Case Report

Primary Brain T-Cell Lymphoma during Pregnancy

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Abstract

The incidence of non-Hodgkin's lymphoma (NHL) during pregnancy is about 0.8 cases per 100,000 women. We describe a case of a 33-year-old woman with primary brain T-cell NHL who was diagnosed at the 32nd week of gestation. She visited the emergency room complaining of a headache, vomiting and drowsiness. Her pregnancy had been uneventful prior to the admission. Brain magnetic resonance imaging (MRI) revealed a mass at the anterior cranial fossa in the mid liner in the front of the lateral ventricle of the brain. She was hospitalised and monitored regarding her complaints until the foetal lungs matured. During her stay, systemic evaluation revealed no other pathology. She delivered a healthy baby in the 34th week of gestation by Caesarean section and the tumour was evacuated while under the same anaesthesia. She underwent another surgery 12 hours after the first operation because of cerebral herniation. Despite aggressive treatment in the neurosurgical intensive care unit, her condition continued to deteriorate and she died on the tenth postpartum day. Examination of the patient at necropsy revealed no other pathology. The lymph nodes and bone marrow were not involved.

Keywords: Pregnancy, Lymphoma, Brain.

Introduction

Malignant brain tumours during pregnancy have a 64% lower occurence in non-pregnant females of a similar age, with an incidence of 3.6 per million live births.[1] The distribution of histological types of brain tumours are similar in both pregnant and non-pregnant women.[2] The most common malignant intracranial tumour is metastasis of systemic malignancies to the brain.[2]

Although primary brain tumours are a relatively infrequent cause of complications and death in women, certain neoplasms e.g. meningiomas, pituitary adenomas, and vestibular schwannomas have a predilection for women.

Lymphoma is diagnosed in approximately 1 in 6000 deliveries and it is the fourth most frequent malignancy diagnosed during pregnancy.[3] Haematological malignancies, as a group, constitute 25% of the cancers complicating pregnancy, behind carcinomas of the breast (26%) and cancer of the uterine cervix (26%).[4] The incidence of lymphomas involving the central nervous system (CNS) has increased substantially. Despite this increase, primary CNS lymphoma still remains rare in women, with an incidence of 0.02 per 100,000 women-years and the incidence of non-Hodgkin's lymphoma (NHL) during pregnancy being 0.8 cases per 100,000 women.[5]

The clinical presentation of intracranial neoplasms depends on several mechanisms; mass and herniation effects secondary to increased intracranial pressure (ICP), local irritation or destruction of cerebral tissue and cranial nerves,

or secondary hormonal effects related to pituitary dysfunction. The presenting symptoms depend on the area of the brain affected by the lesion. Common symptoms arising from a mass effect include headache, nausea and non-specific visual changes. Other symptoms include decreased level of consciousness or changes in mood or personality.

Case Report

We present the case of a 33-year-old woman with primary brain T-cell NHL diagnosed in the 32nd week of gestation. She visited the emergency room complaining of headache, vomiting, and drowsiness. Her pregnancy had been uneventful prior to the episode. Brain magnetic resonance imaging (MRI) revealed a mass at the anterior cranial fossa in

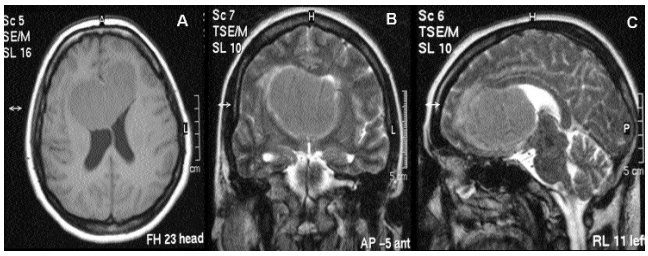

Figure: MRI of the brain tumour including (A) axial, (B) coronal, and (C) sagittal views; there is a 10×5-cm mass (white arrows), localised at the anterior cranial fossa in the midline, in front of the lateral ventricles with compression of the third ventricle and minimal oedema.

the midline, in front of the lateral ventricles with compression of the third ventricle of the brain. She was hospitalised and followed in the neurosurgery unit until the foetal lungs had matured. During this period, a systemic evaluation revealed no other pathology. She delivered a healthy baby by Caesarean section in the 34th week of gestation and the tumour was evacuated under the same anaesthetic. She underwent additional surgery 12 hour after the initial operation because of cerebral herniation. Despite aggressive treatment in the neurosurgical intensive care unit, her condition continued to deteriorate and she died on the tenth postpartum day.

Discussion

The treatment of CNS lymphoma includes high-dose methotrexate combined with radiation. With this treatment regimen, the median survival is 40 months and the 5 year survival rate is 22%.[6] Cyclophosphamide, doxorubicin, vincristine and prednisone, a standard regimen for systemic lymphoma, fails to add any benefit when used after cranial radiation therapy.[7]

Although the stage and immunohistological types of lymphoma may differ in pregnancy, their clinical behaviour, when properly treated, does not seem to differ significantly from that in non-pregnant patients. Treatment choices must be based on the stage, classification, and International Prognostic Index. Recent studies in non-pregnant patients have shown that with aggressive NHL, standard therapy with cyclophosphamide, doxorubicin, vincristine, and prednisone results in 3-year overall survival rates (53-62%) that are not significantly different from those with other intensive chemotherapeutic regimens (48-56%).[8] Similar long-term survival rates in pregnancies complicated by NHL and treated with multi-agent chemotherapy have been reported.[4] For pregnant patients in the first trimester, because of concerns of possible adverse foetal effects of multi-agent chemotherapy (15-25%), the risks of foetal anomalies, delaying chemotherapy and progression of the maternal disease need to be balanced with the patient's desire to avoid potential harm to her foetus.[9] Chemotherapy should be delayed until the second trimester and if possible, delivery should be timed to occur 2-3 weeks after chemotherapy to allow for maternal and foetal blood count recovery.[9] In the third trimester, however, there are a few circumstances in which radiation therapy is used before the delivery. In this situation, steroids should be administered and the baby delivered after 32-34 weeks gestational age, once foetal lung maturation is confirmed by amniocentesis. If spontaneous labour ensues after 32 weeks, it should be allowed to progress, as long as antenatal steroids have been administered, and there are no obstetric indications contra-indicating spontaneous vaginal delivery.[9] Cerebral oedema resulting from brain tumours can be treated with glucocorticoids, although their mechanism of action is unknown.

Conclusions

Fortunately, haematological malignancies occurring during pregnancy are uncommon. It is imperative that physicians and health care providers approach these patients and their families with compassion, empathy and most importantly the knowledge and expertise necessary to optimise the outcome for both mother and baby. A multidisciplinary approach is necessary to manage these cases.

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