs are lymphadenopathy, weight loss and night sweats. As well as reluctance to subject the fetus to radiation from X-rays, the initial diagnosis of mediastinal mass is particularly difficult as signs and symptoms are similar to common complaints during pregnancy.⁵ However, dyspnoea that is progressive, and is associated with a cough and orthopnoea, is suggestive of underlying pathology.⁶

A thorough history and examination is vital, followed by a chest X-ray. A CT scan of the thorax helps to assess the size of the tumour and degree of airway compression. CT scan and X-ray both involve risk of fetal radiation and to limit this, many centres perform chest imaging with subdiaphragmatic lead shielding, which was implemented in our case. Ultrasound scanning can be used as an adjunct to partially stage the abdomen. Magnetic resonance imaging, where available, is the imaging of choice in pregnant patients.⁷ An echocardiogram was performed to assess cardiac output and to rule out cardiac tamponade.

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The treatment decision is based on a combination of tumour grade, the stage of the pregnancy and patient preference. This requires a balance between the fetal and maternal risks posed by the disease and its management on a case-by-case basis.⁷ Treatment of lymphoma diagnosed in the first few weeks of pregnancy may necessitate a difficult decision about termination of the pregnancy. For lymphoma diagnosed in the third trimester, in most cases treatment can be delayed until after delivery. Some of the indolent lymphomas do not require immediate treatment but for higher-grade tumours, such as in our case, such delay may compromise treatment. Treatment with radiotherapy and/or chemotherapy depends on tumour grade and maternal symptomatology. Upward displacement of the diaphragm by the gravid uterus reduces lung size, causing the mediastinal mass to occupy more of the intrathoracic area, which may increase the risk of radiation-induced pulmonary damage if radiotherapy is performed before delivery.⁸ With chemotherapy during pregnancy there are concerns about fetal organogenesis, growth restriction, preterm labour and stillbirth.⁹

Patients with a large mediastinal mass are a challenge for the anaesthetist as there is a high incidence of mortality and morbidity associated with general anaesthesia.¹⁰ As a result of our patient's respiratory symptoms and the size of the mass, chemotherapy was administered to improve lung function and reduce anaesthetic risks. Ideally, delivery should be timed to maximize fetal lung maturity and minimize the risk of neonatal myelosuppression, which may occur in the first few weeks after chemotherapy. Thus, in our case beta-methasone was administered and delivery timed two weeks after chemotherapy. A caesarean section was preferred to a vaginal delivery as it avoided an increase in maternal intrathoracic pressure during pushing. In our case histopathological examination of the placenta was not done, although this is recommended as with haematological malignancies there is a potential risk of maternal-to-fetal transmission. Postpartum haemorrhage can be poorly tolerated as there is a compromised venous return. Pharmacological treatment for uterine hypotonia can cause profound cardiopulmonary disturbances and minimal fluid overload can lead to pulmonary oedema. Neither of these complications was experienced by our patient.

The management of pregnant women with cancer presents a major challenge: it is essential that women with this condition are managed by an MDT for the best outcome.

DECLARATIONS

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