SEPTEMBER 26, 2025

Interhospital Conference

Endocrine unit, Vajira hospital

F1 Puttida Tovirat
F2 Orawan Kongphet

Saharat Leelanuwatkul, M.D.
Praepaiphan Krueworramunee, M.D.

Case

ผู้ป่วยหญิงไทยโสด อายุ 27 ปี

Chief complaint : ปวดศีรษะมากขึ้น 2 เดือนก่อนมาโรงพยาบาล

Present illness

2 เดือนก่อนมาโรงพยาบาล ปวดศีรษะมากขึ้นและมีหน้ามืด ไม่มีตามัว ไม่มีเห็นภาพซ้อน ไม่มีแขนขาอ่อนแรง ไม่มีเจ็บหน้าอก ไม่มีใจสั่น ไปตรวจสุขภาพพบความผิดปกติ จึงส่งตัวมารักษาต่อที่วชิรพยาบาล

Personal history

- ปฏิเสธแพ้ยา หรือแพ้อาหาร
- ปฏิเสธการใช้สารเสพติด ดื่มแอลกอฮอล์ และสูบบุหรี่
- ปฏิเสธการตั้งครรภ์

Family history

- บิดา และพี่ชาย เป็นความดันโลหิตสูง

Physical Examination

BT 36.7 ^{o}C, BP 147/89 mmHg, PR 96 /min, RR 20 /min

Body weight 60 Kg, Height 160 cm, BMI 23.44 kg/m^{2}

- GA: A Thai female, well co-operative
- HEENT: No pale conjunctiva, anicteric sclera, no thyroid gland enlargement
- CVS: Full pulse all extremities, regular pulse, PMI at 5th intercostal space at midclavicular line, normal S1S2, no murmur
- RS: Equal breath sound both lungs, no adventitious sound
- Abdomen: Normoactive bowel sound, soft, not tender, no palpable mass
- Extremities: No pitting edema

Further history and Physical examination required?

Present illness

- 3 ปีก่อนมาโรงพยาบาล ผู้ป่วยให้ประวัติมีอาการปวดศีรษะเป็นๆหายๆ อาการปวดศีรษะมักเป็นตอนออกแรงและเดินขึ้นบันได ลักษณะเป็นปวดบีบรัดทั่วศีรษะ ไม่มีปวดสลับข้าง ครั้งละประมาณ 5 นาที pain score 5/10 นั่งพักอาการดีขึ้น ไม่มีสะดุ้งตื่นเพราะปวดศีรษะ อาการปวดศีรษะไม่สัมพันธ์กับแสง ไม่มีเห็นแสงผิดปกติ ไม่มีตามัว ไม่มีเห็นภาพซ้อน มีหูอื้อ ไม่มีคลื่นไส้อาเจียน ไม่มีปวดตามใบหน้า ไม่มีน้ำตาหรือน้ำมูกไหล ไม่มีแขนขาอ่อนแรง ไม่มีเจ็บหน้าอก ไม่มีใจสั่น มีหน้ามืดบางครั้งเวลาลุกขึ้นจากเตียง มีเหงื่อออกง่ายตามหน้าและมือเวลาอากาศร้อน น้ำหนักเพิ่มขึ้น 10 kg ใน 3 ปี (55kg ⟶ 65kg) ทานอาหารได้ปกติ ไม่มีเบื่ออาหาร ไม่มีหน้าบวมขาบวม ไม่มีผิวหนังแตกลาย ไม่มีผื่นตามตัว ไม่มีขน/สิวขึ้นเยอะผิดปกติ ไม่มีผมร่วง ยังไม่ได้ไปรักษาที่ไหน
- 2 เดือนก่อนมาโรงพยาบาล ปวดศีรษะมากขึ้นและเป็นบ่อยมากขึ้น ไปตรวจสุขภาพพบความผิดปกติ จึงส่งตัวมารักษาต่อที่วชิรพยาบาล

Past history

- Underlying disease:
  - Hypertension diagnosed at age 24-year-old
    - ตรวจเจอจากการไปบริจาคเลือด
- Current medication
  - Amlodipine 10mg/day (start 2 months PTA)
- ปฏิเสธประวัติผ่าตัด

Personal history

- Menarche 13 years old
- Regular menstruation
- ปฏิเสธเพศสัมพันธ์ หรือตั้งครรภ์
- ปฏิเสธแพ้ยา หรือแพ้อาหาร
- ปฏิเสธการใช้สารเสพติด ดื่มแอลกอฮอล์ และสูบบุหรี่
- ปฏิเสธการใช้ยาต้ม ยาหม้อ ยาสมุนไพร และยาลูกกลอน
- ปฏิเสธการเกิดอุบัติเหตุ

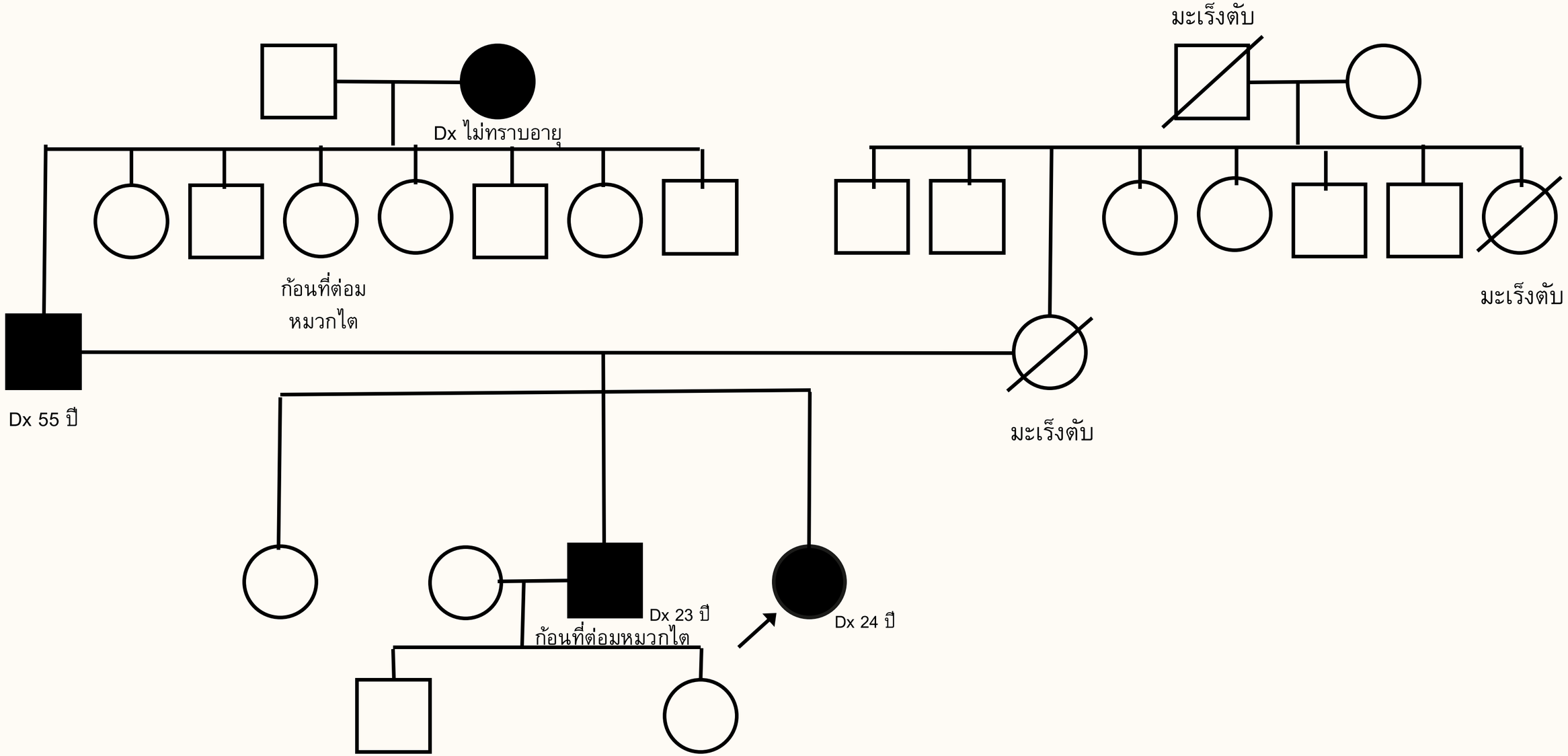
Family history

- ย่า เป็นความดันโลหิตสูง ไม่ทราบอายุตอนวินิจฉัย
- บิดา เป็นความดันโลหิตสูง วินิจฉัยตอนอายุ 55 ปี
- พี่ชาย เป็นความดันโลหิตสูง วินิจฉัยตอนอายุ 23 ปี

- ตา เป็นมะเร็งตับ ไม่ทราบอายุตอนวินิจฉัย
- มารดา เป็นมะเร็งตับ วินิจฉัยตอนอายุ 30 ปี เสียชีวิตแล้ว
- น้า (น้องสาวมารดา) เป็นมะเร็งตับ วินิจฉัยตอนอายุ 32 ปี เสียชีวิตแล้ว

- ปฏิเสธประวัติเส้นเลือดสมองตีบ / แตกในครอบครัว
- ปฏิเสธประวัติเส้นเลือดหัวใจตีบในครอบครัว
- ปฏิเสธประวัติการเสียชีวิตโดยไม่ทราบสาเหตุ / เสียชีวิตจากโรคหัวใจในครอบครัว

Pedigree (Hypertension)



Physical Examination

Vital signs

- Body temperature 36.7 ^{o}C **Blood pressure 147/89 mmHg**

  Pulse rate 96 /min Respiratory rate 20 /min
- Body weight 60 Kg, Height 160 cm, BMI 23.44 kg/m^2

- General appearance: A Thai female, well co-operative, follow to command
- HEENT: No pale conjunctiva, anicteric sclera, **no thyroid gland enlargement, no palpable thyroid nodule**, no palpable superficial lymph node at supraclavicular, clavicular area, **no facial plethora, no moon face, no dorsocervical hump, no loss of supraclavicular fat pad**

Physical Examination

- CVS : No central and peripheral cyanosis, no active precordium, no neck vein engorgement, no carotid bruit, full pulse all extremities, regular pulse, capillary refill <2 second, PMI at 5^{th} intercostal space at midclavicular line, no heaving, no thrill, normal S1S2, no murmur, no distant heart sound
- RS : Normal chest contour, normal chest expansion, trachea in midline, clear and equal breath sound both lungs, no adventitious sound
- Abdomen : Globular shape, no distension, no superficial vein dilatation, no surgical scar, normoactive bowel sound, soft, not tender, no palpable mass
- Extremities : No deformities, no joint swelling, no pitting edema
- Skin : No thin skin, **no purplish striae, no ecchymosis**, no acanthosis nigricans, **normal female hair distribution, no hirsutism**

Neurological Examination

- Mental status : orientate to time, place, person
- Speech: intact naming/repetition/fluency/comprehension
- Cranial nerve
  - CN I : not examined
  - CN II : pupil 3 mm RTL BE, RAPD negative both side, VA normal , VF normal by confrontation
  - CN III, IV, VI : no ptosis , full EOM both eyes, no nystagmus
  - CN V : normal facial sensation, normal muscle of mastication
  - CN VII : no facial palsy
  - CN VIII : normal hearing
  - CN IX,X : intact gag reflex, no uvular deviation
  - CN XI : normal power of sternocleidomastoid and trapezius muscle
  - CN XII : no tongue atrophy or deviation

Neurological Examination

- Motor : Normal muscle tone, no muscle atrophy, muscle power grade V all extremities, **no proximal muscle weakness**
- Sensory : Intact pinprick sensation
- Reflexes : Deep tendon reflex 2+ all extremities, no slow relaxation of DTR
- Clonus : Absent both side
- Meningeal irritation sign: Negative Babinski's sign: plantar flexion both sides
- Cerebellar sign: Intact finger to nose, no dysdiadochokinesia, no truncal ataxia

Problem list

Problem list

A 27-year-old female patient presented with

- Progressive intermittent headache for 2 months
- Significant weight gain 10 kg in 3 years
- Personal history of hypertension in the young (diagnosed at age 24)

- Family history of hypertension in father (at age 55) and older brother (at age 23)
- Family history of liver cancer in grandfather, mother, and aunt
- Family history of adrenal nodule in uncle and older brother

Investigation

Investigation

24hr Urine Normetanephrine/Metanephrine

- Day1: Urine Volume 3600 ml, Urine Cr 1.062 g/24 Hrs (17.7 mg/kg/d)
  - Metanephrine 329 nmol/day (less than 1777)
  - **Normetanephrine 50,361 nmol/day** (less than 3279)
- Day2: Urine Volume 3900 ml, Urine Cr 0.967 g/24 Hrs (16.1 mg/kg/d)
  - Metanephrine 436 nmol/day (less than 1777)
  - **Normetanephrine 59,672 nmol/day** (less than 3279)

8AM cortisol

- 13.3 ug/dl

Thyroid function test

• Free T3	3.13 pg/ml	(2-4.4)
• Free T4	1.45 ng/dl	(0.93-1.7)
• TSH 3rd gen	1.66 uIU/mL	(0.27-4.2)

Blood Chemistry

• BUN	7	mg/dl
• Creatinine	0.55	mg/dL
• Sodium	139	mmol/L
• Potassium	3.88	mmol/L
• Chloride	102	mmol/L
• Carbon dioxide	23	mmol/L
• Calcium	9.5	mg/dL
• Phosphorus	3.3	mg/dL

Complete blood count

- Hb 12.9 g/dl
- Hct 39.8 %
- WBC count 10,020 cells/mm^3
- Neutrophil 68 %
- Lymphocyte 25.3 %
- MCV 76.2 fl
- Plt count 495,000 cells/ mm^3

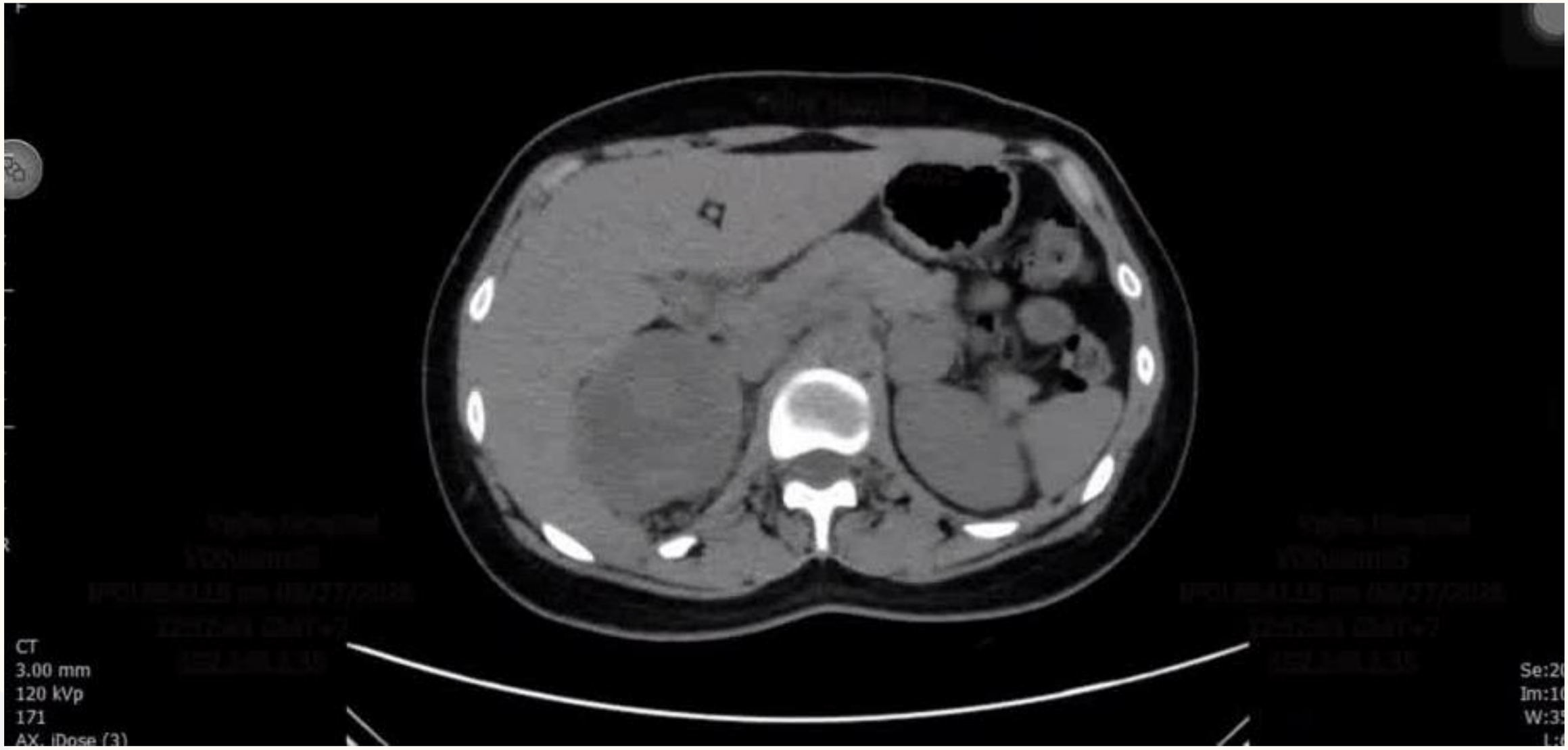
Blood sugar, HbA1c & Lipid profile

• FBS	120	mg/dl
• HbA1C	6.0%	
• Cholesterol	177	mg/dl
• HDL	47	mg/dl
• LDL	122	mg/dl
• Triglyceride	61	mg/dL

Liver function test

• Total Bilirubin	0.55	mg/dl
• Direct Bilirubin	0.24	mg/dl
• AST	4	U/L
• ALT	6	U/L
• ALP	41	U/L
• Albumin	4.4	g/dL

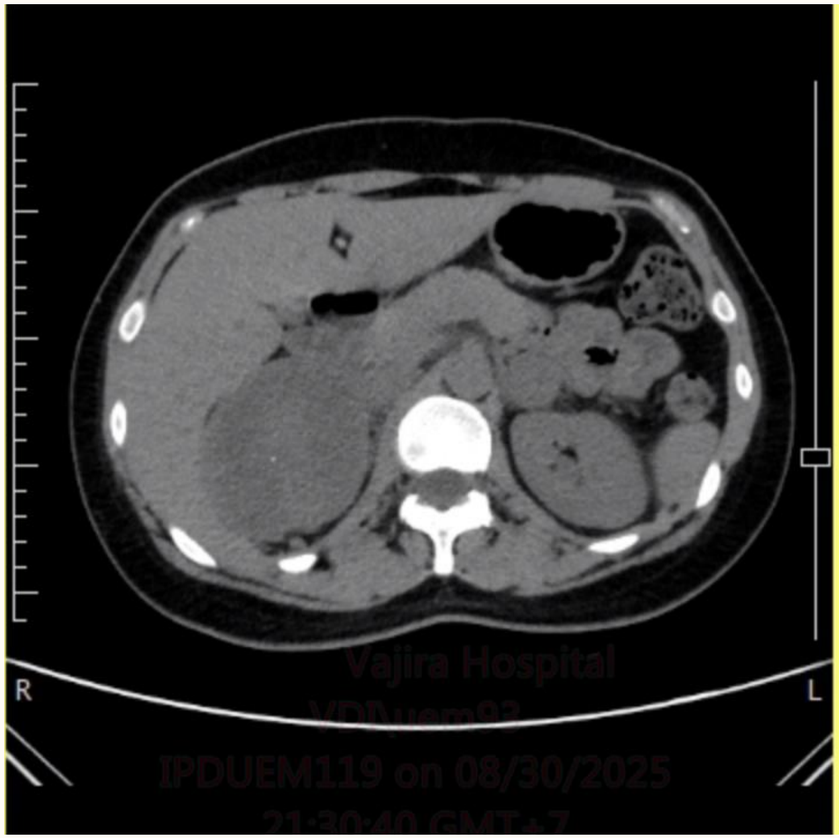
CT whole abdomen (NC)



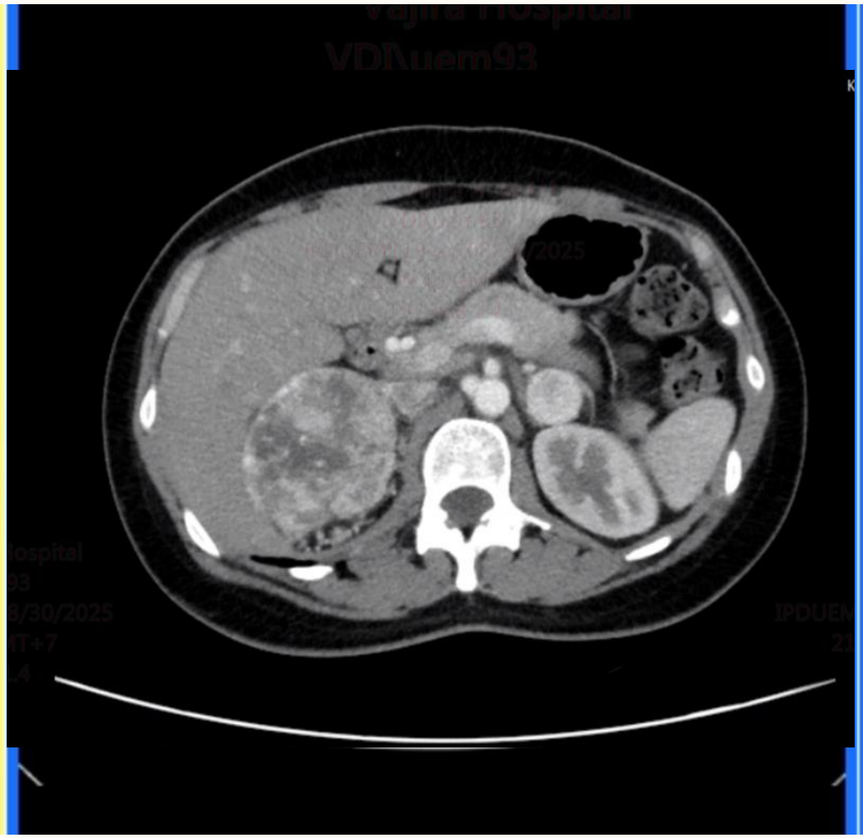
CT whole abdomen (A phase)

CT whole abdomen (V phase)

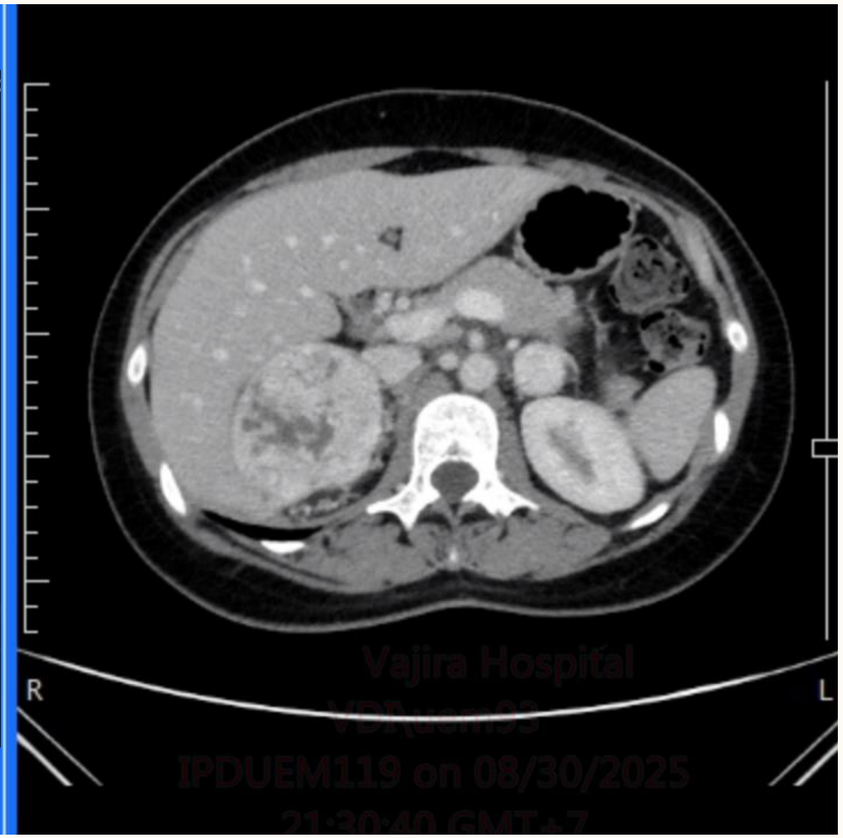
CT whole abdomen



Non contrast



A phase



V phase

CT whole abdomen

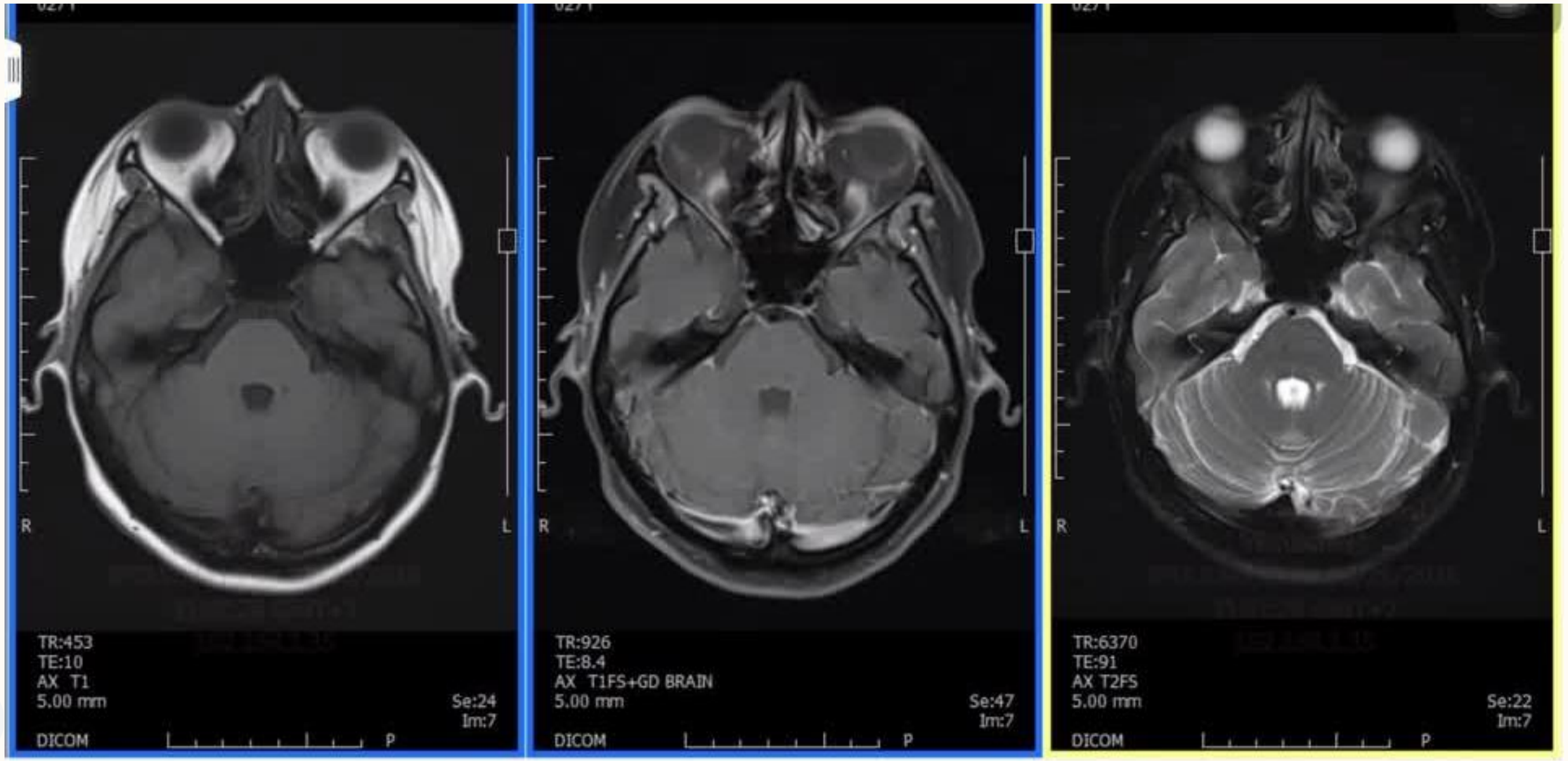
- Large well-defined arterial enhancing mass with progressive enhancement in protovenous and delayed phases, a few tiny internal calcification, and foci of internal necrosis, without adjacent organ invasion, involving medial limb, body, and lateral limb of right adrenal gland, causing inferior displacement of right kidney, size 7.0x7.0x11.0 cm; hypervascular tumor such as pheochromocytoma.

- Arterial enhancing mass with progressive enhancement in protovenous and delayed phases without internal calcification or necrosis at body of left adrenal gland, size 2.7x2.7x3.0 cm.

CT Whole abdomen

- Liver is normal size and no enhancing lesion. Portal vein is normal.
- Gallbladder, pancreas, spleen, both adrenal glands, uterus, left ovary and urinary bladder are normal.
- Both kidneys are normal shape and size with no hydronephrosis or stone.
- Few left renal cysts, size 1 cm or less are noted.
- Both ureters are not dilated.
- Hypodensity lesion with rim enhancement, size 2.5x2.5x2 cm at right ovary is seen. Right ovary is not enlarged.
- No dilatation of biliary and KUB systems.
- No mass at stomach, small bowel and large bowel is detected.
- No lymphadenopathy is seen.
- Abdominal aorta and IVC are unremarkable.

MRI brain and screening whole spine

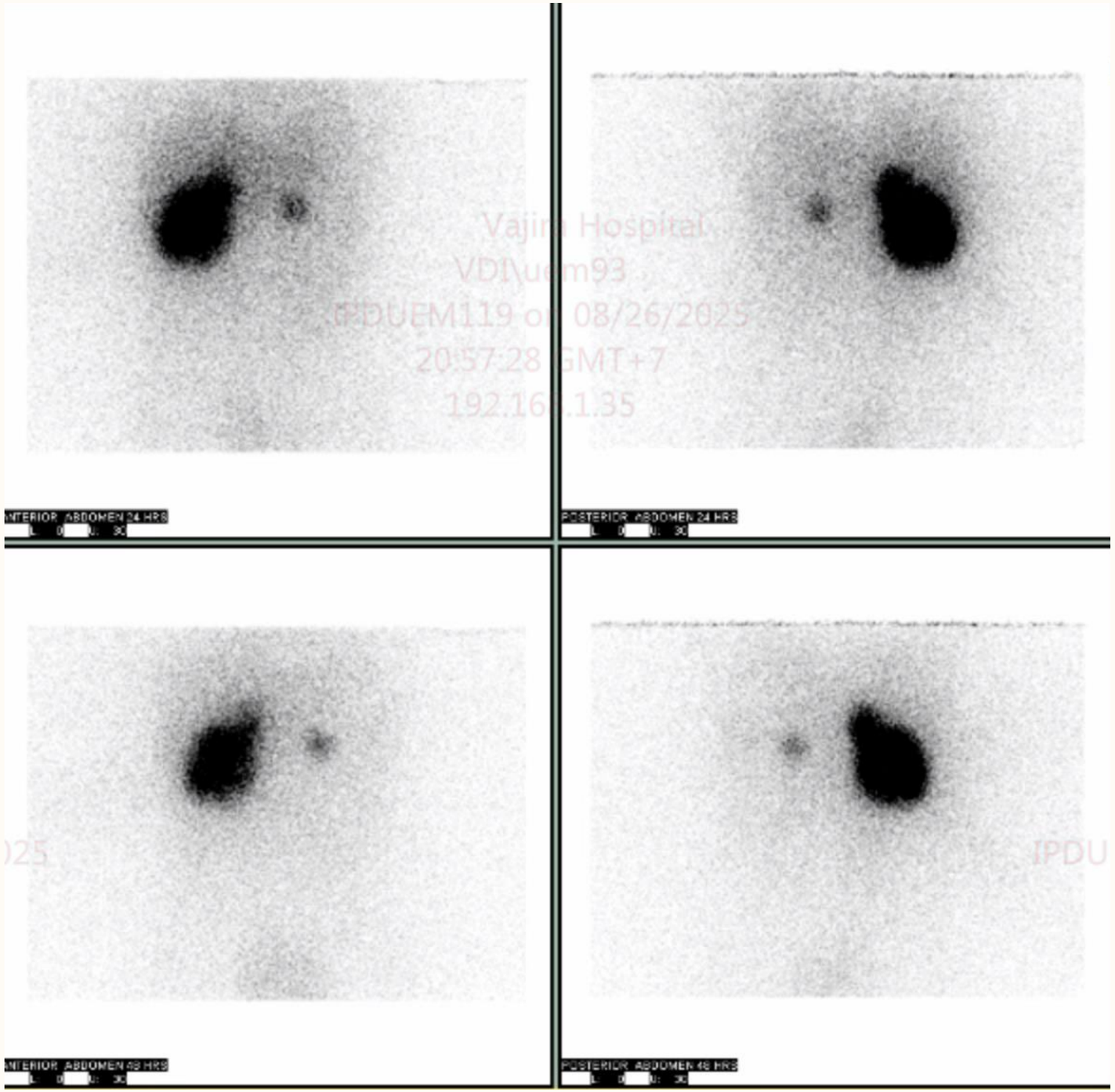


MRI brain and screening whole spine

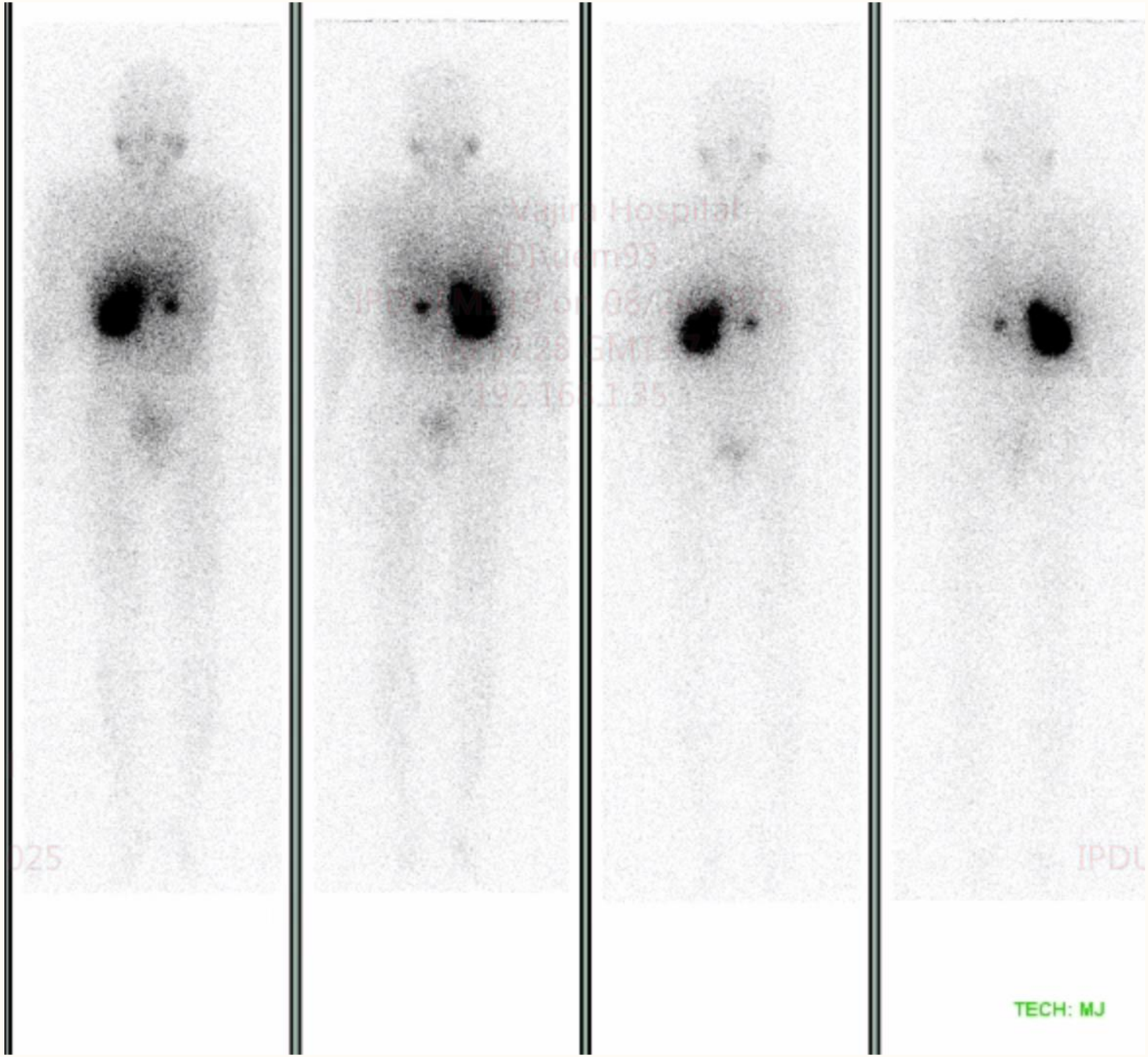
MRI brain and screening whole spine

- No abnormal signal intensity in visualized brain parenchyma
- No abnormal enhancing lesion and leptomeningeal enhancement.
- No demonstrated of intracranial hemorrhage, brain swelling, brain herniation, midline shift or hydrocephalus is seen.
- The cerebellum and brainstem appear normal.
- The pituitary gland shows normal size and signal intensity. Posterior pituitary bright spot is seen.
- No extra-axial space taking lesion is noted.
- Mucous retention cyst at right maxillary sinus.
- The rest of visualized paranasal sinuses, both mastoid air cells and both orbits are unremarkable.
- Screening the whole spine shows no abnormal intramedullary/extramedullary lesion. Normal marrow and disc signal intensity.

I-131 MIBG Total body scan



I-131 MIBG Total body scan

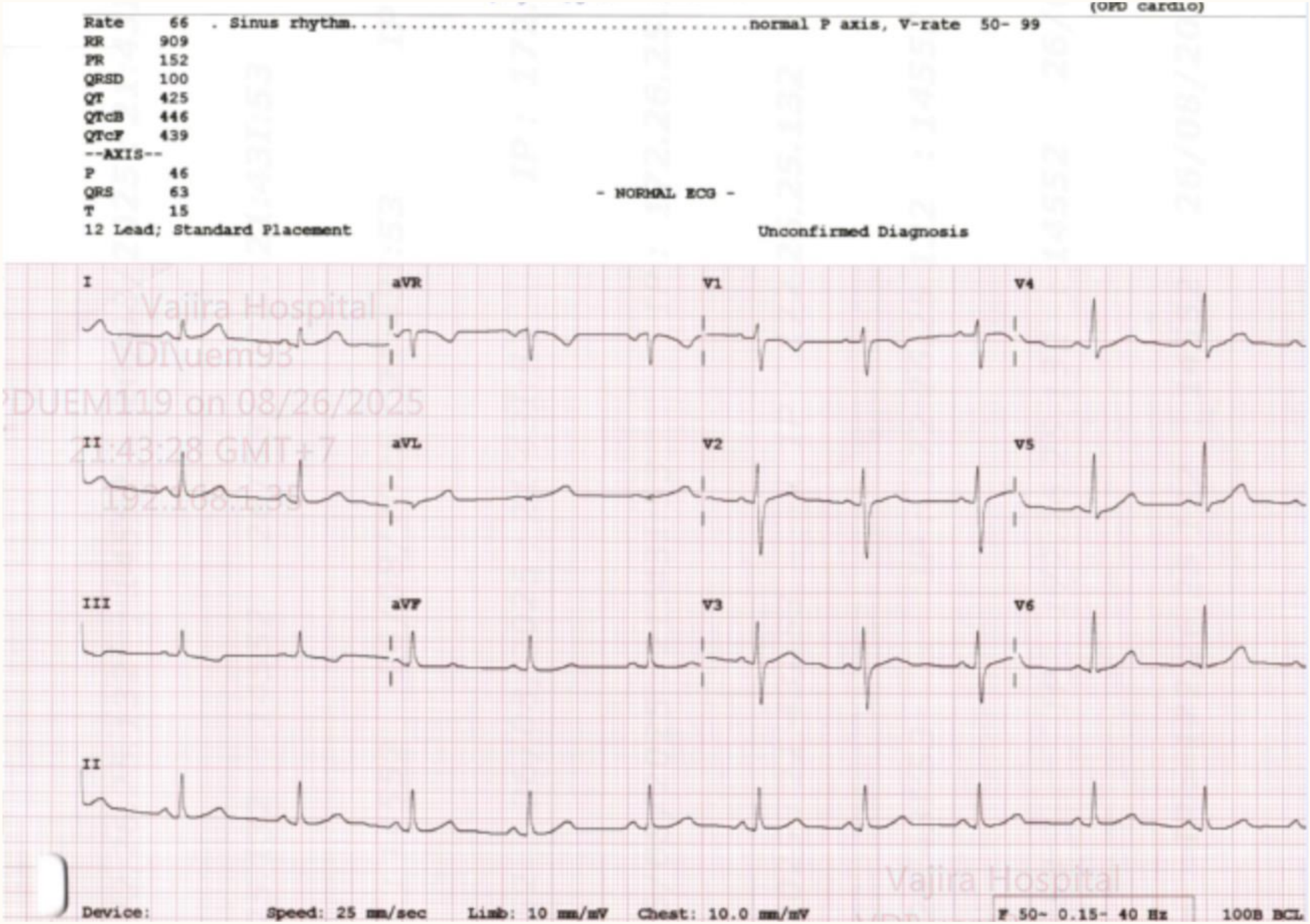


I-131 MIBG Total body scan

- The study reveals **two foci of abnormal MIBG uptake in bilateral sides of upper-medial abdomen, the largest one locates in the right side, another one locates in the left side.**
- Additional SPECT/CT scan shows markedly increased MIBG uptake in right adrenal mass.
- Another small focus of abnormal MIBG uptake in left adrenal mass.
- Otherwise, are unremarkable.

- IMP: Suggestive of **bilateral pheochromocytoma.**

EKG


(OPD cardio)
Rate 66 . Sinus rhythm.....normal P axis, V-rate 50- 99
RR 909
PR 152
QRSD 100
QT 425
QTcB 446
QTcF 439
--AXIS--
P 46
QRS 63
T 15
- NORMAL ECG -
12 Lead; Standard Placement
Unconfirmed Diagnosis
I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
II
Device:
Speed: 25 mm/sec
Limb: 10 mm/mV
Chest: 10.0 mm/mV
F 50- 0.15- 40 Hz
100B BCL

EKG & Echocardiogram

- LV is concentric remodeling. LVEF is normal (64.4% by Biplane's method) with normal wall motion.
- LA is not dilated. There is normal LV diastolic function.
- There is no significant valvular dysfunction
- RA and RV are not dilated. RV systolic function is normal
- Peak PR and Peak TR Doppler velocities could not be obtained for PAP calculation
- With TTE resolution, no intracardiac mass or shunt was seen. Aortic root is not dilated
- There is no pericardial effusion.

Diagnosis & Management

Diagnosis

Bilateral pheochromocytoma

Management

Open right adrenalectomy and

Laparoscopic left adrenalectomy

Preoperative Management

- High salt diet 5g /day
- Doxazosin 8 g/day
- Atenolol 75 g/day
- IV hydration for 2 days before operation
- Hydrocortisone 200 mg IV drip in 24 hr before operation

- Preoperative BP, HR
  - Seated BP 118/71 mmHg HR 70 /min
  - Upright BP 111/72 mmHg HR 80 /min

ANATOMICAL PATHOLOGICAL REPORT

PREVIOUS PATHOLOGY REPORT: S25-03712

PATHOLOGICAL DIAGNOSIS:

A) Mass at right adrenal gland, adrenalectomy:

- Pheochromocytoma, see note.
- Tumor size is 12.0 cm.
- Presence of tumor necrosis.
- No capsular invasion seen.
- No lymphovascular invasion seen.
- No periadrenal soft tissue involvement seen.
- Uninvolved resection margin.

Note: Pheochromocytoma of the adrenal gland scaled score (PASS): Score 6 (Tumor necrosis, Marked nuclear pleomorphism, Large nests and Hyperchromasia). Ki67 stain is suggested in block A2 for grading.

B) Mass at left adrenal gland, adrenalectomy:

- Pheochromocytoma, see note.
- Tumor size is 4.0 cm.
- No tumor necrosis seen.
- No capsular invasion seen.
- No lymphovascular invasion seen.
- No periadrenal soft tissue involvement seen.
- Uninvolved resection margin.

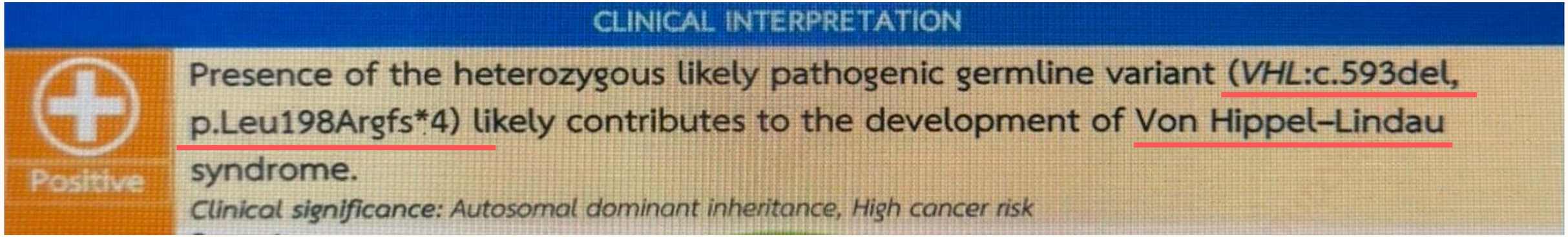
Note: Pheochromocytoma of the adrenal gland scaled score (PASS): Score 4 (Marked nuclear pleomorphism, Large nests and Hyperchromasia). Ki67 stain is suggested in block B1 for grading.

Additional report: (28/05/2025)

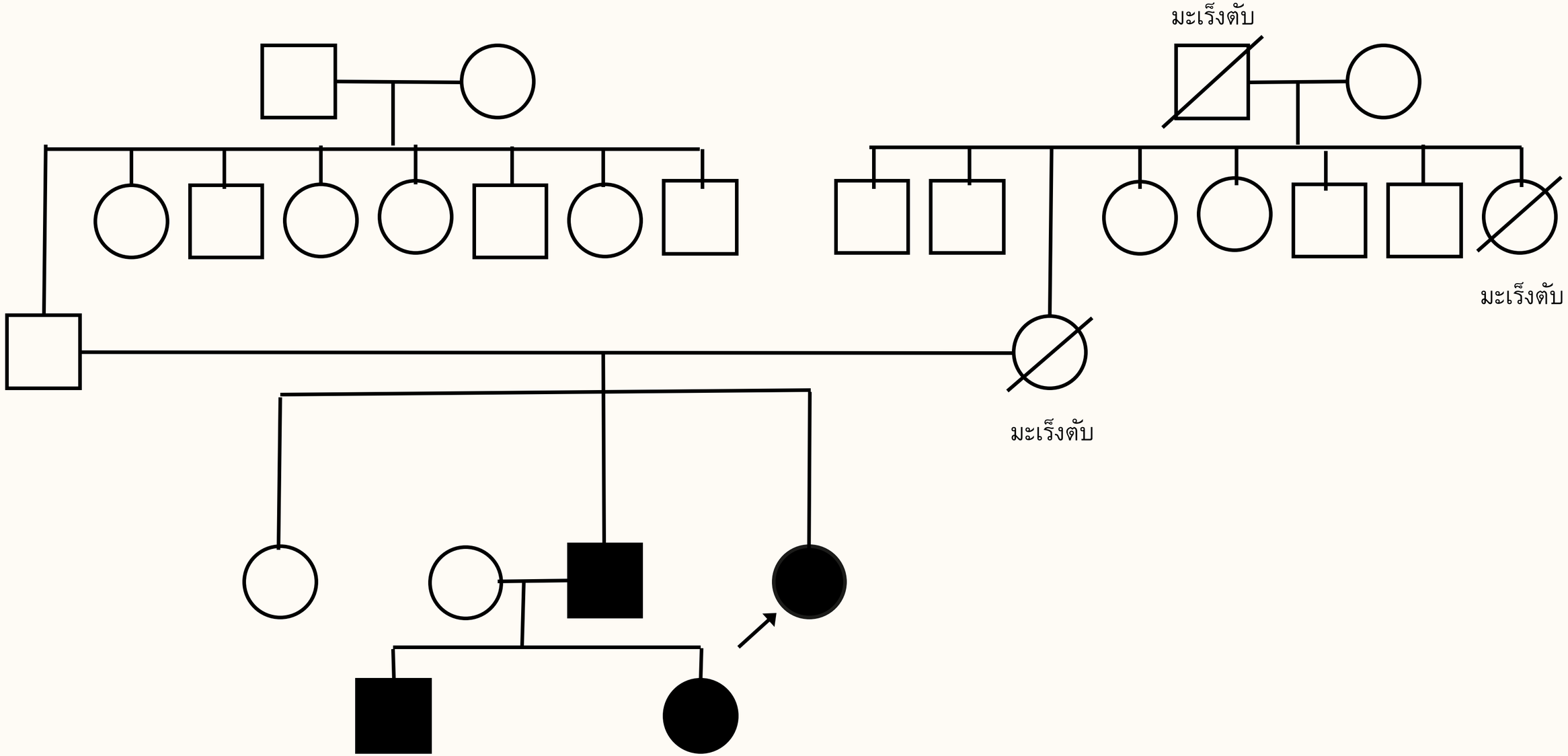
- Ki67 stain reveals positive nuclei approximately >3%.

Pathological report

Genetic testing



Pedigree (VHL confirmed)



Post-op case progression

- Clinical headache improved
- BP 116-127/72-78 mmHg, HR 66-75 bpm
- CBG 92-118mg%

- Off doxazosin, atenolol
- Taper hydrocortisone to prednisolone 10 mg/day
- Add fludrocortisone 0.05mg/day

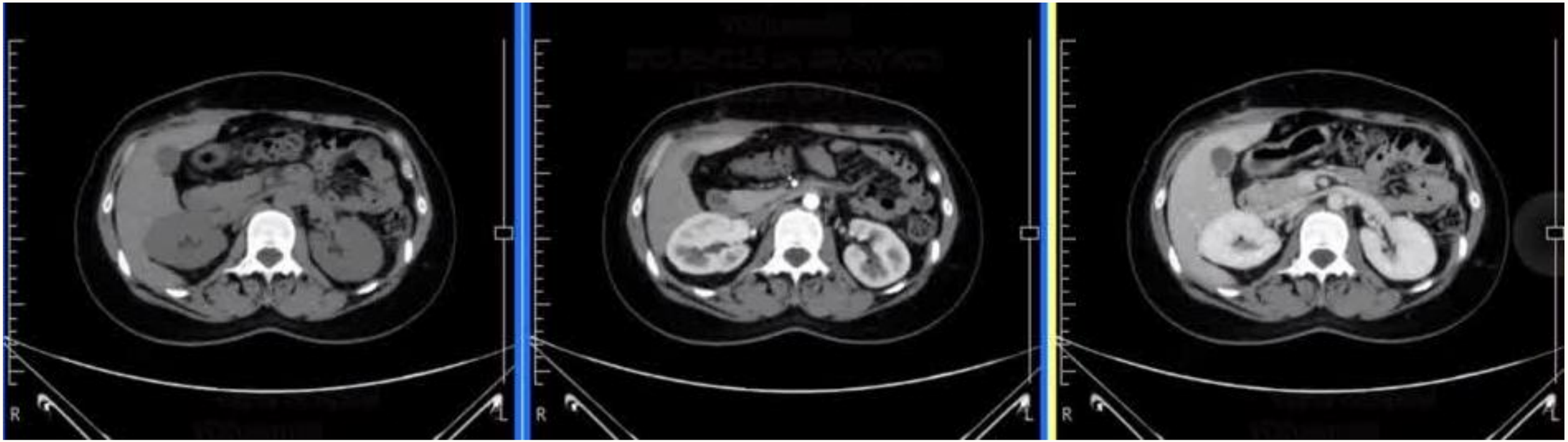
24hr Urine Normetanephrine/Metanephrine

Post-op (16weeks)

- Day1: Urine Volume 2500ml, Urine Cr 0.720 g/24Hrs (11.6mg/kg/d)
  - Metanephrine 254 nmol/day (less than 1777)
  - Normetanephrine 273 nmol/day (less than 3279)

- Day2: Urine Volume 2900ml, Urine Cr 1.343 g/24Hrs (21.6mg/kg/d)
  - Metanephrine 530 nmol/day (less than 1777)
  - Normetanephrine 1,585 nmol/day (less than 3279)

CT upper abdomen Post-op (4weeks)



Non contrast

A phase

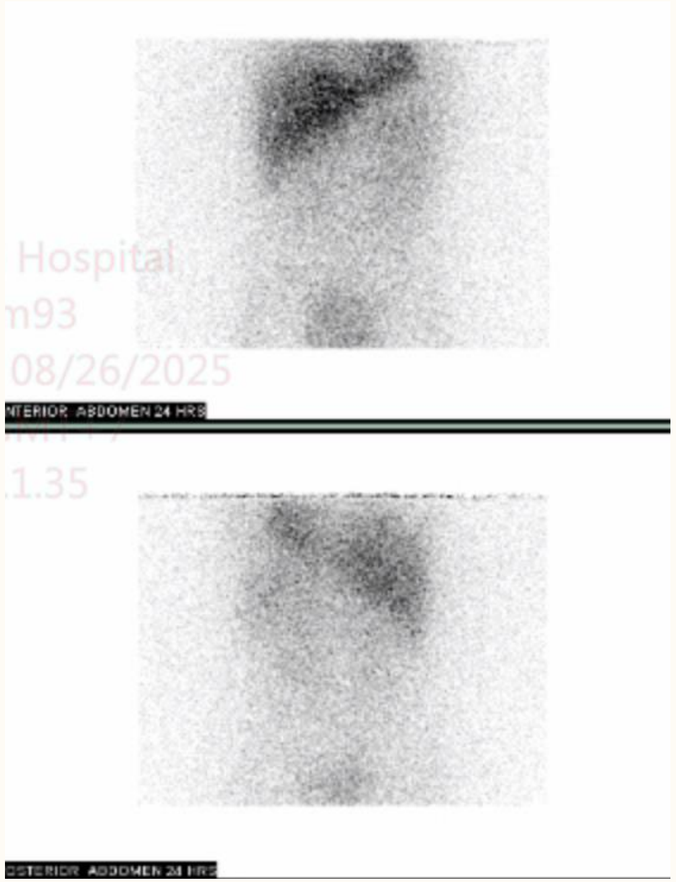
V phase

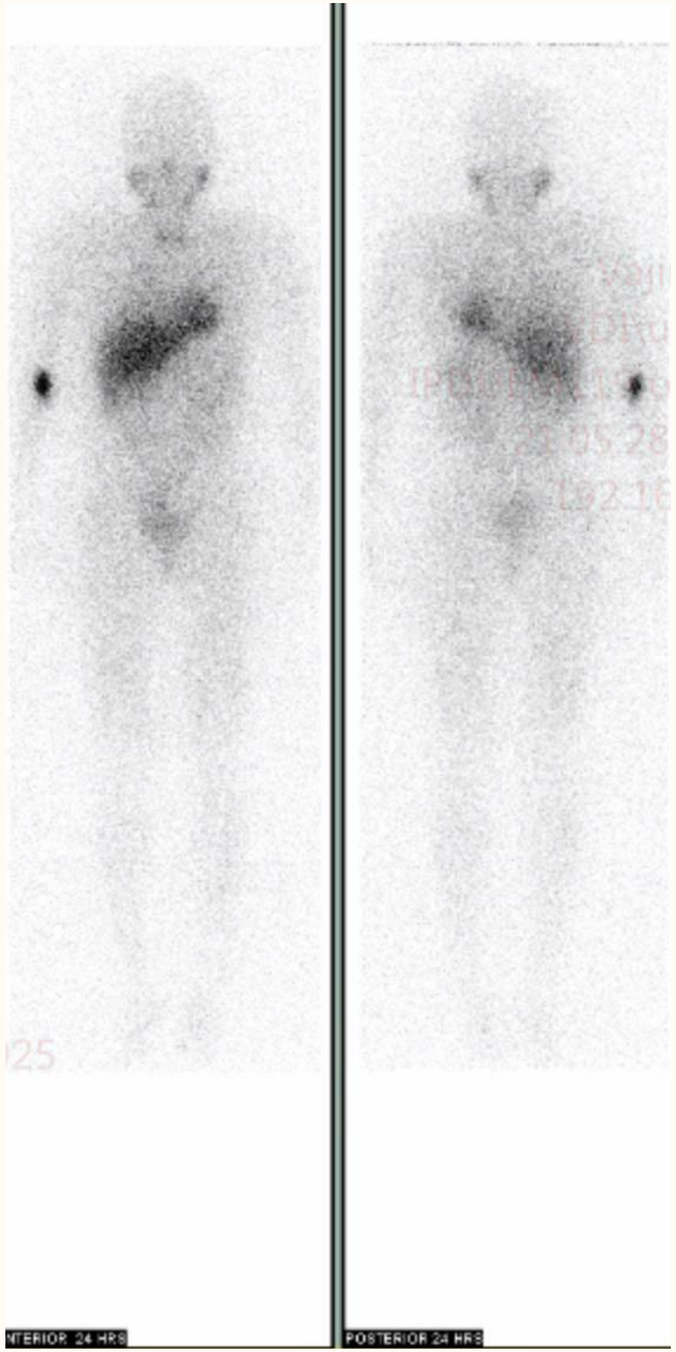
CT upper abdomen Post-op (4weeks)

- **No mass at both adrenal glands and adrenal gland is detected.**
- Liver is normal size and no enhancing lesion. Portal vein is normal.
- Gallbladder, pancreas, spleen, both adrenal glands, uterus, left ovary and urinary bladder are normal.
- Both kidneys are normal shape and size with no hydronephrosis or stone
- Few left renal cysts, size 1 cm or less are no changed.
- No dilatation of biliary and KUB systems
- Few diverticulum at ascending colon and transverse colon are seen.
- No mass at stomach, small bowel and large bowel is detected.
- No lymphadenopathy is seen.

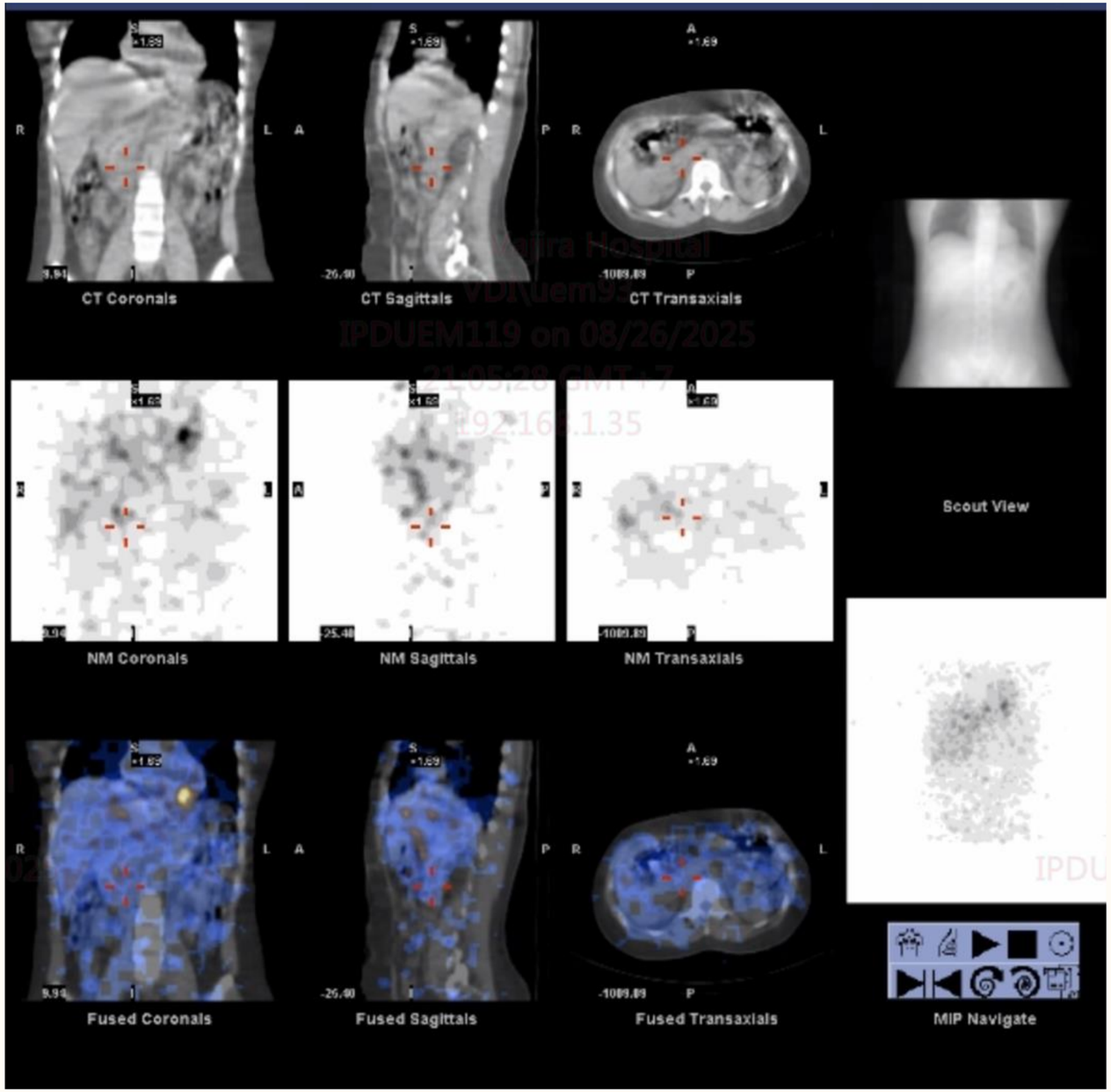
I-131 MIBG

Post-op (16 weeks)





Post-op I-131 MIBG



Post-op I-131 MIBG

The study reveals **no abnormal uptake at surgical bed and elsewhere that suggests residual tumor or recurrent.** Physiologic distribution is noted at salivary glands, thyroid gland, heart, liver, and excrete via urinary system.

IMP: No abnormal uptake at surgical bed and elsewhere that suggest residual tumor or recurrent in this study.

Von Hipple-Lindau (VHL) Syndrome

VHL clinical diagnostic criteria

	Danish criteria	International criteria
Positive family history/ VHL mutation	1 vHL-related manifestation	1 vHL-related manifestation
Negative family history	At least 2 vHL-related manifestations	2 hemangioblastoma (CNS or retinal) or At least one hemangioblastoma (retinal and/or CNS) and a visceral lesion
vHL-related manifestations included in the criteria	Retinal hemangioblastoma hemangioblastoma in cerebellum, the medulla oblongata, or the spinal cord Endolymphatic sac tumor Renal cell carcinoma Pheochromocytoma, paraganglioma, and/or glomus tumor PNET and/or multiple pancreatic cysts	Retinal hemangioblastoma CNS hemangioblastoma Renal cell carcinoma Pheochromocytoma PNET Endolymphatic sac tumor Pancreatic cysts* Epididymal cystadenomas*

Binderup ML, et al. Dan Med J 2018;65(3):B5461

Clinical manifestation

Table 3. Frequency of lesions and average age range of presentation in VHL patients

Clinical Feature	Average (range) of presentation (years)	Frequency (%)	Reference
CNS hemangioblastoma	30 (9–78)	60–80%	1
Retinal hemangioblastoma	25 (1–67)	49–62%	4, 6
Endolymphatic sac tumors	31 (12–50)	6–15%	7, 15
Renal cell carcinoma or cysts	39 (16–67)	30–70%	6
Pheochromocytoma	30 (5–58)	10–20%	6
Pancreatic neuroendocrine tumors or cysts	36 (1–70)	35–70%	8, 15
Epididymal cystadenomas	Unknown (16–40)	25–60%	6, 15
Broad ligament cystadenomas	Unknown (16–46)	Unknown	8

Varshney N. J Kidney Cancer VHL. 2017 Aug 2;4(3):20-29

VHL-related PPGLs

Molecular Cluster

- Cluster 1B : pseudohypoxia signaling related mutations
- Driven by impaired degradation of HIF-α
- Leading to angiogenesis and tumorigenesis

Clinical manifestations :

- Sustained hypertension due to the continuous release of norepinephrine
- Tumor Location
  - **Pheochromocytomas (PCCs), about 50% bilateral adrenal tumors.**
  - Less frequently paragangliomas (PGLs), typically in the head and neck.

Endocrine Reviews,2022 vol43, No 2.

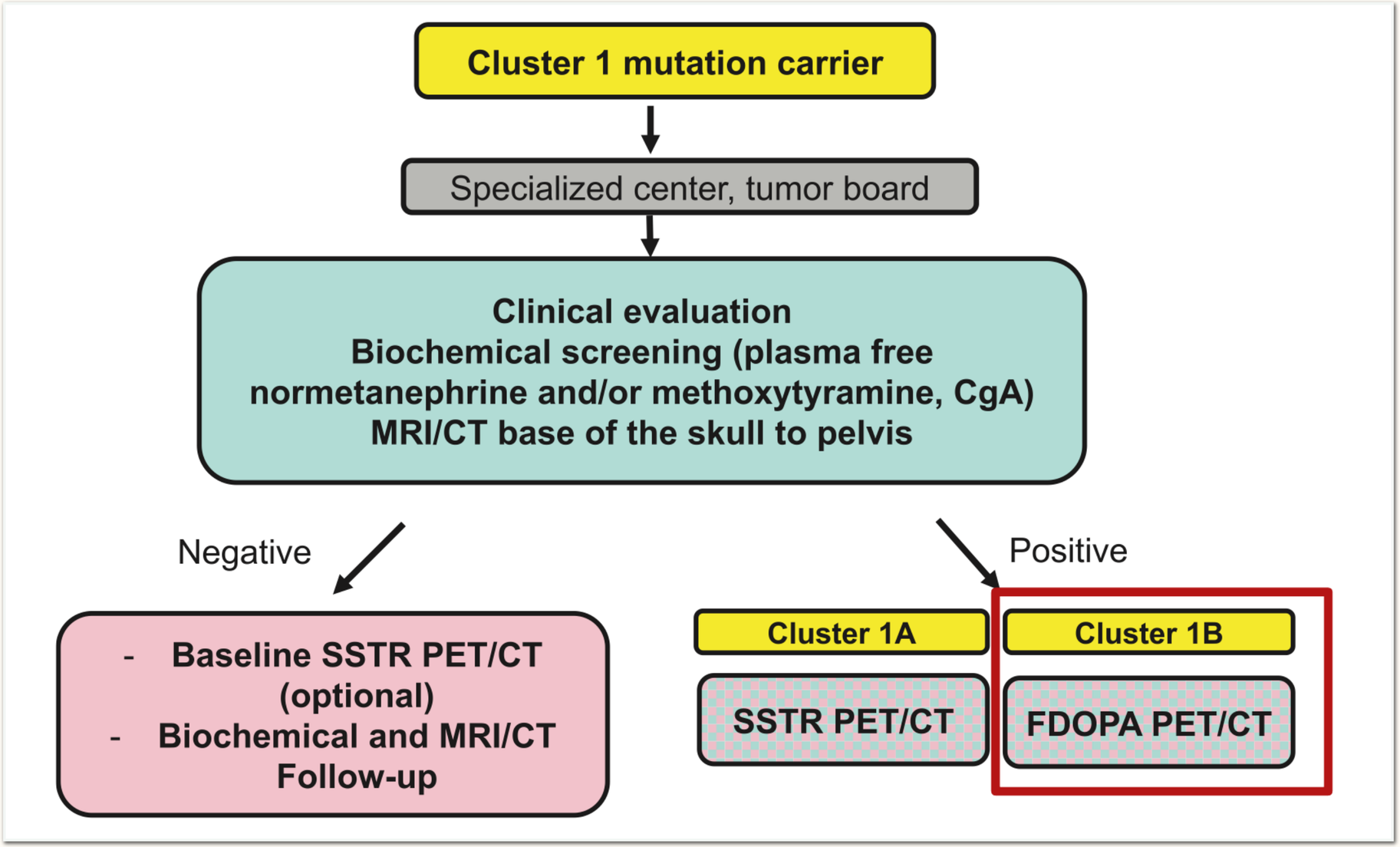
VHL-related PPGLs

Penetrance

- Lifetime penetrance of VHL ~15–20% for developing PPGLs.
- Risk varies by mutation type
  - Missense mutations (VHL type 2) ⟶ higher PPGL risk.
  - Exon deletions/truncating mutations (VHL type 1) ⟶ lower PPGL risk.
- Metastatic risk : 5-8 %

Endocrine Reviews,2022 vol43, No 2.

VHL-related PPGLs



Endocrine Reviews,2022 vol43, No 2.

VHL-related PPGLs

Treatment : Cortical-sparing adrenalectomy

- Preferred to preserve adrenal function.
- Risk of recurrence (~13%) but no reduction in overall survival.
- Considered appropriate for less aggressive VHL-related PCCs.

Endocrine Reviews,2022 vol43, No 2.

VHL-related PPGLs

Table 12. Individualized follow-up of patients *with a history* of a PPGL depending on the underlying mutation status and disease characteristics

Follow-up (*history* of a PPGL)	*High risk*	*Intermediate risk*	*Low risk*
	History of metastatic PPGL, history of sympathetic PGL, *SDHA, SDHB,* and *FH* (*FH* limited data), *HIF2A/EPAS1*	History of head and neck PGL, history of high-risk PCC (noradrenergic, ≥5 cm, recurrent, multiple) *SDHAF2, SDHC, SDHD, VHL, NF1, MAX, TMEM127, RET* with high/moderate risk for PCC	History of low-risk PCC (adrenergic, <5 cm), *RET* with low risk for PCC
Clinical, biochemistry	6-12 months (for *HIF2A*/EPAS1including hematocrit)	12 months (6 months for high-risk PCC)	12 months
Imaging (MRI base of the skull to pelvis/ MRI base of the skull, neck, abdomen, pelvis plus low-dose contrast-enhanced chest CT, alternating, for cluster 1; MRI abdomen/pelvis for cluster 2)	12-24 months (with history of disease initially 12, then 12-24 months) 6-12 months for history of very large primary PPGLs or those with large necrosis, high Ki67, and vascular and lymphatic invasion	24-36 months for *SDHAF2, SDHC, SDHD* (24 months for S*DHD*),*VHL* At least every 5 years for *NF1, MAX, TMEM127, RET* (only abdominal/pelvic MRI)	Optional
Special cases	For *HIF2A/EPAS1:* optic fundus examination every 12 months; PCC ≥5 cm: preoperative staging with additional contrast-enhanced chest CT or functional imaging History of nonfunctioning PPGL: Alternating, MRI (base of the skull to pelvis)/MRI base of the skull/ neck/abdomen/pelvis plus low-dose contrast-enhanced chest CT every 24 months History of metastatic PPGL/ sympathetic PGL: functional imaging 3-6 months postsurgery, afterwards, alternating, yearly MRI (base of the skull to pelvis)/MRI base of the skull/neck/ abdomen/pelvis plus low-dose chest CT, possibly functional imaging every 24-36 months	*VHL* mutations: risk of renal cell cancer, consider abdominal MRI every 12 months; optic fundus examination every 12 months; CNS tumors, CNS MRI every 24-36 months; *RET* mutations: risk of primary hyperparathyroidism and medullary thyroid carcinoma (every 12 months calcitonin, calcium, PTH if applicable)	
Postsurgery	Clinical and biochemical follow-up 3-6 weeks after surgery (after recovery)		

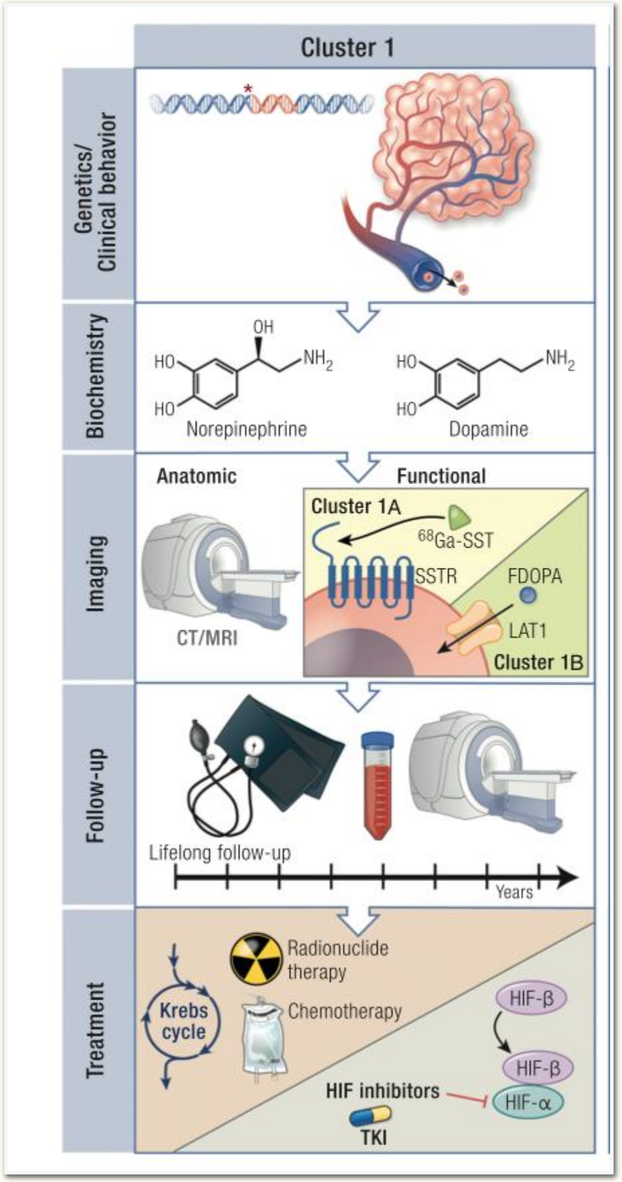
Endocrine Reviews,2022 vol43, No 2.

Surveillance of VHL manifestations

Surveillance Modality (Tumors being screened)	**AGE[1]**						**Pregnancy[11]**
	<5 years	**Beginning at age 5y**	**Beginning at age 11y**	**Beginning at age 15y**	**Beginning at age 30y**	**Beginning at age 65y[1]**	
History and Physical Examination[2]	Yearly from age 1 year	Yearly	Yearly	Yearly	Yearly	Yearly	Prior to conception[11]
Blood Pressure and Pulse (Pheochromocytomas/paragangliomas)	Yearly from age 2 years	Yearly	Yearly	Yearly	Yearly	Yearly	Prior to conceptionr[11]
Dilated Eye Examination[3] (Retinal Hemangioblastomas)	Every 6-12 months, beginning before age 1 year	Every 6-12 months	Every 6-12 months	Every 6-12 months	Yearly	Yearly	Prior to conception, then Every 6-12 months[11]
Metanephrines[4] (Pheochromocytomas/paragangliomas)	——	Yearly	Yearly	Yearly	Yearly	Stop routine[1]	Prior to conception[11]
MRI Brain and Spine w/wo Contrast[5,6,7] (CNS Hemangioblastomas)	——	——	Every 2 years[8]	Every 2 years[8]	Every 2 years[8]	Stop routine[1]	Prior to conception[11]
Audiogram (Endolymphatic sac tumors)	——	——	Every 2 years	Every 2 years	Every 2 years	Stop routine[1]	——
MRI Abdomen w/wo Contrast[5,6,7] (Renal cell carcinomas, Pheochromocytomas/paragangliomas, Pancreatic neuroendocrine tumors/cysts)	——	——	——	Every 2 years[9]	Every 2 years[9]	Stop routine[1]	Prior to conception[11]
MRI Internal Auditory Canal[10] (Endolymphatic sac tumors)	——	——	——	Once	——	——	No specific changes

VHL active-surveillance-guidelines , update 10/9/2020

VHL-related PPGLs



Cluster 1B: Pseudohypoxia signaling

Norepinephrine

Anatomical : CT/MRI
Functional : FDOPA PET/CT

Life long follow up

Surgery

Endocrine Reviews,2022 vol43, No 2.

Thank you