038 Pregnancy unmasking a pelvic sympathetic paraganglioma: a case report

Vanwonderghem Yuran, Vandendriessche Barbara, Randon Caren, Roets Ellen, **Creytens David**, T'Sjoen Guy & Lapauw Bruno Department of Endocrinology, Ghent University Hospital, Belgium.

Introduction: Paraganglioma (adrenal or extra-adrenal) in pregnancy is a rare condition with a high maternal and foetal mortality rate. A multidisciplinary approach is important as it will influence the maternal and foetal outcome. This is illustrated in the following case report. Case report and results
A 32-year-old woman was referred because of persistent hypertension at 33 weeks of pregnancy. There were no spontaneous complaints of headache, palpitations or sweating. She did mention presyncope and tachycardia when making bowel movements. Biochemical evaluation was performed on a 24-hour urine collection, which showed elevated levels of adrenaline (204.5 mg/24h OK20 mg/d, noradrenaline (2736 mg/24h 10K100 mg and normetanephrines (8694 mg/ 24h, 100K610 mg). Whole body MRI showed a pararectal mass (6 cm), suggestive of an extra-adrenal paraganglioma, explaining the specific occurrence of complaints. At diagnosis the patient was 33 weeks into her second pregnancy. The obstetric history showed one uncomplicated vaginal delivery. On clinical examination there were no syndromic features. No significant family history was noted. Since the patient had already reached her third trimester of pregnancy, resection of the paraganglioma was postponed until after delivery. She was pretreated with alpha-adrenergic receptor blockers, beta-adrenergic receptor blockers and calcium agonists, and responded well to medical treatment. An elective Caesarean section was performed at 34 weeks. The delivery was uneventful with stable hemodynamics. Both mother and child were admitted to the intensive care unit, but no hemodynamic complications occurred. One week before the second surgery she was put on a salt-rich diet and intravenous fluids to restore the intravascular volume. Because of the size of the tumour and its location close to the right internal iliac artery, surgery consisted of an open resection. Histologically, the tumour was confirmed to be a paraganglioma. Genetic analysis showed a complete loss of SDBH expression. Follow up was arranged four weeks after discharge from hospital, at which time biochemical re-evaluation by measuring plasma and urinary metanephrines will be performed.

Discussion: Paraganglioma during pregnancy is a rare condition. Establishing a correct diagnosis is important as it will influence both maternal and foetal outcome. High concentrations of catecholamines can result in a compromised uteroplacental circulation, leading to intrauterine growth restriction or intrauterine hypoxia (1). Paraganglioma during pregnancy can be harder to diagnose as its most common presenting feature is hypertension. In this situation it can be difficult to differentiate between a paraganglioma and pregnancy related hypertension. The latter usually develops after 20 weeks of pregnancy. Hypertension due to paraganglioma can present at any time during pregnancy, though it is more paroxysmal in nature and hypotensive episodes can also occur. Diagnosis of a paraganglioma is similar in pregnant and non-pregnant patients and includes measuring plasma and urinary fractionated metanephrines (1). There are no specific recommendations for the treatment of paraganglioma during pregnancy and each case should be evaluated by a multidisciplinary team at centres with appropriate expertise (2).

Conclusion: Currently, optimal treatment of paraganglioma during pregnancy remains debatable. This case illustrates the importance of a timely diagnosis as well as the need for a multidisciplinary team that will make decisions concerning the timing and method of delivery and the treatment for the underlying paraganglioma.

References 1. Van der Weerd, K., van Noord, C., Loeve, M., Knapen, M.F.C.M., Visser, W., de Herder, W.W., Franssen, G., 2017. ENDOCRINOLOGY IN PREGNANCY: Pheochromocytoma in pregnancy: case series and review of literature. *European Journal of Endocrinology* 177, R49–R58. doi:10.1530/EJE-16-0920. 2. Lenders, J.W.M., Duh, Q.-Y., Eisenhofer, G., Gimenez-Roqueplo, A.-P., Grebe, S.K.G., Murad, M.H., 2014. Pheochromocytoma and Paraganglioma: An Endocrine Society Clinical Practice Guideline. *The Journal of Clinical Endocrinology & Metabolism* 99, 1915–1942. doi:10.1210/jc.2014-1498.

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