Adrenal
Pathology

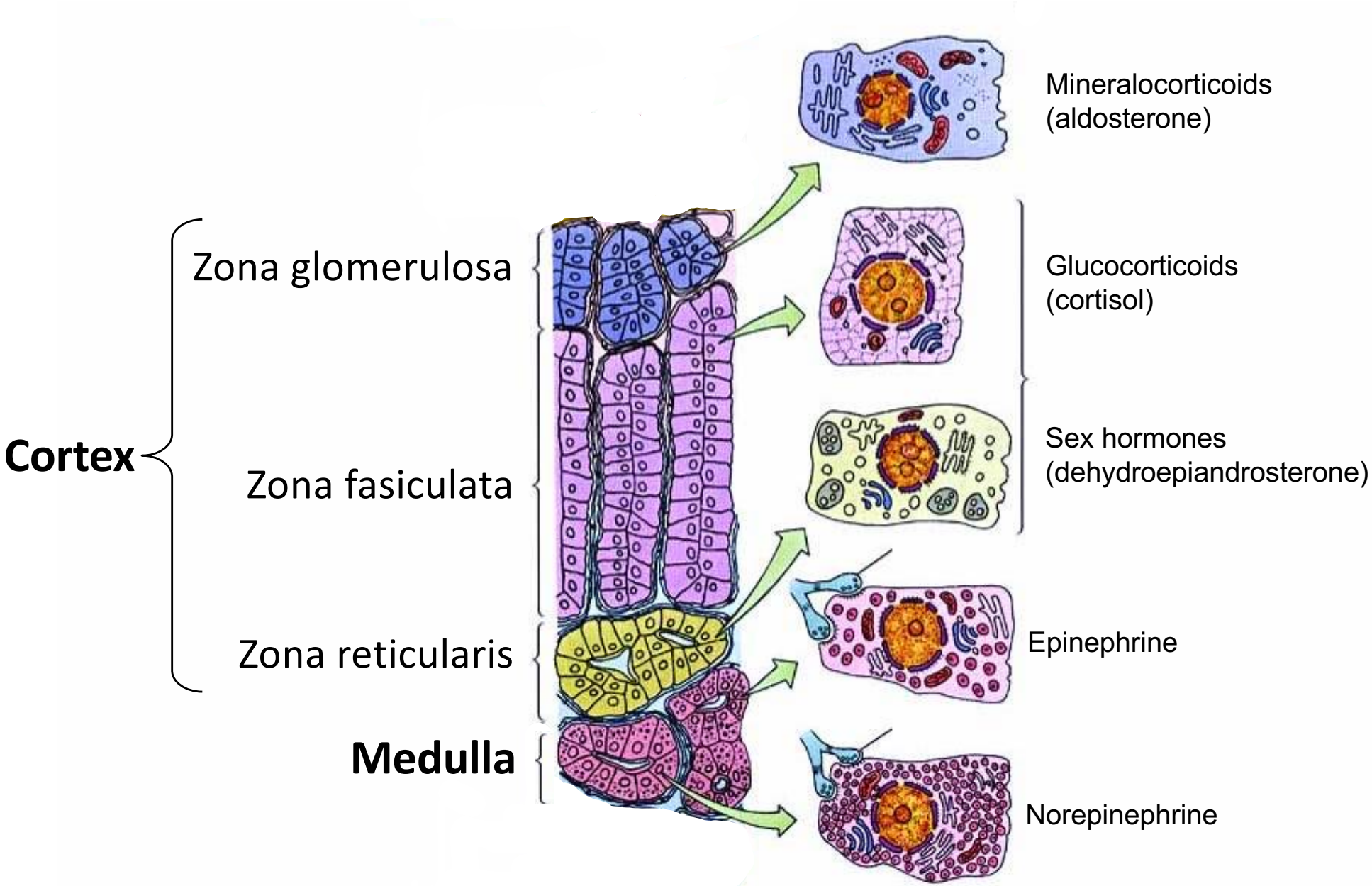
Introduction

Too much hormone

Too little hormone

Tumors

Introduction



Adrenal gland histology and hormones

Introduction

Too much hormone

Introduction

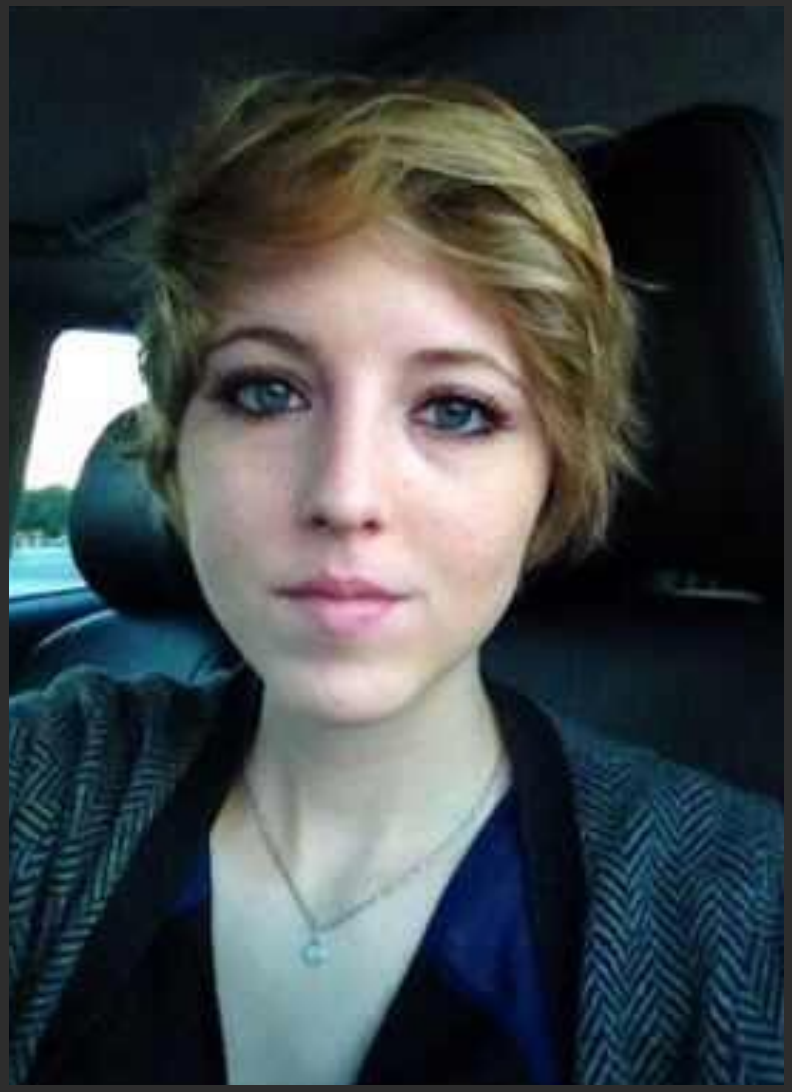
Too much hormone

- Cushing syndrome

Cushing Syndrome

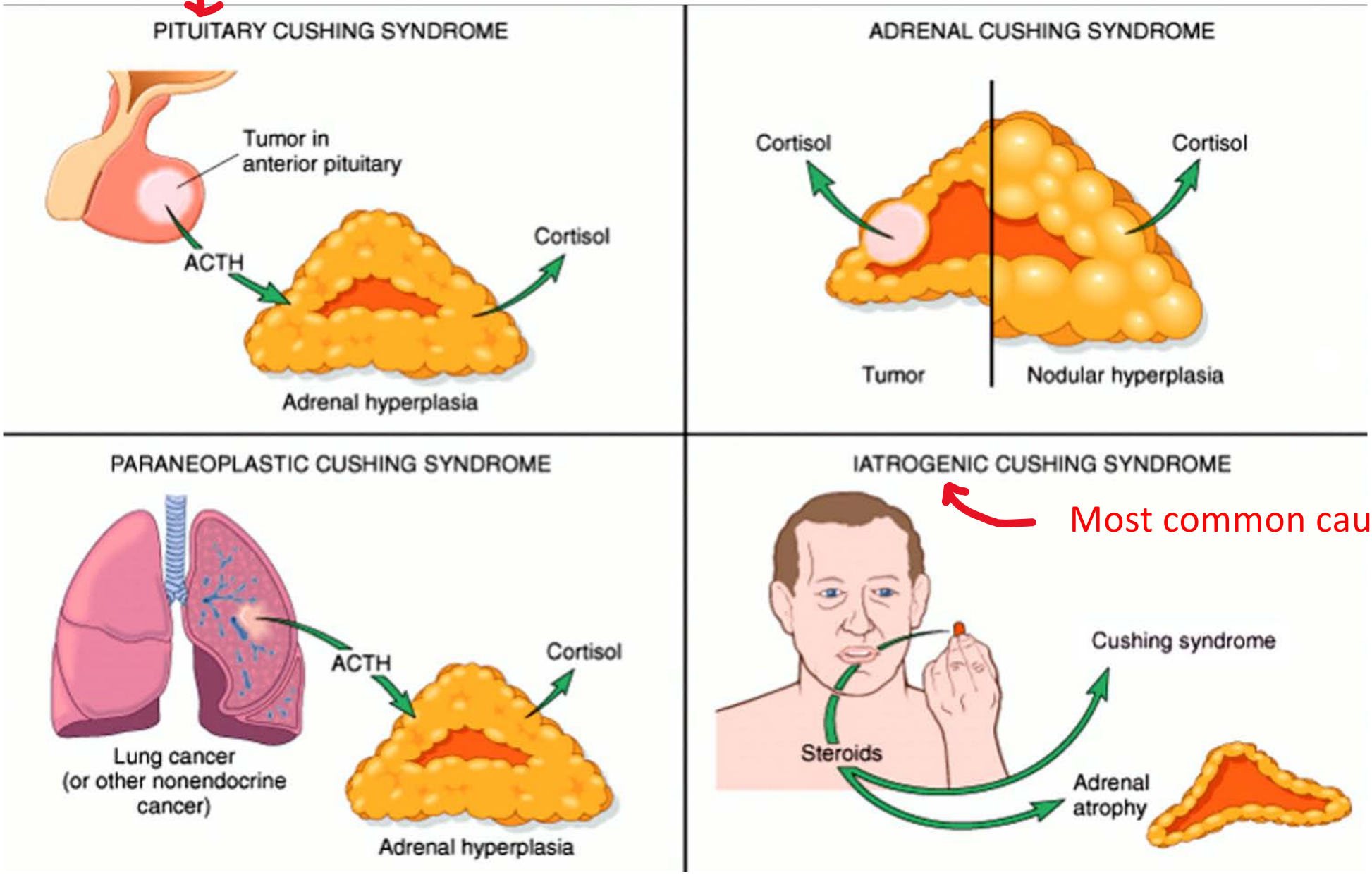
- Hypertension
- Characteristic pattern of weight gain
- Glucose intolerance
- Infection

Cushing syndrome: buffalo hump



Cushing syndrome: moon facies

Second most common cause



Causes of Cushing syndrome

Introduction

Too much hormone

Too little hormone

- Adrenal insufficiency

Primary Chronic Adrenal Insufficiency

- Also called Addison disease
- Too little cortisol and mineralocorticoids
- Usual cause: autoimmune destruction
- Symptoms
  - Slow onset
  - Weakness, fatigue, GI complaints
  - Hypotension
  - Skin hyperpigmentation

Hyperpigmentation in Addison disease

Hyperpigmentation in Addison disease

Introduction

Too much hormone

Too little hormone

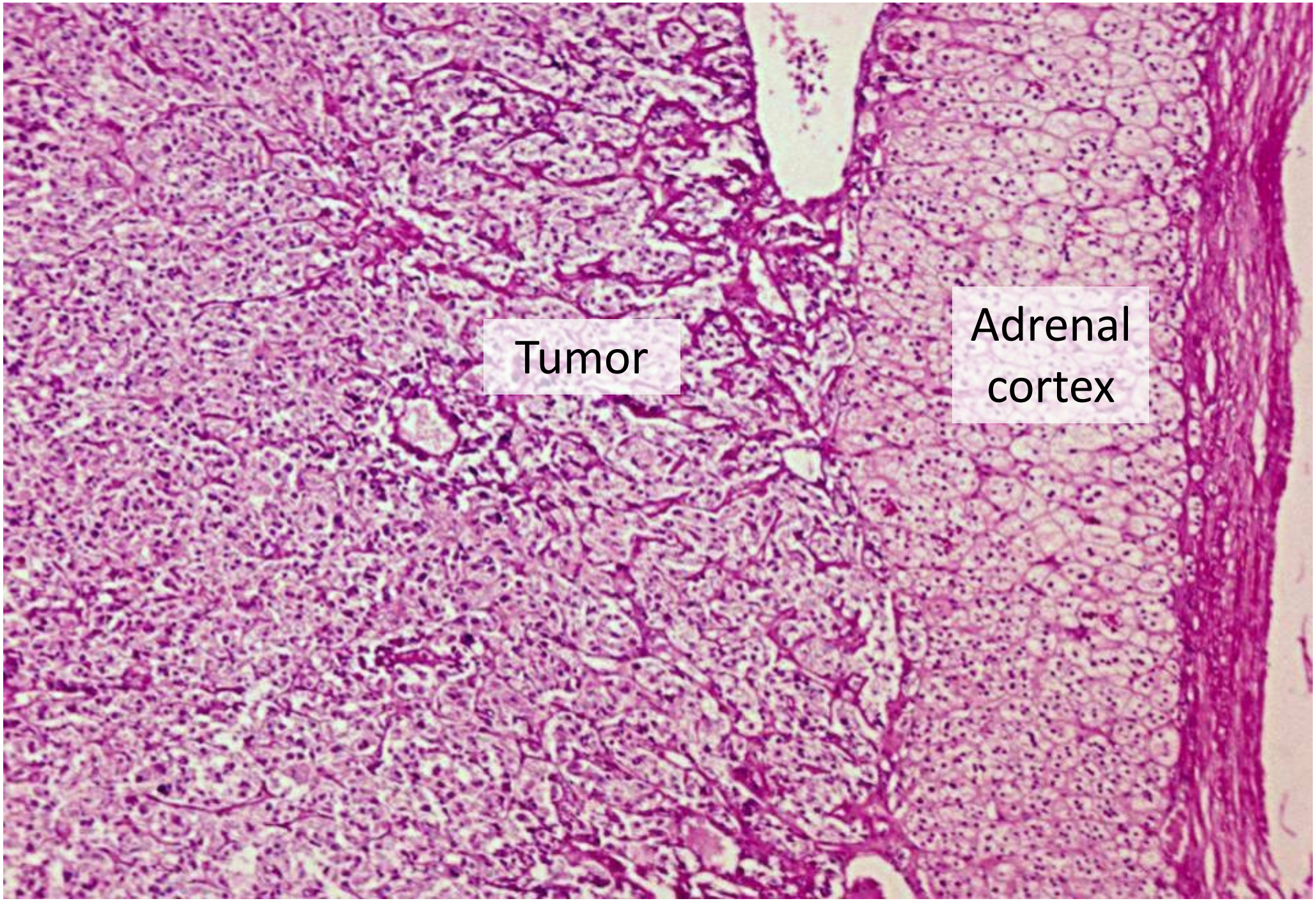
Tumors

Pheochromocytoma

- Neoplasm of catecholamine-producing cells
- Causes hypertension!
- Urine: catecholamines, VMA and metanephrines
- The 10% tumor!
  - 10% extra-adrenal (“paraganglioma”)
  - 10% bilateral
  - 10% (or more) familial
  - 10% malignant
  - 10% don’t have hypertension

Symptoms of pheochromocytoma:

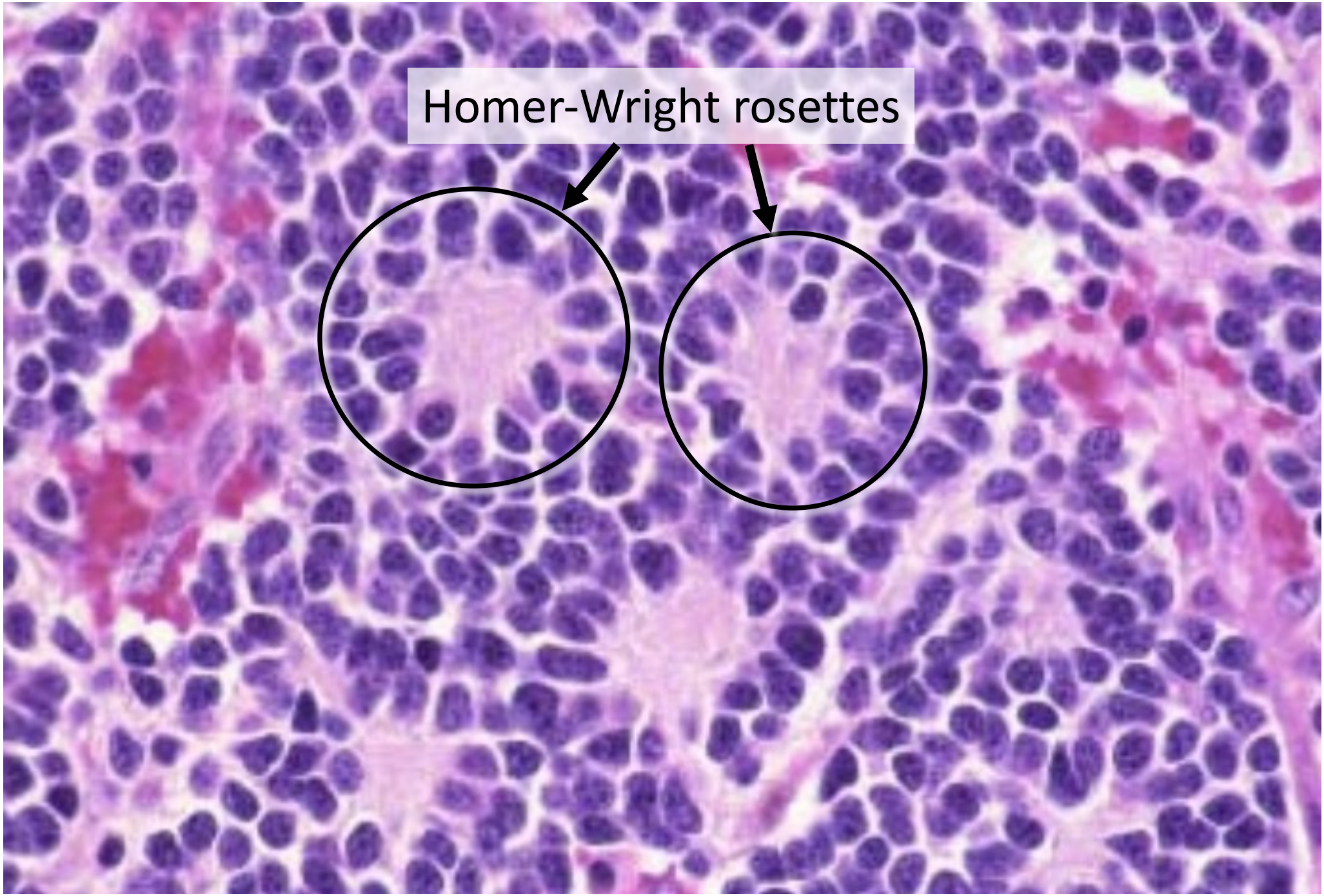
- Pressure (increased blood pressure)
- Pain (headache)
- Perspiration
- Palpitations (tachycardia)
- Pallor
- Paroxysms!



Pheochromocytoma

Neuroblastoma

- Derived from neural crest cells
- Relatively common childhood tumor
- Prognosis better in:
  - Children < 18 months
  - Lower stage/grade
  - Hyperdiploid tumors
  - Fewer copies of *N-myc*



Neuroblastoma

MEN Syndromes

Brad Pitt vs. John Cleese

No contest!

MEN Syndromes

- Genetic disorders
- MEN-1 and MEN-2
- Patients get multiple endocrine tumors
- Patients young; tumors aggressive

MEN tumors are worse than sporadic ones

- Younger age
- Multiple organs
- Aggressive

MEN Syndromes Ridiculously Simplified

	MEN-1	MEN-2
Thyroid	Not much	Medullary carcinoma
Other Endocrine Organs	Hyperplasia, adenoma, carcinoma	Not much

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MEN I

The three P's

- Parathyroid hyperplasia
- Pancreatic endocrine tumors
- Pituitary adenoma

MEN I Genetics

- MEN1 gene mutation
- Tumor suppressor gene (like most other tumor-promoting syndromes)
- Mutation inactivates the gene

MEN-1

Pitt-uitary adenoma

MEN1 gene

- run-of-the-mill
- inactive
- turn off

MEN Syndromes Ridiculously Simplified

	MEN-1	MEN-2
Thyroid	Not much	Medullary carcinoma
Other Endocrine Organs	Hyperplasia, adenoma, carcinoma	Not much

MEN 2A

- Medullary thyroid carcinoma
- Pheochromocytoma
- Parathyroid hyperplasia

MEN 2B

- Medullary thyroid carcinoma
- Pheochromocytoma
- Marfanoid habitus

MEN 2 Genetics

- RET gene mutation
- Proto-oncogene (unusual!)
- Mutation turns gene on

MEN-2

Medullary thyroid carcinoma
(a cancer of the Cleese-cells)

BRETon gene

one of a kind

always turned on