

PHEOCHROMOCYTOMA – THE RARE REASON OF CUSHING’S SYNDROME DUE TO ECTOPIC CORTICOTROPIN SECRETION.

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OBJECTIVES

Cushing syndrome due to ectopic (adrenocorticotrophic hormone) ACTH secretion (EAS) constitutes approximately 10% of Cushing’s syndrome (CS). In this group only in about 5% cases pheochromocytoma is the source of ACTH.

CASE 1

A 70 year-old woman with 3-months history of malaise, weakness, abdominal pain, loss of weight and appetite, hypertension and diabetes mellitus. One month earlier she had an episode of acute renal insufficiency after coronarography. Patient revealed mental confusion, cachexia, swelling of legs, skin with increased pigmentation, echymoses and petechiae. The laboratory data showed elevated leukocytosis, hyperglycemia, severe hypokalemia, and metabolic alkalosis (pH 7.661 [7.380 – 7.420], pCO₂ 34.7 mmHg [32.5 – 43.7], pO₂ 42.2 mmHg [75.0 – 95.0], HCO₃ 38.3 mmol/l [22.0 – 26.0], BE 16.7 mmol/l [-1.0 – 3.0]), markedly elevated serum cortisol concentration and ACTH without cortisol suppression after dexamethasone. Measurements of Metanephrine and Normetanephrine in urine were increased. MRI and somatostatin receptor scintigraphy (SRS) disclosed 40 mm mass in right adrenal gland. After pharmacological treatment patient underwent laparoscopic surgery. Histological examinations confirmed EAS and pheochromocytoma. Unfortunately in the 9-th day after surgery patient died because of ARDS syndrome.

parameter	WBC 10 ³ /ul [4.00 – 10.00]	Potassium mmol/l [3.5 – 5.1]	Sodium mmol/l [136.0 – 145.0]	ALT U/l [5 – 33]	GGTP U/l [5 – 36]	Ch-E U/l [5320 – 12920]	Urea mmol/l [2.76-8.07]	Creatynine Umol/l [44-80]
value	16.99	2.4 – 2.9 – 4.3	134.0 – 143.0	128	115	2537	5.8	75.0

parameter	Morning cortisol ug/dl [2.3- 20.9]	Midnight cortisol ug/dl [2.3- 20.9]	ACTH pg/ml [6.0 – 56.1]	VMA umol/24h [9-35]	MN ug/24h [52-341]	NMN ug/24h [88-440]	CgA nmol/l [0.00 – 6.00]
value	79 -80	70 -78	360 – 376	135.2	2997.2	1928.3	58.6

Tab.1. Laboratory results.

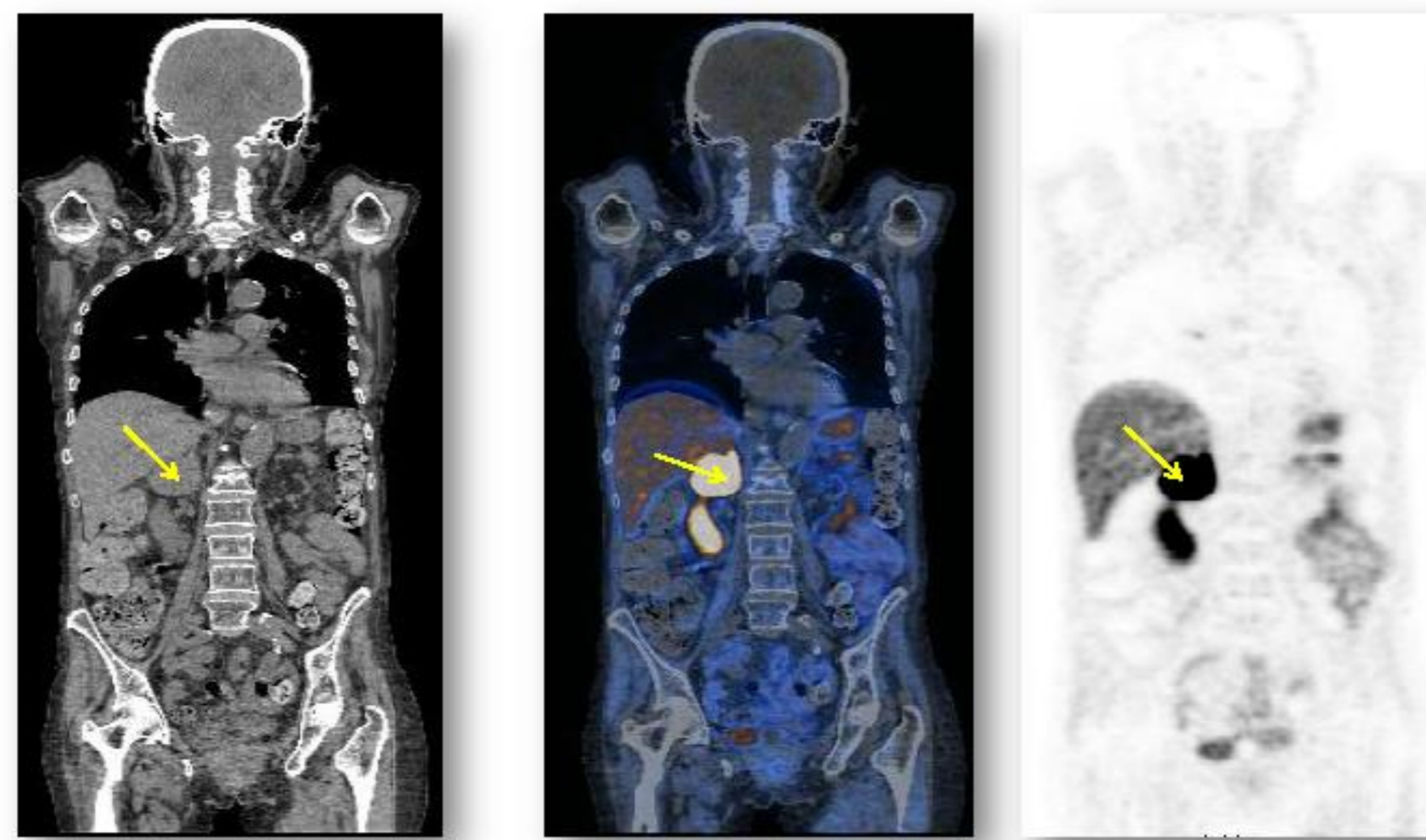


Fig.1. Scintigraphy examination with Ga-68-labelled somatostatin analogues.



Fig.2. MRI of the abdomen.



Fig. 3. MRI of the abdomen.



Fig.4. CT scan.

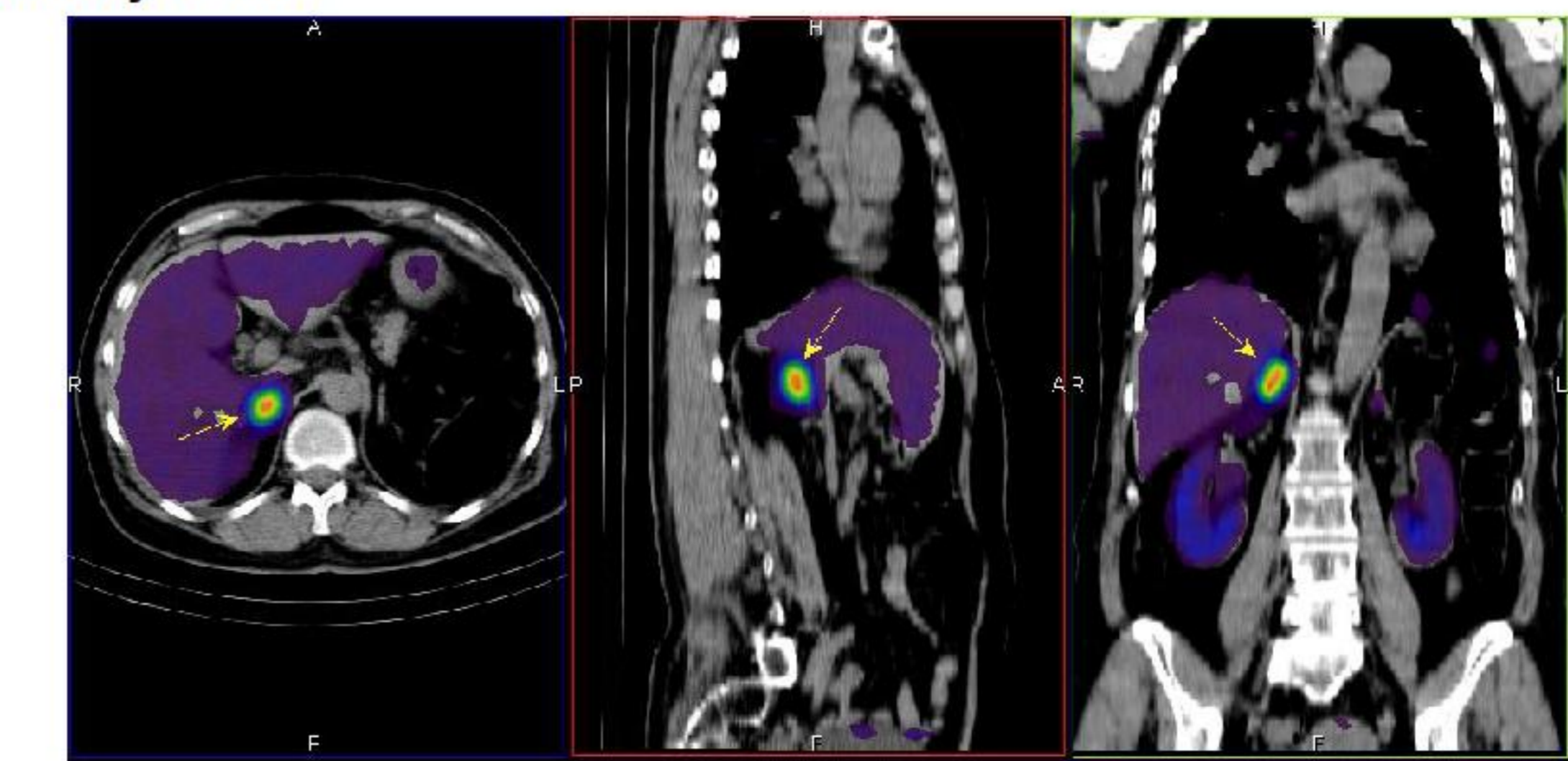


Fig.6. Scintigraphy examination with 99mTc-labelled somatostatin analogues.

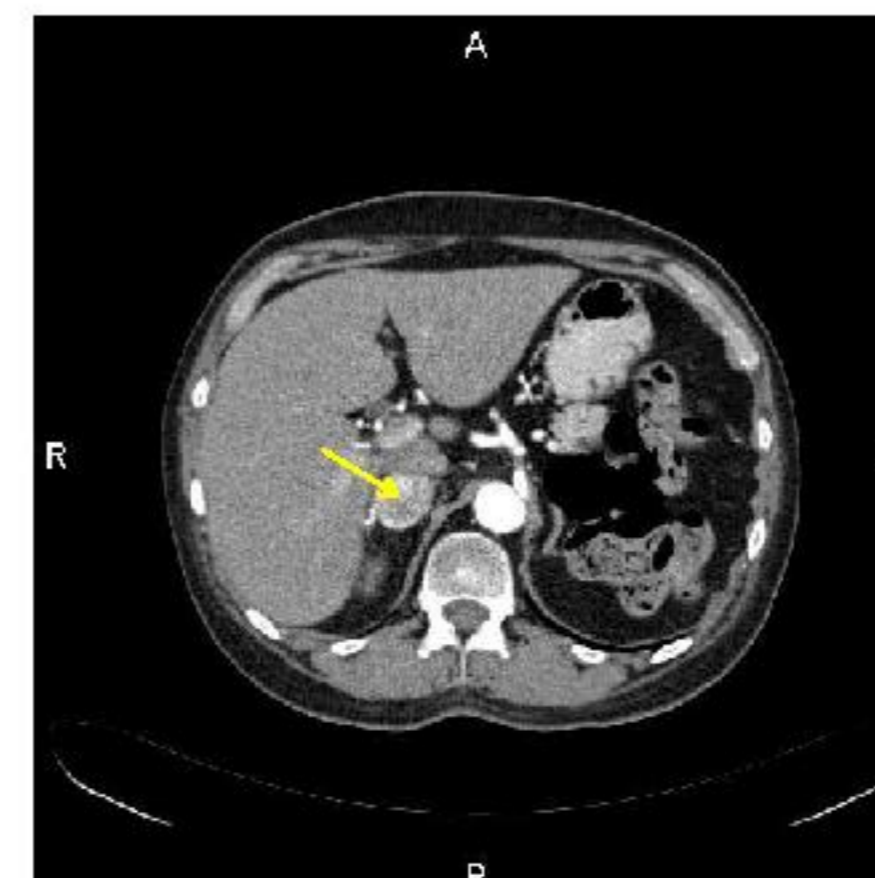


Fig.7. CT of the abdomen.

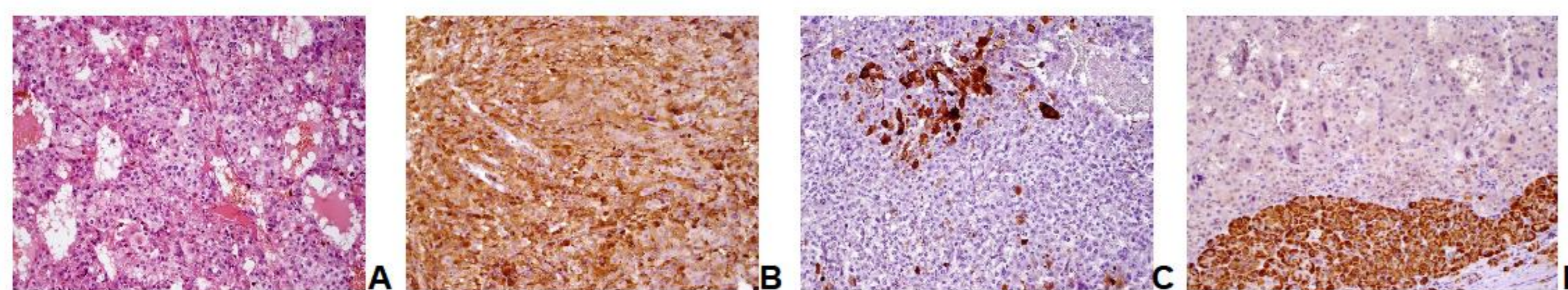
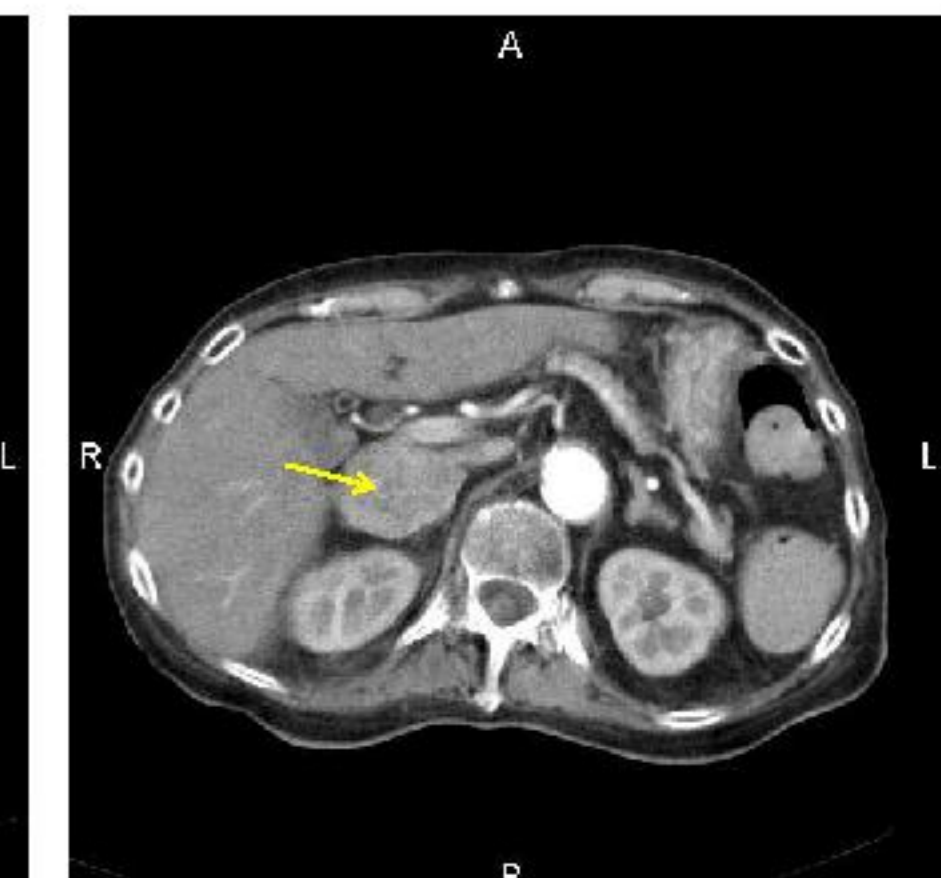
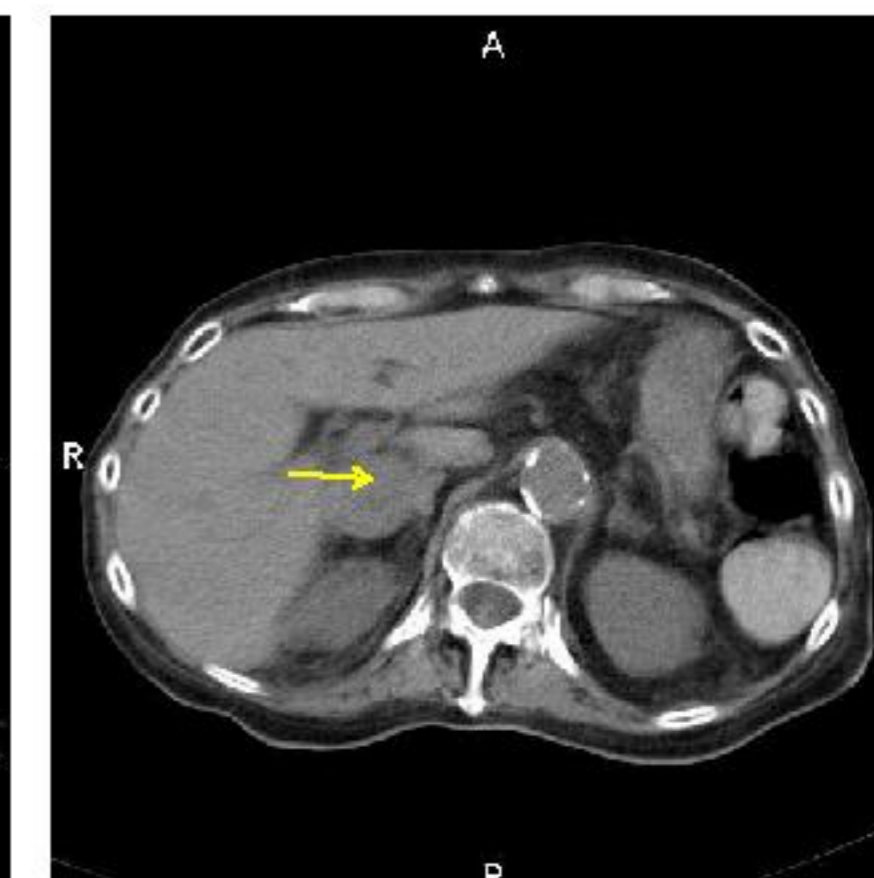


Fig.5. Histological examinations. A: HE, B: Chromogranin-A, C: reaction to ACTH, D melan-A

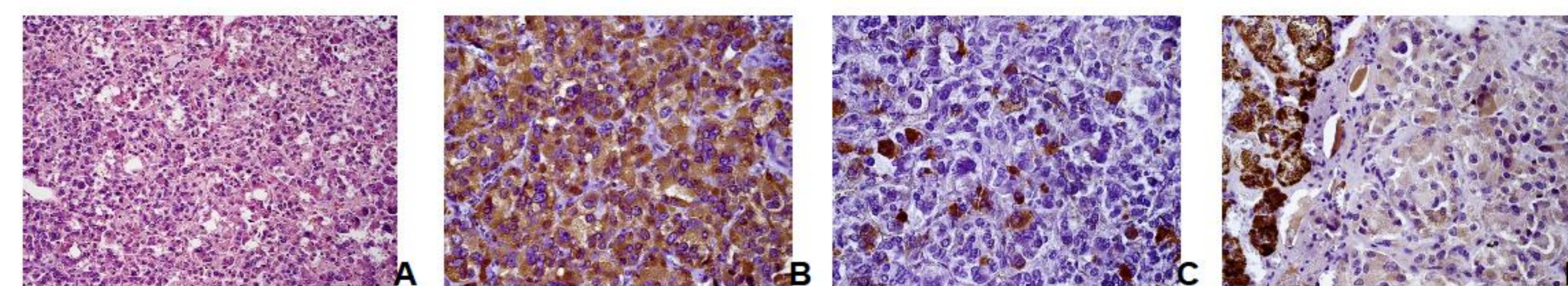


Fig.8. Histological examinations. A: HE, B: Chromogranin-A, C: reaction to ACTH, D melan-A

METHODS

We present two clinical cases of patients with EAS by pheochromocytoma.

CASE 2

A 61 year-old woman with 2-months history of weight gain, proximal myopathy, depressive disorders, abdominal pain, diabetes mellitus of recent onset and worseness of hypertension control. Patient had facial and leg edema, plethoric face, dermal and muscle atrophy and moderate central obesity. The laboratory data showed hyperglycemia, moderate hypokalemia, moderate leukocytosis, disturbed circadian cortisol rhythm and increased ACTH level. Measurements of Metanephrine and Normetanephrine in urine were increased. Abdominal CT scan and SRS revealed 30 mm mass in right adrenal gland and radiological signs suggested pheochromocytoma. After pharmacological treatment patient was successfully laparoscopic operated and all signs of Cushing’s syndrome regressed. Histological examinations confirmed EAS and pheochromocytoma.

parameter	WBC 10 ³ /ul [4.00 – 10.00]	Potassium mmol/l [3.5 – 5.1]	Sodium mmol/l [136.0 – 145.0]	ALT U/l [5 – 33]	Urea mmol/l [2.76-8.07]	Creatynine Umol/l [44-80]
value	15.58	3.6	143.0	68	6.0	68.0

parameter	Morning cortisol ug/dl [2.3- 23.3]	Midnight cortisol ug/dl [2.3- 20.9]	ACTH pg/ml [6.0 – 56.0]	5HIA umol/24h [10.0 – 40.0]	MN ug/24h [52-341]	NMN ug/24h [88-440]	CgA nmol/l [6.00 – 98.00]
value	32.5	25.87	182	69.1	2490	2526.8	605

Tab.2. Laboratory results.

CONCLUSIONS

EAS is difficult to diagnose. Other organs lesions make the prognosis worse.

References

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