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INTRODUCTION

Pheochromocytomas are rare neuroendocrine tumors that can occur sporadically or, in about 30% of cases, in the context of family syndromes. Ten percent are malignant. Type 1 neurofibromatosis is an autosomal dominant disease that is associated with the occurrence of these tumors. Pheochromocytomas appear in 0.1 to 5.7 % of patients with type 1 neurofibromatosis, and are usually solitary and benign lesions.

CASE REPORT

We present a case of a 50-years-old female, with a past history of type 1 neurofibromatosis, arterial hypertension, depressive syndrome, divergent strabismus and tabagism. Medicated with irbesartan 300mg, sertraline and alprazolam.

She was referenced to our service of Endocrinology due to a 6 cm nodular lesion in the right adrenal gland, detected in a renal CT.

On physical examination, she presented high blood pressure, *cafe-au-lait* spots and cutaneous neurofibromas. (image 1)



1. Cafe-au-lait spots and cutaneous neurofibromas

Adrenal functional study revealed high levels of plasma metanephrines, urinary fractionated catecholamines and metanephrines, and vanillylmandelic acid. (image 2)

The MRI showed a massive heterogeneous right adrenal gland mass with 48x48x63 mm, with hyperintense signal on T2-weighted images, internal cystic areas and hyperintensity on T1-weighted images that may correspond to hemorrhagic areas. This mass presents frank capture of contrast and marked restriction in diffusion weighting. These findings favour the diagnosis of pheochromocytoma. (images 3 and 4)

The body scintigraphy with I131-MIBG showed an image compatible with right adrenal pheochromocytoma. (image 5)

2. Adrenal functional study	Before surgery	After surgery (6 months)	Normal
Plasma metanephrines (nmol/L)			
Metanephrine	5,27	0,36	<0,46
Normetanephrine	6,13	0,44	<1,09
Vanillylmandelic acid (mg/24hr)	20,15	4,21	1,39-6,53
Urinary fractionated catecholamines (24hr urine)(µg/24hr)			
Adrenaline	308,82	1,17	0,01-19,96
Noradrenaline	199,42	14,73	15,06-80,03
Dopamine	611,94	18,82	64,93-400,0
Urinary fractionated metanephrines (24hr urine)(µg/24hr)			
Total metanephrines	4615,86	254,76	179,0-651,0
Metanephrine	2425,77	66,53	74,0-297,0
Normetanephrine	2190,09	188,23	105,0-354,0

Laparoscopic right adrenalectomy was performed, after appropriate preoperative therapy.

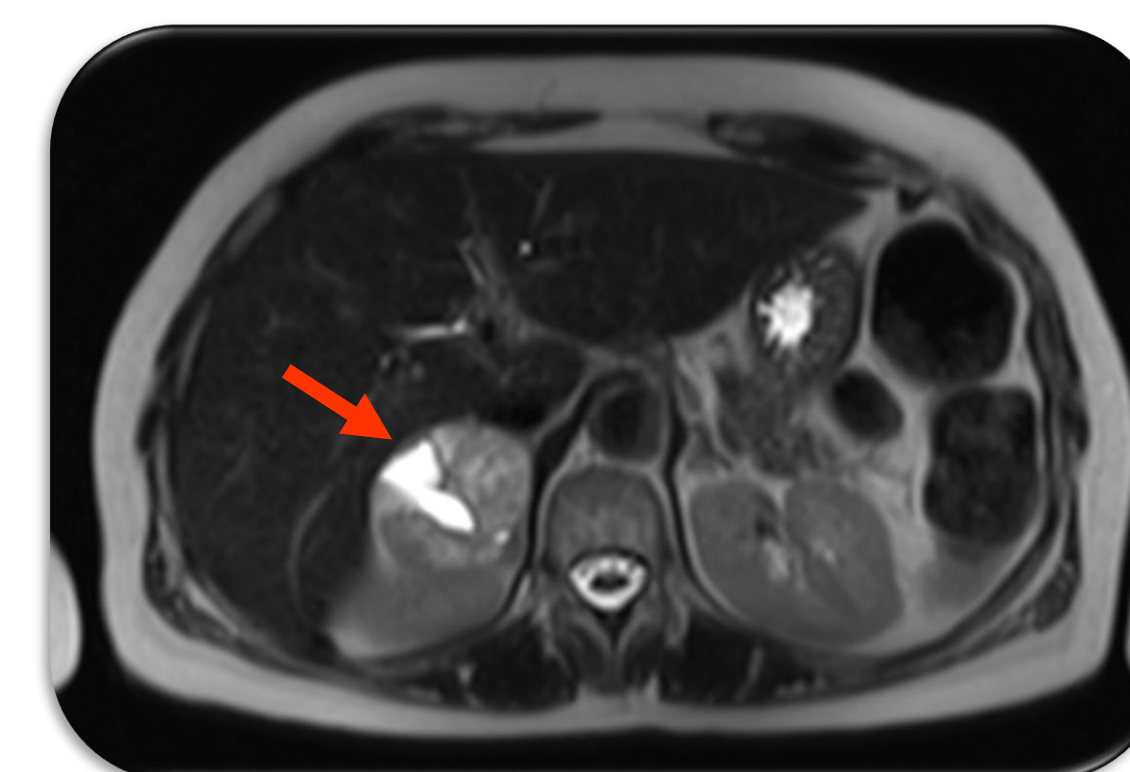
Histological examination revealed a pheochromocytoma with capsule invasion, adjacent adipose tissue invasion, but without vascular invasion.

After surgery, levels of plasma metanephrines, urinary fractionated catecholamines and metanephrines, and vanillylmandelic acid became normal.

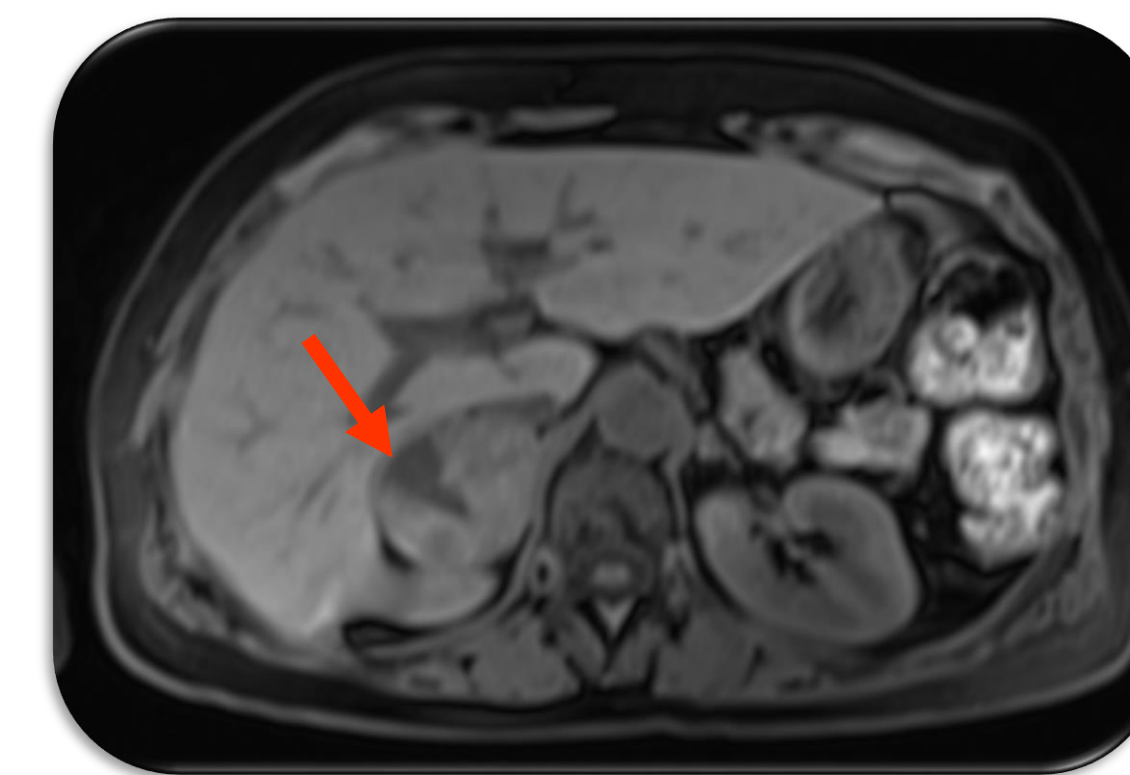
Currently, she has her arterial blood pressure controlled without medication.

CONCLUSION

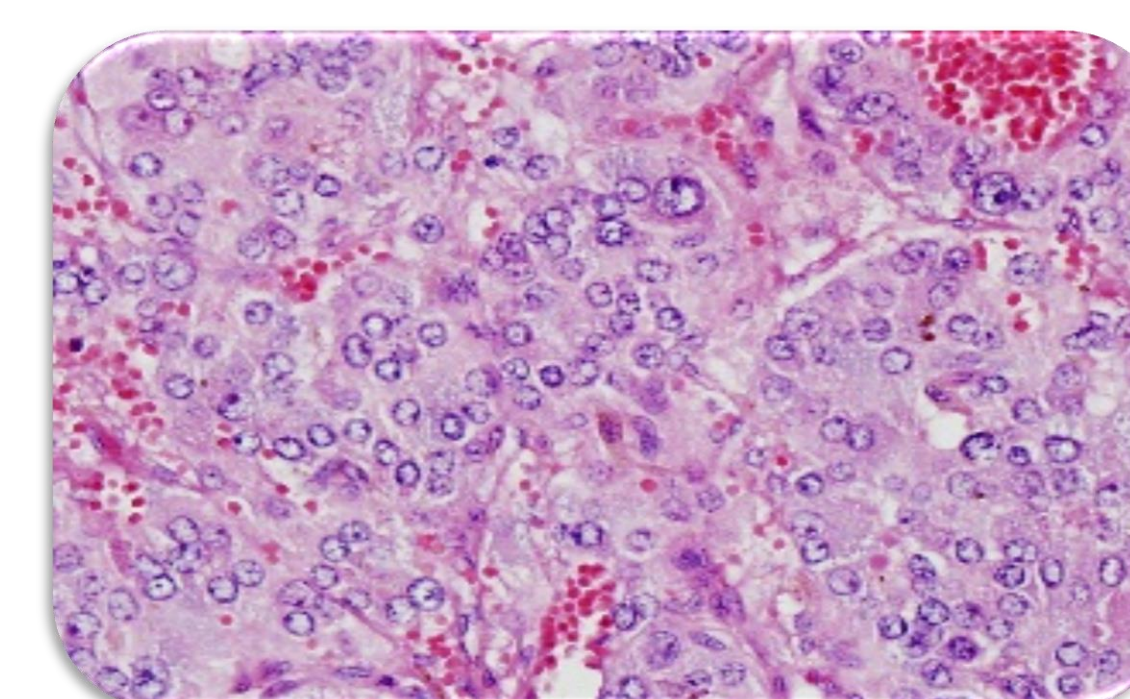
We consider important to present this case due to the rarity of the case, the possibility of malignancy, a hypothesis strengthened by tumor size, and the challenge of long-term follow up of these tumors.



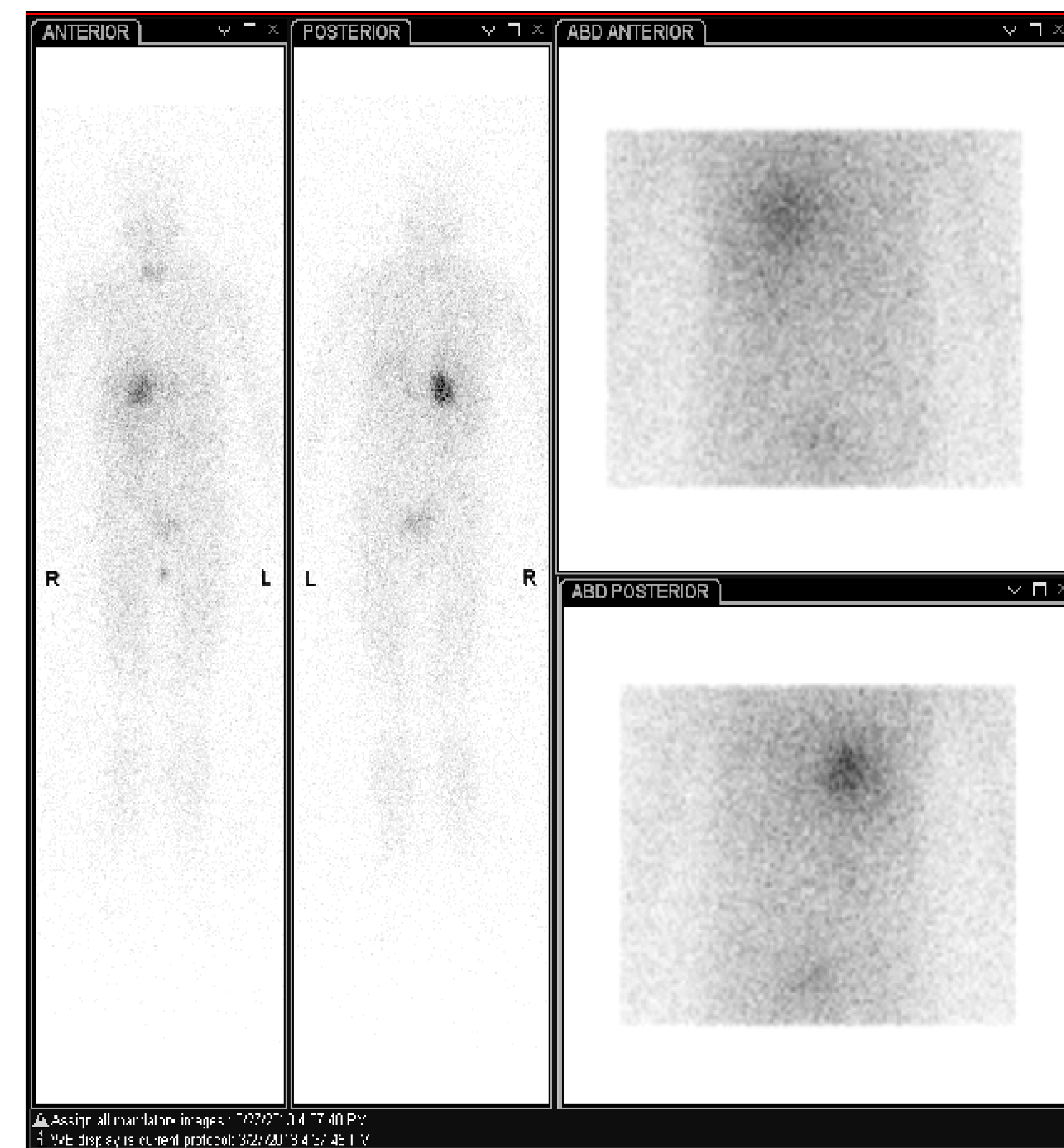
3. T2-weighted MRI upper abdomen



4. T1-weighted MRI upper abdomen



6. Histological examination (hematoxylin and eosin staining; magnification 400x) compatible with pheochromocytoma



5. Body scintigraphy with 131I-MIBG