

Case Report

ESIM 2015

Felix Machleidt
University Hospital Lübeck
Germany



UNIVERSITÄT ZU LÜBECK





51 year old male

➤ **History of present illness:**

- palpitations
- headache
- dyspnoea
- high blood pressure
- chest pain

➤ **Since about one year:**

- weakness
- constipation
- night sweat, no weight loss
- paroxysmal hypertensive episodes, sometimes with palpitations

➤ **Past medical history: hypertension, smoker**

➤ **Current medication: Bisoprolol**

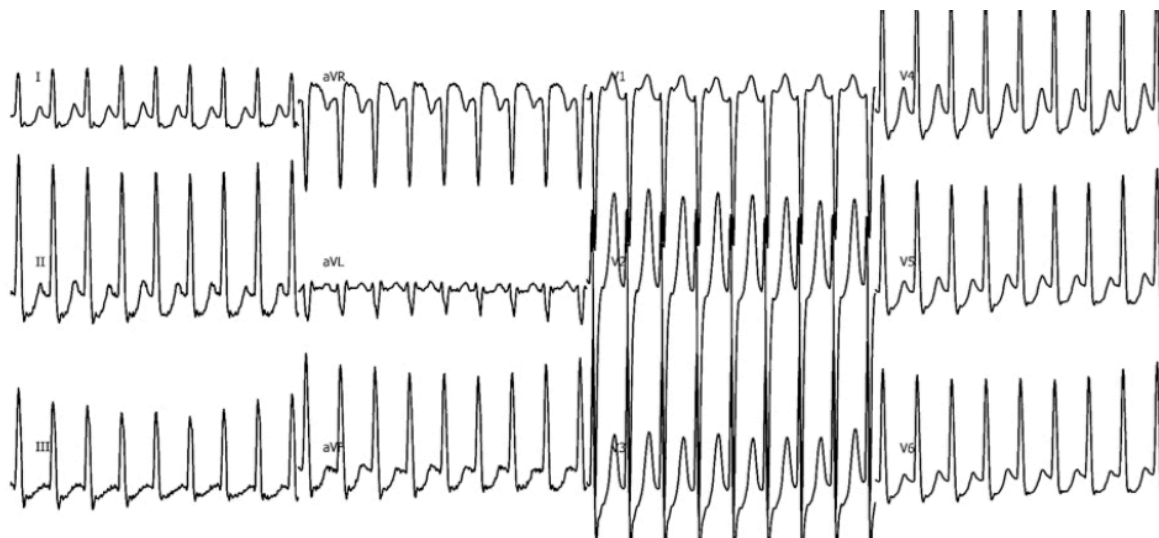


Findings

- physical examination
 - cold + clammy skin, HR 100/min, hypoactive bowel sounds
 - after physical exam: anxiety, palpitations + headache

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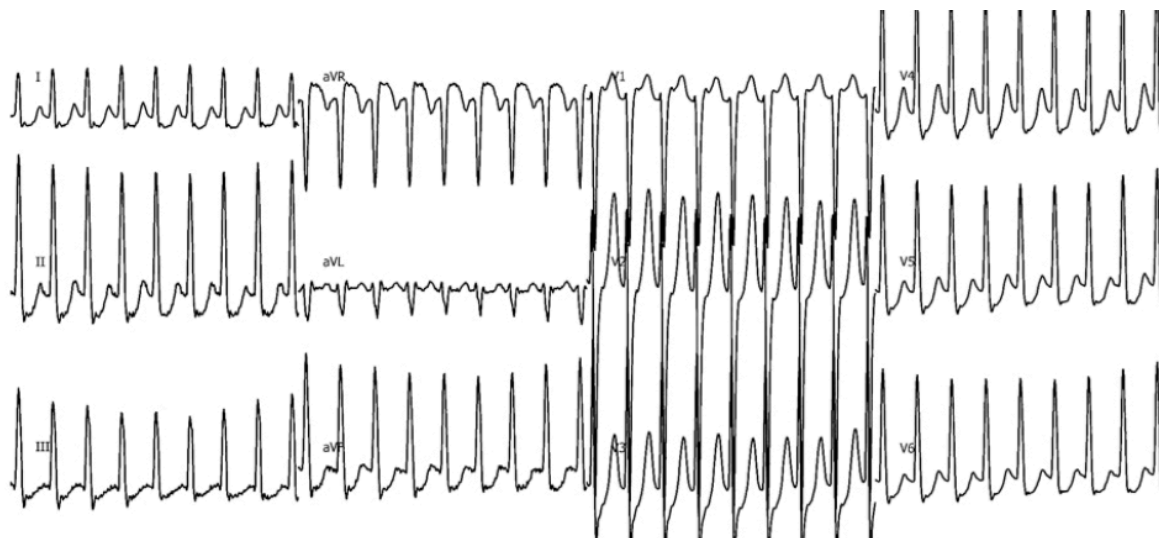
➤ physical examination

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- blood pressure 230/120 mmHg

➤ lab results

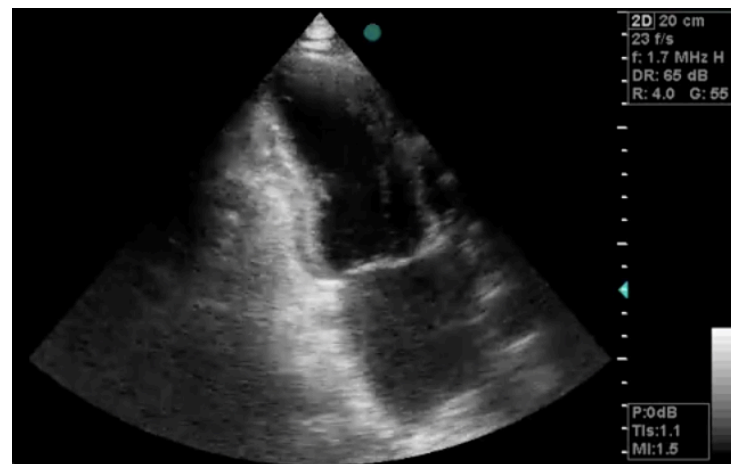
- blood sugar 360 mg/dl (20 mmol/l), potassium 2.9 (norm 3.5-5 mmol/l)
- leukocytosis and lymphocytopenia, ABG: metabolic alkalosis
- troponin t 30 pg/ml (<14 pg/ml)

HAEMATOLOGIE	
Leukozyten	23260 +
Erythrozyten	4.23 -
Hämoglobin	124 -
Hämatokrit	0.36 -
MCV	86.1
MCH	29.3
MCHC	341
Thrombozyten	189
MPV	11.3

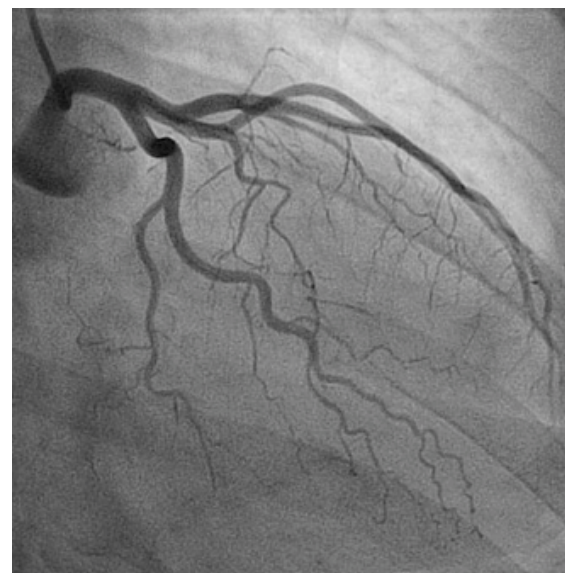


Findings

- Echo
 - slight apical hypokinesis
 - small pericardial effusion



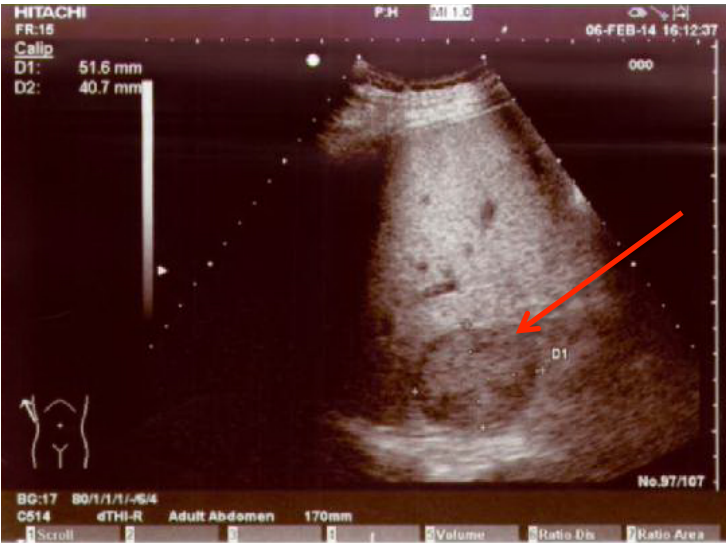
- Coronary Angiography
 - no coronary heart disease



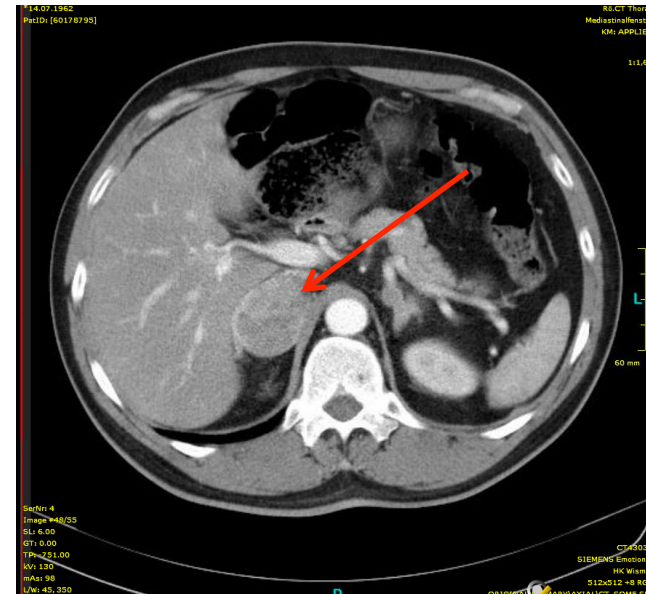
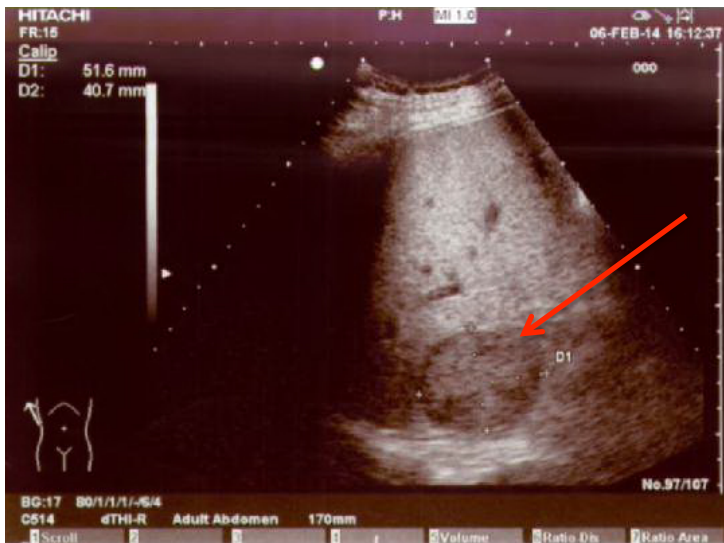
Imaging



Imaging



Imaging



- right adrenal mass of 5.4 cm in largest dimension

???



Pheo?

Hypertension, headache, AVNRT

???



Pheo?

Hypertension, headache, AVNRT

Cushing?

Diabetes, hypertension, low potassium, met. alkalosis,
headache

???



Pheo?

Hypertension, headache, AVNRT

Cushing?

Diabetes, hypertension, low potassium, met. alkalosis, headache

BC + metastasis + paraneoplastic hormone secretion?

Smoker

Lab results

PROTEINE						
Albumin i.PI.	32.1	-		33 - 50	g/l	Colorimetrie
C-reaktives Prot. i.PI.	1.00			- 5	mg/l	Turbidimetrie
Hb A1C i.BI.		7.7	+	4.3 - 6.0	% Hb	HPLC
Hb A1C i.BI.		60.7	+	28 - 42	mmol/mol	HPLC
Protein-El'phorese i.S.	Extr.Bef.				Hb	
ENDOKRINOLOGIE						
fr. Trijodthyr. (fT3) i.S.		1.52	-	2.8 - 7.1	pmol/l	ECLIA
fr. Thyroxin (fT4) i.S.		8.30	-	12 - 22	pmol/l	ECLIA
TSH i.S.	0.167			0.27 - 4.20	mIU/l	ECLIA
Cortisol i.S.		6130	++	171 - 535	nmol/l	ECLIA
Cortisol i.S.		222.1			µg/dl	berechnet
ACTH i.PI.		371.9	++	10 - 60	ng/l	ECLIA
Renin i.PI.		50.0	++ 3)	3.0 - 21.0	ng/l	ELISA
Aldosteron i.S.		302	++ 3)	29 - 145	ng/l	ELISA
A/R-Quotient		6.04		0 - 50		

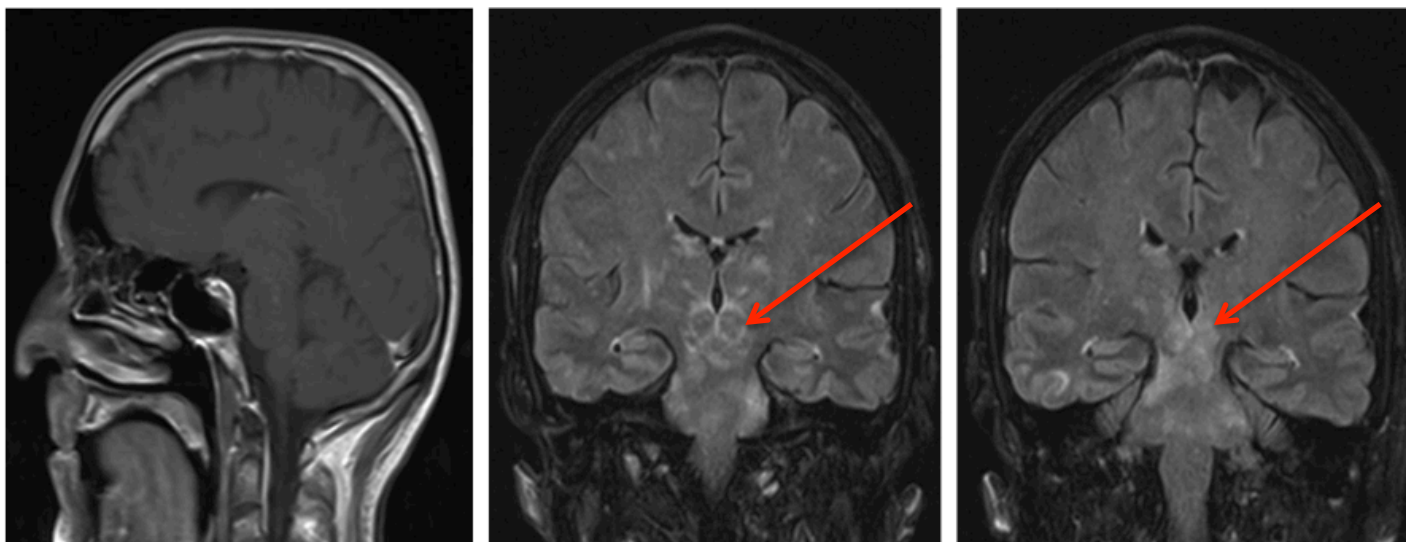
➤ ACTH-dependent Cushing Syndrome

Lab results

PROTEINE						
Albumin i.PI.	32.1	-		33 - 50	g/l	Colorimetrie
C-reaktives Prot. i.PI.	1.00			- 5	mg/l	Turbidimetrie
Hb A1C i.BI.		7.7	+	4.3 - 6.0	% Hb	HPLC
Hb A1C i.BI.		60.7	+	28 - 42	mmol/mol	HPLC
Protein-El'phorese i.S.	Extr.Bef.				Hb	
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- ACTH-dependent Cushing Syndrome
- No decrease in high dose dexamethasone suppression test
- Ectopic ACTH-dependent Cushing Syndrome

More imaging



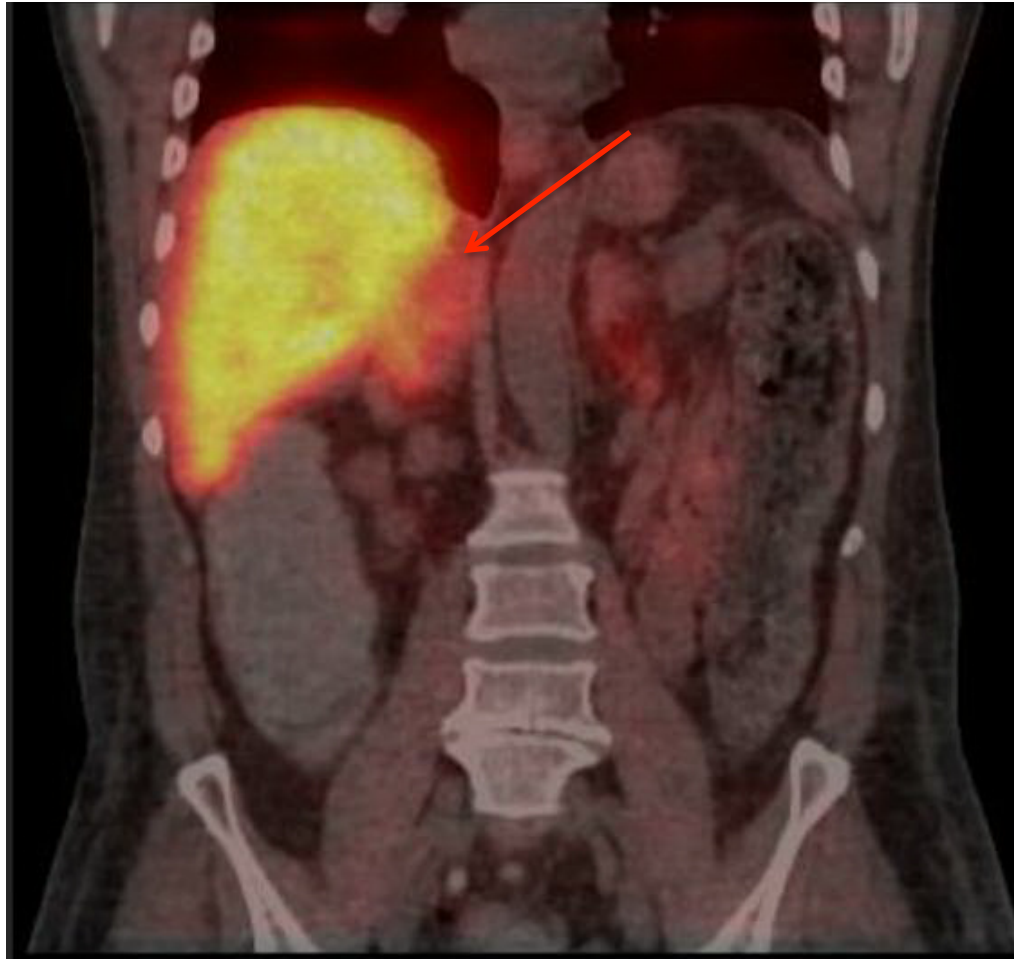
- no pituitary tumor was evident by MRI
- Perifocal signal alterations in the brainstem
- diffuse metastases? encephalitis? posterior reversible encephalopathy syndrome (PRES)

More lab results

- elevated plasma metanephrine and normetanephrine levels
- pheochromocytoma!

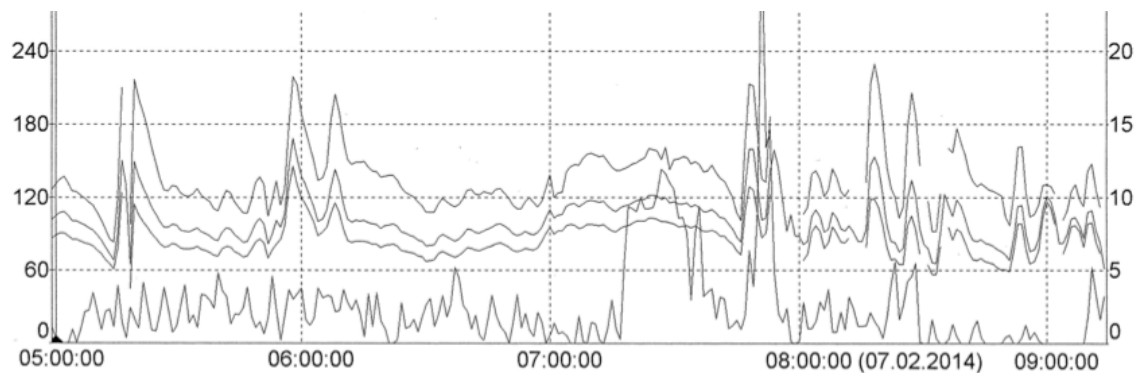
ENDOKRINOLOGIE			
fr. Trijodthyr. (fT3) i.S.		1.52	-
fr. Thyroxin (fT4) i.S.		8.30	-
TSH i.S.	0.167	-	
Cortisol i.S.		6130	++ 2555
Cortisol i.S.		222.1	92.6
ACTH i.PI.		371.9	++
Renin i.PI.		50.0	++ 3)
Aldosteron i.S.		302	++ 3)
A/R-Quotient		6.04	
Adrenalin i.PI.			
Noradrenalin i. PI.			
Dopamin i.PI.			
Metanephrin i.PI.		3305	4)
Normetanephrin i.P.		3230	5)

^{123}I od-MIBG Scintigraphy



Treatment

- Blood pressure
 - oral alpha-blocking agent (phenoxybenzamin)
 - i.v. alpha-blocking agent (urapidil)



- AVNRT
 - Carotid sinus massage
 - Adenosin
 - Metoprolol i.v.



Treatment

- Blood sugars
 - insulin i.v.
 - marked insulin resistance; high insulin doses

- Hypercortisolism
 - intravenous etomidate in a low, non-hypnotic dose

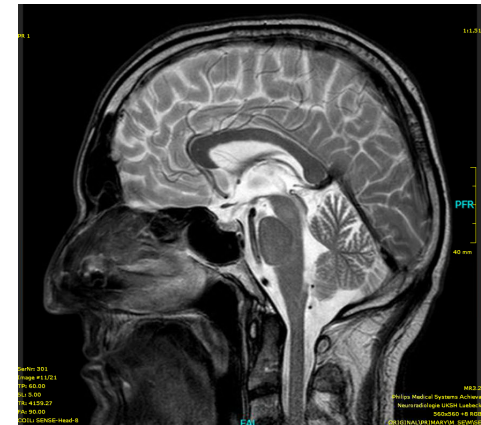
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TSH i.S.	0.467						0.27 - 4.20 mIU/l ECLIA
Cortisol i.S.		6130	++	2555		1242	++ 171 - 535 nmol/l ECLIA
Cortisol i.S.		222.1		92.6		45.0	µg/dl berechnet
ACTH i.PI.		571.5	++				10 - 65 ng/l ECLIA
Renin i.PI.		50.0	++ 3)				3.0 - 21.0 ng/l ELISA
Aldosteron i.S.		302	++ 3)			125	3) 29 - 145 ng/l ELISA
A/R-Quotient		6.04					0 - 50
Adrenalin i.PI.					13530	6)	s. Bemerk. Fremdlabor
Noradrenalin i. PI.					26570	7)	s. Bemerk. Fremdlabor
Dopamin i.PI.					480	8)	s. Bemerk. Fremdlabor
Metanephrin i.PI.		3305	4)		2700	4)	s. Bemerk. Fremdlabor
Normetanephrin i.P.		3230	5)		2850	5)	s. Bemerk. Fremdlabor

Therapy

- Early performed adrenalectomy

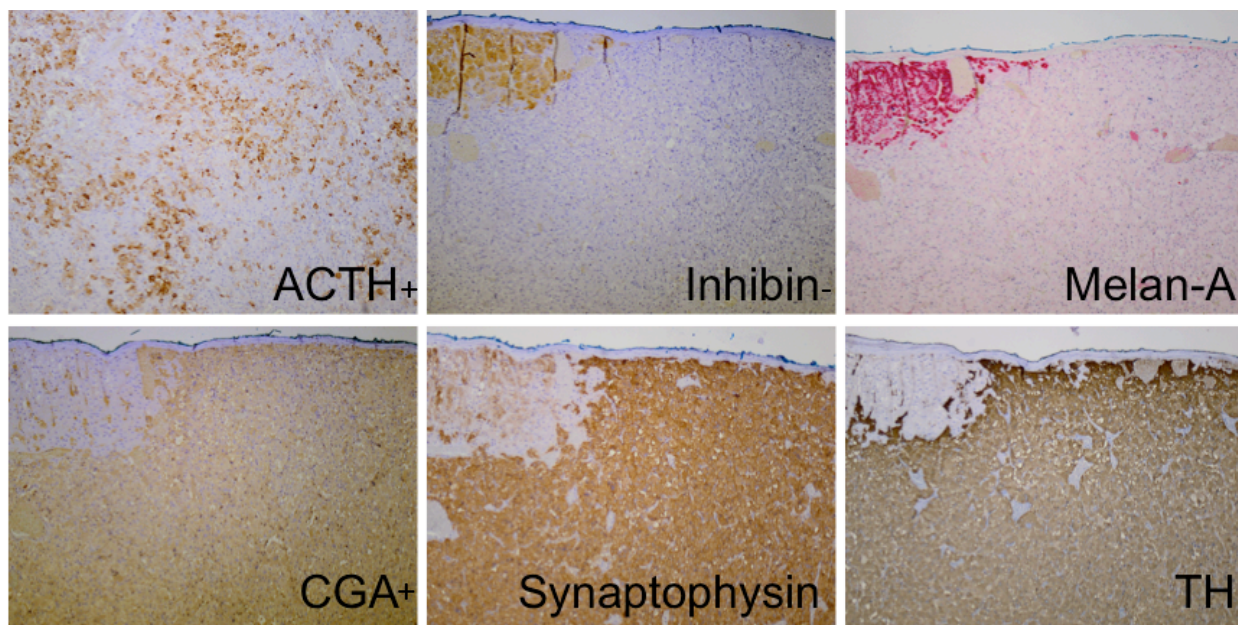
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Eingangszeit:	05:53	10:07	08:29	10:06	09:48				
						Referenz-	Einheit	Verfahren	
						bereich			
PROTEINE									
Albumin i.Pl.	10.1	--			26.7	-	33 - 50	g/l	Colorimetrie
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ENDOKRINOLOGIE									
Metanephrin i.Pl.					13.7	2)			s. Bemerk. Fremdlabor
Normetanephrin i.P.					50.9	3)			s. Bemerk. Fremdlabor

HZ	108	112	112	112	112	Pat. angeleitet: <input type="checkbox"/> BZ-Test <input type="checkbox"/> Spritzen
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HZ	108	112	112	112	112	Pat. angeleitet: <input type="checkbox"/> BZ-Test <input type="checkbox"/> Spritzen
Zeit	11:28	11:36	11:37	11:42	11:54	
HZ	112	112	112	112	112	Pat. angeleitet: <input type="checkbox"/> BZ-Test <input type="checkbox"/> Spritzen
Zeit	12:05	12:05				



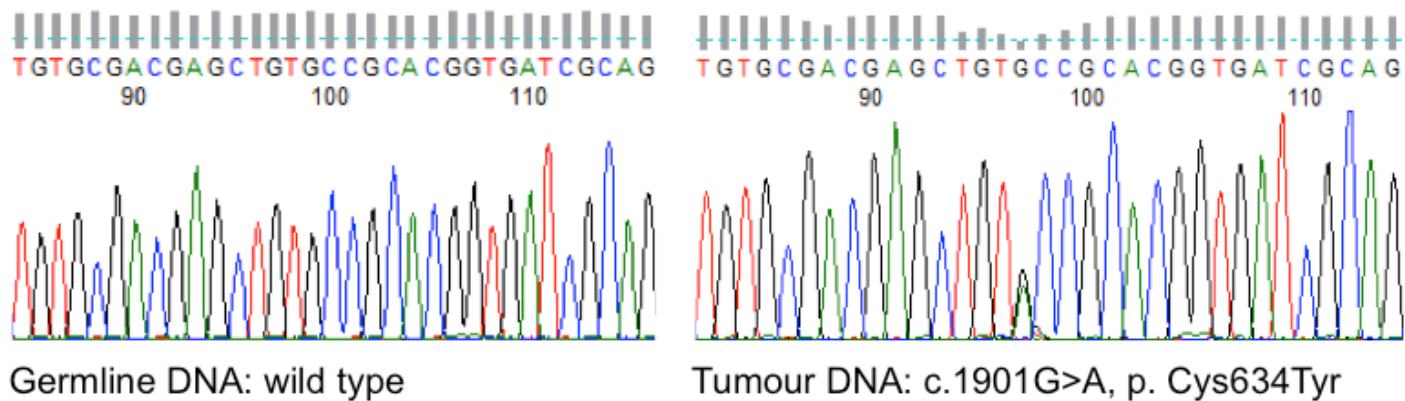
- Normal blood pressure without any antihypertensive medication
- No further tachycardic episodes
- Cerebral changes interpreted as PRES seen on MRI completely normalized
- The patient fully recovered and was discharged without complaints

Immunohistochemical staining



- Immunohistochemical staining of the resected tumor confirmed ACTH-producing PHEO
 - Images were taken with 40x objective
(CGA: chromogranin A, TH: tyrosine hydroxylase)

Genetics



- Sequence analysis in tissue and blood revealed a point mutation in the RET gene, which is commonly found in PHEOs



- two pathologies can occur simultaneously
- patients with adrenal masses should always be tested on PHEO and Cushing
- symptoms can present as overlapping syndrom without being specific for each condition alone

Visit Lübeck

You are very welcome!

