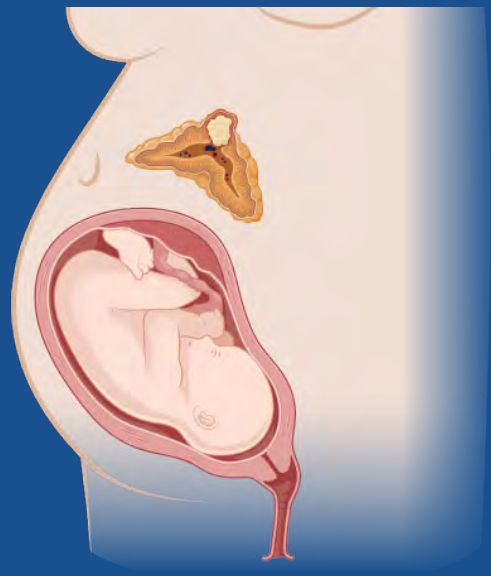
BỆNH VIỆN ĐẠI HỌC Y DƯỢC TPHCM®
UNIVERSITY MEDICAL CENTER HCMC
*Thấu hiểu nỗi đau - Niềm tin của bạn*

ASEAN NETWORK of ADRENAL HYPERTENSION

# Paraganglioma in Pregnancy




Moderator: **Thang V Tran**
Deputy Head, Department of Endocrinology, University Medical Center Ho Chi Minh City, Vietnam.
Secretary, Asean Network of Adrenal Hypertension.

Presenter: **Hieu T N Tran**
Department of Endocrinology, University Medical Center Ho Chi Minh City, Vietnam.
Resident Physician, Department of Endocrinology, Changi General Hospital, Singapore.
PhD in CTS year 2, Duke-NUS Medical School, Singapore.

# Abbreviations

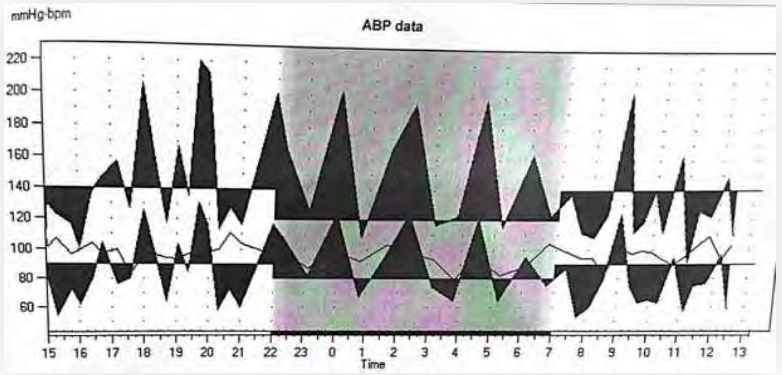
- PPGL: Pheochromocytoma and Paraganglioma
- HT: Hypertension
- IUGR: intrauterine growth restriction
- Hx: History
- Dx: Diagnosis
- Rx: treatment
- BP: Blood pressure, HR: heart rate

# **32-year-old woman**, **G1P0**, generally well before pregnancy

- Referred from Cardiology for evaluation of a suspicious **right adrenal tumor** identified on abdominal ultrasound during the assessment for young-onset hypertension diagnosed at 7 weeks of pregnancy.
- 3-month prior: reported **intermittent episodes of non-exertional dyspnea** lasting ~ 5-15 mins. No BP check. → Dx: Post-covid-19 syndrome without treatment. 1–month later: conceived a baby.
- Prenatal check-up at 7 weeks of pregnancy noted **BP 160/100 mmHg** → Dx: Hypertension.
  → Rx: metoprolol 50mg od. with no improvement.
- At 20 weeks of pregnancy: reported headache, nausea, dizziness, and several episodes of fainting lasting 2-3 mins.
  → Follow-up: SBP 110-120 mmHg (on metoprolol).
  → Rx: stopped metoprolol.
  → Patient self-referred to the outpatient cardiology at our hospital seeking for a second opinion.

24-hour Holter: **BP fluctuations**

- highest **220/108 mmHg**, mostly at night;
- lowest **131/53 mmHg**, mostly in the morning;
- HR ~ **79-111 bpm**.



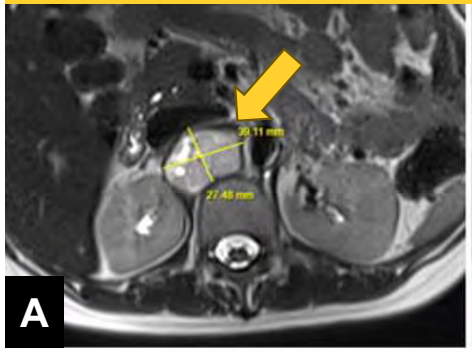
# **32-year-old woman**, **G1P0**, young HT, at second-third trimester

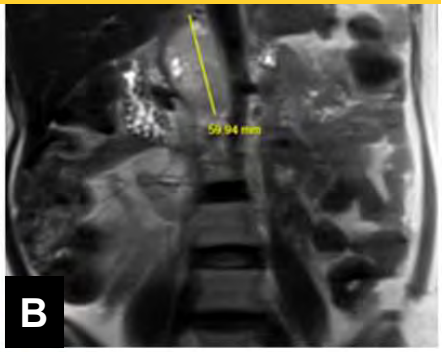
- At cardiology outpatient clinic: BP 219/130 mmHg, HR 99 bpm → Rx: added nifedipine for BP control. Also initiated work-up for young hypertension.
  - Fasting BSL 5.61 mmol/L.
  - Sodium 138 mmol/L (normal 136-146).
  - Potassium 4.2 mmol/L (normal 3.4-5.1).
  - Creatinine 61.3 umol/L (normal 58-96), eGFR 98.
  - FBC, LFTs, TFTs: no abnormality detected.
  - No proteinuria.
  - Plasma metanephrine 104 pg/mL (normal <90).
  - **Plasma normetanephrine 4279 ↑ pg/mL** (normal <196).
  - Plasma aldosterone > 100 ↑ ng/dL.
  - Plasma direct renin concentration 69.9 ↑ uIU/mL.
  - Random serum cortisol 21.25 ug/mL (normal 6.02-18.4).
  - Renal Artery Doppler Ultrasound: No renal artery stenosis.
  - Abdominal Ultrasound: suspicious right adrenal tumor.
- Family Hx: younger brother: Papillary thyroid carcinoma; mother: uterine fibrosis; grandmother (mother side): sudden death at age 60.
- Patient was referred to ENDO for further evaluation and management.
  → admitted to the hospital for BP control and MRI abdomen.

# **32-year-old woman**, **G1P0**, young HT, at third trimester. At ENDO ward.

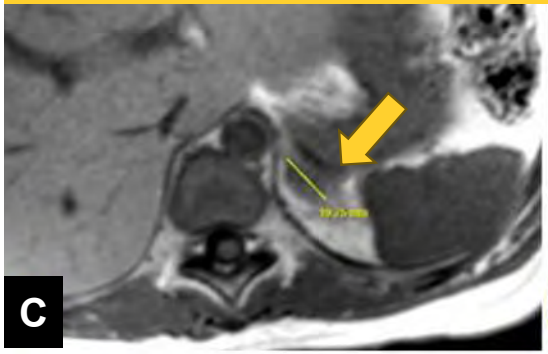
- On examination: HR 91 bpm, BP 100/60 - 213/130 mmHg. No postural hypotension. No Cushingoid features. No proximal myopathy. No pedal edema. Other systematic examinations: unremarkable.
- Fetal ultrasound: sign of intrauterine growth restriction (at 29 weeks).
- Impression: PPGL, diagnosed at second/third trimester. Uncontrolled BP. IUGR.
  → Rx: initiated doxazosin 1mg bd.
- **MRI abdomen**: 2 detected lesions. Both are heterogeneously high signal intensity on T2W.

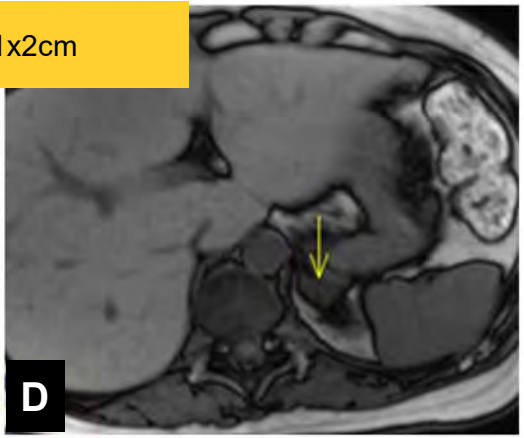
A heterogenous retroperineal mass, located at ~L1-L2, 4x3x6 cm, displaying the IVC and right renal artery anteriorly.




Left adrenal mass, measuring ~1x2cm



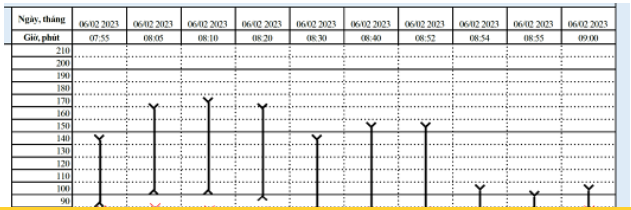
## **32-year-old woman**, **G1P0**, PPGL at third trimester

- Alpha-blocker: Doxazosin 2 mg stat → optimized the dosage based on BP.
- Beta-blocker: Added propanolol.
- Other antihypertensive meds: Continued Nifedipine 30 mg od.
- Encouraged salty diet and fluid intake.

- Adrenal MDM (endocrinologist, cardiologist, gynecologist, urologist, radiologist, interventional radiologist):
  ➔ Aimed for elective C-section at 36-38 week of pregnancy, followed by removal of PPGL at 6-8 week postpartum.

# Uneventful elective C-section at 36 weeks of pregnancy

- Baby boy: 1950 grams, Apgar score 7/8 (no respiratory distress).
- Mother (patient): better BP control post-delivery. Discharged home 3 days post C-section

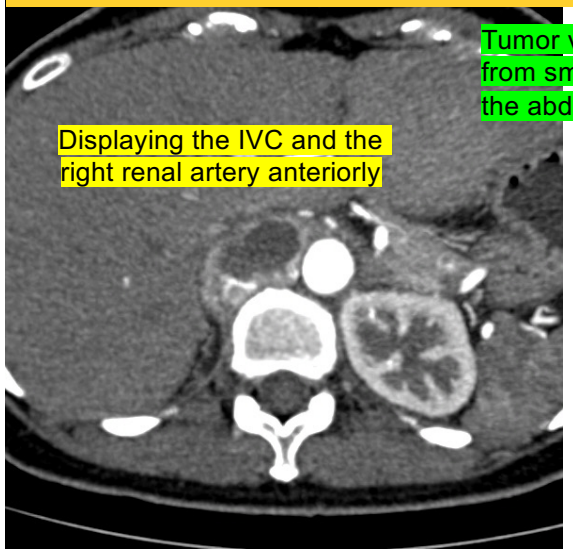


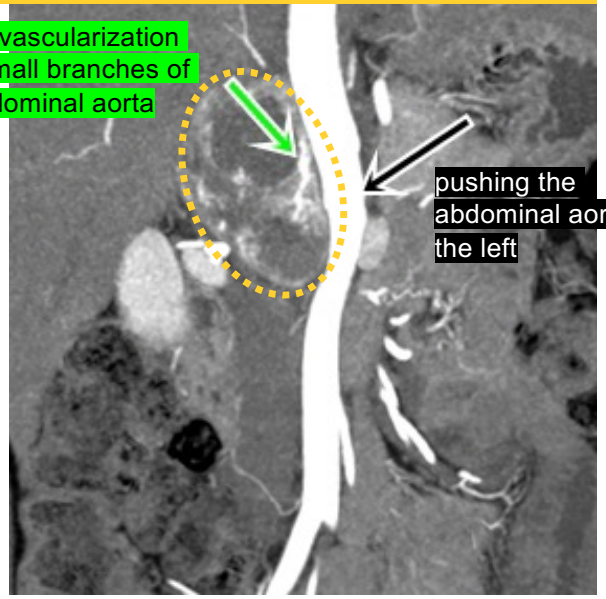

**Follow-up 1-month later:**

- BP: 90-130/70-50 mmHg. No episodes of faiting or elevated BP (on doxazosin 2 mg tds, propanolol 20 mg tds, nifedipine 30 mg on).

- **CT chest-abdo-pelvis scan** (scheduled at 8 weeks post delivery): **confirmed the retroperitoneal mass**. Previous MRI finding of left adrenal tumor was an **illusion**.

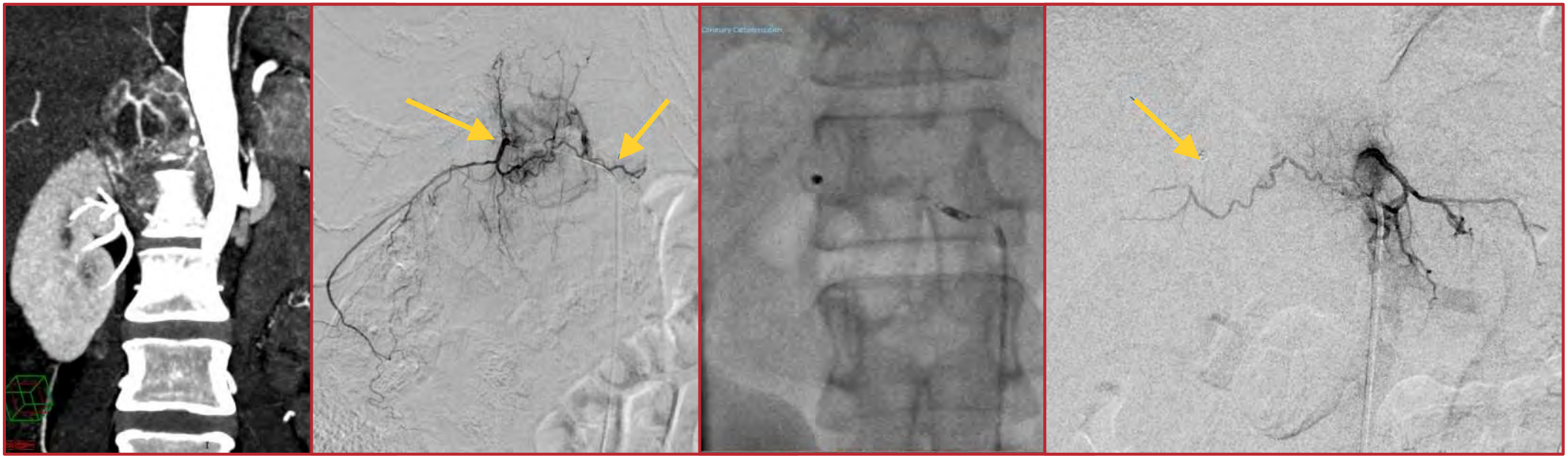
heterogenous enhancement, well-defined lesion, 4x4x7 cm, at L1-L2



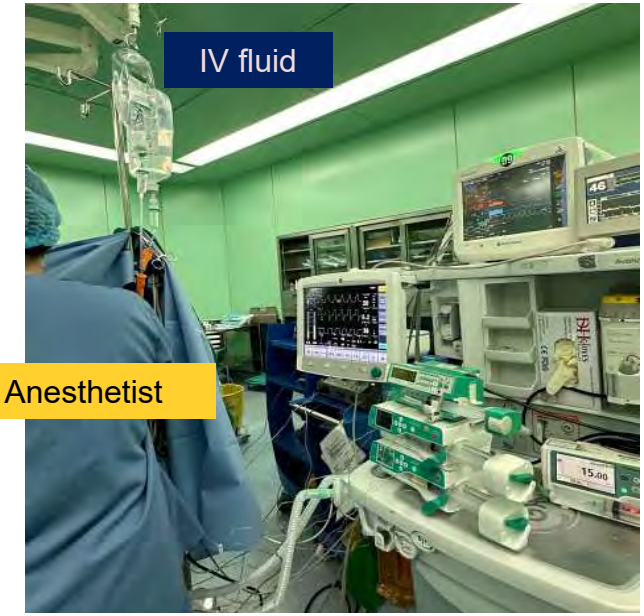



# Transcatheter artery embolization

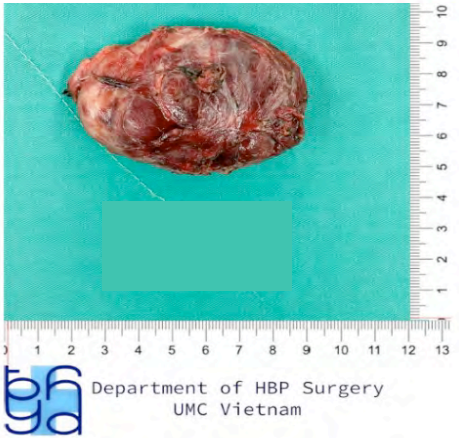
- Adrenal MDM decision: Because of the size, position, and nature of the tumor, **transcatheter artery embolization** was performed **2 days prior** to the open removal of PPGL to minimize the potential blood loss during the removal of PPGL.

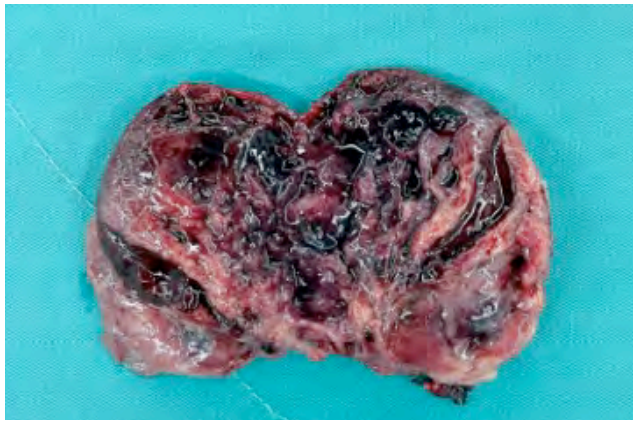


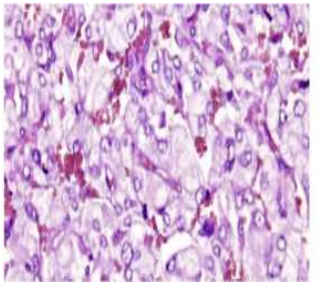
# Open removal of PPGL

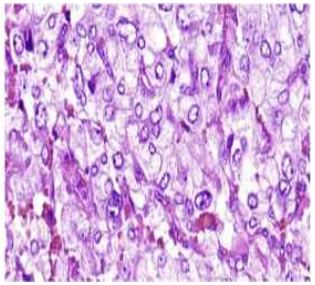










- Total blood loss: 300mL.
- Operation time: 180 mins.

## Post-op follow-up

- BP normalized without antihypertenvie medications.
- Genetic testing: **no germline mutation**.

(Tested genes: *PRKAR1A, MEN1, CDKN1B, CDKN1A, CDKN2B, CDKN2C, IFNG, TSC1, TSC2, HRAS, VHL, RET, SDHA, KIF1B, FH, SDHAF2, TMEM127, NF1, SDHC, SDHD, SDHB, MAX, DNMT3A, EPAS1, CCND1*)

- Plasma metanephrines normalized

  Plasma metanephrine: 31.8 pg/mL (normal: < 90).

  Plasma normetanephrine : 57.87 pg/mL (normal: < 196).

→ annual follow-up with plasma metanephrines.

## Discussion points

- Diagnosis of PPGL in pregnancy.
  - Differentiate between gestational HT, pre-eclampsia, and chronic HT.
  - Confirmation of PPGL in pregnancy.
- Antepartum management and timing of delivery:

Pros and Cons of:

  - elective C-section, followed by the removal of PPGL
  - ***or*** combined C-section and tumor resection