Pheochromocytoma diagnosed during pregnancy: lessons learned from a series of ten patients

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Résumé en anglais	BACKGROUND: Pheochromocytoma (PHEO) in pregnancy is a life-threatening condition. Its management is challenging with regards to the timing and type of surgery. METHODS: A retrospective review of the management of ten patients diagnosed with pheochromocytoma during pregnancy was performed. Data were collected on the initial diagnostic workup, symptoms, treatment, and follow-up. RESULTS: PHEO was diagnosed in ten patients who were between the 10th and the 29th weeks of pregnancy. Six patients had none to mild symptoms, while four had complications of paroxysmal hypertension. Imaging investigations consisted of MRI, CT scan and ultrasounds. All had urinary metanephrines, measured as part of their workup. Three patients had MEN 2A, one VHL syndrome, one suspected SDH mutation. All patients were treated either with α/β blockers or calcium channel blockers to stabilize their clinical conditions. Seven patients underwent a laparoscopic adrenalectomy before delivery. Three out of these seven patients had a bilateral PHEO and underwent a unilateral adrenalectomy of the larger tumor during pregnancy, followed by a planned cesarean section and a subsequent contralateral adrenalectomy within a few months after delivery. Three patients had emergency surgery for maternal or fetal complications, with C-section followed by concomitant or delayed adrenalectomy. All newborns from the group of planned surgery were healthy, while two out three newborns within the emergency surgery group died shortly after delivery secondary to cardiac and pulmonary complications. CONCLUSIONS: PHEO in pregnancy is a rare condition. Maternal and fetal prognosis improved over the last decades, but still lethal consequences may be present if misdiagnosed or mistreated. A thorough multidisciplinary team approach should be tailored on an individual basis to better manage the pathology. Unilateral adrenalectomy in a pregnant patient with bilateral PHEO may be an option to avoid the risk of adrenal insufficiency after bilateral adrenalectomy.
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