

level limitations. Addressing these barriers is essential to optimize subtype-directed management and improve long-term cardiovascular outcomes in patients with PA. Despite this, clinical outcomes were favorable. Strengthening adherence to diagnostic pathways, particularly subtype confirmation, may further improve patient outcomes.

EP_A004

SINGLE-STAGE ADRENALECTOMY AND HYSTERECTOMY FOR PHEOCHROMOCYTOMA WITH GIANT UTERINE FIBROID: A MULTIDISCIPLINARY PERIOPERATIVE CHALLENGE

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INTRODUCTION

Pheochromocytoma is a catecholamine-secreting adrenal tumor associated with major perioperative hemodynamic instability. When concurrent major pelvic pathology requires surgery, operative planning becomes particularly challenging. We describe the successful single-stage management of pheochromocytoma and a giant uterine fibroid, highlighting the importance of multidisciplinary coordination and perioperative optimization.

CASE

A 49-year-old female with symptomatic uterine fibroid was found to have proliferative endometrium on a pipelle biopsy. Computed tomography (CT) abdomen incidentally detected a right adrenal mass alongside a large posterior uterine fibroid (11.9 × 17.4 × 14.6 cm). CT adrenal protocol demonstrated a heterogeneously enhancing right adrenal mass (6.9 × 6.9 × 9.6 cm) with high unenhanced attenuation. Biochemical evaluation revealed markedly elevated 24-hour urinary metanephrine (4.7× upper limit) and normetanephrine (2.4× upper limit).

Following multidisciplinary discussions, a single-stage surgical approach was planned after careful assessment of feasibility and perioperative risk in view of the uncertain

malignant potential of the pelvic mass and to minimize repeated exposure to anesthesia. Preoperative optimization included transitioning from terazosin to phenoxybenzamine, with subsequent addition of bisoprolol for hemodynamic control. The operative strategy prioritized pheochromocytoma resection first, given its potential for significant hemodynamic instability. Progression to hysterectomy was contingent upon achieving adequate intraoperative hemodynamic stability following adrenalectomy, with continuous reassessment by the anesthetic and surgical teams.

Right adrenalectomy was performed first, followed by total abdominal hysterectomy with bilateral salpingo-oophorectomy. Significant hemodynamic lability occurred during tumor manipulation, with hypertensive surges managed using sodium nitroprusside and remifentanyl infusions. Following adrenal vein ligation and tumor removal, hypotension was managed with noradrenaline and additional adrenaline support as required. Total operative time was approximately 6 hours. Postoperatively, transient noradrenaline support was required but was rapidly weaned as hemodynamic stability was achieved.

CONCLUSION

Single-stage adrenalectomy and major pelvic surgery can be safely performed in selected patients with pheochromocytoma when guided by meticulous preoperative optimization, clear intraoperative sequencing, and close multidisciplinary coordination.

EP_A005

MIMICKING PHEOCHROMOCYTOMA: HYPERTENSIVE CRISIS FROM ADRENAL HEMATOMA IN JAK2-POSITIVE POLYCYTHEMIA RUBRA VERA

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INTRODUCTION

Hemorrhagic suprarenal masses presenting with hypertensive emergency pose a significant diagnostic challenge, particularly when biochemical and radiological findings are inconclusive. The clinical presentation may mimic catecholamine-secreting tumors, necessitating consideration of a broad differential diagnosis, including pheochromocytoma, adrenocortical carcinoma, retroperitoneal hemorrhage, and hematological-related extramedullary lesions. Accurate diagnosis is essential, as management strategies differ significantly.

CASE

We report a 48-year-old male smoker with no known prior medical illness who presented with sudden left-sided chest pain radiating to the epigastrium, associated with vomiting. On arrival, he was markedly hypertensive (207/131 mmHg). He reported a 1-year history of paroxysmal palpitations, headaches, migraines, and intermittent diaphoresis. Computed tomography angiography excluded aortic dissection but demonstrated a left retroperitoneal hemorrhage with non-visualization of the adrenal gland, suggestive of adrenal or tumor-related hemorrhage.

He was initially managed empirically as a pheochromocytoma while undergoing biochemical evaluation; however, urinary metanephrines were only mildly elevated. Repeat imaging demonstrated interval enlargement of a non-enhancing suprarenal mass, raising concern for tumor-related hemorrhage. Subsequent ultrasonography, however, favored a liquefied hematoma, and percutaneous drainage yielded 750 mL of sanguineous fluid, resulting in marked clinical improvement.

Notably, an elevated hematocrit prompted further evaluation for erythrocytosis. Subsequent testing confirmed JAK2 mutation-positive polycythemia rubra vera, providing a unifying explanation for both the erythrocytosis and spontaneous adrenal hemorrhage. The patient was commenced on hydroxyurea and referred for hematology follow-up.

CONCLUSION

Adrenal hemorrhage may closely mimic pheochromocytoma in hypertensive emergencies. A systematic, multidisciplinary approach integrating clinical, biochemical, and imaging findings is essential to avoid misdiagnosis and guide appropriate management, particularly in patients with underlying hematological disorders.

EP_A006

TWO HEARTBEATS, ONE TUMOR: NON-FUNCTIONING ADRENOCORTICAL TUMOR IN PREGNANCY

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INTRODUCTION

Adrenocortical carcinoma (ACC) is a rare and aggressive malignancy, with an incidence of 1–2 cases per million annually. Its occurrence during pregnancy is exceptionally

uncommon, presenting significant diagnostic and management challenges due to overlapping physiological changes and concerns for both maternal and fetal outcomes. While most cases are hormonally functional, non-functioning ACC during pregnancy is particularly rare and may result in delayed diagnosis.

CASE

A 35-year-old Malay female was referred following ultrasonography for persistent back and left flank pain, which revealed a left adrenal incidentaloma and a concurrent 10-week intrauterine pregnancy. There were no clinical signs of hormone excess. Physical examination revealed a normotensive patient with a large, palpable left abdominal mass, without Cushingoid or virilizing features.

Magnetic resonance imaging (MRI) demonstrated a 12.1 × 10.3 × 12.8 cm heterogeneous left adrenal mass with cystic and necrotic components, displacing adjacent structures. Hormonal evaluation was within normal limits: 24-hour urinary cortisol 320.2 nmol/24 hours (reference range [RR] 11.8–350), midnight salivary cortisol <3 and 4.2 nmol/L (RR <11.3), plasma metanephrine <0.2 nmol/L (RR <0.5), normetanephrine 0.6 nmol/L (RR <0.9), and 17-hydroxyprogesterone 223 ng/dL (RR <285), consistent with a non-functioning tumor.

Following multidisciplinary consultation, the patient underwent open left adrenalectomy at 16 weeks' gestation. Histopathological analysis confirmed ACC (Weiss score 8/9) without extra-adrenal extension. Surveillance MRI at 32 weeks demonstrated no recurrence or residual mass. At 40 weeks' gestation, she delivered a healthy infant, and both mother and child remain well on follow-up.

CONCLUSION

Non-functioning ACC during pregnancy is rare and poses significant diagnostic challenges. Early imaging, comprehensive hormonal assessment, and timely surgical intervention during the second trimester are essential. Multidisciplinary management is crucial to optimizing maternal and fetal outcomes.