Pheochromocytoma and Pregnancy: a Case Report

S.Belaazri, FZ.Lamine, A.Siati, N.Zeraidi, A.Baidada, A.Kharbach
Gynecology and obstetrics Service M1-M3, Maternity Souissi, University Hospital Ibn Sina, Rabat, Morocco
Corresponding Author: Sofiane Belaazri
sbelazri23@hotmail.com

ABSTRACT

Pheochromocytoma is the tumor of neural crest cells of adrenal gland secreting catecholamines. We report a case of a patient diagnosed with pheochromocytoma during her 2nd pregnancy at 17 week period of gestation. An obstetrical ultrasound scan confirmed fetal viability. The outcome was favorable after surgical resection. Pheochromocytoma diagnosed during pregnancy is rare. If undiagnosed, the materno-fetal mortality is high. The diagnosis should be considered if there are evocative clinical signs or resistant gestational hypertension. The specific treatment is surgical resection.

Keywords: Pheochromocytoma, pregnancy, gestational hypertension

1. INTRODUCTION

The pheochromocytoma is a rare tumor, with a prevalence of approximately 1 per 50,000 pregnancies\(^{(1)}\). Diagnosis is difficult: first, the occurrence of hypertension in pregnancy context oriented primarily toward pre-eclampsia; secondly, the symptomatology is atypical in 50% of cases. In the absence of a diagnosis, the risk of fetal death and/or mother is high, especially in the peripartum. In recent years, advances in medical and surgical management have greatly improved prognosis\(^{(2)}\).

We report the case of a pheochromocytoma treated successfully in the first trimester of pregnancy

2. CASE REPORT

Ms. M.B, 25 years of patient G2P1, the first pregnancy: vaginal delivery, a child living 7 years and a half, the second is estimated at 16 weeks’ gestation present. Having as antecedent hypertension for 10 months under Nicardipine 50 mg 3 times a day, admitted to emergency care for a peak hypertensive 230/120 mmHg.

Examination on admission showed a conscious patient, afebrile, pale, headache, blurred vision, sweating, and palpitation. The blood pressure was 230/130 mm Hg, heart rate 90 bpm.

The obstetrical examination revealed that the uterine height corresponds to a pregnancy of 16-17 weeks’ gestation with no bleeding and contracture. Obstetric ultrasound 17 SA objectified scalable mono-fetal pregnancy biparietal of 36 mm, a length of 26 mm femoral and abdominal circumference of 131 mm.

Abdominal ultrasound showed the presence at the right adrenal lodge a very limited lesional tissue process hypoechoic, homogeneous measuring 22 × 15 mm. There was no anomaly contralateral adrenal or abnormality associated in retroperitoneal (fig 1).
Pheochromocytoma was very likely given the clinical context: Triad HTA--sweats Palpitations.
The electrocardiogram showed left ventricular hypertrophy. Doppler ultrasound of the renal arteries was unremarkable same for echocardiography.
Laboratory tests confirmed the diagnosis with elevated plasma and urinary metanephrines levels. He also showed anemia and leukocytosis. Renal function, liver function tests, blood glucose, hemostasis and urinary protein were normal.
The patient was placed on 1 mg Methylodopa 500 cp throughout the 8 hours + Nicardipine 1.5 cc / h with a transfer surgical ICU for surgical excision.
The right adrenalectomy was performed under general anesthesia by laparotomy (right subcostal incision) 17 WA 3J. The intraoperative bleeding was minimal, no hemodynamic instability was observed during the intervention.
Histopathological analysis confirmed the diagnosis of pheochromocytoma adrenal right 2.5 cm long axis, without histological signs of malignancy.
The postoperative course was favorable. The patient was discharged on the postoperative day 7 service. On the obstetrical, there were no uterine contractions, nor abnormal fetal heart rate, the control ultrasound found a eutrophic fetus normal vitality. For hypotension figures were balanced by Methylodopa the 500 mg every 8 hours.

3. DISCUSSION

Pheochromocytomas are tumors derived from cells from the neural crest. In 80 to 85% of cases, they are developed at the expense of the adrenal medulla, the others sitting at the autonomic nervous system, the latter being sometimes called paraganglioma(3). The symptoms are secondary to the production of catecholamines from chromaffin cells that make up the pheochromocytoma. The problem during pregnancy is especially diagnostic, because as a "great pretender," pheochromocytoma mime pre-eclampsia is the most common(4).
The clinical diagnosis of pheochromocytoma is not always easy during pregnancy (Table I).
Clinical signs of pheochromocytoma features in pregnant women

<table>
<thead>
<tr>
<th>Clinical Signs</th>
<th>Characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>Triad of Headache, Profuse sweating, and Palpitations</td>
<td>- Characterize the disease</td>
</tr>
<tr>
<td></td>
<td>- Frequency is 50% of cases</td>
</tr>
<tr>
<td>hypertensive crises</td>
<td>- Often associated with the triade</td>
</tr>
<tr>
<td></td>
<td>- Paroxysmal and labile</td>
</tr>
<tr>
<td></td>
<td>- Spontaneous or triggered by different stress stimuli, palpitations, uterine</td>
</tr>
<tr>
<td></td>
<td>contractions, anesthesia, pushing effort during childbirth</td>
</tr>
<tr>
<td></td>
<td>- Hypertension resistant to medical treatment.</td>
</tr>
<tr>
<td></td>
<td>- Occurring before 20 weeks of amenorrhea (differential diagnosis with</td>
</tr>
<tr>
<td></td>
<td>preeclampsia)</td>
</tr>
<tr>
<td></td>
<td>- Frequent association with diabetes (catecholamines cause insulin resistance)</td>
</tr>
<tr>
<td>Palpitations, nausea, anxiety, visual disturbances, chest pain, or abdominal</td>
<td>- Atypical signs often associated with polyendocrinopathies</td>
</tr>
<tr>
<td>pain</td>
<td></td>
</tr>
<tr>
<td>Hereditary context</td>
<td>- Search for polyendocrinopathy (NEM2)</td>
</tr>
<tr>
<td></td>
<td>- Diagnosis often preconception</td>
</tr>
<tr>
<td>Cardiac decompensation (OAP, cardiopulmonary insufficiency, shock)</td>
<td>- A major complication in the case of late diagnosis</td>
</tr>
<tr>
<td></td>
<td>- Maternal and fetal worse prognosis</td>
</tr>
</tbody>
</table>

In 50% of cases, there is the classic triad headaches, palpitations, sweating and hypertensive crises; they can be triggered by various stimuli such as palpation of the abdomen, the uterine contractions, fetal movements, anesthesia and thrust forces during delivery\(^5\). It should also suggest the diagnosis before a resistant hypertension treatment or appear only Trendelenburg position\(^6\). High blood pressure cannot be part of the picture, and the symptoms may be limited to palpitations, nausea, anxiety, blurred vision, chest or abdominal pain. These atypical presentations are frequent when pheochromocytoma is associated with multiple endocrine disease type II (MEN 2): the pressure is then present in a third of cases against 72% in sporadic forms\(^5\). In pregnant women, the detection of hypertension does not lead to the search for a pheochromocytoma first line. The frequency of argument, the differential diagnosis is preeclampsia. Be aware reject the diagnosis of preeclampsia when blood pressure is not accompanied or proteinuria or edema, knowing that proteinuria does not exclude the diagnosis of pheochromocytoma. Similarly, diagnosis of pheochromocytoma should be considered when hypertension occurs before 20 weeks of gestation, or when it is associated with diabetes, the excess catecholamine-induced insulin resistance\(^7\). Confirmation of the diagnosis of pheochromocytoma is made by the determination of metanephrines and urinary normetanephrine of 24 hours, which is not altered by pregnancy. The collection should be done, if possible when a hypertensive crisis. The iconographic reference examination during pregnancy is MRI. Ultrasound has good sensitivity to locate adrenal tumors but does not explore the ectopic locations, and this review is technically difficult as and as the pregnancy progresses. The MIBG scan is, of course, against-indicated\(^8\).

The fetal prognosis is conditioned by impaired maternal-fetal circulation related to the secretion of \(\alpha\)-adrenergic catecholamines by the tumor while inducing vasoconstriction. The uteroplacental insufficiency may be responsible for premature birth, miscarriage, fetal growth retardation, a retroplacental hematoma or death in utero. On the breast level, the massive release of catecholamines can lead to cardiovascular decompensation and even death to the mother. Tumor compression by the gravid uterus, uterine contractions, active fetal movements, pain, and expulsive efforts of delivery are elements capable of
causing decompensation blood pressure especially in the absence of diagnosis\(^{(7)}\). The treatment is surgical matching adrenalectomy. The intervention must be supervised by medical treatment stabilizing blood pressure to reduce the perioperative risk. The β-blockers are the most used. Calcium channel blockers can be used and have the advantage of not inducing orthostatic hypotension. Diuretics may increase hypovolemia and therefore should be avoided. In pregnant women, the difficulty is to control hypertension by preventing maternal hypotension, deleterious to the fetus. Caesarean section is recommended because its mortality rate is lower than vaginal delivery (19% versus 31%). Gestational age chosen depends on the clinical response to treatment, accessibility of the tumor and the presence or absence of fetal distress. Some recommend a first tumor resection before 24 WA (it is the case for our patient). Beyond 24-28 WA pending fetal maturity is recommended to perform the same operation if possible, fetal extraction by caesarean middle path and the excision of the tumor\(^{(9)}\). General anesthesia is preferred\(^{(10)}\). After treatment, the risk of recurrence is high (17%), even more in malignant forms, extrasurrénaliennes or family. Annual monitoring for life is recommended for these malignant forms and at least ten years for benign forms\(^{(5,7)}\). The good management of pheochromocytoma during pregnancy thus requires a trained multidisciplinary team of obstetrician, visceral surgeon, endocrinologist, radiologist and anesthesiologist.

4. CONCLUSION

The pheochromocytoma is a rare disease revealed by pregnancy. Given its poor prognosis for the mother and the fetus, it is essential to remember that early diagnosis is critical to improving maternal and fetal prognosis; we must think about whenever hypertension in pregnancy is accompanied by clinical signs or in the case of resistance to treatment. The most sensitive diagnostic test and the more specific is the measurement of urinary metanephrines. Adrenal MRI in pregnancy has a place in the tumor localization strategy. The specific treatment is surgical excision always supervised by medical treatment.

REFERENCES