

ADRENAL MALIGNANT MELANOMA MASQUERADING AS A PHEOCHROMOCYTOMA

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Introduction

- Pheochromocytomas represent tumors arising from the adrenal medulla. Their principal characteristic is the secretion of catecholamines (epinephrine, norepinephrine, dopamine).
- The adrenals are a common metastatic site of lung, breast, ovarian and skin cancer. Malignant melanoma originates principally from the skin and rarely from other tissues, like eye's choroid tissue, esophagus, colon, while sporadic cases of primary adrenal melanoma have also been described.

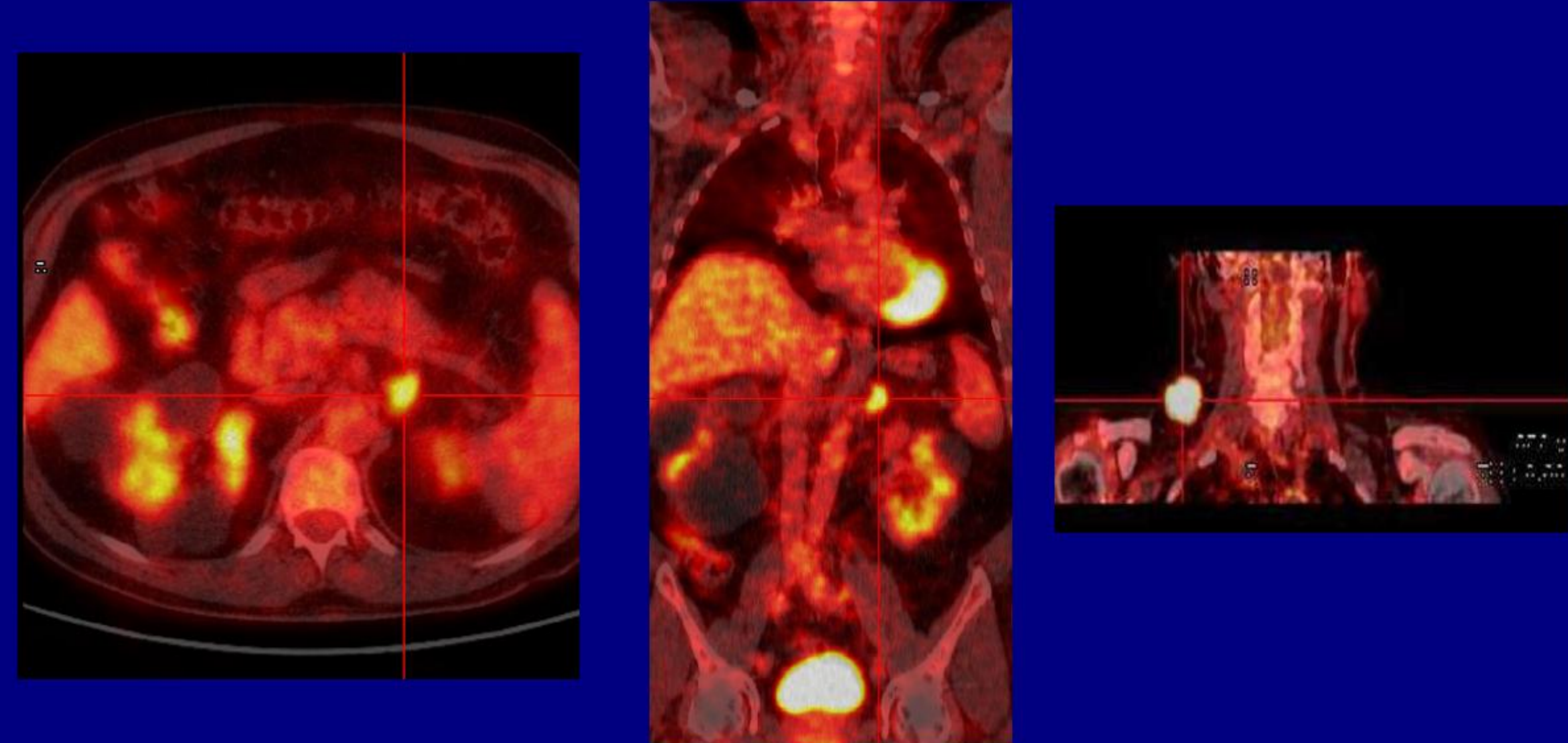
Case Description

- A 61-year-old male was referred for evaluation of a 5x5 cm, well defined, firm mass in the right supraclavicular region and another 88,5 mm inhomogeneous lesion in the left adrenal.
- He had a medical history of total thyroidectomy with cervical lymph node dissection because of a multifocal papillary and medullary thyroid carcinoma three years before presentation.
- Post-thyroidectomy imaging had been negative as was genetic testing for RET proto-oncogene mutation.
- The patient had no symptoms or signs of catecholamine excess. Apart from the palpable neck mass, the rest of the physical examination was unremarkable.

Laboratory tests (before and after adrenal dissection)

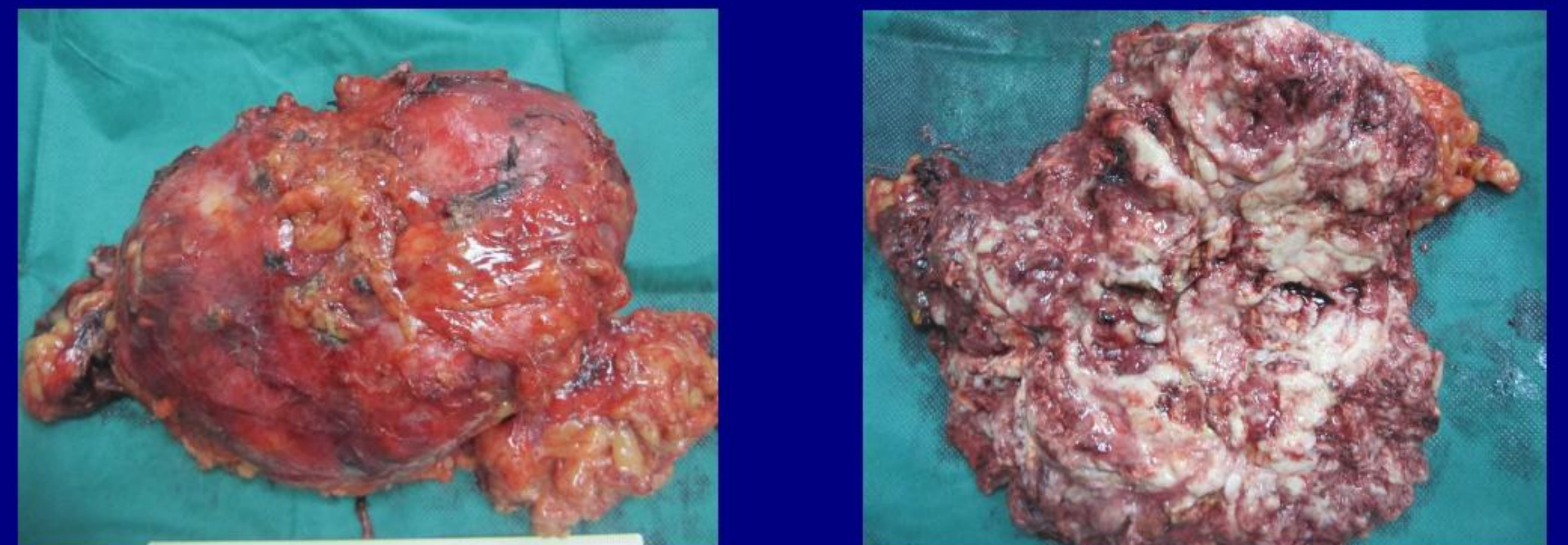
	Before	After	Reference range
Calcitonin	217 pg/ml	351 pg/ml	(<18)
Thyroglobulin	0,26 ng/mL		(<78)
AntiTg	<2,2 U/ml		(<20)
Chromogranin A	2,2 nmol/l		(<4)
Neuron Specific Enolase	79 µg/l	11,9 µg/l	(<16)
24h urinary catecholamine	817 µg/24h	432 µg/24h	(80- 515)
24h urinary epinephrine	8,8 µg/24h	10,8 µg/24h	(4- 25)
24h urinary norepinephrine	105,1 µg/24h	88,9 µg/24h	(20- 105)
24h urinary dopamine	703 µg/24h	332 µg/24h	(65- 400)
24h urinary VMA	6,2 mg/24h	3,9 mg/24h	(1- 11)

- Dopamine-secreting pheochromocytomas are extremely rare and have been associated with lack of the characteristic symptoms indicative of catecholamine excess and with greater rates of malignancy.

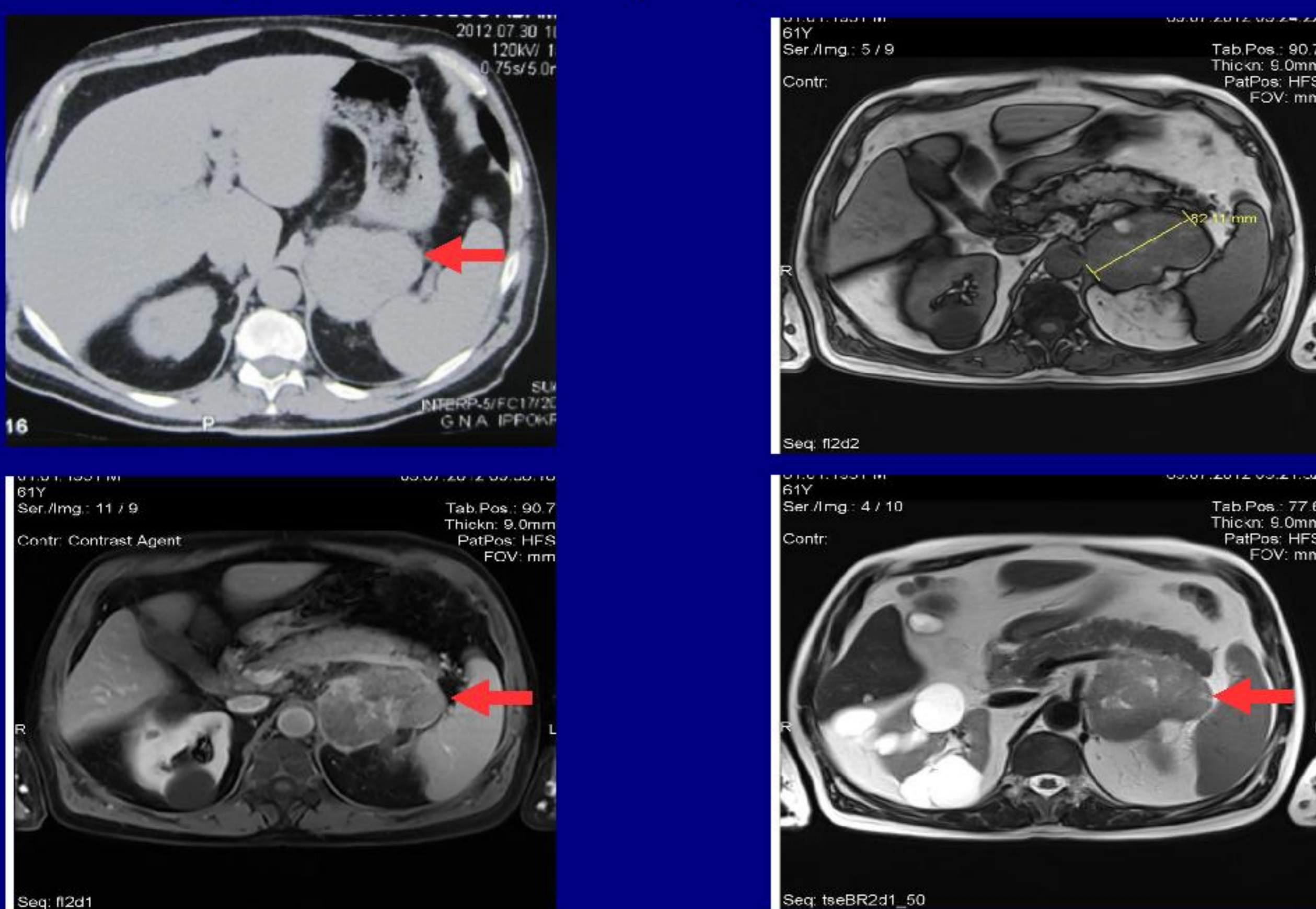


F18- FDG PET/CT 11,5 mCi:

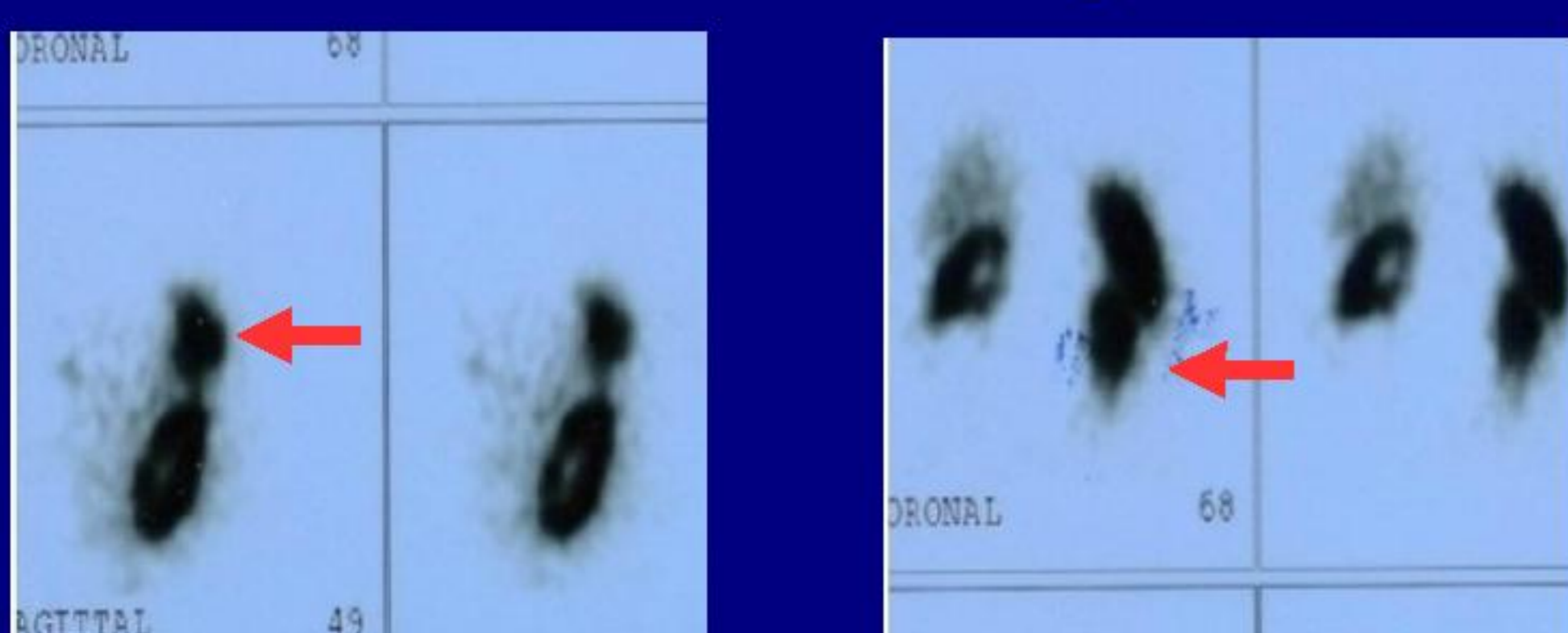
- Left adrenal gland (SUV max: 4,7).
- Right supraclavicular region (SUV max: 10,0).



- Tumor 16,5 x 10,7 x 10 cm in diameter which occupies the left adrenal gland.
- No infiltration of the capsule, the surrounding lymph nodes and organs.
- Poorly differentiated malignant neoplasm composed of spindle shaped or epithelioid cells, with eosinophilic cytoplasm, markedly atypical and pleomorphic nuclei with prominent nucleoli, abundant mitoses and areas of necrosis.
- Strong positivity for immunohistochemical markers S- 100 and Melan- A.

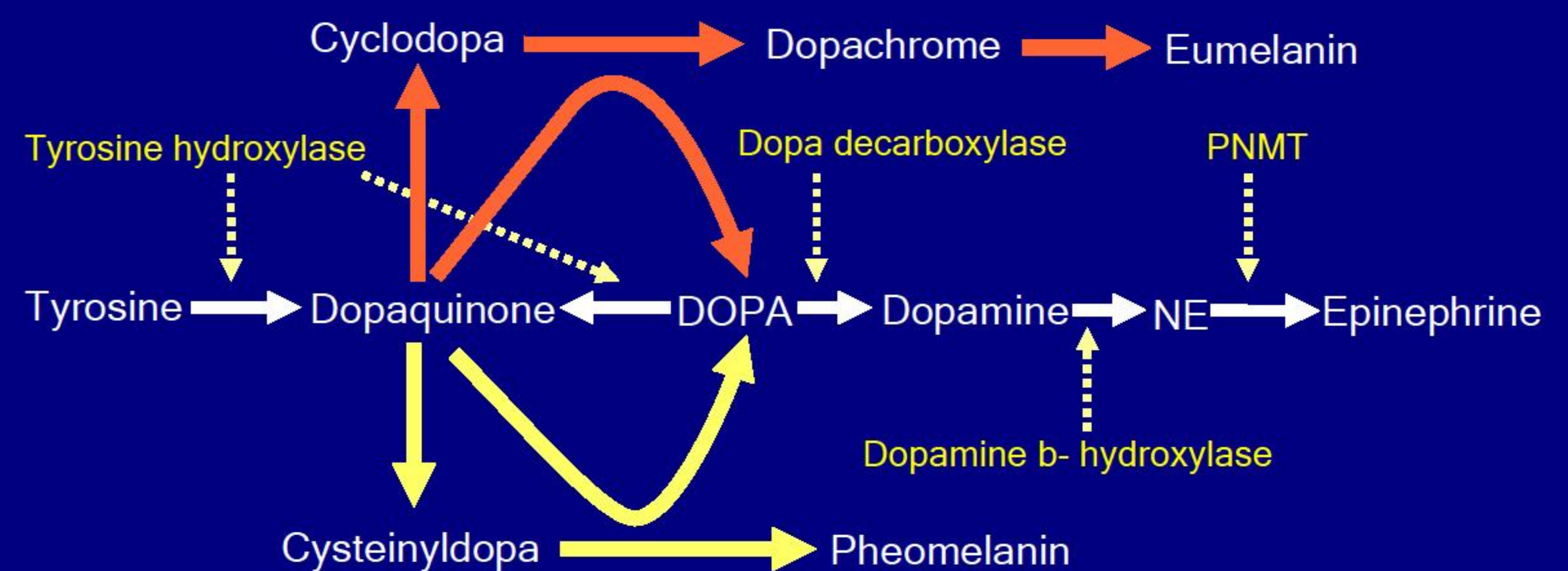


CT (top left): Inhomogeneous solid mass 8,5 cm in diameter with diffuse hemorrhagic and necrotic areas and increased tracer uptake .
MRI: large, solid mass 88,5 mm in diameter of the left adrenal gland, with inhomogeneous enhancement of the contrast agent and a low diffusion rate.



111In- Pentetreotide (OCTREOSCAN):

- Increased tracer uptake in the left adrenal gland
- ¹³¹I- MIBG : negative (not displayed).



Biosynthetic pathway of melanins and catecholamines:

- In melanoma tissue there is induction of tyrosine hydroxylase leading to production of DOPA and melanins. Activity of DOPA decarboxylase converts part of DOPA to 3,4- dihydroxyphenylethylamine (dopamine, DA).

Conclusions

This rare case illustrates the difficulties in the differential diagnosis of a dopamine-secreting adrenal mass. Both adrenal melanomas and pheochromocytomas should be considered in the differential diagnosis of an adrenal mass, especially when the primary biochemical finding is an increased urinary dopamine excretion in order to determine the best diagnostic approach to the patient.

References:

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