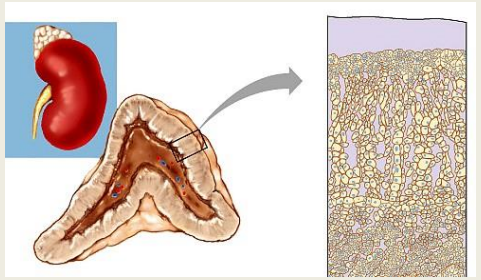
Endocrine pathology II.

Dr. Attila Zalatnai



Adrenal gland

Malformations: ectopic islands (kidney, ovary)

Apoplexia: Waterhouse-Friderichsen syndrome

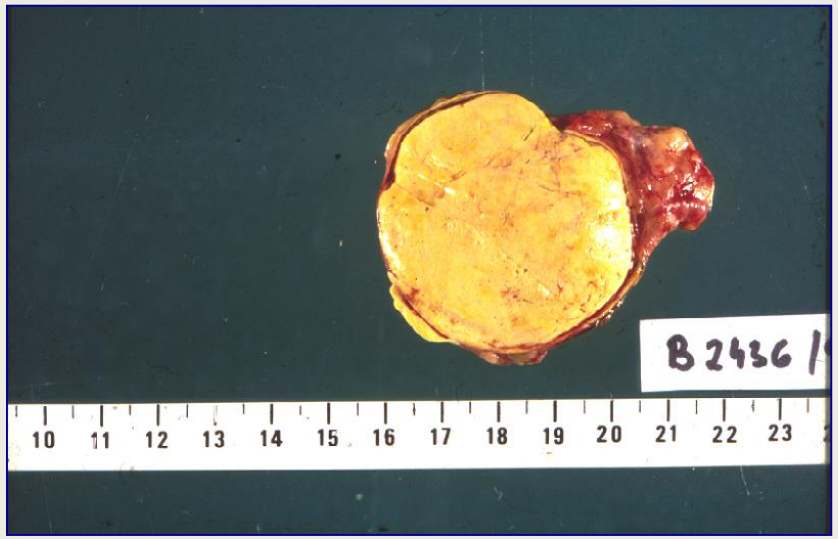
Congenital adrenal hyperplasia (adrenogenital syndrome)

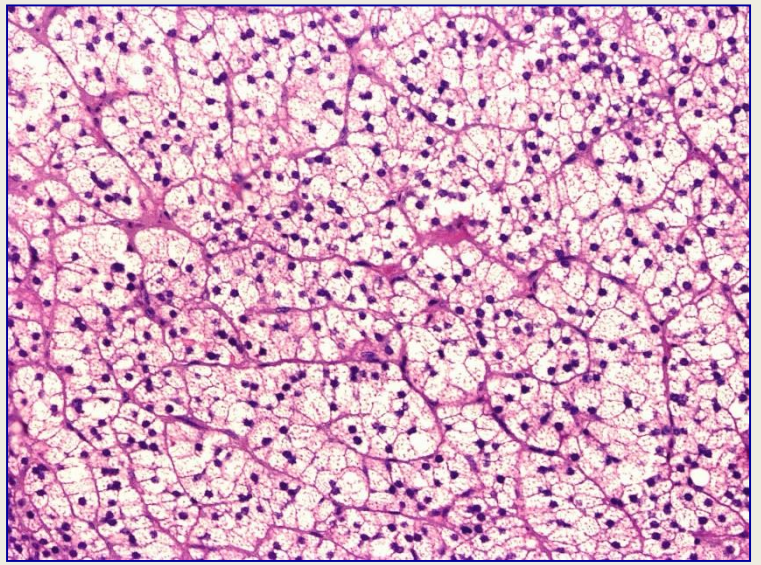
Hyperplasia, nodular hyperplasia

Adenomas: functioning (hormone producing) - nonfunctioning

- glycocorticoids: Cushing syndrome
- mineralocorticoids: Conn syndrome

Myelolipoma






CM
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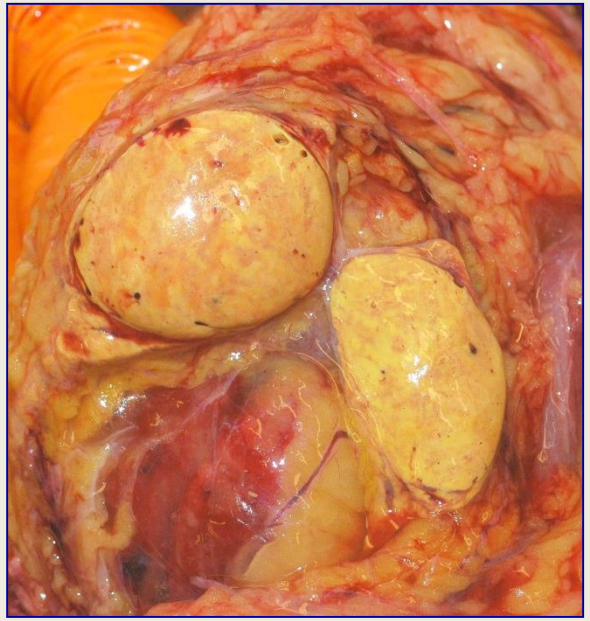
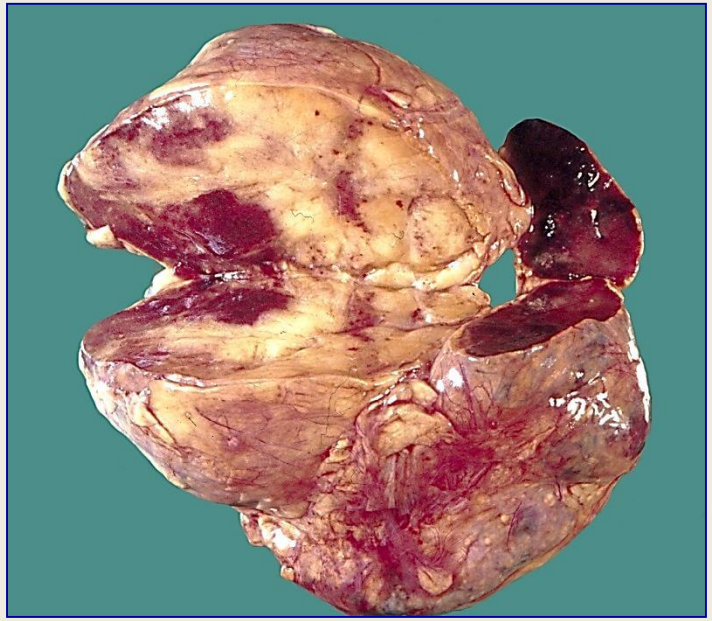
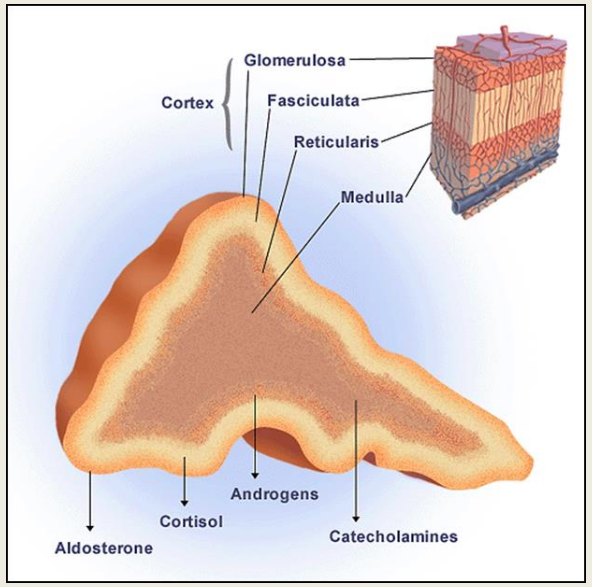
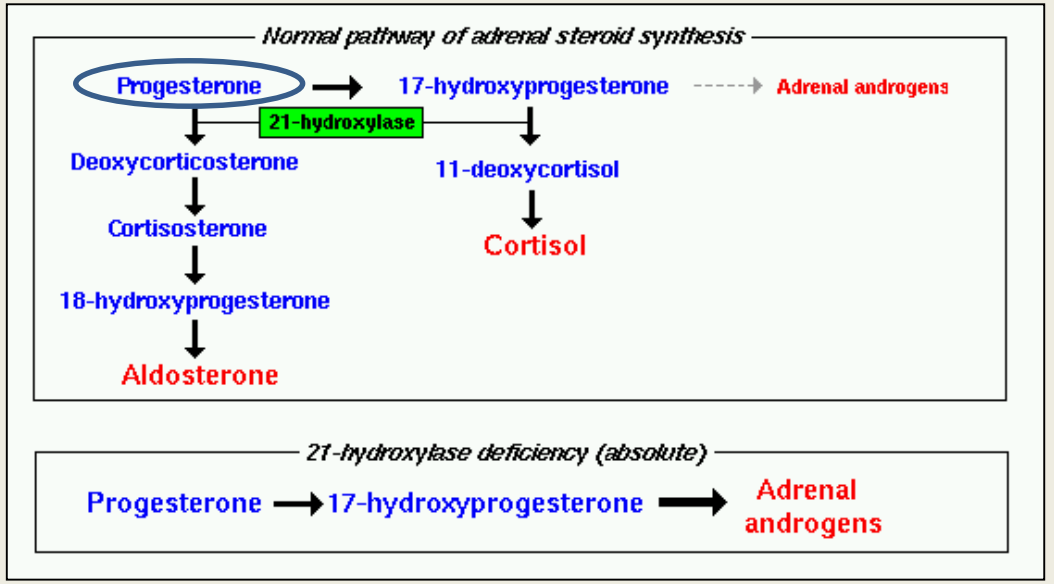
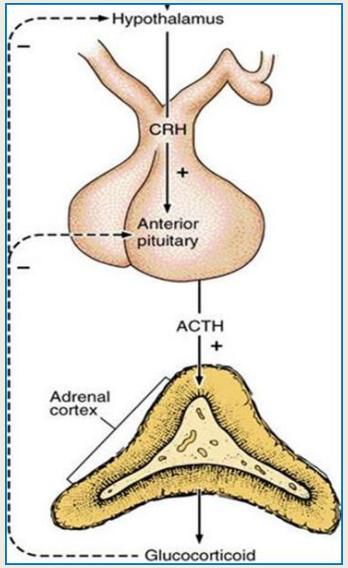


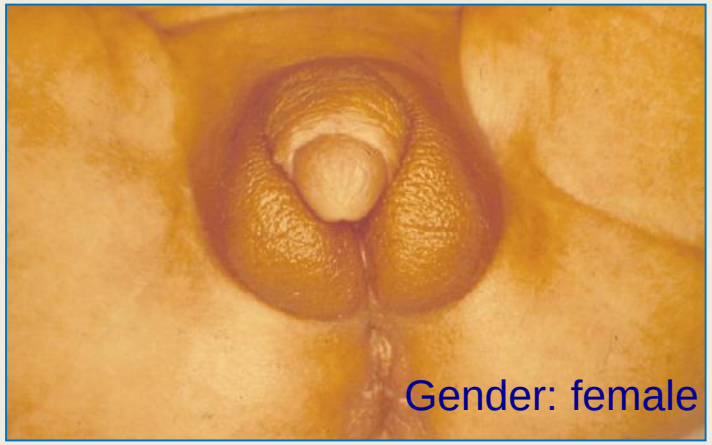


Congenital adrenal hyperplasia
(adrenogenital syndrome) – AR disease







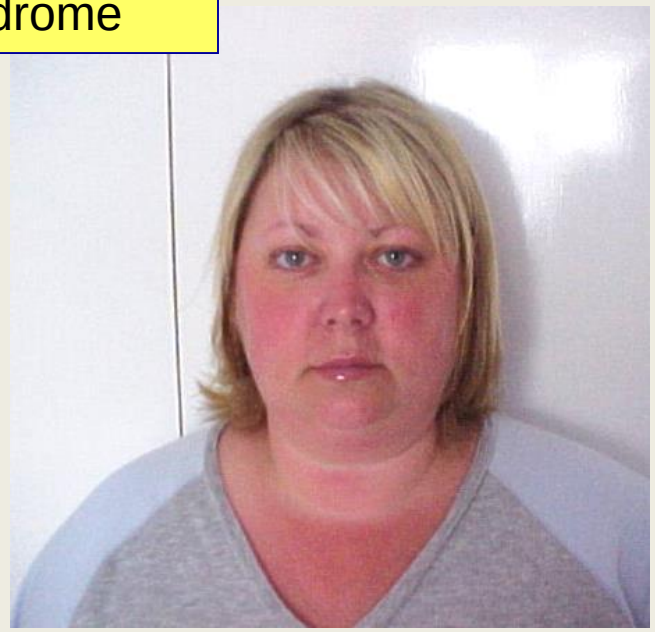


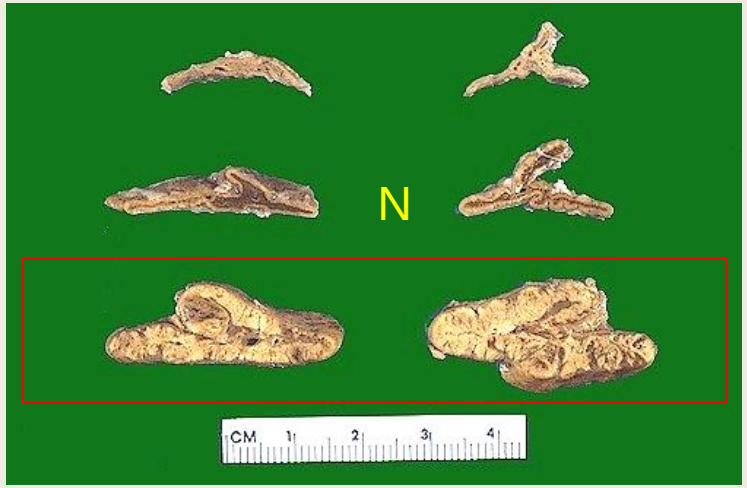
Cushing syndrome
(cortisol overproduction)

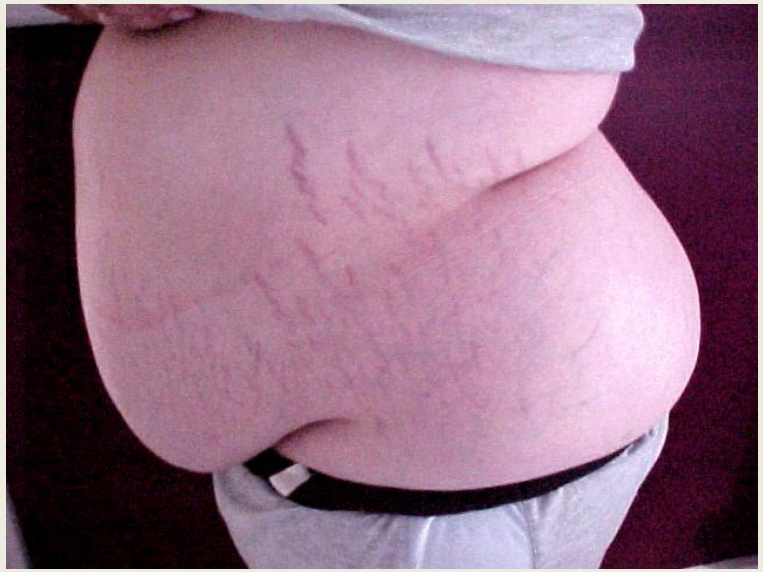
- Pituitary ACTH-adenoma (Cushing's disease)
- Adrenocortical adenoma
- Adrenocortical hyperplasia
- Paraneoplastic (lung cancer…
- Pancreatic islet cell tumor
- Exogeneous, prolonged corticosteroid treatment

osteoporosis, ulcer of stomach, delayed wound healing, immunosuppression

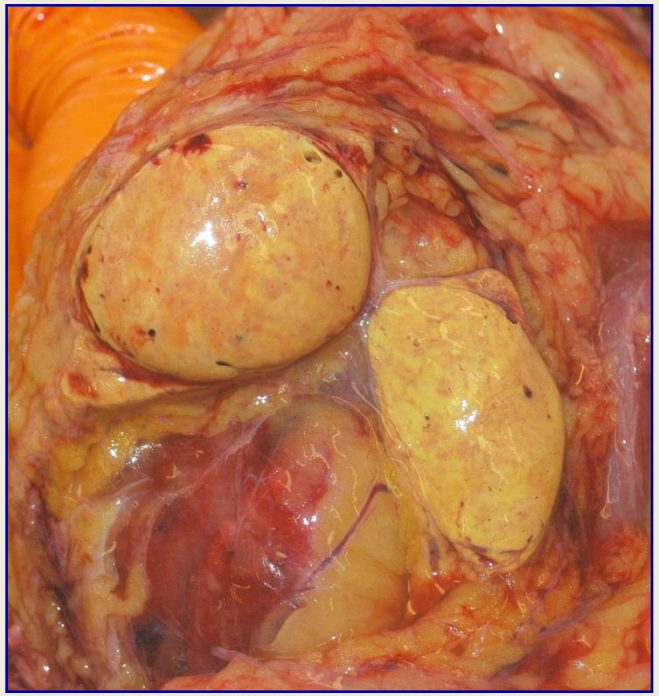
Cushing syndrome







Conn syndrome
(aldosterone overproduction)



- secondary hypertension!

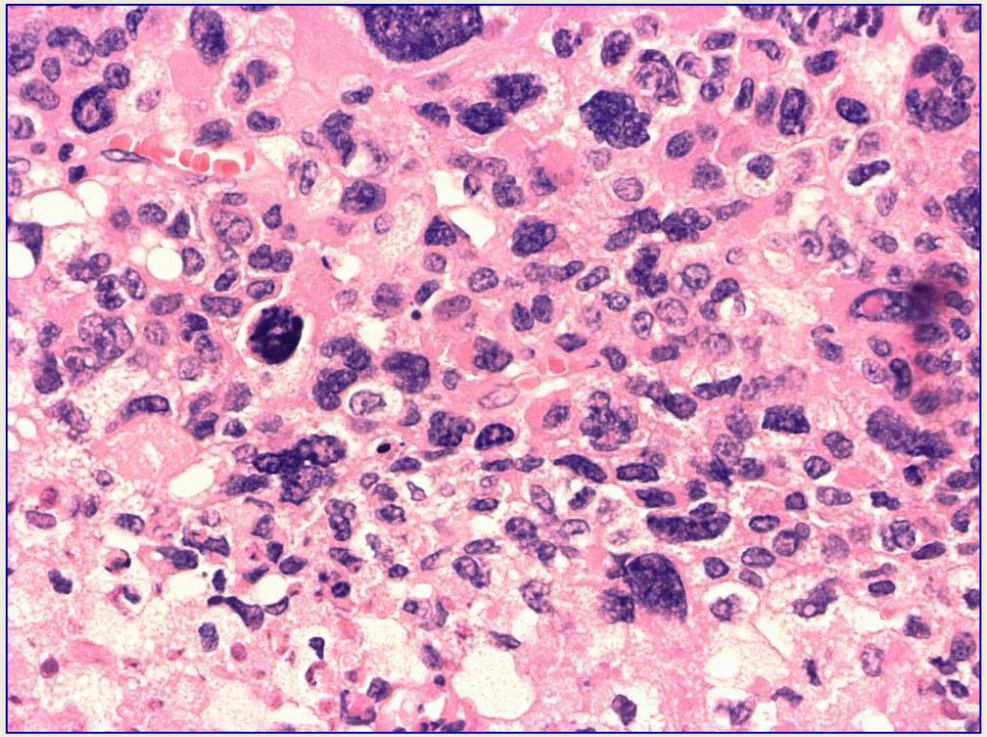
- high serum sodium level

- hypokalemia

Adrenal cortex – malignant tumors

Adrenocortical-carcinoma:

- Large, the prognosis is grave
- Hormone producing, or inactive
- Striking polymorphism!
- Metastases: liver, lung

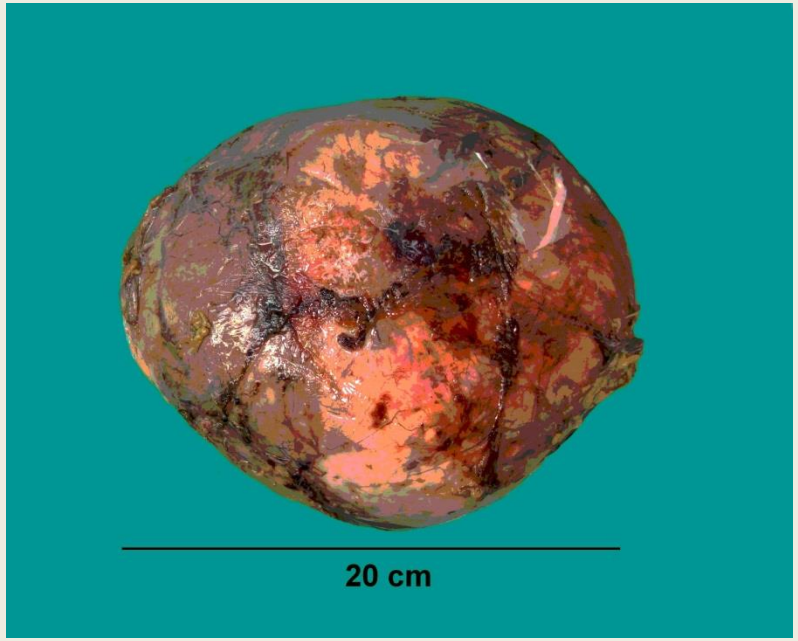


Metastatic tumors:

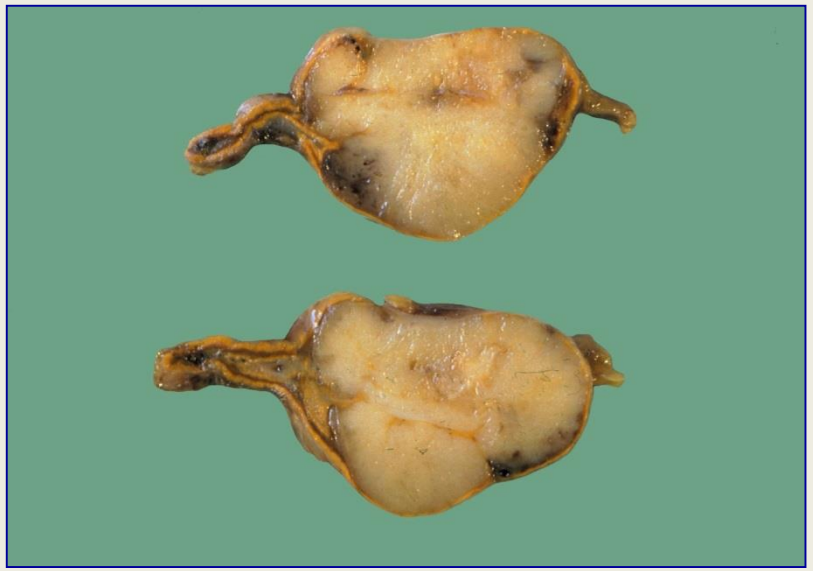
- mostly bilateral
- **lung**, stomach, pancreas
- rarely produce adrenal insufficiency

Primary adrenal insuffciency (Addison disease)

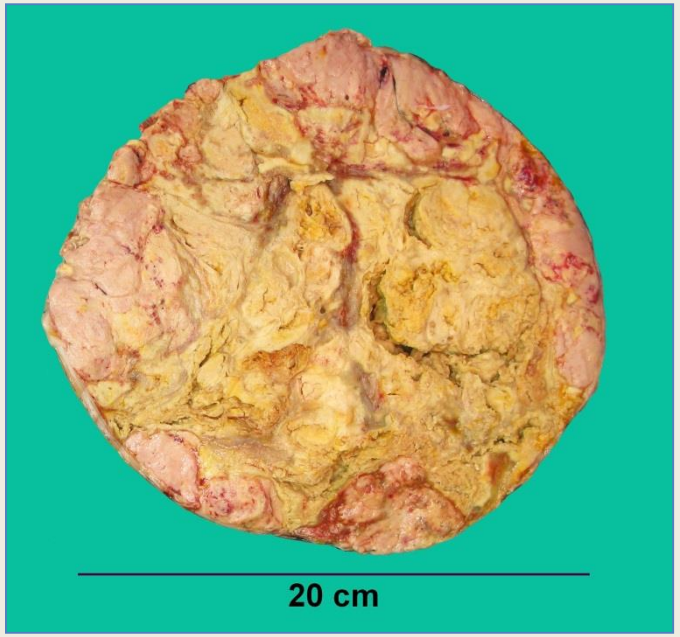
- > 90% of the cortex is destroyed
- mainly of autoimmune origin
- other causes: metastases, amyloidosis, hemorrhage, tbc
- hyperpigmented skin, hypotonia, hyponatremia, hyperkalemia

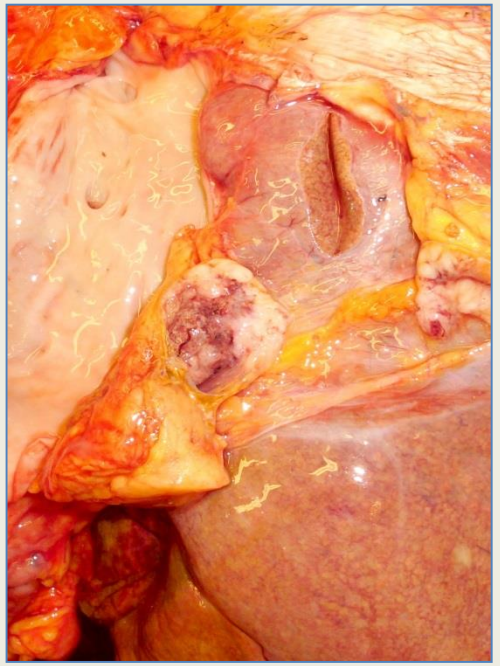


Adrenocortical carcinoma



Metastasis





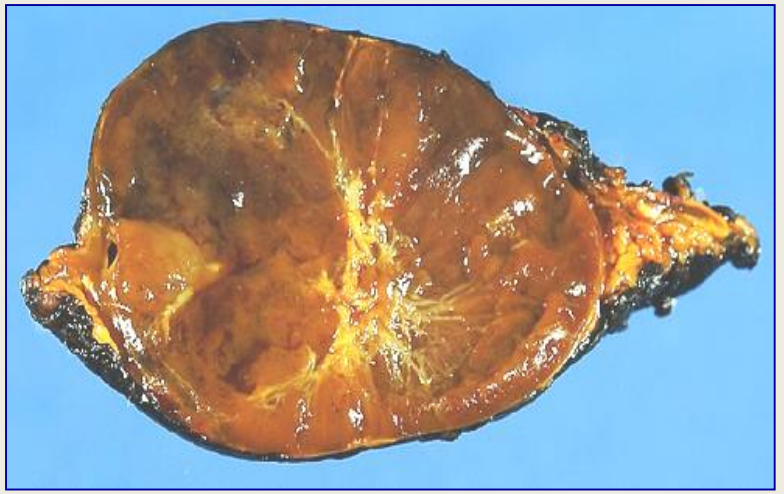
Adrenal medulla

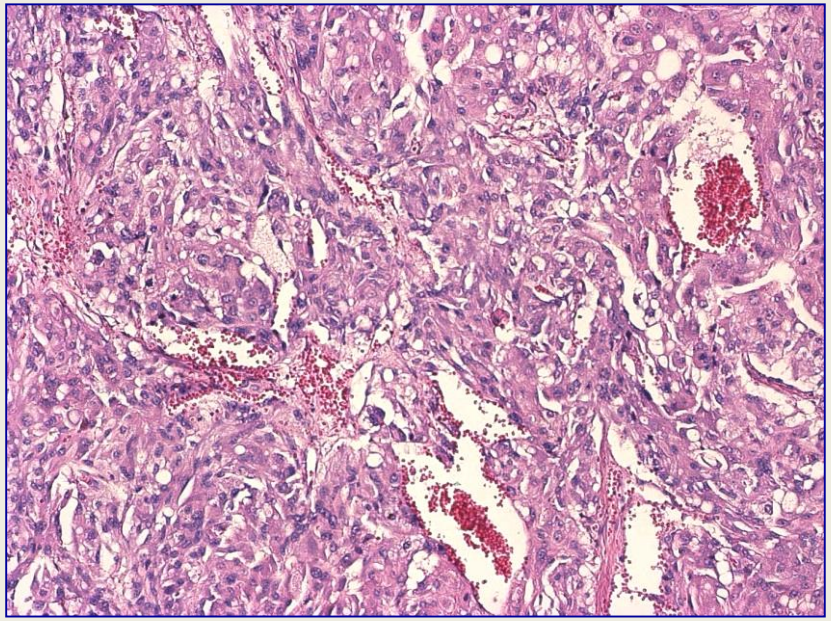
Benign: pheochromocytoma

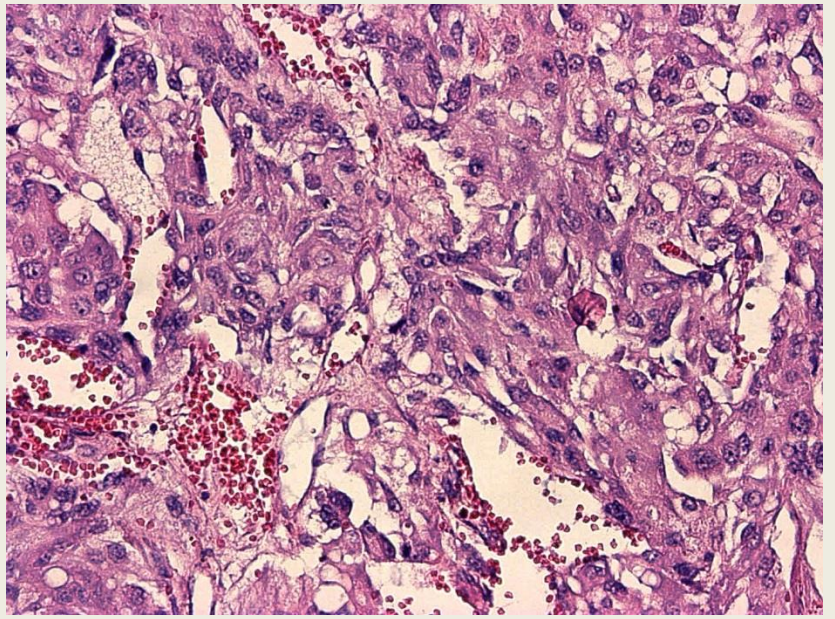
- catecholamine (adrenalin, NA) overproduction
- paroxysmal or permanent hypertension
- sharply circumscribed

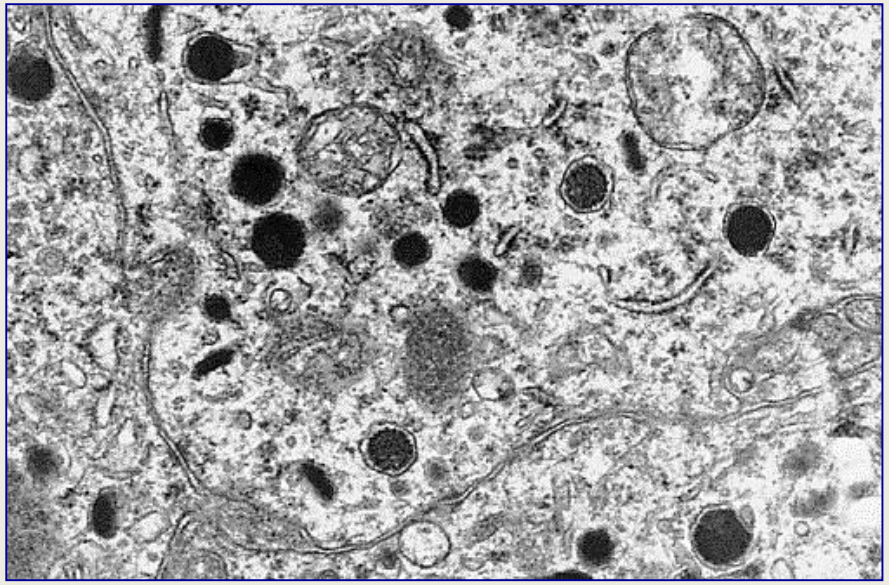
„10% rule”

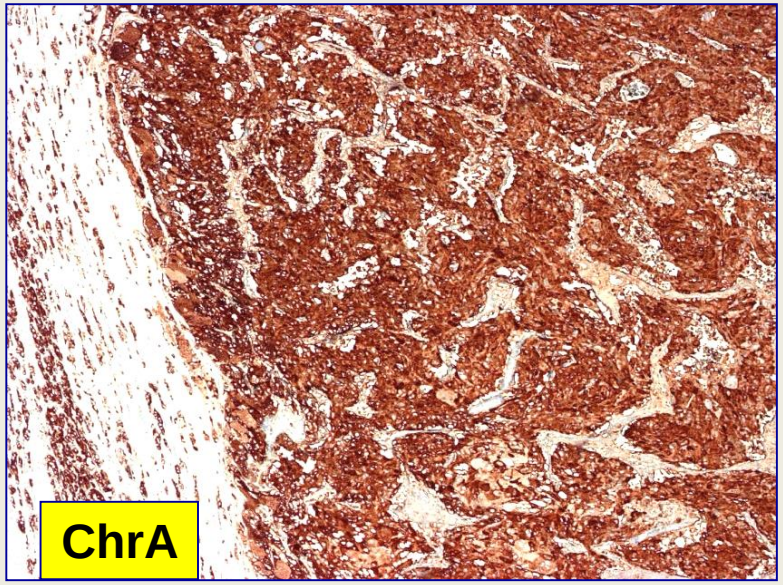
(10% bilateral, 10% part of MEN-2 syndrome,
10% extraadrenal, 10% malignant)

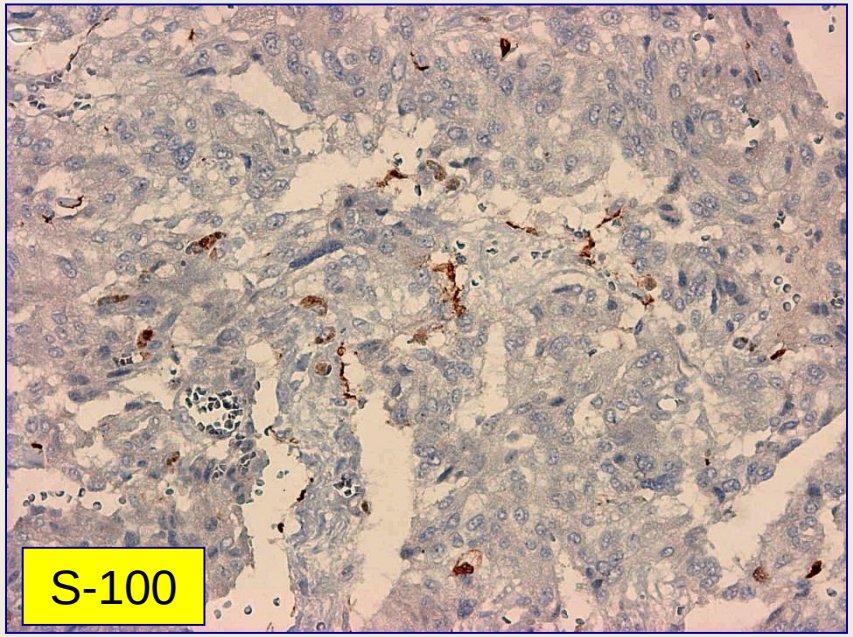






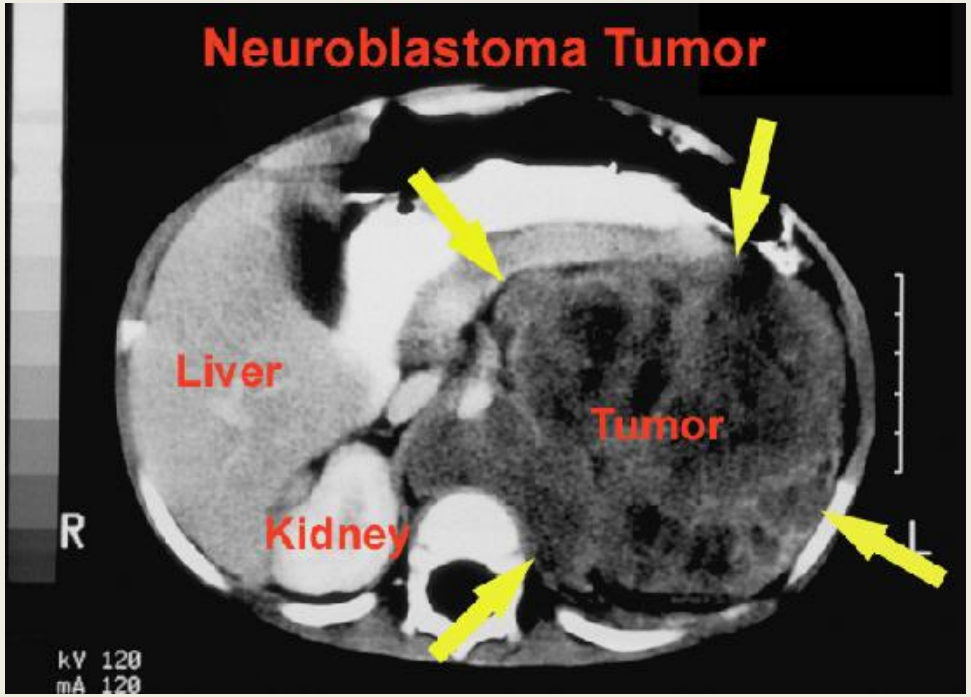


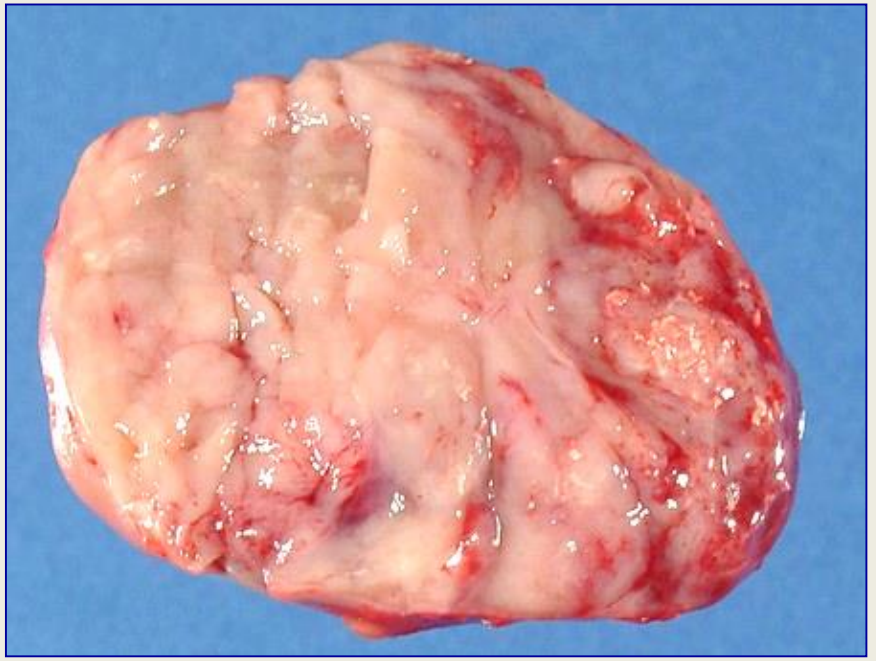

ChrA

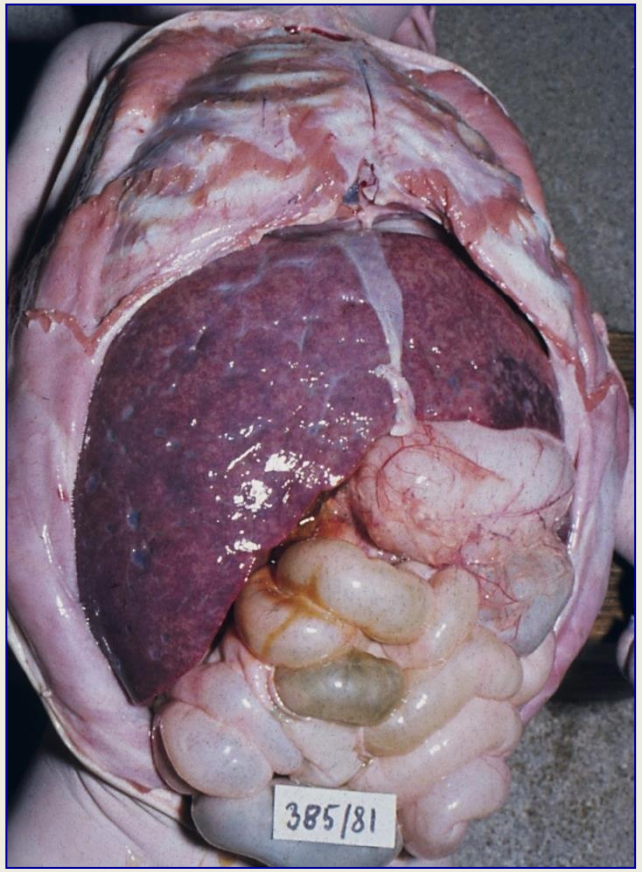

S-100

Adrenal medulla - neuroblastoma

- 2nd most frequent childhood malignant tumor
- Peak incidence: year 1-2
- Presents as a palpable abdominal mass
- Belongs to the small round blue cell tumor family („penaut” tumors)
- Lymphogenic and hematogenic metastases



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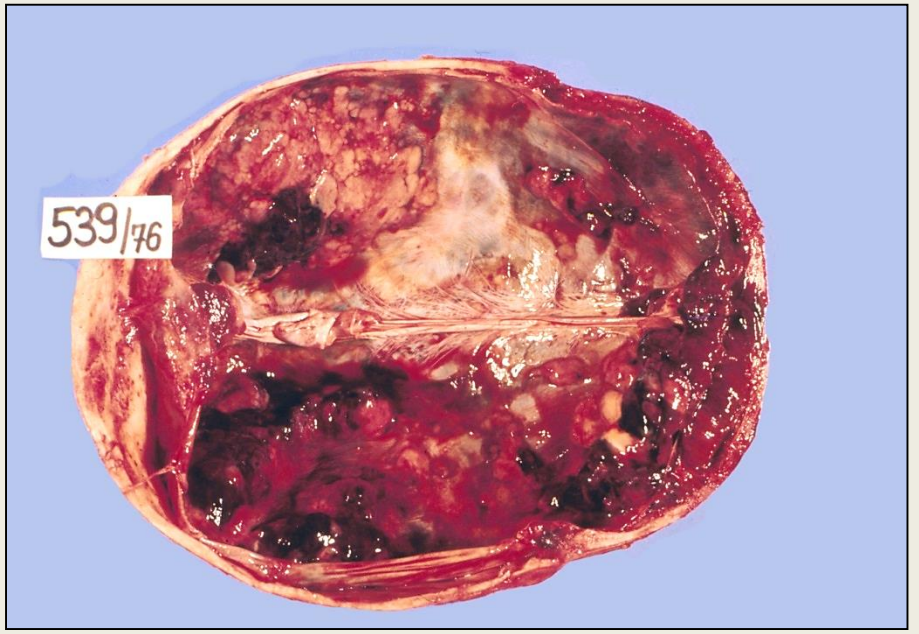
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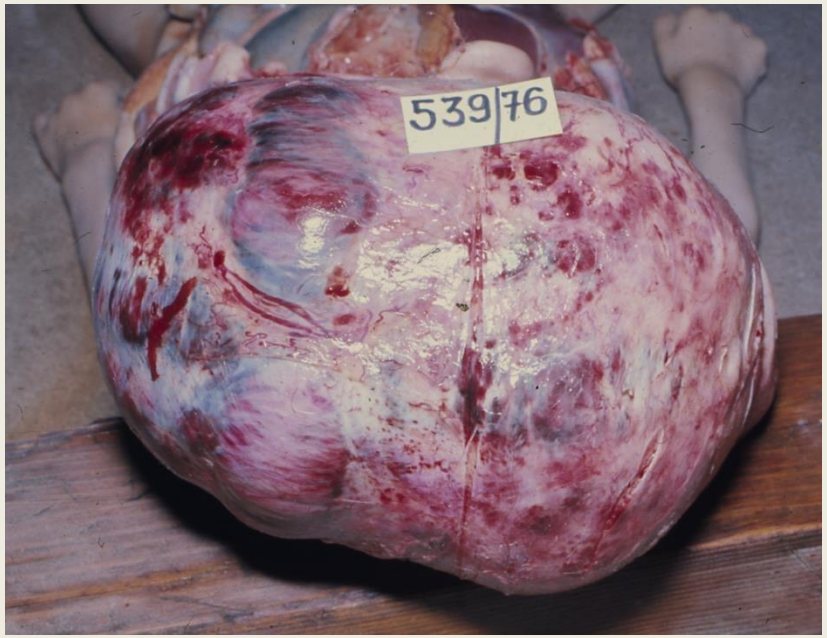
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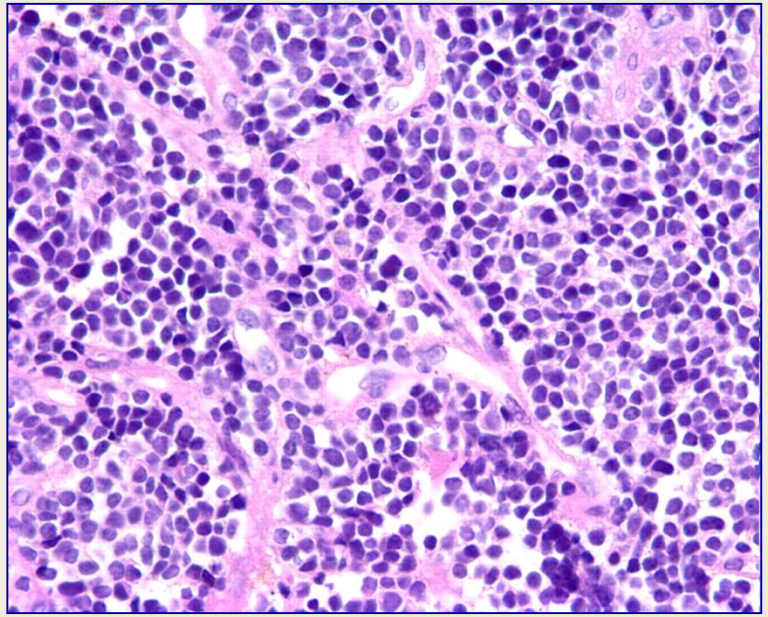
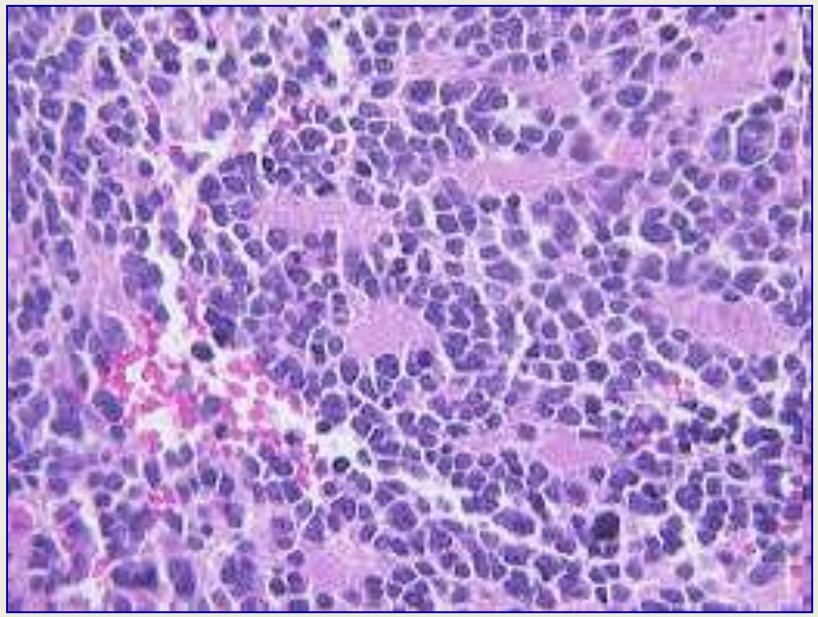
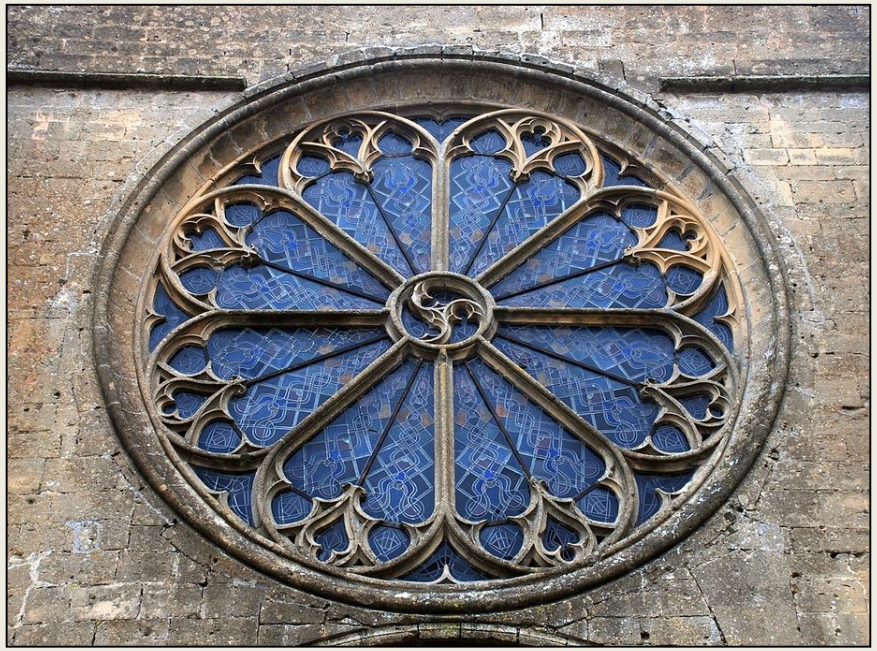
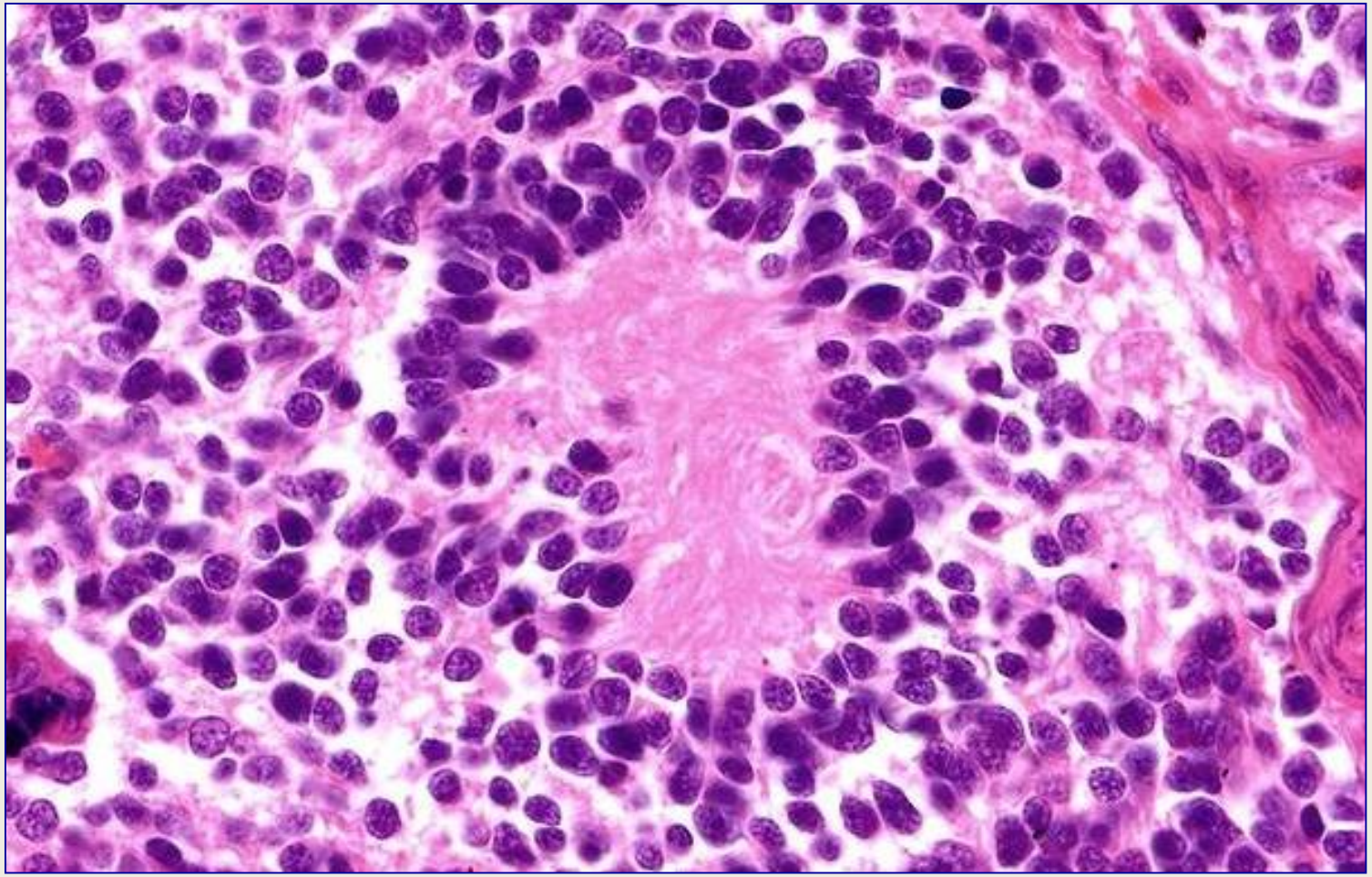




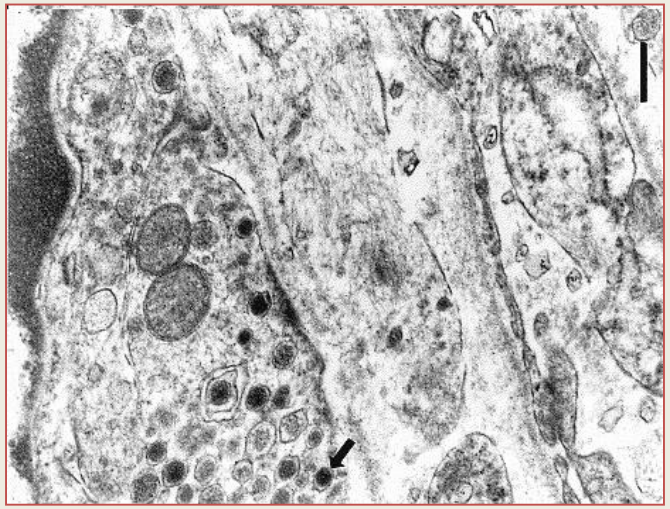
Homer Wright
pseudorosettes

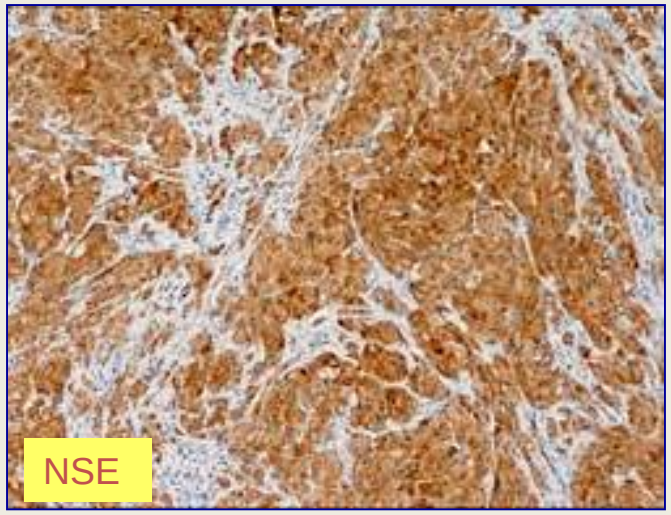


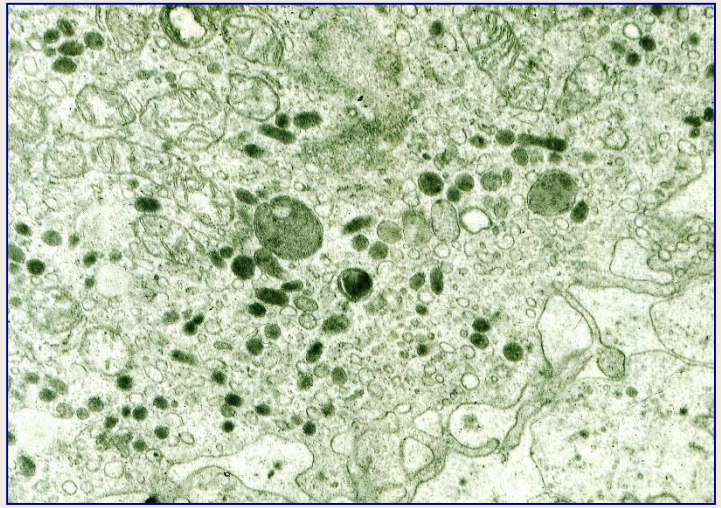
rosette



Adrenal medulla - neuroblastoma





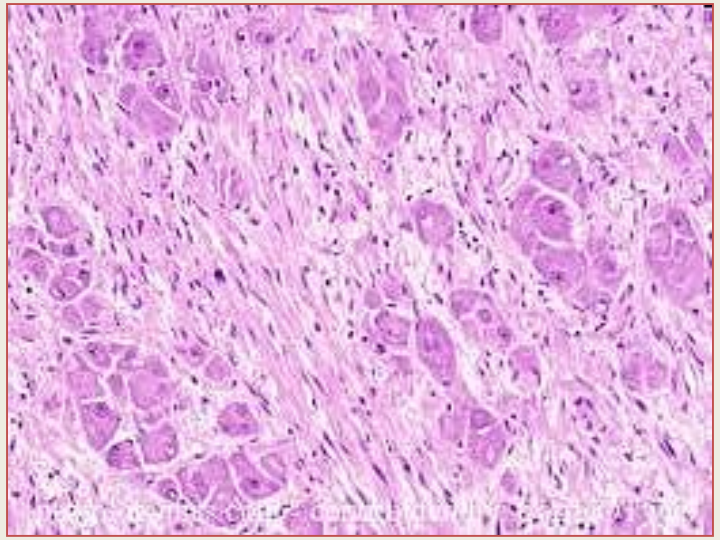


Adrenal medulla - neuroblastoma

Expected prognosis:

- stroma-rich (Schwann-cell differentiation; favorable prognosis) - stroma-poor
- mitotic activity and calcification
- overexpression of N-myc protooncogene (25 % - unfavorable prognosis)

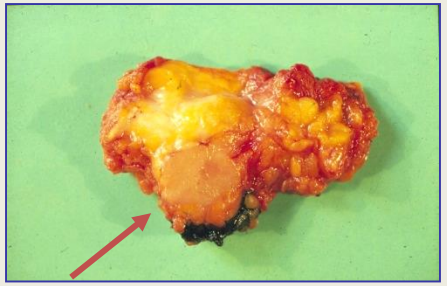
Neuroblastoma → ganglioneuroblastoma → ganglioneuroma



MEN SYNDROMES

MEN-1 (Wermer syndrome)

- autosomal dominant trait (MEN-1 gene inactivation)
- pituitary adenoma + parathyroid hyperplasia + pancreatic islet cell tumor
  (prolactinomas, GH-adenoma) nonfunctioning, PPoma, frequently multicentric

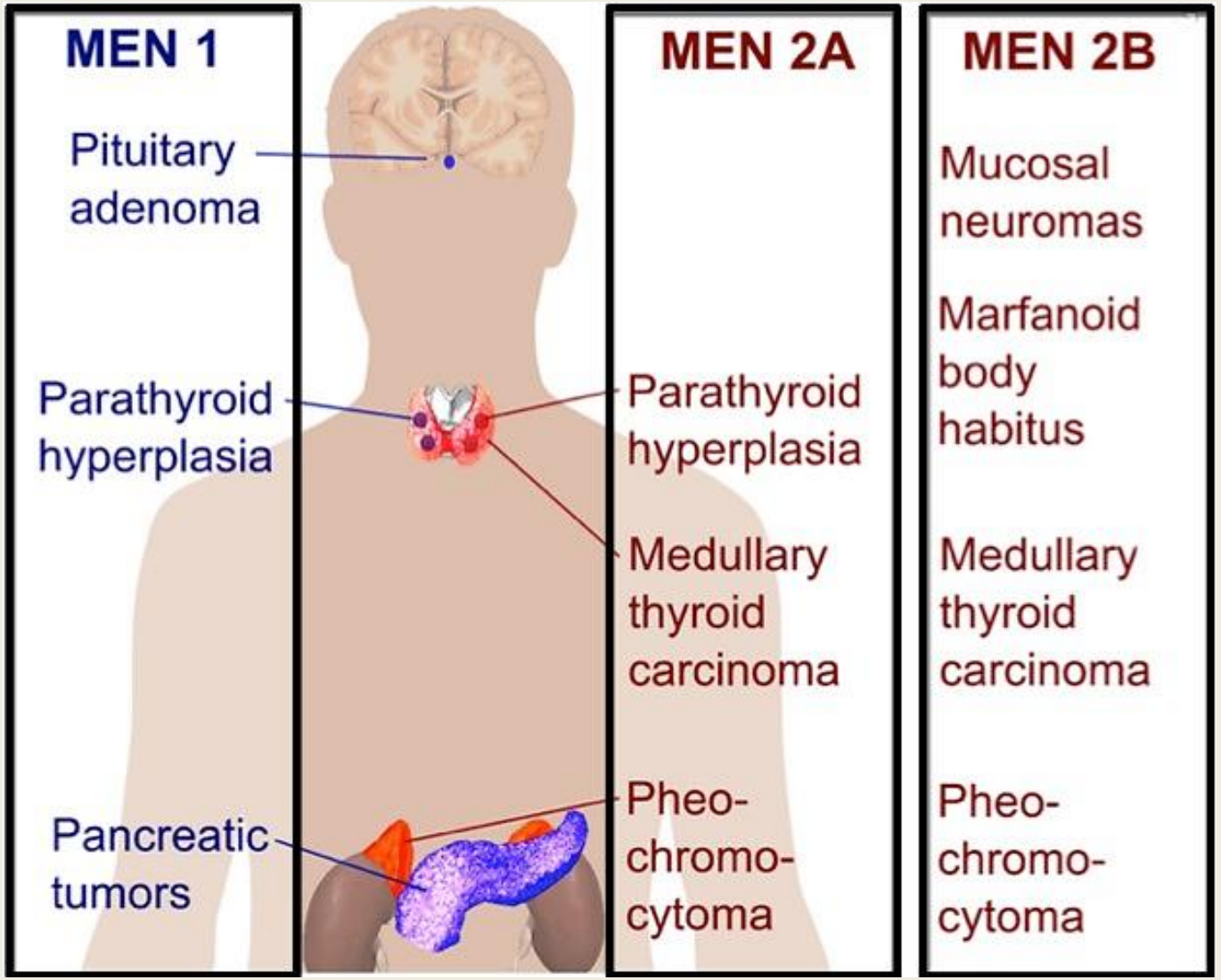


MEN-2A (Sipple-syndrome)

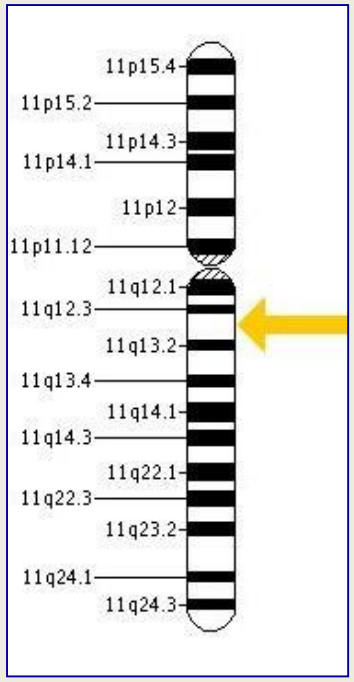
- autosomal dominant
- RET-protooncogen (germline mutation) activation
- bilateral pheochromocytoma + bilateral thyroid medullary cancer + parathyroid hyperplasia

MEN-2B

- autosomal dominant
- pheochromocytoma + medullary thyroid cancer + intestinal ganglioneuromas + mucosal neuromas

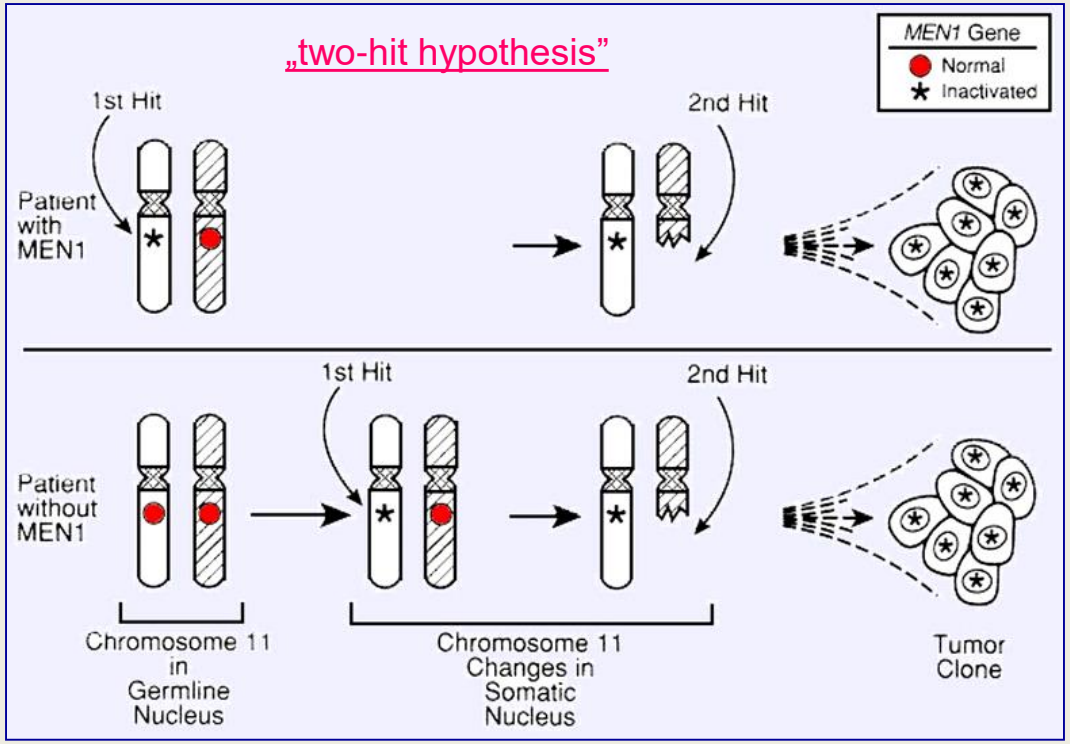

MEN 1
Pituitary adenoma
Parathyroid hyperplasia
Pancreatic tumors
MEN 2A
Parathyroid hyperplasia
Medullary thyroid carcinoma
Pheo-chromo-cytoma
MEN 2B
Mucosal neuromas
Marfanoid body habitus
Medullary thyroid carcinoma
Pheo-chromo-cytoma

MEN-1 GENE

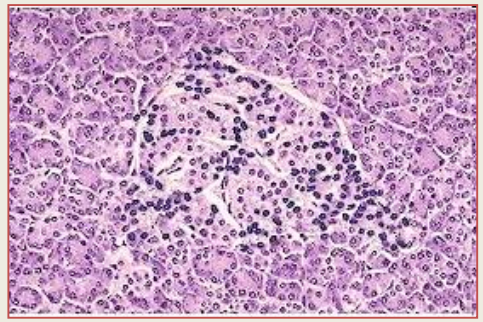


Chr. 11

- MEN-1 tumor suppressor gene
- menin protein
- downregulates AKT-induced kinase activity
- suppresses proliferation and anti-apoptosis
- over 1300 mutations (LOH)



Diabetes mellitus



Type I. (insulin dependent; juvenile) - autoimmun origin

Type II. (non insulin dependent; adult) - receptor insensitivity

Gestational diabetes

Secondary diabetes forms (hemochromatosis, hemorrhagic pancreatitis, chronic fibrotizing pancreatitis, cystic fibrosis, glucagonoma…)

Late compllications

accelerated atherosclerosis

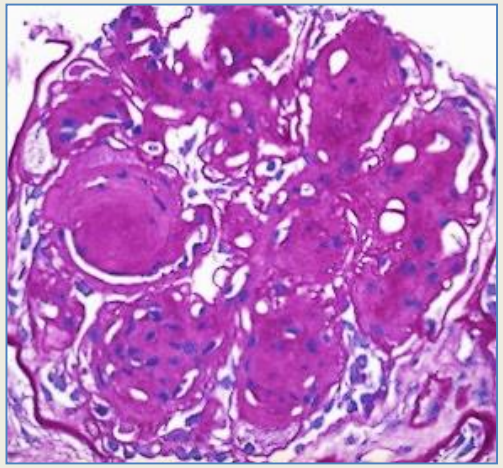
microangiopathy (retinal microaneurysm, proliferative lesions)

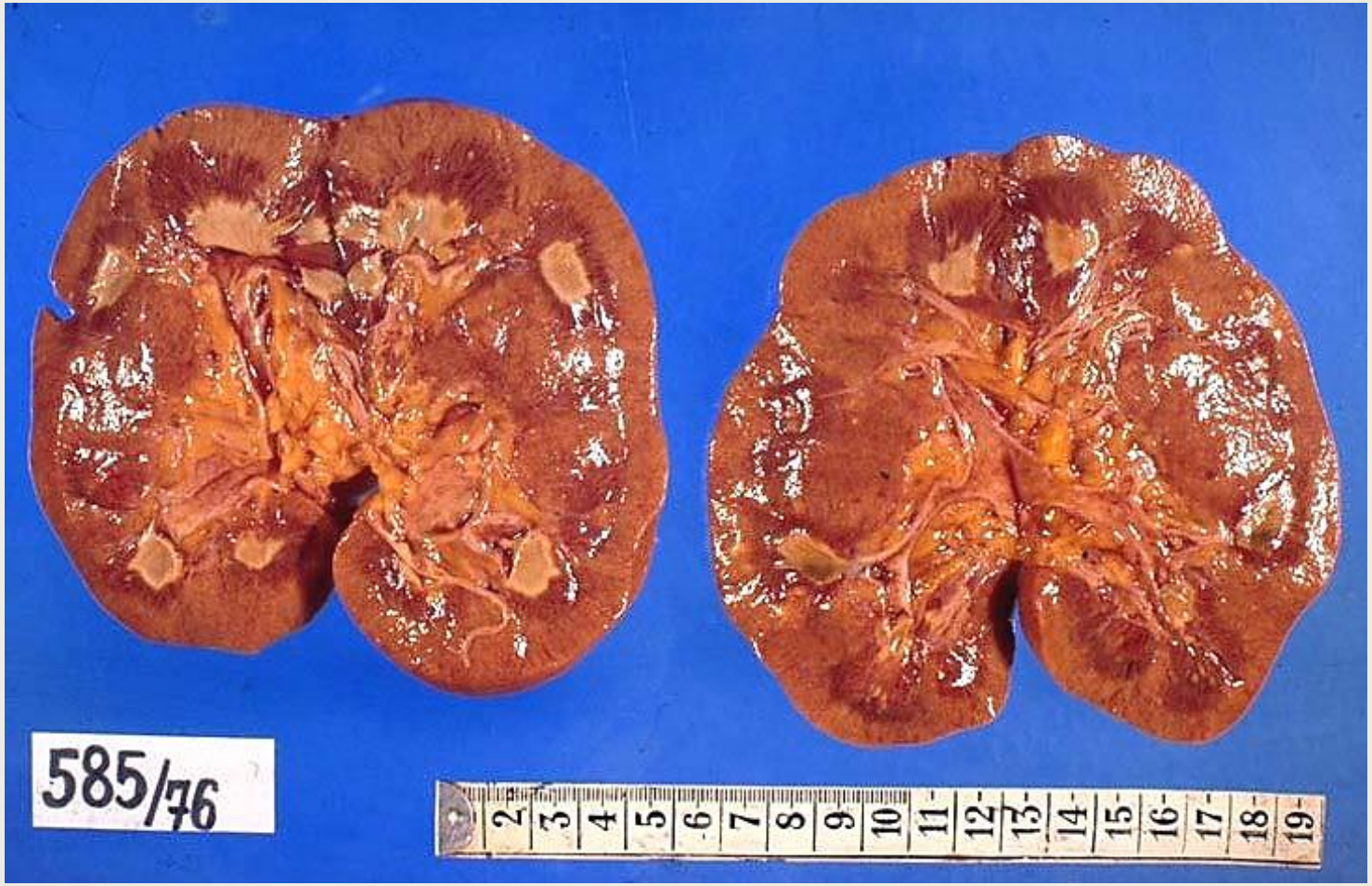
papillanecrosis

Kimmelstiel-Wilson syndrome

diabetic foot

recurrent pyodermas, Candidiasis

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