Case Report

U TUYẾN THƯỢNG THẬN TÁI PHÁT

BS Lê Văn Tài
Khoa Siêu Âm **MEDIC**

Tổng quan

- Tế bào ưa crôm (pheochromocyte) là các tế bào nội tiết thần kinh, có nguồn gốc từ mào thần kinh phân bố nhiều ở tủy thượng thận (90%) và tế bào cận hạch giao cảm (paraganglion).
- Pheochromocytoma (u tế bào ưa crôm) là u thần kinh nội tiết, u ở tủy thượng thận thường được gọi là pheochromocytoma, u ngoài tuyến thượng thận có tên là u tế bào cận hạch (paraganglioma) (pheochromocytoma ngoài tuyến thượng thận).
- U tiết quá mức catecholamine (adrenaline, oradrenaline) gây ra các triệu chứng nguy hiểm: CHA kịch phát, tam chứng điển hình: nhức đầu, vã mồ hôi, hồi hợp do tim đập nhanh.

- 30 – 40% liên quan đột biến di truyền: Đa u tuyến nội tiết type 2 (MEN 2A &2B), Von Hippel – Lindau, bệnh u sợi thần kinh loại 1 (NF1), H/C Paraganglioma – pheochromocytoma gia đình (PGL/PCC).
- U Ngoài tuyến thượng thận (paraganglioma): 10%, có chức năng & không có chức năng, ở cạnh cột sống, gặp nhiều dưới hoành, trên hoành khoảng 10%.
- Pheochromocytoma: được gọi là “u 10%”: 10 % ác tính, 10% ngoài tuyến thượng thận, 10% u có 2 bên, 10% có tính gia đình, 10% ở trẻ em.

Bệnh án

CÔNG TY TNHH Y TẾ HÒA HẢO - PHÒNG KHÁM ĐA KHOA
(Tên cũ: TRUNG TÂM CHẨN ĐOÁN Y KHOA - MEDIC)
254 Hòa Hảo, P.Vườn Lài, TP. Hồ Chí Minh
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PHIẾU KHÁM BỆNH

PHÒNG KHÁM CẤP CỨU - PHÒNG: CẤP CỨU 2

Họ tên: [redacted] Năm sinh: **1991 - Nữ** ĐT: **0902599799**

Địa chỉ: **911/1/6 LẠC LONG QUÂN- F. BÀY HIỀN- TPHCM**

Nghề nghiệp: Số thẻ BHYT:

HA - Mạch: / - ; Nhiệt độ: **37** °C;

Lý do khám: Kiểm tra sức khoẻ

CHỈ ĐỊNH:

CẬN LÂM SÀNG: (TỔNG SỐ: 2 mục)

Siêu Âm Bụng Tổng Quát Máu (Xin chú ý tuyến thượng thận)
; Siêu Âm Tuyến Giáp màu

XÉT NGHIỆM: (TỔNG SỐ: 9)

Giờ	Người lấy mẫu
____:____	BA XÉT NGHIỆM K. Phượng

☐ Máu ☐ Nước tiểu ☐ Đàm ☐ Phân ☐ Dịch ☐ Khác

HÀNG MỸ MỸ 100269 1991 F 29/11/25 09:02 PID: 8482628 Tăng lưng

Ion đồ chung	Free T4	Test bộ 3 Catecholamines/máu	
T3	Renine activity / máu	Cortisol / Máu	
TSH (Thế hệ 3)	Aldosterone (máu) (Liaison)	ACTH	

CĐ BỔ SUNG:

(Số HĐ: 5066660, Số tiền: 2,390,000 đ)

Ngày 29 tháng 11 năm 2025 - 08:58

Bác sĩ

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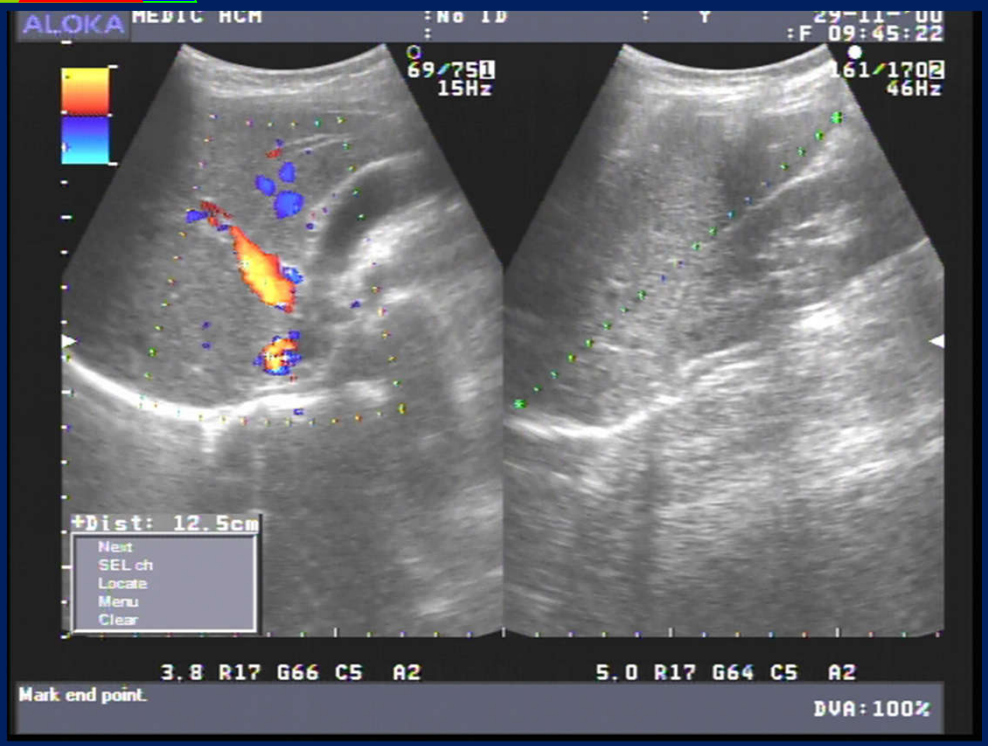
- Bệnh nhân nữ 34 tuổi, ở thành phố HCM.
- Lâm sàng:

 - Mệt, nhức đầu, tim đập nhanh, HA tăng (16/12 cmHg).

 - Tiền sử: Mổ nội soi u tuyến thượng thận (P) 18 năm (BV Chợ Rẫy).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69/75
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161/170
46Hz
+Dist: 12.5cm
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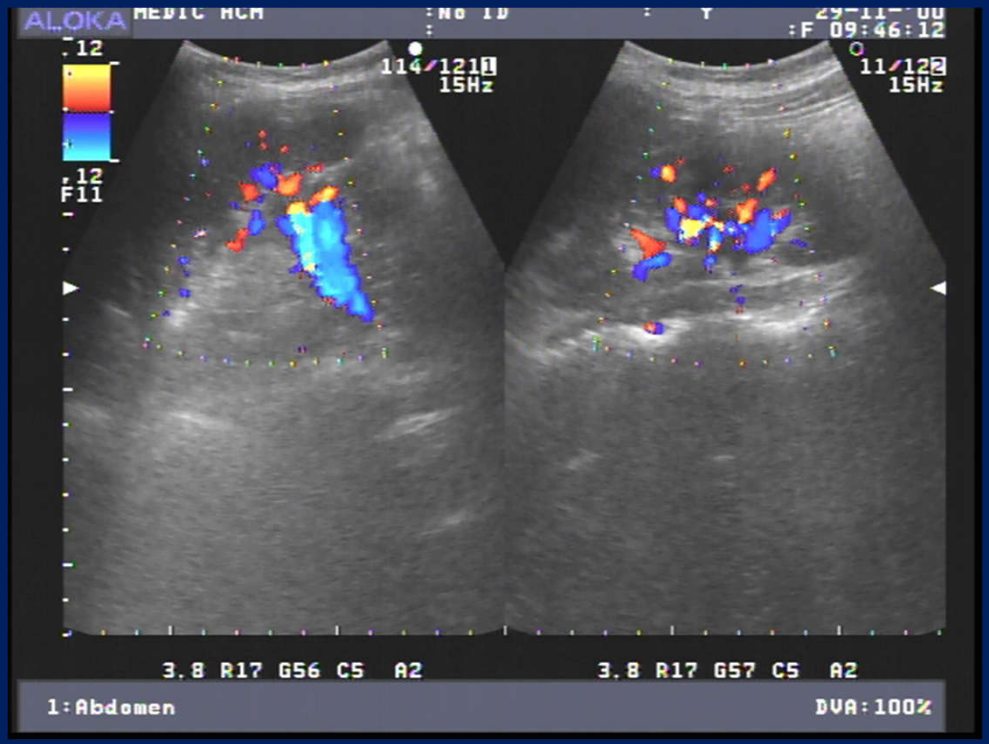
5.0 R17 G64 C5 A2
Mark end point.
DVA:100%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114/121
15Hz
11/12
15Hz
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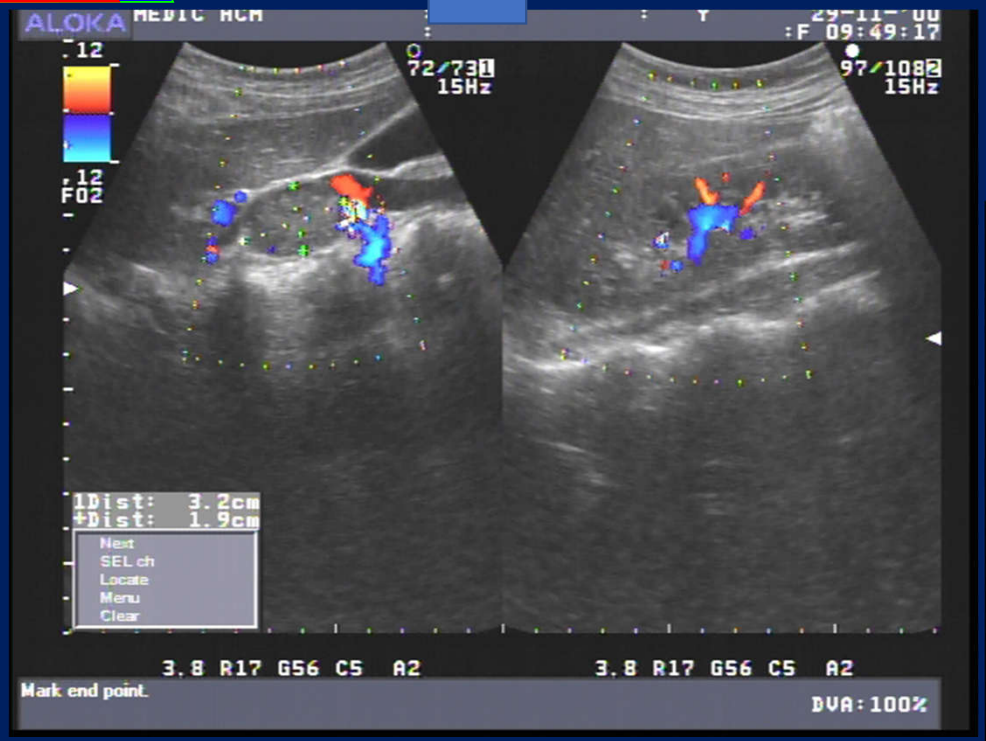
DVA:100%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72/73 1
15Hz
97/108 2
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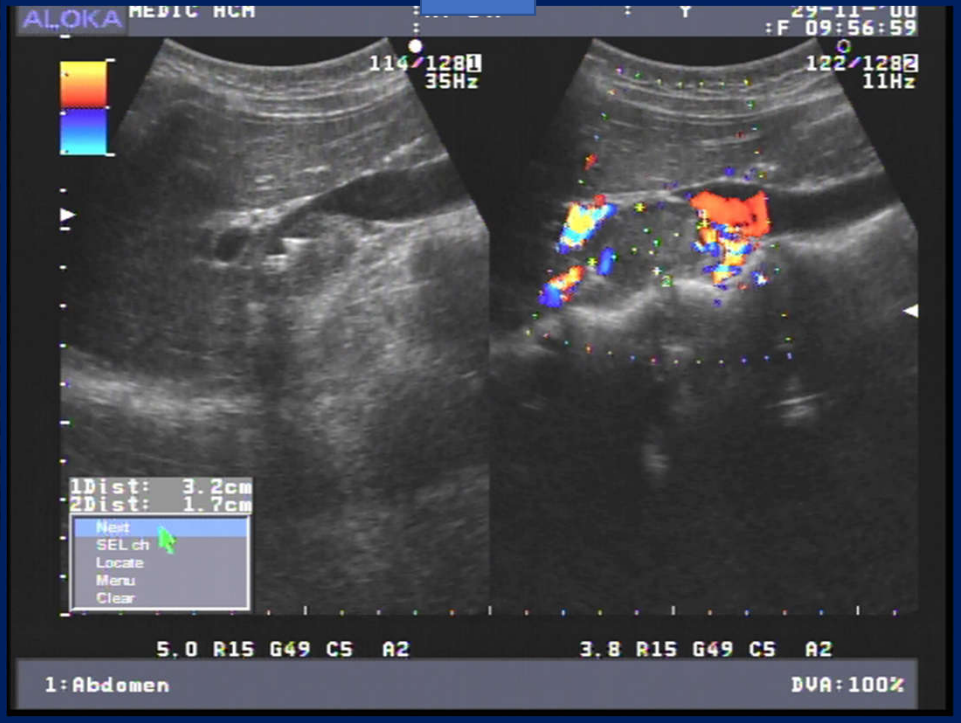
DVA:100%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35Hz
122/128 2
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2Dist: 1.7cm
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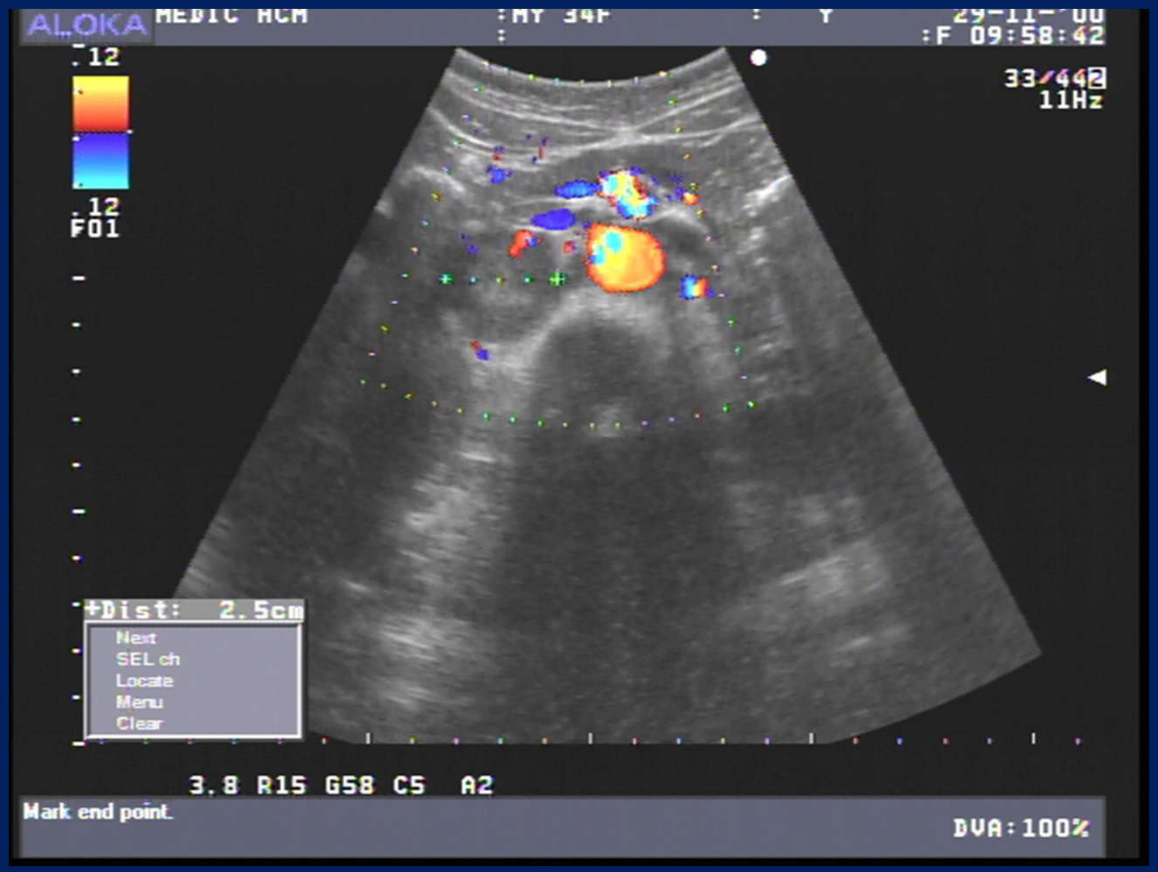
1:Abdomen
DVA:100%



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8482628

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Máy: **ALOKA -ProSound SSD4000**

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Họ và tên : **34** tuổi **Nữ**
Địa chỉ :
Chẩn đoán sơ bộ : NHỨC ĐẦU, TIM ĐẬP NHANH MỆT, THA 16/12 CMHG. TC: MỔ U TUYẾN THƯỢNG THẬN (P) 18 NĂM.
BS chỉ định : BS. LÝ VĂN KHẢI BV chỉ định : MEDIC

VÙNG KHẢO SÁT : SIÊU ÂM BỤNG TỔNG QUÁT MÀU

- GAN: Không to, bờ đều, cấu trúc đồng dạng, không sang thương khu trú.
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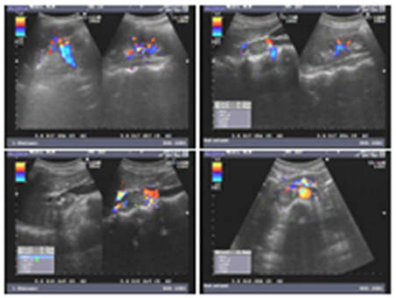
- TUY: Cấu trúc, kích thước bình thường. LÁCH: không to, đồng dạng.
- THẬN P: không sỏi, không ứ nước. THẬN T: không sỏi, không ứ nước.
- CÓ U TUYẾN THƯỢNG THẬN (P) TÁI PHÁT ECHO HỖN HỢP D = 32 X 19 MM, U NẰM GẦN ĐƯỜNG GIỮA, SÁT U CÓ VÀI CLIP.
- BÀNG QUANG: không sỏi, không bướu, vách mỏng.
- TỬ CUNG: Đồng nhất. BUỒNG TRỨNG (P), (T): Không u.
- Động mạch chủ bụng không phình.
- Ascites (-). Không hạch ổ bụng. Không tràn dịch màng phổi.



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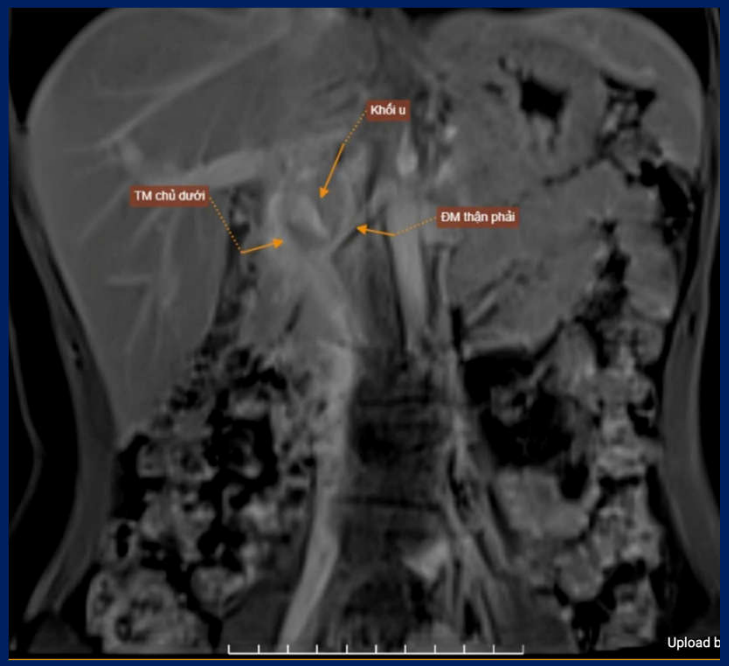
Đề nghị : **MSCT BỤNG.**

Tp. Hồ Chí Minh, ngày 29/11/2025 09:51
(Bác sĩ đã ký)

Bs. CKI. Lê Văn Tài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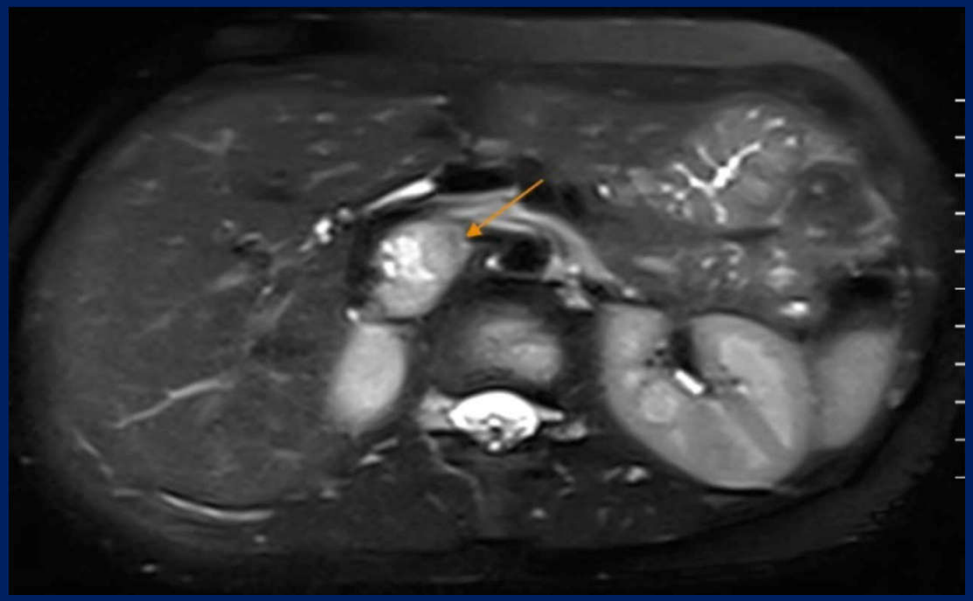

Bàn luận

- Tỉ lệ tái phát 6,5 – 23 % sau phẫu thuật 5 -15 năm.
- Tái phát tại chỗ đa phần do ung thư (không do ung thư: mổ làm rơi tế bào u, cắt không hết u), di căn xương, phổi, hạch.
- Chưa có phương pháp nào để xác định chính xác lành ác. Do đó phải theo dõi suốt đời sau mổ, tái khám hàng năm.
- Yếu tố nguy cơ tái phát:

- U ở người trẻ, kích thước > 5 cm.
- Tỉ lệ ác tính: paraganglioma (> 30%) cao hơn pheochrocytoma (< 10%).
- U do đột biến gen (SDHB: Succinate Dehydrogenase complex iron sulfur subunit B).

• Chẩn đoán:

- xét nghiệm máu, nước tiểu đo catecholamine, metanephrine.

- Siêu âm, CT, MRI, PET

• Điều trị:

- Phẫu thuật, hóa trị, xạ trị, điều trị trúng đích

- Chuẩn bị tiền phẫu: thuốc hạ áp (chẹn alpha), kiểm soát nhịp tim (ức chế beta).

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Diagnosis

Diagnosing pheochromocytoma involves a combination of biochemical tests and imaging studies. The primary biochemical tests include measuring plasma free metanephrines or urinary metanephrines, which are metabolites of catecholamines. Plasma-free metanephrines have a sensitivity of up to 99% and are considered the most reliable initial test.
Imaging studies are used to localize the tumor once biochemical tests confirm the diagnosis. Common imaging modalities include:

- **Computed Tomography (CT) Scan:** Provides detailed images of the adrenal glands and surrounding tissues.
- **Magnetic Resonance Imaging (MRI):** Preferred for children and pregnant women due to the absence of ionizing radiation.
- **Metaiodobenzylguanidine (MIBG) Scintigraphy:** Uses a radioactive compound that is taken up by pheochromocytoma cells, helping to identify the tumor location.
- **Positron Emission Tomography (PET) Scan:** Emerging as a promising technique for detecting and localizing pheochromocytomas.

Genetic testing is also recommended, especially for patients with a family history of pheochromocytoma or related genetic disorders such as MEN2, VHL syndrome, and NF1.

Cleveland Clinic

Most pheochromocytomas are benign (not cancerous). Approximately 10% to 15% of pheochromocytomas may be malignant (cancerous). There's no standard staging system for pheochromocytoma if it's cancerous. Instead, it's described as the following:

- **Localized pheochromocytoma:** The tumor is in one or both adrenal glands only.
- **Regional pheochromocytoma:** The cancer has spread to lymph nodes or other tissues near your adrenal glands.
- **Metastatic pheochromocytoma:** The cancer has spread to other parts of your body, like your liver, lungs, bone or distant lymph nodes.
- **Recurrent pheochromocytoma**: The cancer has recurred (come back) after it has been treated. It may come back in the same place or in another part of your body.

What causes pheochromocytoma?

In most cases of pheochromocytoma, the exact cause is unknown, and it occurs randomly.

Approximately 25% to 35% of people who have pheochromocytoma have a hereditary condition (passed through the family) that's linked to pheochromocytoma, including:

- Multiple endocrine neoplasia 2 syndrome, types A and B (MEN2A and MEN2B).
- Von Hippel-Lindau (VHL) disease.
- Neurofibromatosis type 1 (NF1).
- Hereditary paraganglioma syndrome.
- Carney-Stratakis dyad [paraganglioma and gastrointestinal stromal tumor (GIST)].
- Carney triad (paraganglioma, GIST and pulmonary chondroma).

Pheochromocytomas may also be caused by mutations (changes) of one of at least 10 different genes.

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Cureus. 2020 May 28;12(5):e8328. doi: 10.7759/cureus.8328

Recurrence of Pheochromocytoma With Metastases After Resection of Primary Tumor

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Editors: Alexander Muacevic, John R Adler

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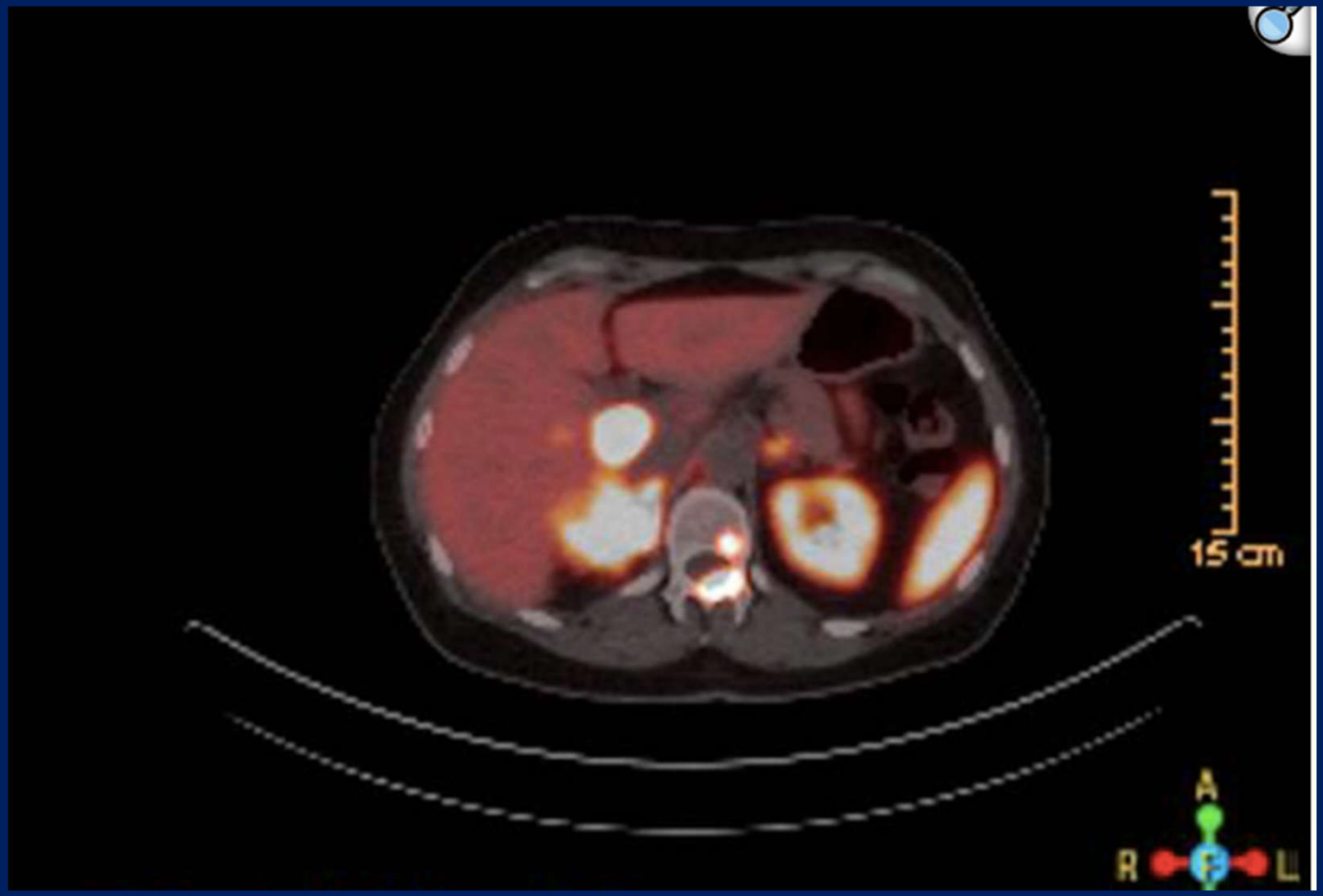
PMCID: PMC7255549 PMID: 32489750

Abstract

Pheochromocytomas and paragangliomas are rare tumors that arise from the chromaffin cells of the adrenal medulla or sympathetic paravertebral ganglia, respectively. Long-term surveillance is recommended regardless of the thoroughness of surgical resection. Here, we present a patient who was diagnosed with pheochromocytoma who underwent right adrenalectomy and was lost to follow up. She presented 15 years later with recurrence and was found to have multiple metastases. Subsequent genetic testing was also negative.

A small study of 192 patients showed that age, familial disease, tumor site (right-sided and extra-adrenal tumors), and size were independent predictors of recurrence. In the same study, pheochromocytoma recurred in 29 patients, out of which only 15 cases were malignant [5]. Timing of recurrence is also extremely variable, with some recurrences of metastatic disease having been discovered as long as 53 years after the initial surgery [6].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Hypertension Research (2020) 43:500–510
https://doi.org/10.1038/s41440-019-0339-y

ARTICLE

Predictors of recurrence of pheochromocytoma and paraganglioma: a multicenter study in Piedmont, Italy

Mirko Parasiliti-Caprino[1] · Barbara Lucatello[1] · Chiara Lopez[1] · Jacopo Burrello[2] · Francesca Maletta[3] · Marinella Mistrangelo[4] · Enrica Migliore[5] · Francesco Tassone[6] · Antonio La Grotta[7] · Anna Pia[8] · Giuseppe Reimondo[8] · Roberta Giordano[9] · Giuseppe Giraudo[10] · Alessandro Piovesan[11] · Giovannino Ciccone[5] · Dèsirée Deandreis[12] · Paolo Limone[13] · Fabio Orlandi[14] · Giorgio Borretta[6] · Marco Volante[15] · Paolo Mulatero[2] · Mauro Papotti[3] · Gianluca Aimaretti[16] · Massimo Terzolo[8] · Mario Morino[10] · Barbara Pasini[17] · Franco Veglio[2] · Ezio Ghigo[1] · Emanuela Arvat[11] · Mauro Maccario[1]

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Abstract

The available data on the natural history of pheochromocytomas and paragangliomas after radical surgery are heterogeneous and discordant. The aim of our retrospective multicenter study was to find predictors of recurrence in patients with pheochromocytomas and sympathetic paragangliomas submitted to radical surgery in Piedmont (a region in northwest Italy). We collected data from 242 patients diagnosed between 1990 and 2016. Forty-two patients (17.4%) had disease recurrence. Multivariate analysis showed that genetic mutation (HR = 3.62; 95% CI 1.44−9.13; $p = 0.006$), younger age (HR = 0.97; 95% CI 0.95−0.99; $p = 0.031$) and larger tumor size (HR = 1.01; 95% CI 1.00−1.02; $p = 0.015$) were independently associated with a higher recurrence risk of pheochromocytoma and paraganglioma; in pheochromocytomas, genetic mutation (HR = 3.4; 95% CI 1.00−11.48; $p = 0.049$), younger age (HR = 0.97; 95% CI 0.94−0.99; $p = 0.02$), higher tumor size (HR = 1.01; 95% CI 1.00−1.03; $p = 0.043$) and PASS value (HR = 1.16; 95% CI 1.03−1.3; $p = 0.011$) were associated with recurrence. Moreover, tumor size was the only predictor of metastatic pheochromocytoma and paraganglioma (HR = 4.6; 95% CI 1.4−15.0; $p = 0.012$); tumor size (HR = 3.93; 95% CI 1.2−16.4; $p = 0.026$) and PASS value (HR = 1.27; 95% CI 1.06−1.53; $p = 0.007$) were predictors of metastatic pheochromocytoma. In conclusion, our findings suggest that the recurrence of pheochromocytoma and sympathetic paraganglioma develops more frequently in younger subjects, patients with a family history of chromaffin tissue neoplasms, mutations in susceptibility genes, larger tumors and higher values of PASS. We recommend genetic testing in all patients with PPGL and strict follow-up at least on an annual basis.

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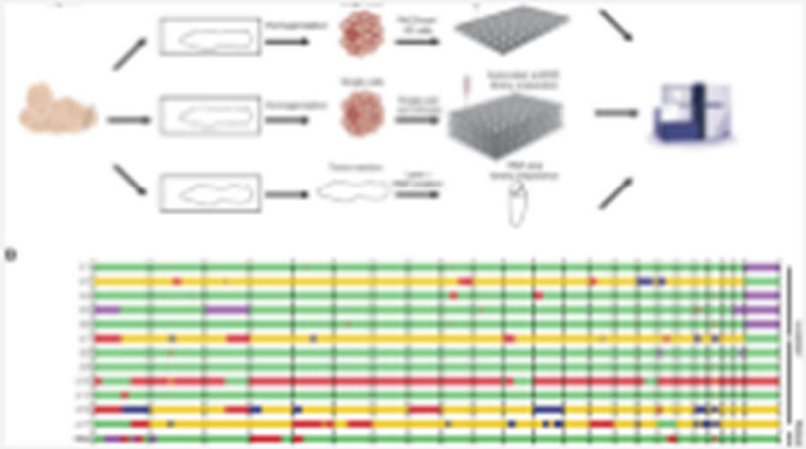

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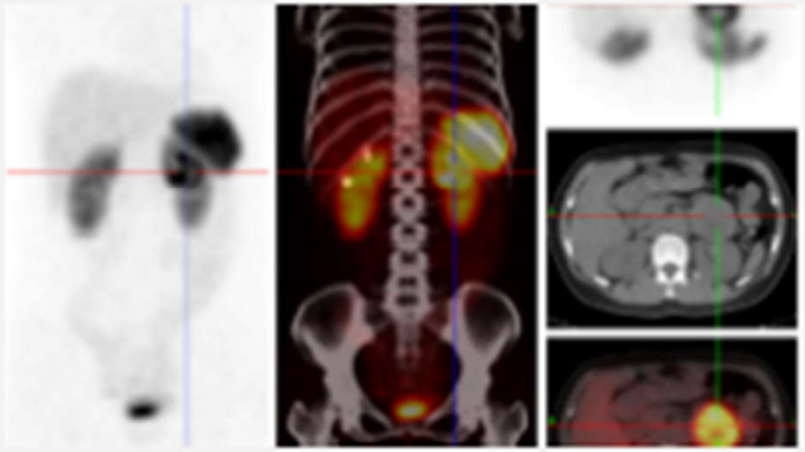

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Sec. Adrenal Endocrinology

Volume 14 - 2023 | https://doi.org/10.3389/fendo.2023.1279828

Local recurrence and metastatic disease in pheochromocytomas and sympathetic paragangliomas

Marta Araujo-Castro [1,2*†] Iñigo García Sanz [3]

César Mínguez Ojeda [4] Felicia Hanzu [5] Mireia Mora [5]

Purpose: To evaluate the rate of recurrence among patients with pheochromocytomas and sympathetic paragangliomas (PGLs; together PPGLs) and to identify predictors of recurrence (local recurrence and/or metastatic disease).

Methods: This retrospective multicenter study included information of 303 patients with PPGLs in follow-up in 19 Spanish tertiary hospitals. Recurrent disease was defined by the development of local recurrence and/or metastatic disease after initial complete surgical resection.

Results: A total of 303 patients with PPGLs that underwent 311 resections were included (288 pheochromocytomas and 15 sympathetic PGLs). After a median follow-up of 4.8 years (range 1-19), 24 patients (7.9%) had recurrent disease (3 local recurrence, 17 metastatic disease and 4 local recurrence followed by metastatic disease). The median time from the diagnosis of the PPGL to the recurrence was of 11.2 months (range 0.5-174) and recurrent disease cases distributed uniformly during the follow-up period. The presence of a pathogenic variant in *SDHB* gene (hazard ratio [HR] 13.3, 95% CI 4.20-41.92), higher urinary normetanephrine levels (HR 1.02 per each increase in standard deviation, 95% CI 1.01-1.03) and a larger tumor size (HR 1.01 per each increase in mm, 95% CI 1.00-1.02) were independently associated with disease recurrence.

Conclusion: The recurrence of PPGLs occurred more frequently in patients with *SDHB* mutations, with larger tumors and with higher urinary normetanephrine levels. Since PPGL recurrence may occur at any time after the initial PPGL diagnosis is performed, we recommend performing a strict follow-up in all patients with PPGLs, especially in those patients with a higher risk of recurrent disease.

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Case Report

Recurrent Pheochromocytoma With Bone Metastasis Eight Years After Bilateral Adrenalectomies in a Patient With Neurofibromatosis Type 1

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pheochromocytoma recurrence

ABSTRACT

Background/Objective: Pheochromocytoma can recur years after curative surgical resection. Rarely, it may reoccur as metastasis. Here, we present a case of metastatic pheochromocytoma to the bones in a patient with neurofibromatosis type 1 (NF1), 8 years after initial resection of primary bilateral adrenal pheochromocytomas without metastases.

Case Report: A 44-year-old woman presented with diffuse body pain and palpitations. Her past medical history included NF1 and hypertension. Eight years prior to her current presentation, she had undergone a bilateral adrenalectomy for the management of bilateral adrenal pheochromocytomas. Her plasma metanephrines normalized after surgery and remained normal at her 1-year postoperative visit. She was subsequently lost to follow-up until her current presentation. Our evaluation revealed significantly elevated urine and plasma metanephrines as well as innumerable DOTATATE avid lesions along the axial and perpendicular spine compatible with a metastatic neuroendocrine tumor. She was started on doxazosin and metoprolol and discharged home with a plan to be seen by Oncology to discuss systemic therapy.

Discussion: Predicting malignant disease in patients with primary tumors without metastases is challenging. There is no single factor that can reliably predict tumor behavior. It is unknown if individuals with NF1, who have a genetic predisposition for developing pheochromocytomas, are at an increased risk of malignant disease.

Conclusion: Due to a lack of accurate predictors, annual biochemical testing is recommended after primary tumor resection and in patients with a genetic predisposition. Strict lifelong follow-up should be strongly considered due to a possible higher risk of malignant disease.

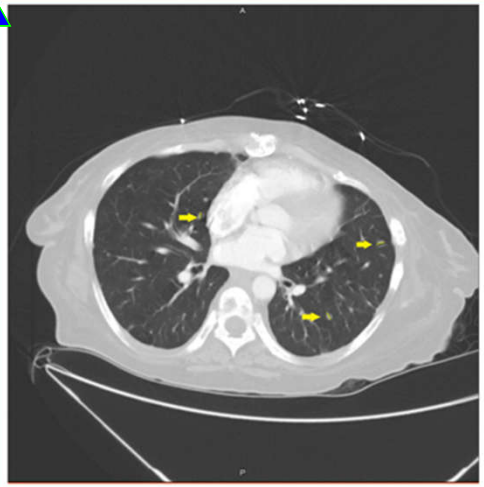

Fig. 1. CT scan of the chest demonstrated multiple pulmonary nodules (yellow arrows). *CT* = computed tomography.

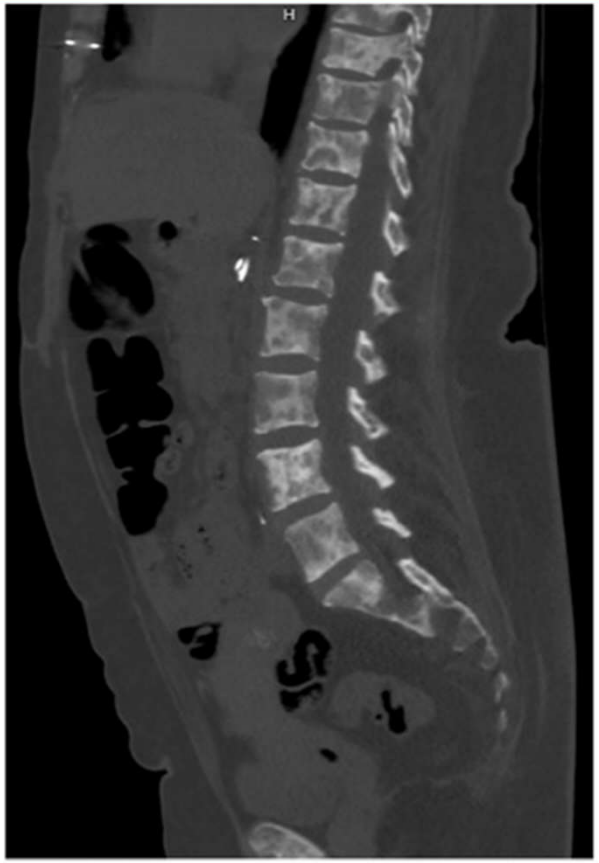

Fig. 2. Mixed lucent and sclerotic lesions Involving the vertebrae noted from CT scan. *CT* = computed tomography.

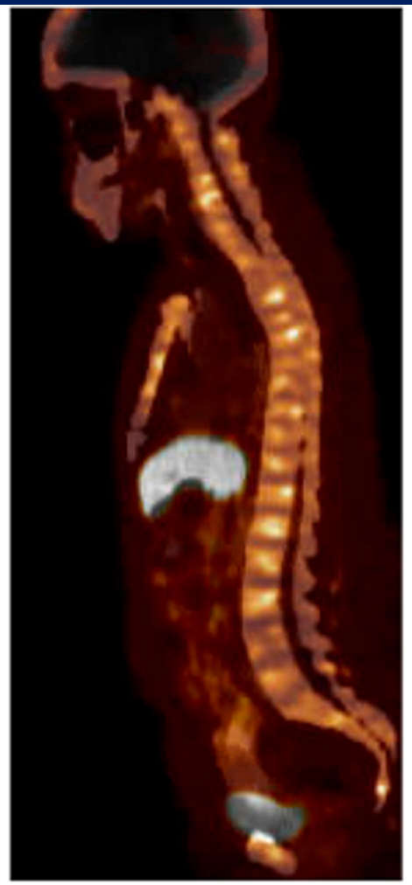

Fig. 3. Ga-68-DOTA PET/CT with innumerable DOTATATE avid lesions along the bones. *CT* = computed tomography.

Kết luận

- Cần nghĩ đến pheochromocytoma tái phát tại chỗ hay di căn khi khám bệnh nhân đã phẫu thuật u trước đó, nay có triệu chứng lâm sàng gợi ý u tái phát.
- Siêu âm chi phí thấp, luôn sẵn có tuy nhiên dễ bỏ sót u, nên kết hợp CT, MRI nhất là trong trường hợp lâm sàng nghi u tái phát, siêu âm âm tính.
- Cần theo dõi định kỳ hàng năm sau mổ u để phát hiện sớm u tái phát.


CÁM ƠN!