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Case Report

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PHEOCHROMOCYTOMA IN PREGNANCY: A CASE REPORT AND LITERATURE REVIEW

Benmouna Imane*[1], Harmouchi Othmane[1], Pr Lakhdar Amina[1], Pr Zeraidi Najia[1], Pr Baidada Aziz[1], Zebbakh Hajar[2], Pr El Haddad Siham[2], Pr Allali Nazik[2], Pr Chat Latifa[2]

[1]Department of Gynecology and Obstetric and Endoscopy, Maternity Souissi Hospital, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco.

[2]Maternal and Child Imaging Department., Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco.

***Corresponding Author: Dr. Benmouna Imane**

Department of Gynecology and Obstetric and Endoscopy, Maternity Souissi Hospital, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco,

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ABSTRACT

Pheochromocytoma is a rare tumor which causes increased levels of circulating catecholamines. it is associated with very high maternal and fetal mortality. Early diagnosis and proper management can lessen its mortality and morbidity. We report a case of a patient whose first pregnancy was complicated by foetal demise in a context suggestive of preeclampsia. She was diagnosed with pheochromocytoma as she was beginning a second pregnancy. The treatment consisted of preoperative medical preparation followed by right adrenalectomy. Our case demonstrated that delay in diagnosis could result in catastrophic results.

KEYWORDS: Pregnancy, Pheochromocytoma.

INTRODUCTION

Pheochromocytoma is a rare tumor and a rare cause of secondary hypertension; The prevalence is in the order of 1 per 50,000 pregnancies.[1] In the absence of a diagnosis, the risks of fetal and/or maternal mortality are high, especially in the peripartum period. Adverse maternal and fetal effects associated with pheochromocytoma are thought to be secondary to the effects of catecholamines on maternal blood pressure and uteroplacental blood flow.

Patient and Observation

A 30-year-old woman at 22 weeks of gestation presented to our hospital with high blood pressure 170/100 mm Hg, palpitations, and recurring headaches.During her prior pregnancy, she obtained no prenatal care. She had presented with severe preeclampsia at term, and was delivered by emergent cesarean section for fetal distress. She had no additional history of hypertension and was never on antihypertensive medications. She denied alcohol, smoking, or drug use. Physical examination revealed a blood pressure of 1170/100 mm Hg and a heart rate of 138 bpm, Laboratory tests at baseline showed negative protein, Her liver enzyme, creatinine, and platelet counts were all in normal range.ultrasound examinations revealed intrauterine fetal demise. A treatment with alpha-methyldopa at a dose of 750 mg/day was initiated. After induction the patient gave birth to a male stillborn of 350 g. Over the next 24 h, the patient's blood pressure abruptly increased to 240/120mm, and heart rate increased to 160 bpm, also she presented a pain in her right flank pain. So we did ultrasound imaging, Ultrasound imaging revealed no problems with her kidneys, but we found a solid mass with fine cystic component above her kidney, highly suspicious for an adrenal-originating tumor (Figure 1). The abdominal computerized tomography (CT) showed a well-defined 98 × 82 ×77 mm heterogeneous mass in the right adrenal gland which contained some necrosis areas without calcifications (Figure 2).

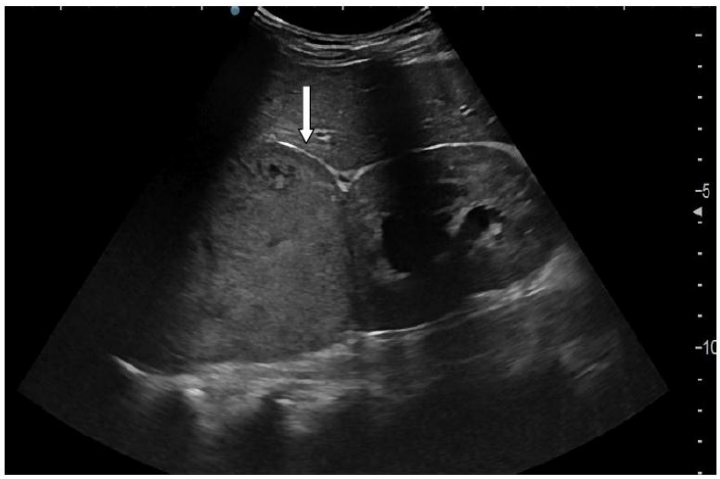

Figure 1: Abdominal ultrasound demonstrating right adrenal pheochromocytoma; arrow indicate large heterogeneous mass.

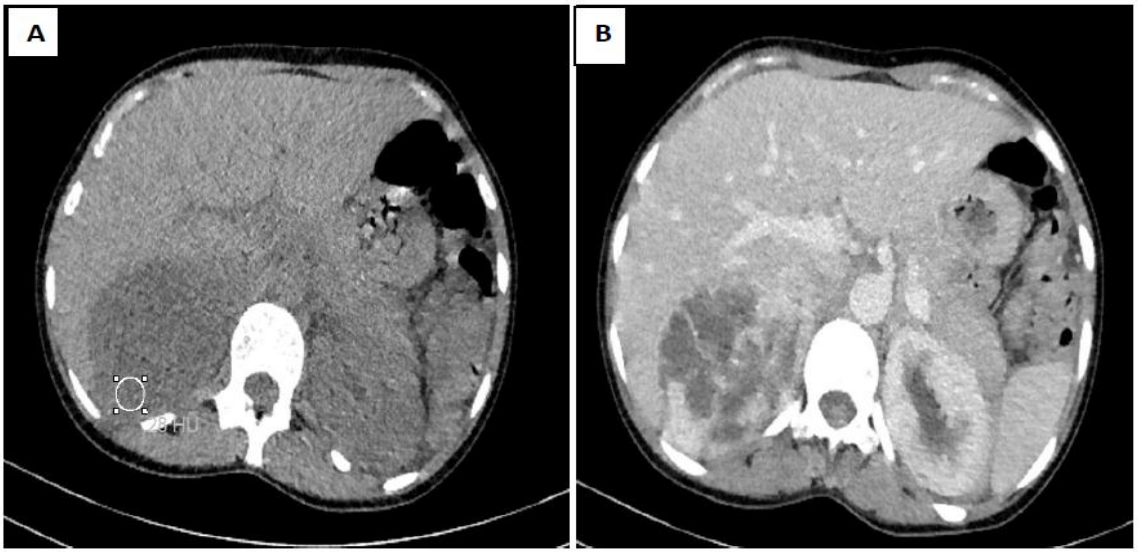

Figure 2: Axial section of an abdominal CT scan showing a right adrenal mass, oval, well circumscribed, heterogeneous with areas of necrosis without calcifications, coming into contact with the liver anteriorly and the diaphragmatic pillar posteriorly (A), enhanced early and heterogeneously after injection (B).

The urine collection revealed a markedly elevated excretion of normetanephrine (1809 mg; reference range, 110 à 320 mg) and métanéphrines à (88 mg ;reference range, 40 à 150 mg).

Considering the clinical picture and radiological and laboratory test results, pheochromocytoma (PCC) of the left adrenal gland was suspected. he patient was transferred to our endocrinology ward and treatment with alpha- and beta-blockade with phenoxybenzamine and propranolol was started, methyldopa was discontinued.BP was reduced and remained between the range of 100/60 mm Hg to 140/80 mm Hg, with a HR of 90 to 110 bpm. Following a multidisciplinary team assessment a laparotomy was performed. A right adrenal phaeochromocytoma was removed, Microscopic examination confirmed the diagnosis of pheochromocytoma. She was taken to our intensive care unit after the surgery. Her hemodynamic condition was monitored for 24 hours. She was discharged 6 days after surgery. Postoperative catecholamine levels were norepinephrine 664 pg/ml, epinephrine 22 pg/ml, and dopamine 49 pg/ml. The patient has felt well and has had normal blood pressure after discharge.

DISCUSSION

Pheochromocytoma is a chromaffin cell tumor producing excess catecholamines and located in 90% of cases in the adrenal medulla. It is a rare disease that can be revealed by pregnancy. The rarity of this association and its similarity to gestational hypertension explain the frequency of unrecognized diagnoses during pregnancy. Several symptoms have been associated with pheochromocytoma, but none of them are specific. Certain syndromic groupings, the paroxysmal nature of the symptoms, and their association with hypertension are suggestive. The triad "headache/palpitations/sweating" is found in almost 90%

of pheochromocytomas.[2,3] The diagnosis must also be made in the presence of treatment-resistant hypertension, even in the absence of vasomotor disorders. Other clinical signs may be found: tremors, dizziness, diaphoresis, anxiety, nausea, vomiting and/or visual changes.These paroxysms typically last less than 15 min. An abnormality in carbohydrate metabolism may occur because of increased catecholamines secretion.[1] Some factors can cause clinically overt pheochromocytoma such as increase in intraabdominal pressure, fetal movement, uterine contraction, delivery (vaginal or surgical), and even general anesthesia. Once suspected, the diagnosis is generally easy as pregnancy, either normal or complicated by preeclampsia, does not alter the reference values of catecholamines or their metabolites in maternal plasma or urine.[4] The metabolites most commonly measured include vanilmadelic acid (VMA) and metanephrine. The sensitivity of urinary VMA for diagnosing a pheochromocytoma is 98%. Urinary metanephrines have at least a 90% sensitivity with a positive predictive value of 83%.[5]

After biochemical test, The iconographic examination of reference during pregnancy is MRI. Ultrasound has a good sensitivity to localize adrenal tumors, However, this test has some limitations, such as the inability to detect ectopic localizations, and this examination becomes technically difficult as the pregnancy progresses. Meta-iodobenzyl-guanidine scintigraphy is of course contraindicated.[6]

The main goal of pheochromocytoma management is preventing hypertension crisis. Thus, medical treatment with alpha blocker should be initiated at the time of diagnosis and continued for at least 10 to 14 days before surgery. Phenoxybenzamine is the drug of choice (pregnancy class c), which allow the control of hypertension, with proven benefit in reducing mortality.[7,8] Beta-blockade may be used to control the tachycardia and arrhythmia, but should never be attempted before alpha blockade is established because it may lead to unopposed alpha stimulation, causing severe vasoconstriction and even death. The definitive treatment of pheochromocytoma is surgical removal of the tumor.[9] The timing of tumor resection remains controversial. If the diagnosis is made before 23 weeks of gestation, the tumor should be removed once adrenergic blockade has been achieved.[10,11] The prognosis seems to be better with the laparoscopic approach. The first laparoscopic adrenalectomy performed outside pregnancy was described by Gagner et al.[12] After 24 weeks gestation uterine size makes abdominal exploration and access to the tumor difficult unless the woman is first delivered. Therefore, alpha-blockade is continued until the fetus is mature and then an elective Cesarean section should be performed, followed immediately by exploration for the tumor.[13] However, when the maternal prognosis is at risk, early intervention is necessary whatever the term. In very severe cases, a delay in surgery to surgery leads to higher risks of intratumoral hemorrhage[2] and fetal death secondary to blood pressure instability.[14] Monitoring after surgery is the same as for any pheochromocytoma. The proper management of pheochromocytoma during pregnancy therefore requires a trained multidisciplinary team, composed of the obstetrician, the endocrinologist, radiologist and anaesthetist.

CONCLUSION

pheochromocytoma is a rare tumor, especially during pregnancy. It can be misdiagnosed as pregnancy-induced hypertension. Untreated PCC significantly increases maternal and fetal mortality. our case demonstrated that delay in diagnosis could result in catastrophic results.

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