

# Ectopic ACTH syndrome (EAS) - diagnostic and therapeutic challenge



Katarzyna Piątek, Małgorzata Kałużna, Katarzyna Ziemnicka, Marek Ruchała

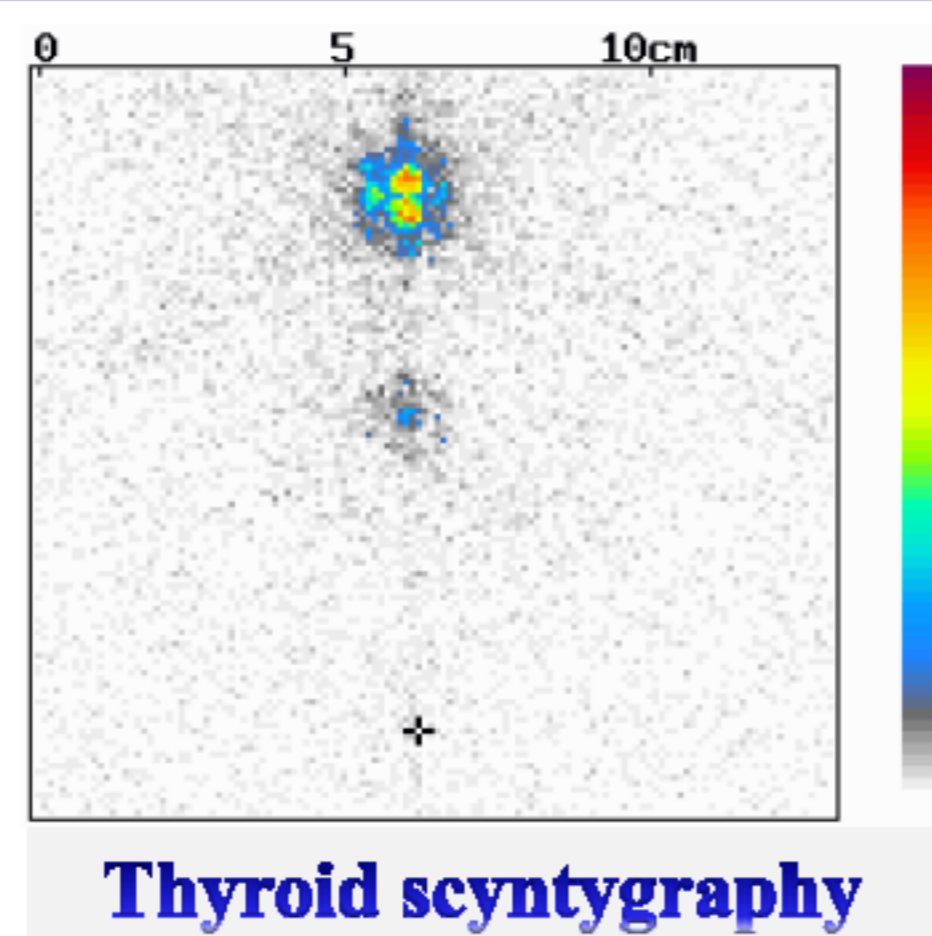
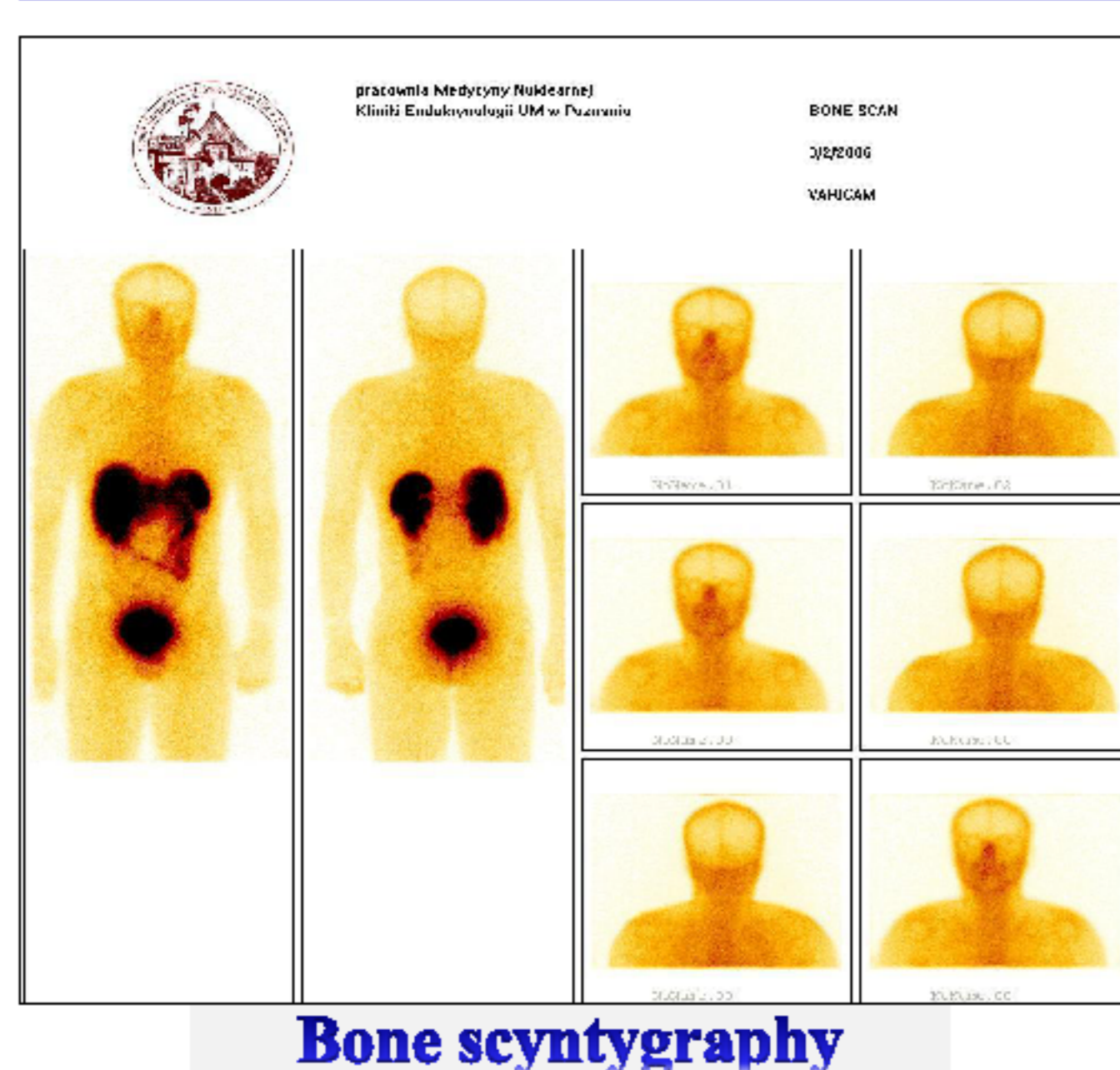
Department of Endocrinology, Metabolism and Internal Diseases,  
Poznan University of Medical Sciences, Poznan, Poland

e-mail: katarzyna.piatek.kp@gmail.com

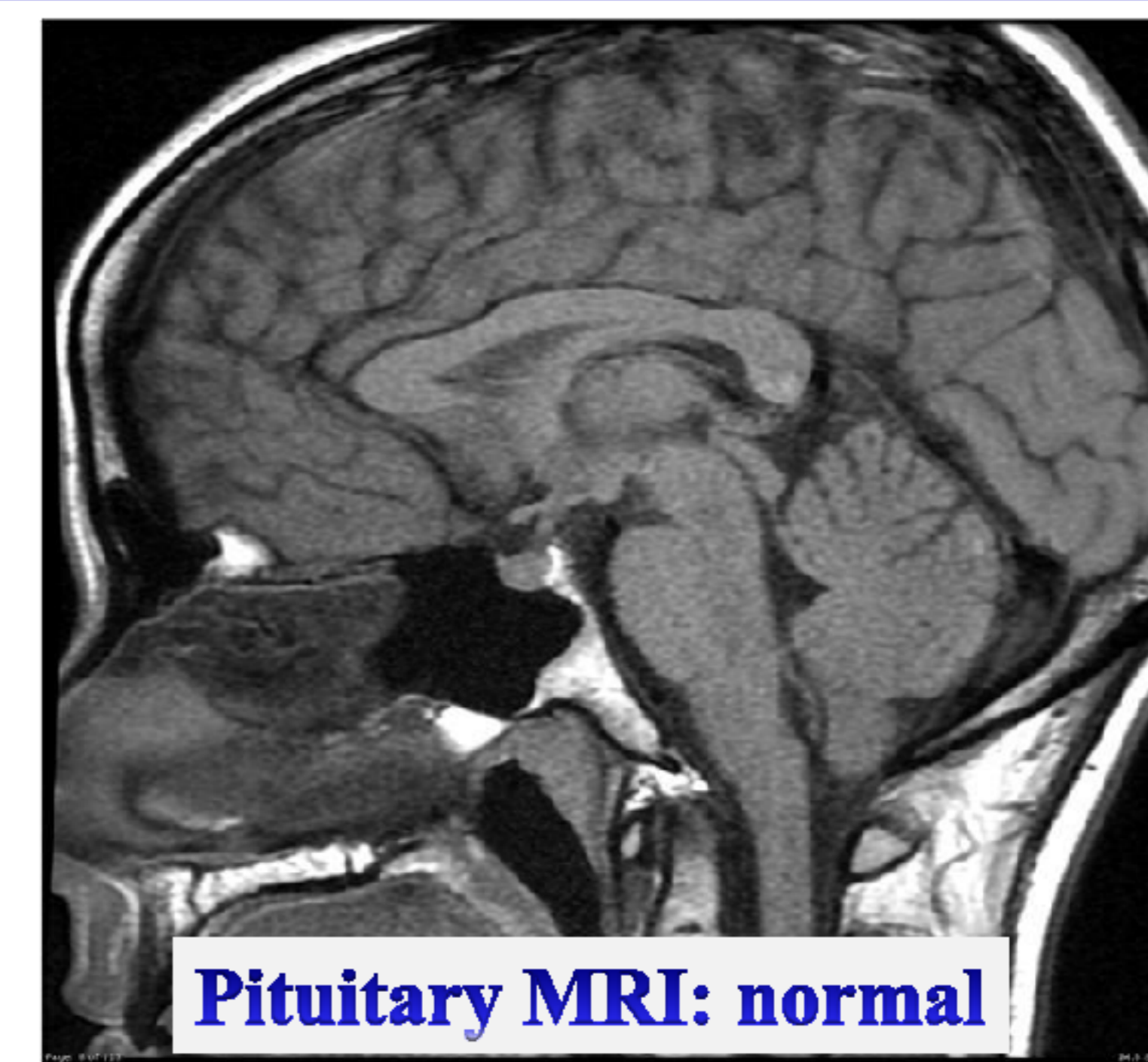
## Introduction:

Rapid deterioration of health condition in patient with diagnosed neoplastic disease, especially metastatic one, requires consideration of cancer progression. However other rare severe complications may occur. In 0.6-0.7% patients with medullary thyroid cancer (MTC) the ectopic ACTH syndrome (ECS) is observed. Hereby, we present a case of ECS in patient with MTC.

## Case report



- 09.2004 Thyroid FNAB: *cellulae atypice*
- 01.2005 Total thyroidectomy: *Medullary carcinoma*
- 08.2005 Neck exploration
- 05.2006 Neck lymphadenectomy

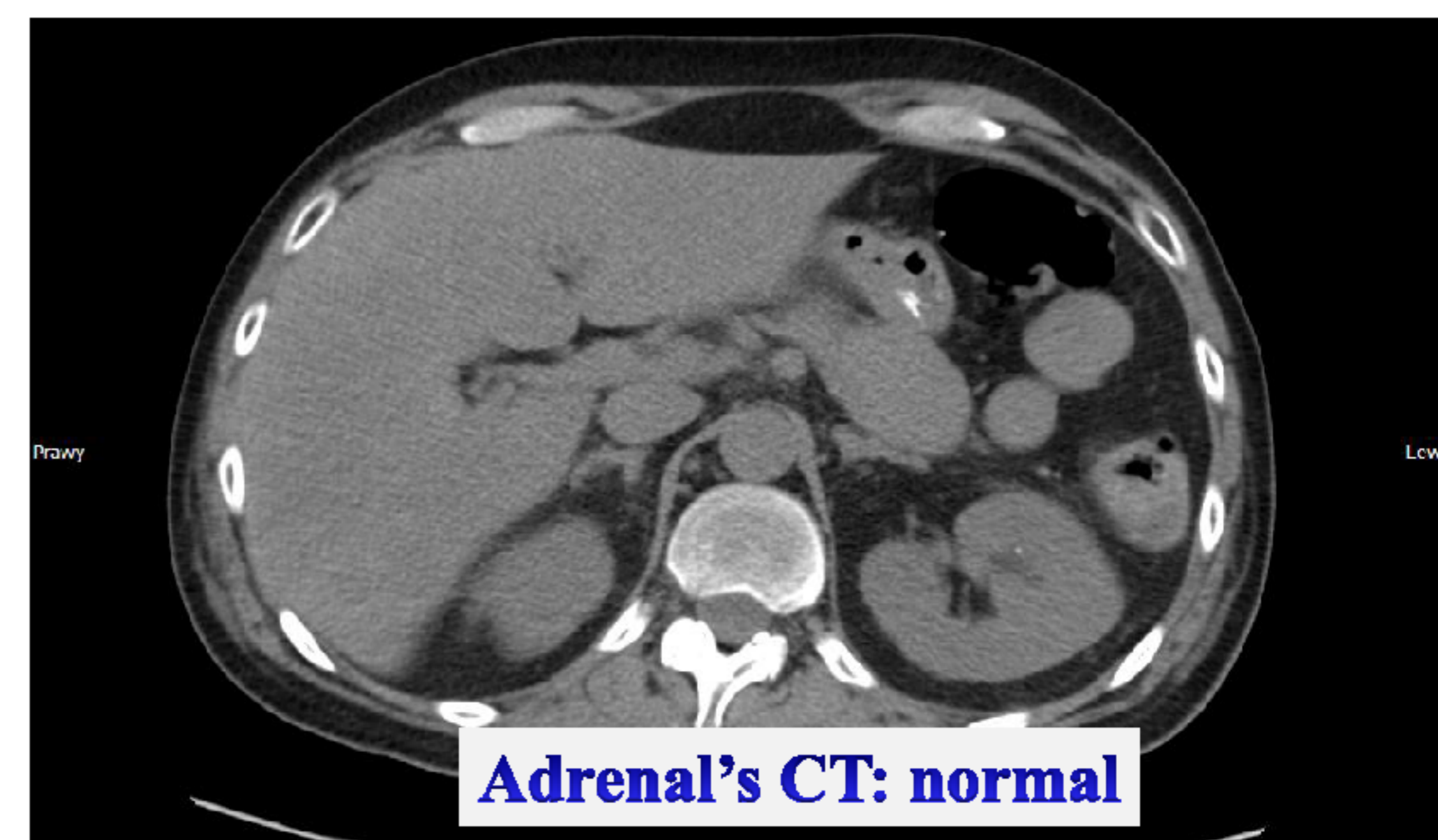


A 37-year-old man was admitted to the Department of Internal Medicine in serious clinical condition with general fatigue and chest pain. Myocardial infarction has been excluded. Patient's past medical history was remarkable for medullary thyroid carcinoma with numerous reoperations. Routine laboratory test showed *de novo* diabetes mellitus. Calcitonin serum level was 499,53 pg/ml (N 0-10 pg/ml). Additional tests revealed severe hypercortisolemia (cortisol level - 2100 nmol/l) (N 7-10 am: 171-536 nmol/l).

WBC:  $15.2 \times 10^3/\mu\text{l}$  [3.90 — 11.00]      Glucose profile  
 Erythrocytes:  $4.88 \times 10^6/\mu\text{l}$  [3.50 — 5.80]      08:00: 217 mg/dl  
 Hemoglobin: 15.6 g/dl [12.0 — 17.2]      11:00: 380 mg/dl  
 Hematocrit %: 45.2 % [33.0 — 50.5]      17:00: 259 mg/dl  
 PLT:  $223 \times 10^3/\mu\text{l}$  [130 — 400]      20:00: 346 mg/dl

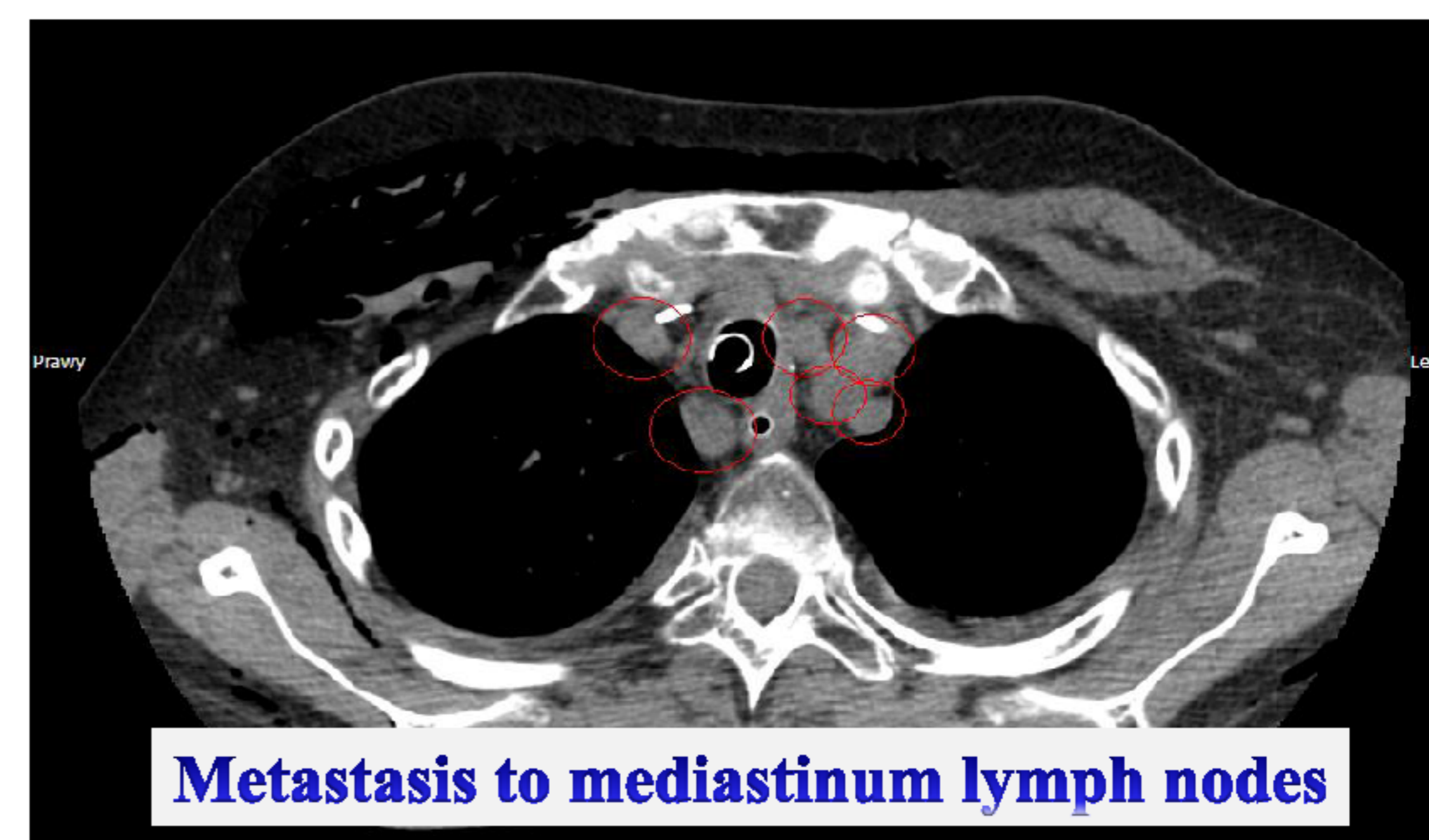
CRP: 1,40 mg/l [ $< 5,00$ ]  
 Calcitonin: 499.53 pg/ml [0 — 10]  
 Cortisol: 2100 nmol/l [7-10am: 171-536]  
 ACTH: 329,0 pg/ml [ $< 60,0$ ]

- 09.2010 Excisio of the thyroid remnants in left thyroid bed
- 08.2011 Rexcisio of the thyroid remnants in left thyroid bed
- 10.2011 Excisio of the metastatic chest lymph nodes
- 11.2012 Qualification to tyrosine kinase inhibitors treatment



The patient was referred to the Department of Endocrinology for further diagnostics and treatment. ACTH level was 329,0 pg/ml (N $<60,0$ ). There was no suppression of cortisol secretion after 1 mg of Dexamethasone. Magnetic resonance imaging (MRI) of the pituitary gland showed no signs of macro or microadenoma. Computer tomography (CT) of the chest revealed metastasis to the mediastinum lymph nodes. The adrenal glands in CT were normal size and shape. **The EAS was diagnosed.**

- 12.2012 A&E department
- 12.2012 **Diagnose of Ectopic ACTH Syndrome**



Daily rhythm of cortisol excretion:  
 08.00 2192 nmol/l  
 10.00 1750 nmol/l  
 18.00 1667 nmol/l  
 22.00 2084 nmol/l

Test with 8 mg of dexamethasone  
 cortisol: 1748 nmol/l

- 14.01.2013 Bilateral adrenalectomy
  - 02.02.2013 Internal bleeding, *megacolon toxicum*
  - 05.02.2013 Internal bleeding
  - 11.02.2013 Fecal peritonitis
  - 23.02.2013 Eventration
  - 09.03.2013 **Death**
- Due to the rapid deterioration of general condition despite of ketoconazole treatment, bilateral adrenalectomy was performed. The patient in severe condition was transferred to the Intensive Care Unit. Four reoperations were conducted due to the: internal bleeding, megacolon toxicum, fecal peritonitis and eventration. On the 41th day after first surgery the patient died due to sepsis.

## Conclusions

The EAS is a rare condition in patients with medullary thyroid cancer (MTC), but our patient's history indicates necessity of intensive search for EAS in case of advanced MTC, especially in rapid health deterioration.