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(CASE REPORT)

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Pheochromocytomas and paragangliomas during pregnancy: A case report and review of the literature

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Abstract

Pheochromocytoma is a rare, potentially lethal tumor that can be revealed by pregnancy, often mimicking a clinical picture of pre-eclampsia. We report the case of a pheochromocytoma that occurred in the 1st trimester, presenting with episodes of arterial hypertension (AHT). Treatment consisted of preoperative medical preparation followed by surgery. The maternal and fetal outcome was favorable. The occurrence of pheochromocytoma during pregnancy poses a problem in both diagnosis and blood pressure control. The therapeutic strategy depends on the gestational age, the maternal-fetal outcome (retention), and the response to medical treatment.

Keywords: Pheochromocytoma; Pregnancy; Anesthesia; Prognosis

1. Introduction

Pheochromocytomas (PCCs) and paragangliomas (PGLs) are rare neuroendocrine tumors that produce excess catecholamines [1]. They are classified into two types: pheochromocytomas with an adrenal localization (77%) and paragangliomas, which are extra-adrenal (23%). PGLs are located along the neurovascular axes, predominantly in the abdomen (para-aortic), and rarely in the chest, pelvis, or neck [2, 3]. All pheochromocytomas and some paragangliomas can secrete norepinephrine, epinephrine, and dopamine. This compromises organ function through alpha or beta-adrenergic action on the sympathetic nervous system receptors. The cardiovascular consequences are sometimes dramatic: AHT, rhythm disorders, acute pulmonary edema (APE), cardiomyopathy, or even heart failure and maternal death. Hereditary tumor forms are described in 20 to 30% of cases [3, 4, 5].

Maternal and fetal mortality is high in the absence of treatment and monitoring when the diagnosis is missed. The effects on the mother depend on the lability of blood pressure, the duration and severity of hypertensive peaks, as well as any pre-existing vascular conditions. Fetal consequences result from the vasoconstrictor effect of catecholamines on the uteroplacental circulation, sometimes leading to fetal hypoxia, placental abruption, growth restriction, or even *in utero* fetal death [4, 6].

2. Patient and Observation

A 26-year-old patient, followed for Type 1 diabetes for 5 years on insulin, with a hypertensive father who was also being followed for pulmonary neoplasia, was admitted as a primigravida for the management of arterial hypertension (AHT) at 14 weeks of gestation (14SA). Biological and radiological investigations returned results in favor of a bilateral pheochromocytoma with inter-aorto-caval lymph node involvement (Table 1).

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In the syndromic context, the patient underwent a MEN (Multiple Endocrine Neoplasia) workup (Table 1), which was unremarkable (phospho-calcic balance, PTH, calcitonin, and a normal cervical ultrasound). A search for VON HIPPEL-LINDAU disease (VHL) (ophthalmic examination was performed, which was unremarkable, with no renal or pancreatic cysts on abdominal ultrasound) was also conducted.

Table 1 Biological and Radiological Workup

Workup	**Results**
MEN Workup(Multiple Endocrine Neoplasia)	
Calcemia	87 mg/L
Albuminemia	43 g/L
Phosphorus	39 mg/L
Vitamin D	6.7 ng/L (Deficiency)
PTH	86.9 pg/L
Calcitonin	5.72
Urinary Metanephrines Measurement	** **
Metanephrine	Below 20 nmol/24h
Normetanephrine	49 (20 times the normal range)
Thyroid Workup	Normal
Abdominal Ultrasound	Bilateral adrenocortical tumor associated with thoraco-aortic lymphadenopathy
Abdominal MRI	MRI appearance suggesting bilateral pheochromocytoma associated with a pre-aortic ganglionic mass
Cervical Ultrasound	Two left lobar thyroid nodules classified EU-TIRADS III
CTAP (Computed Tomography of the Abdomen and Pelvis)	- Two bilateral adrenal masses primarily suggesting pheochromocytoma. - Parietal digestive thickening of the 3rd duodenal portion associated with a locally advanced tumor mass: malignant GIST? Pancreatic body tumor? - Suspect inter-aorto-caval lymphadenopathy

For her AHT, the patient was started on dual therapy based on methyldopa (500mg/8h) and a calcium channel blocker (50mg/12h), and the evolution was marked by the improvement of blood pressure readings.

Following patient preparation, including embolization of the three tumor lesions (GIST: Gastrointestinal Stromal Tumor, and the two adrenal masses), she underwent a laparotomy at 18SA: exploration revealed a locally advanced pancreatic head tumor + two adrenal tumors. The procedure was a caudal duodenopancreatectomy + bilateral adrenalectomy. The immediate postoperative course was uneventful.

Anatomopathological analysis of the surgical specimens showed:

- Pheochromocytoma with a PASS score of 4 for the two adrenal tumors.
- Well-differentiated neuroendocrine tumor of grade 2 for the duodenopancreatic specimen.
- Pre-aortic lymphadenopathy: histological appearance of a paraganglioma.

The pregnancy continued to progress until 30 days post-operation, when the ultrasound control showed negative cardiac activity (pregnancy stopped at 22SA). The patient underwent induction with misoprostol, which was effective after three doses.

3. Discussion

The incidence of pheochromocytoma in pregnant women is approximately 1/54000 [3, 7]. Paraganglioma is even rarer.

These tumors are responsible for paroxysmal symptoms: the most classic is AHT (87% of pheochromocytomas and 86% of functional secreting paragangliomas), sometimes associated with headaches, sweating, and palpitations (Menard's Triad) [8, 9]. Hypertension from a pheochromocytoma or paraganglioma is either permanent or paroxysmal and can start at any stage of the pregnancy. It is essential to screen for endocrine hypertension in the presence of severe, resistant AHT or with suggestive clinical, biological, or imaging findings. The diagnostic approach must be rigorous to rule out differential diagnoses through a renal, hormonal, and radiological workup [10].

In some cases, the condition is revealed by sinus tachycardia or rhythm disorders, sometimes by acute adrenergic cardiomyopathy or even a state of shock due to acute heart failure.

Biological diagnosis in pregnant women relies on measuring plasma and 24-hour urinary metanephrines, which are derivatives of catecholamines [11, 12]. For topographic diagnosis, ultrasound and MRI are preferred during pregnancy due to their safety [13]. MRI classically shows a heterogeneous image in T2 hypersignal with intense enhancement after gadolinium injection [14]. Isotopic examinations are contraindicated during pregnancy due to their fetal toxicity.

Maternal mortality is as high as 9% with no difference between pheochromocytoma and paraganglioma, versus 14% for the fetus [3]. These risks are increased if the tumor is discovered postpartum. The prognosis essentially depends on the time of diagnosis.

The optimal therapeutic strategy must be discussed in a multidisciplinary consultation meeting (MCM) involving obstetricians, anesthetists, intensivists, cardiologists, endocrinologists, and endocrine surgeons. Medical treatment is initiated upon diagnosis through alpha-adrenergic receptor blockade. This treatment is started at least 10 to 14 days before delivery to stabilize blood pressure and reduce perioperative complications [15]. Calcium channel blockers limit hypertensive peaks through their vasodilatory action.

According to older literature, cesarean section was the preferred route of delivery due to an excess mortality rate during vaginal delivery (33% versus 19% via cesarean section) [4, 19-22]. This increased risk may be linked to the stress and pain of labor stimulating the sympathetic system, which can lead to a detrimental catecholaminergic surge. However, there are recent reports of vaginal deliveries under epidural analgesia with good outcomes in patients whose diagnosis was unknown during delivery [15, 23]. Recent analyses suggest that vaginal delivery is not systematically associated with an unfavorable outcome, although cesarean section appears to be twice as frequent when the diagnosis is made before or during pregnancy [16]. Thus, vaginal delivery could be proposed to patients based on the context for certain selected endocrine tumors.

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Tumor excision during cesarean section will only be performed in cases where the maternal or fetal prognosis is compromised, as was the case with our patient. Before 24 weeks of gestation (SA), tumor removal via laparoscopy is discussed in the literature, with limited insufflation pressures, despite the risk of spontaneous abortion [21, 22, 24, 25]. After 24 SA, resection will be scheduled remotely from the delivery.

A recent analysis demonstrates that surgery during pregnancy is not associated with better outcomes [16].

4. Conclusion

The presence of paroxysmal AHT without proteinuria in pregnant women should raise the possibility of a pheochromocytoma or paraganglioma. The prognosis is even worse if the diagnosis is missed. Pregnancy presents a real challenge in the management of these tumors by limiting topographic diagnosis and medical/surgical treatments. Multidisciplinary management is based on antihypertensive treatment (calcium channel blockers, alpha-blockers) and anticipation of management, particularly for delivery. Cesarean section is often favored, but the route of delivery can be discussed. The development of a follow-up protocol allows for appropriate monitoring and management of these patients during pregnancy.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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