Primary Uterine Leiomyosarcoma and Primary Atypical Meningioma Diagnosed during Pregnancy

BARBARA BODNER-ADLER, PLAMEN LOZANO, KLAUS BODNER and HARALD ZEISLER

Department of Obstetrics and Gynecology, University of Vienna Medical School, Vienna, Austria

Abstract. Objective: Uterine leiomyosarcoma during pregnancy is an extremely rare event. The incidence of meningioma during pregnancy is comparable with that in nonpregnant women of the same age group. We report a case of both – a primary uterine leiomyosarcoma and additionally an atypical meningioma of the brain both diagnosed during pregnancy. Case Report: The patient was admitted with generalised seizures at 31 weeks of gestation. A tumoral mass was detected and initial conservative treatment was started. The patient delivered her infant via caesarean section, at 34 weeks of gestation. During caesarean section a pedunculated uterine fibroid was removed and total gross resection due to the brain tumour was also performed. Histopathological diagnosis of both tumours revealed an atypical meningioma of the brain and a uterine leiomyosarcoma. The patient underwent laparotomy and received six cycles of adjuvant chemotherapy. Conclusion: We are the first to report a case of a woman with two separate primary neoplasms both diagnosed during pregnancy. Treatment options seem to be reduced in pregnant women and mainly depend on the patient’s condition as well as the gestational age at presentation.

Uterine leiomyosarcomas (LMS) are rare, highly malignant neoplasms that make up about 1% of all uterine malignancies (1). The risk of local recurrence and metastasis is high, with reported 5-year survival rates ranging between 12% and 25%. The median age for women with LMS is after childbearing age (between 43 and 53 years). Total abdominal hysterectomy and bilateral salpingo-oophorectomy are considered the standard therapy for LMS of the uterus (2, 3). The role of conservative fertility-sparing surgery in young women remains controversial (4). Uterine LMS associated with pregnancy is even rarer and only a few cases are reported in the literature (4, 5). The incidence of meningioma in pregnant women is comparable with that in nonpregnant women of the same age group (6).

Here we report a case of a patient with both uterine LMS and atypical meningioma of the brain diagnosed during the second trimester of pregnancy.

Case Report

We report the case of a 39-year-old woman, gravida 1 para 0, who presented in her 31st week of pregnancy at the Department of Neurology with a first-ever documented seizure, deteriorating neurological status and vertigo. Her past surgical history was uneventful and her family history was not significant. The ultrasound examination at this time revealed a normal pregnancy at 31 weeks of gestation.

Contrast-enhanced magnetic resonance imaging (MRI) was performed immediately and revealed a 5×4.5 cm well-defined mass in the right parietooccipital lobe causing mass effect and elevated intracranial pressure. After improvement of the patient’s condition and with the brain tumour suspected of being of benign origin, an urgent neurosurgical intervention was not necessary. Conservative treatment with antiepileptic medicaton and fortefortin was started.

At 34 weeks of gestation, the patient was administered 2×12 mg dexamethasone to achieve fetal lung maturation and a caesarean section (C/S) was performed under spinal anesthesia. During C/S, a large pedunculated and solitary fibroid of the fundus uteri was detected and was easily removed. Gross pathological examination of the uterine fibroid revealed a tumour measuring 12 cm in diameter. The external surface was irregularly nodular, the core appeared homogeneous, soft, grey-red brown in colour as a sign of a necrotic process.

A male baby was delivered with a birth weight of 2,917 g with APGAR score 9/9/10 at 1, 5 and 10 minutes respectively. The arterial cord pH was 7.28. The follow-up of the newborn showed him to be a healthy infant.
Total gross resection of the brain tumour was performed via a right frontal craniotomy. Histological analyses of the tissue showed monomorphic, predominantly meningotheliomatous cells with focal mitotic activity exceeding 5 mitoses per 10 high-power fields fulfilling the criteria for atypical meningioma.

The histopathological and immunohistochemical analysis of the removed uterine fibroid revealed a uterine LMS. The tumour was characterised by high cellularity and nuclear polymorphism, as well as patchy necrosis and large areas of haemorrhage. Mitotic activity exceeded 10 mitoses per 10 high-power fields with many atypical mitoses. Coagulative tumour cell necrosis occupied as much as 2/3 of the tumour. Well-documented invasion of the pre-existing myometrium and infiltration of blood vessels was analogous to an LMS. Immunohistochemically, the tumour showed strong positive staining reaction for muscle actin, alpha-smooth muscle actin and neuron-specific enolase, as well as a weak positive reaction for vimentin.

The patient underwent a total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy and paraaortic lymphadenectomy. The postoperative management was uneventful. Adjuvant chemotherapy consisting of six cycles of epirubicin and ifosfamide was administered. The patient was followed up at three-month intervals by means of vagino-rectal palpation, blood analysis and computed tomography of the chest and abdomen and MRI of the brain. At 19 months of follow-up after operations and chemotherapy, the patient was well and without any sign of recurrent disease.

Discussion

We report the case of a patient with uterine LMS as well as atypical meningioma, both diagnosed during pregnancy.

All reported cases of uterine LMS associated with pregnancy were found incidentally in pregnancy. Kyodo and colleagues reported the only case in which they had observed the progress of the tumour during pregnancy (5).

Similarly, the diagnosis of our patient was made histologically after operation. The fibroid was detected at the beginning of pregnancy and follow-ups with ultrasound examinations were performed. Neither any progress nor any clinically suspicious indications could be detected during pregnancy and therefore the tumour was mistakenly diagnosed as a leiomyoma. As the fibroid was solitary and pedunculated, it was easily removed during C/S. The indication for elective C/S at 34 weeks of pregnancy was a brain tumour also diagnosed during this pregnancy. Tumour growth during pregnancy is not unusual and has been demonstrated for uterine leiomyoma as a result of high estrogen and progesterone levels. In our patient, no progression was documented.

The appropriate management of uterine LMS in pregnancy remains unclear. Since the patients are of a childbearing age, there is a desire to conserve fertility if possible. Uterine LMS is generally considered to be more aggressive than other types of uterine tumour and is well known for its frequent recurrence and resistance to chemotherapy. Therefore, an aggressive surgical procedure is usually considered to be the treatment of choice at the time of diagnosis (1). Our patient underwent a total abdominal hysterectomy with bilateral salpingo-oophorectomy, omentectomy and paraaortal lymphadenectomy 6 weeks after CS. An adjuvant chemotherapy consisting of six cycles of epirubicin and ifosfamide was given postoperatively. Fortunately, the diagnosis of uterine LMS as well as menigioma was made at the end of the second trimester and the pregnancy was not influenced by these diseases.

Brain tumours in pregnancy are extremely rare events with only a few reports existing in the literature (6-9). Isla and colleagues reported e.g. seven cases of pregnant women who presented with neurological symptoms during pregnancy and subsequent diagnosis included 2 meningiomas, 2 ependymomas and 2 gliomas (7). Cushing and Eisenhardt in 1938 were the first to describe the relation between pregnancy and the rapid increase of neurological symptoms in women with meningiomas (8). It is undisputable that a hormonal relationship may play a role in the appearance of some tumours, particularly meningiomas (9). A case-control study in Iowa, USA, examined the effect of parity and age at first birth on the risk for various types of carcinomas (10). The authors reported that the risk of brain cancer was slightly higher in women older than 25 years. This is in line with our observation, as our patient was 39 years old when the tumour was diagnosed.

Treatment generally consists of surgical resection. The management strategy for brain tumours during pregnancy should be tailored to the individual case. In our patient, conservative management was possible for 3 weeks to allow foetal lung maturation and at the end of 34 weeks of gestation an elective C/S was performed. In conclusion, we are the first to describe a case with two primary unassociated tumours, both diagnosed during pregnancy. Symptoms such as headache, mental status changes and seizures are common manifestations of eclampsia but the knowledge of intracerebral tumours during pregnancy as differential diagnosis is also important.

Treatment options seem to be reduced in pregnant women and mainly depend on the patient’s condition as well as on the gestational age at presentation. In a multidisciplinary approach, an optimal therapy schedule should be assessed depending on these two conditions.

References

Bodner-Adler et al: Primary Uterine Leiomyosarcoma and Primary Atypical Meningioma Diagnosed during Pregnancy


Received March 20, 2008
Revised June 10, 2008
Accepted June 16, 2008