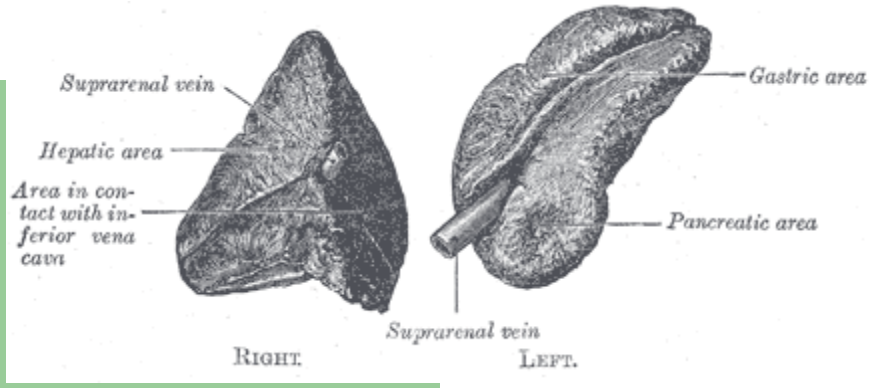


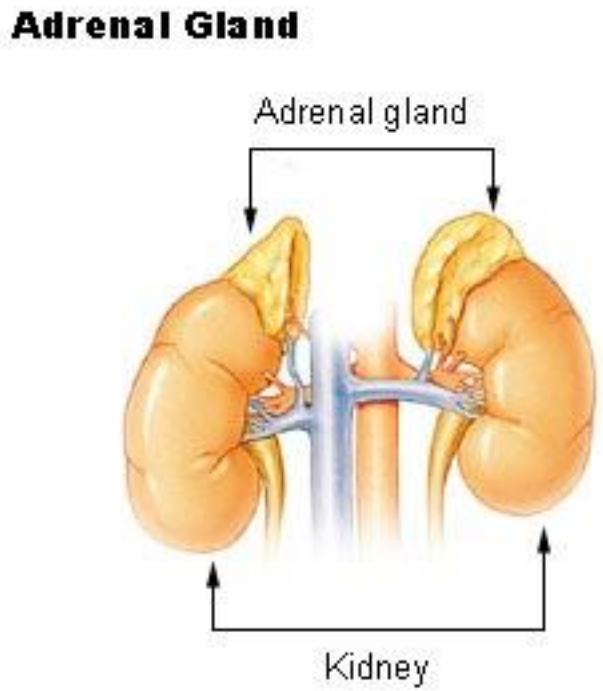
THE HIGHS AND LOWS OF ADRENAL GLAND PATHOLOGY

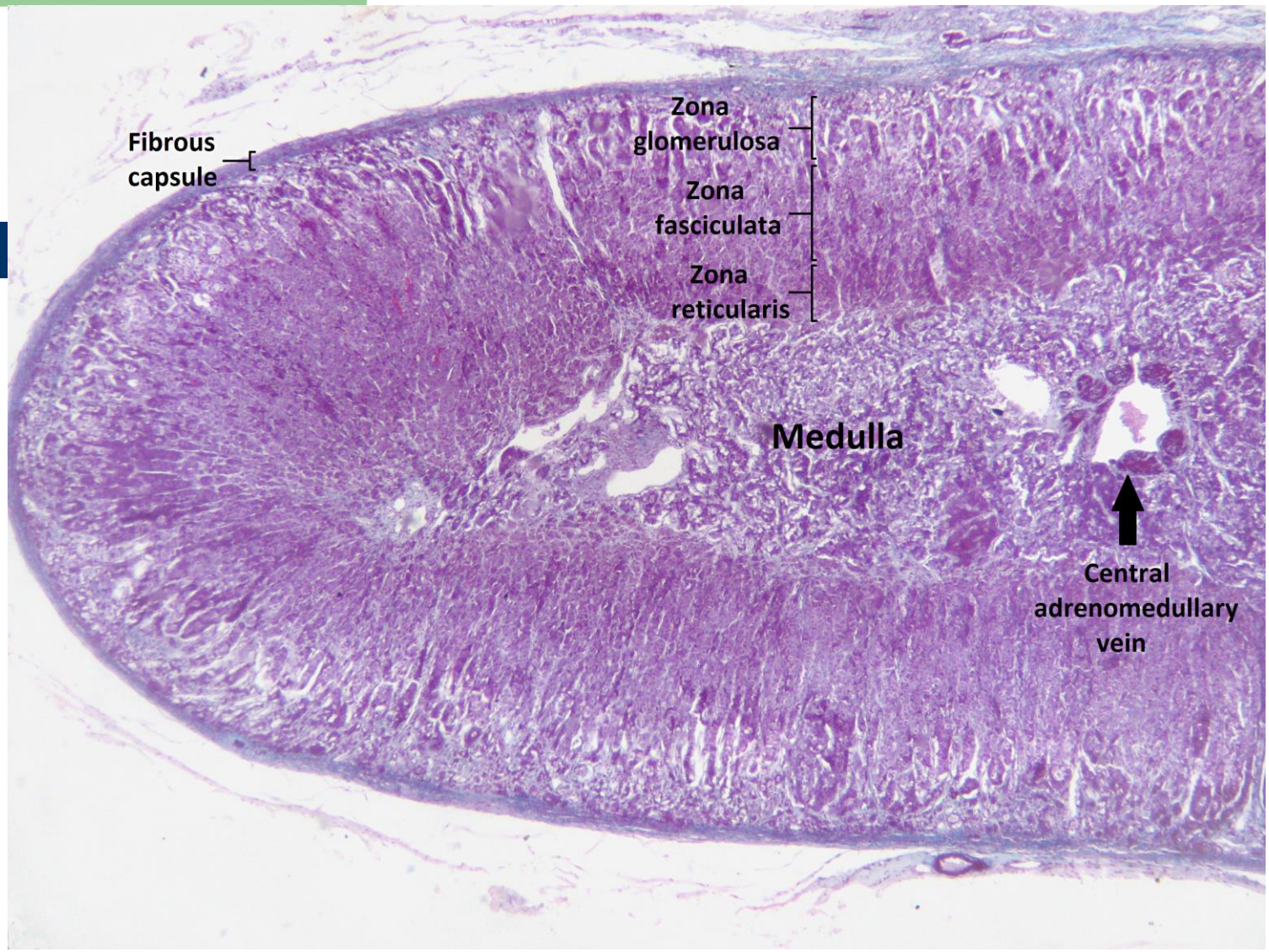
Symptoms of Adrenal Gland Disorders

- Depends on whether it is making too much or too little hormone
- And on what you Google!
- Symptoms include obesity, skin problems, high blood pressure, moodiness, depression,muscle and bone weakness, high blood sugar, headache, excessive urination,rapid heart rate, anxiety, fatigue, hair thinning, dizziness, low blood pressure, patches of darker skin

The Adrenals

- Sit in the retroperitoneal tissues at upper poles of kidneys at level of 12th vertebra
- Normally weighs 4g at surgery or sudden death but can range between 2g and 6g
- At hospital autopsy the average weight is 6g due to the stress of terminal illness

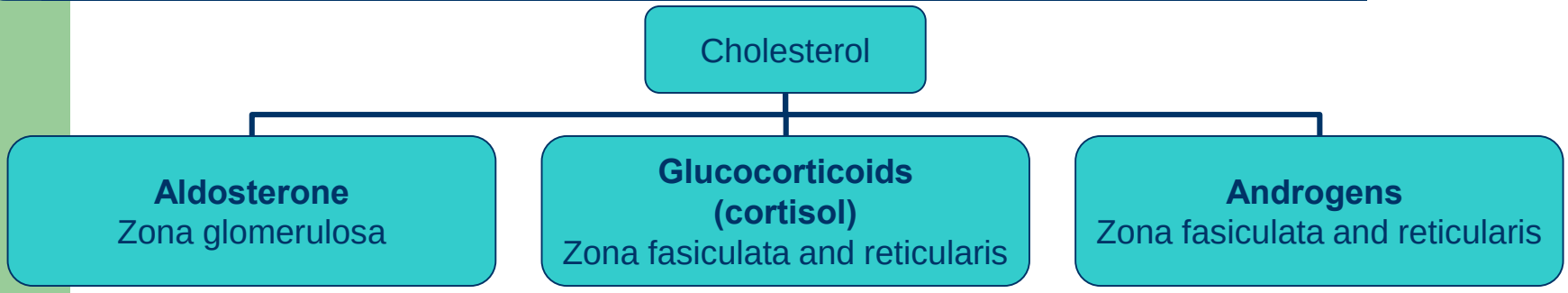



Fibrous capsule
Zona glomerulosa
Zona fasciculata
Zona reticularis
Medulla
Central adrenomedullary vein

Hormone Synthesis

- The cortex synthesises 3 main groups of hormones which are all derived from cholesterol
- GLUCOCORTICOIDS eg cortisol
- MINERALOCORTICOIDS eg aldosterone
- ANDROGENS
- The pattern of hormone production differs between the three zones due to differential expression of enzymes

Steroidogenesis

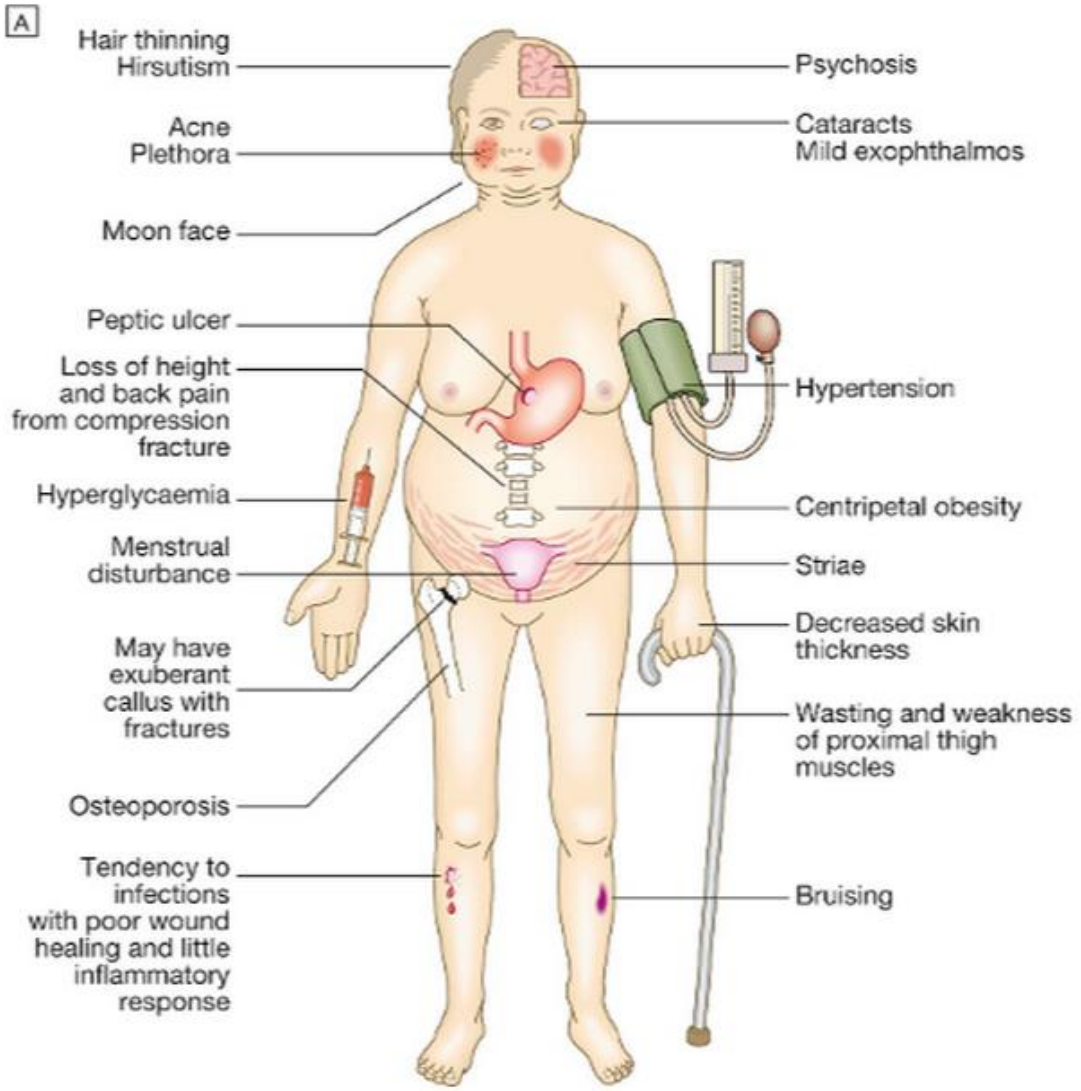


High levels of cortisol (Cushings Syndrome)

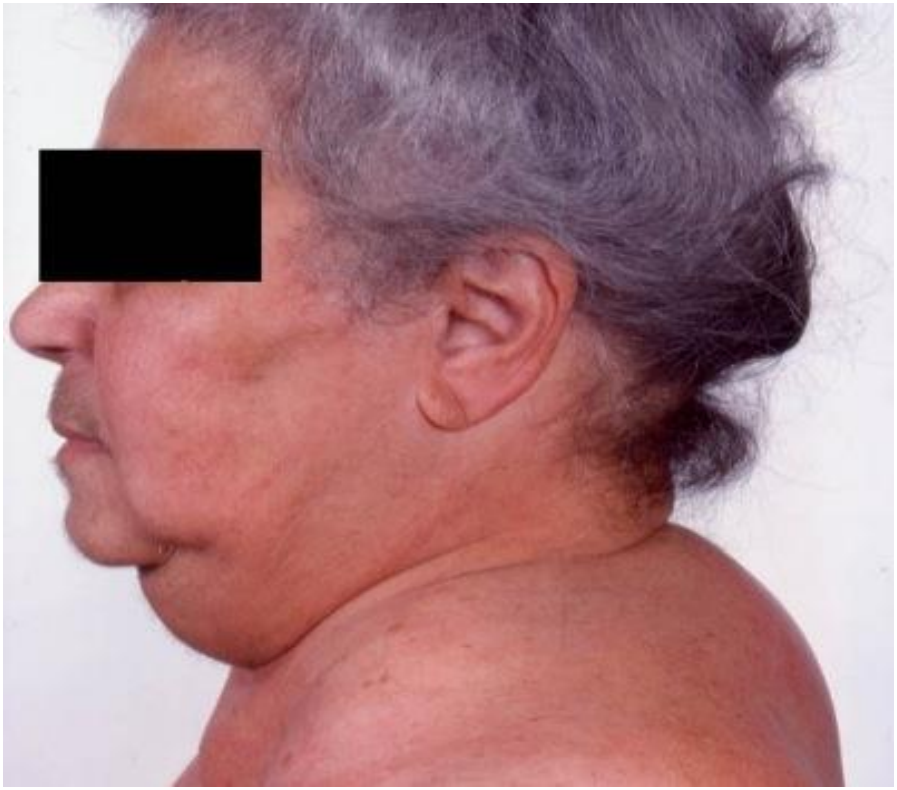
- First described by Harvey Cushing in 1932
- Most cases (80%) are due to increased hormone production from a pituitary adenoma (Cushing's disease).
- The remaining cases (20%) are due to cortisol secretion from an adrenal tumour

Cushing's Syndrome

- Weight gain, central obesity, facial rounding, plethora, easy bruising, thin skin, poor wound healing, purple striae, proximal muscle weakness, emotional changes, hypertension, fungal infections, hirsutism.


A
Hair thinning
Hirsutism
Acne
Plethora
Moon face
Peptic ulcer
Loss of height and back pain from compression fracture
Hyperglycaemia
Menstrual disturbance
May have exuberant callus with fractures
Osteoporosis
Tendency to infections with poor wound healing and little inflammatory response
Psychosis
Cataracts
Mild exophthalmos
Hypertension
Centripetal obesity
Striae
Decreased skin thickness
Wasting and weakness of proximal thigh muscles
Bruising

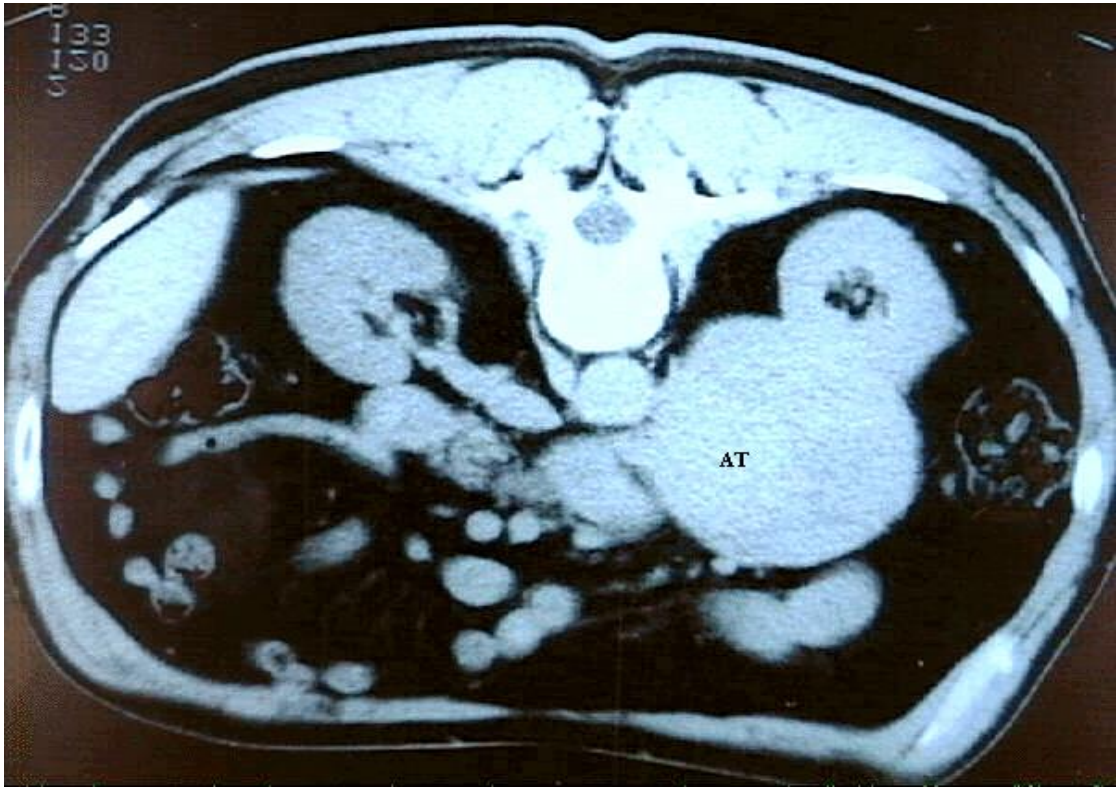
Cushing's syndrome



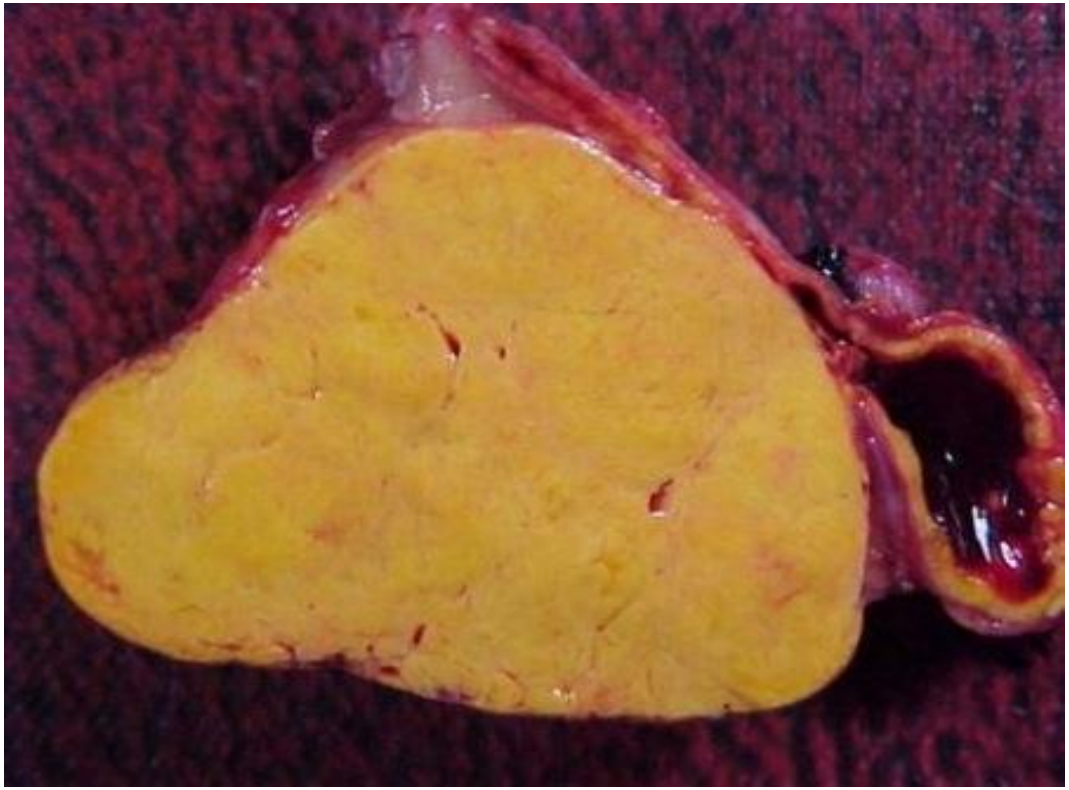
High levels of aldosterone (Conn's syndrome)

- Aldosterone overproduction results in high blood pressure and low potassium levels.
- Symptoms include muscle weakness, cramping, headaches, increased thirst and urination.

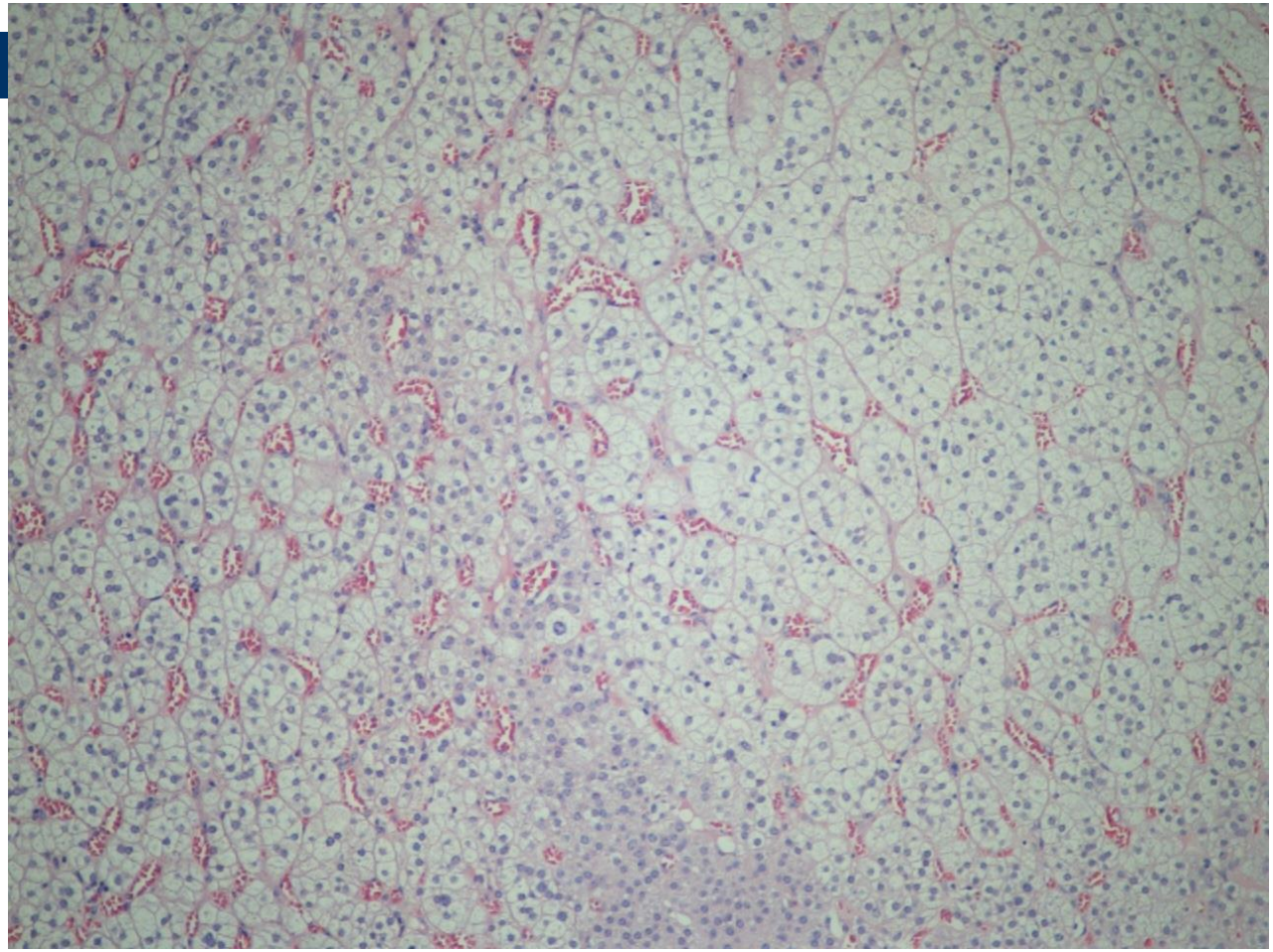
CT of adrenal mass



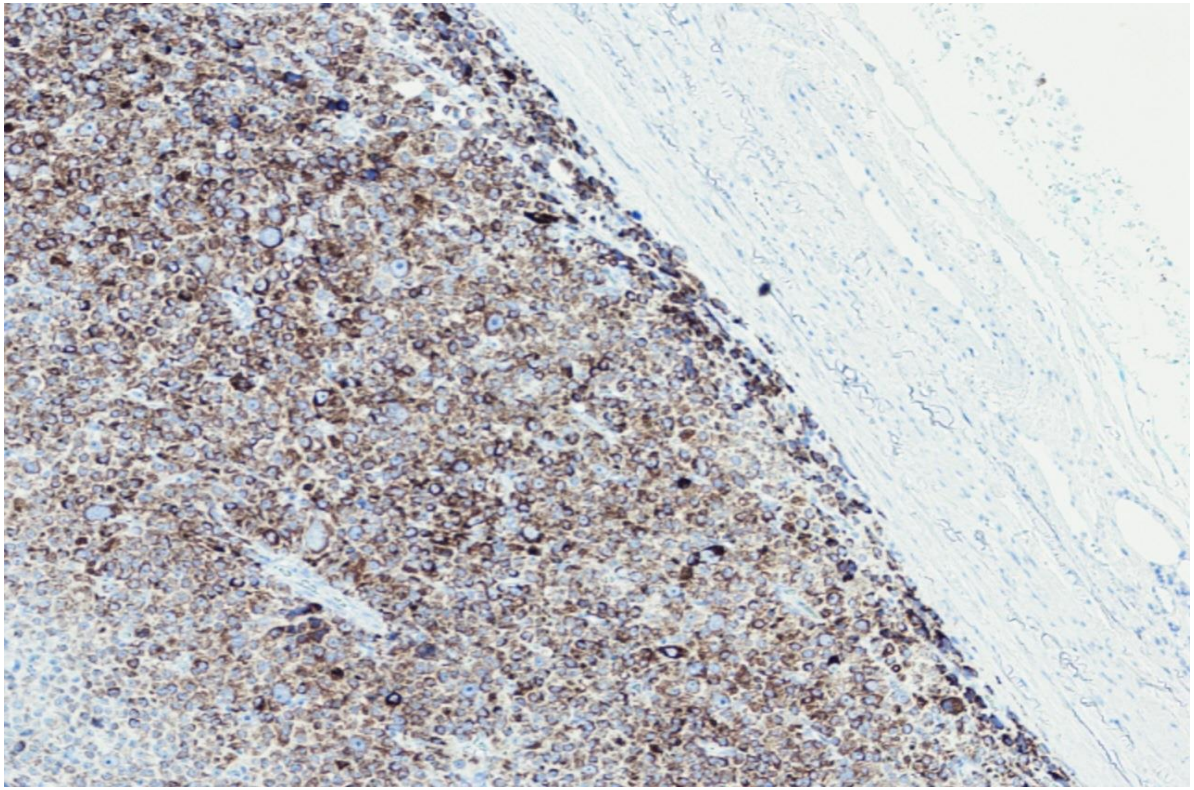
Adenoma in Conn's Syndrome



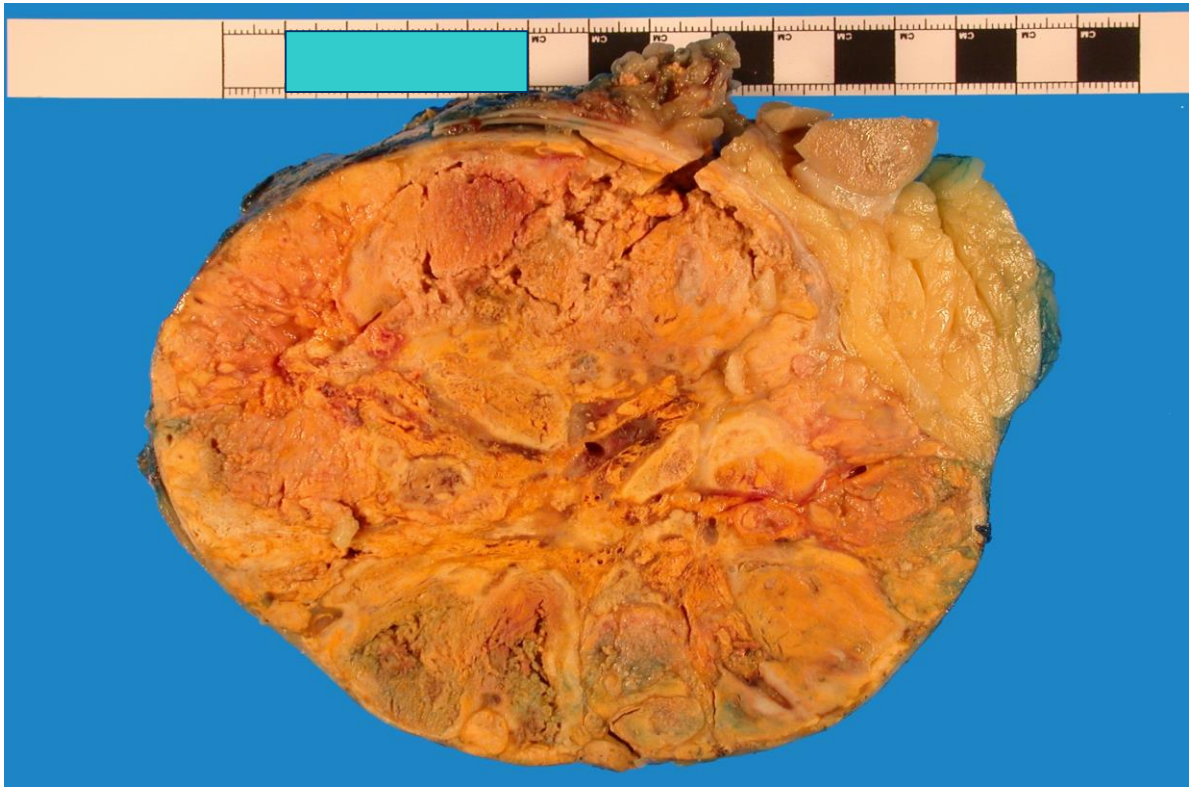
Cortical adenoma



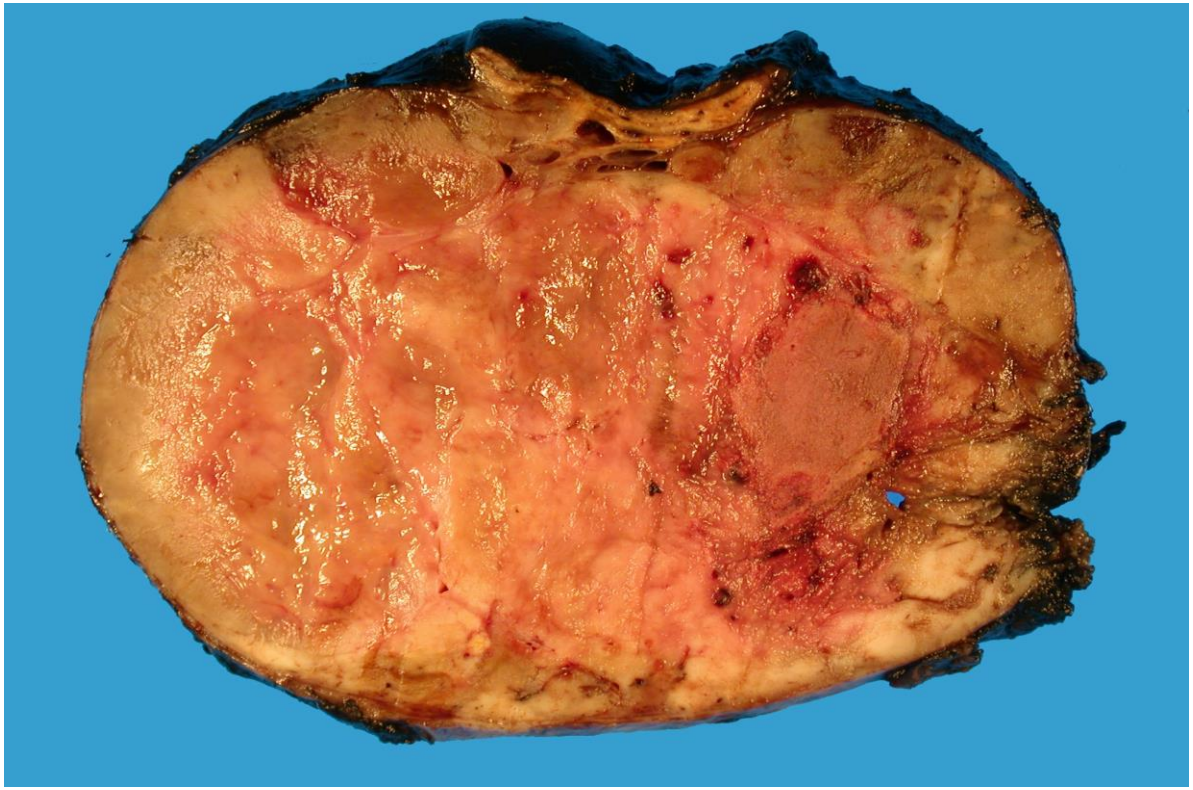
Melan A



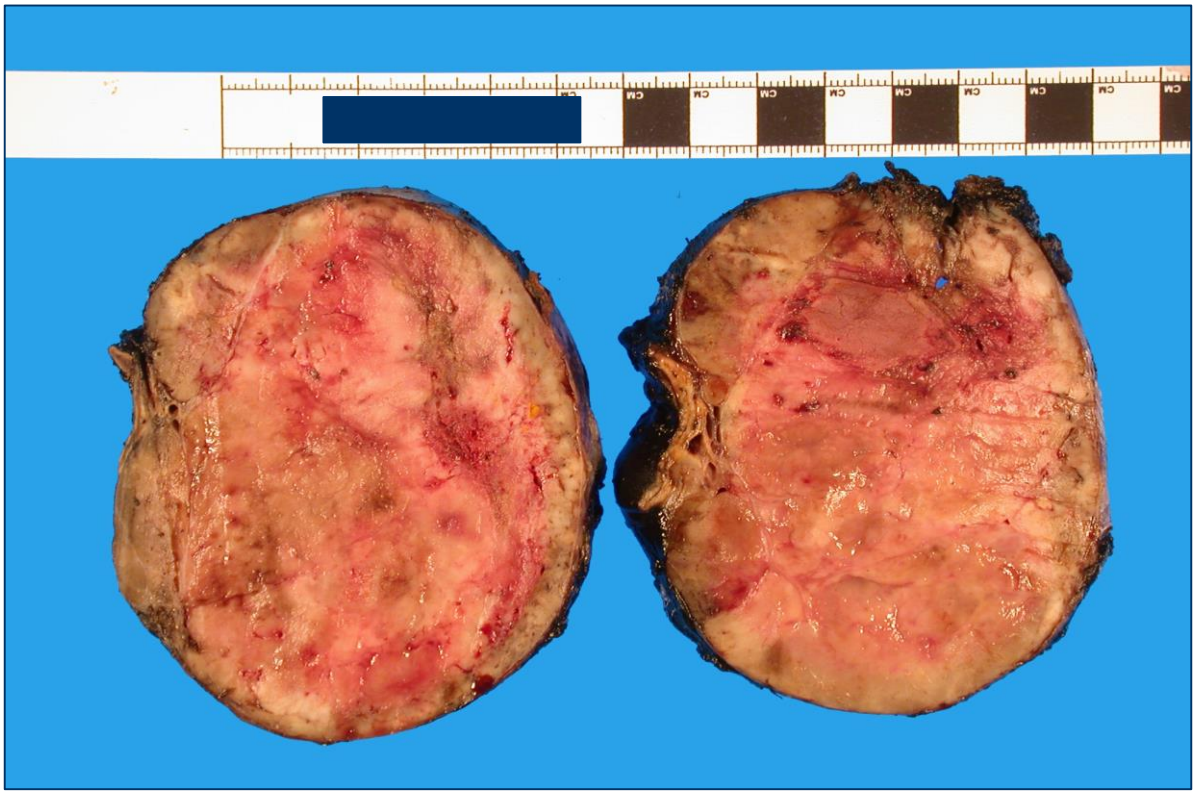
Cortical carcinoma



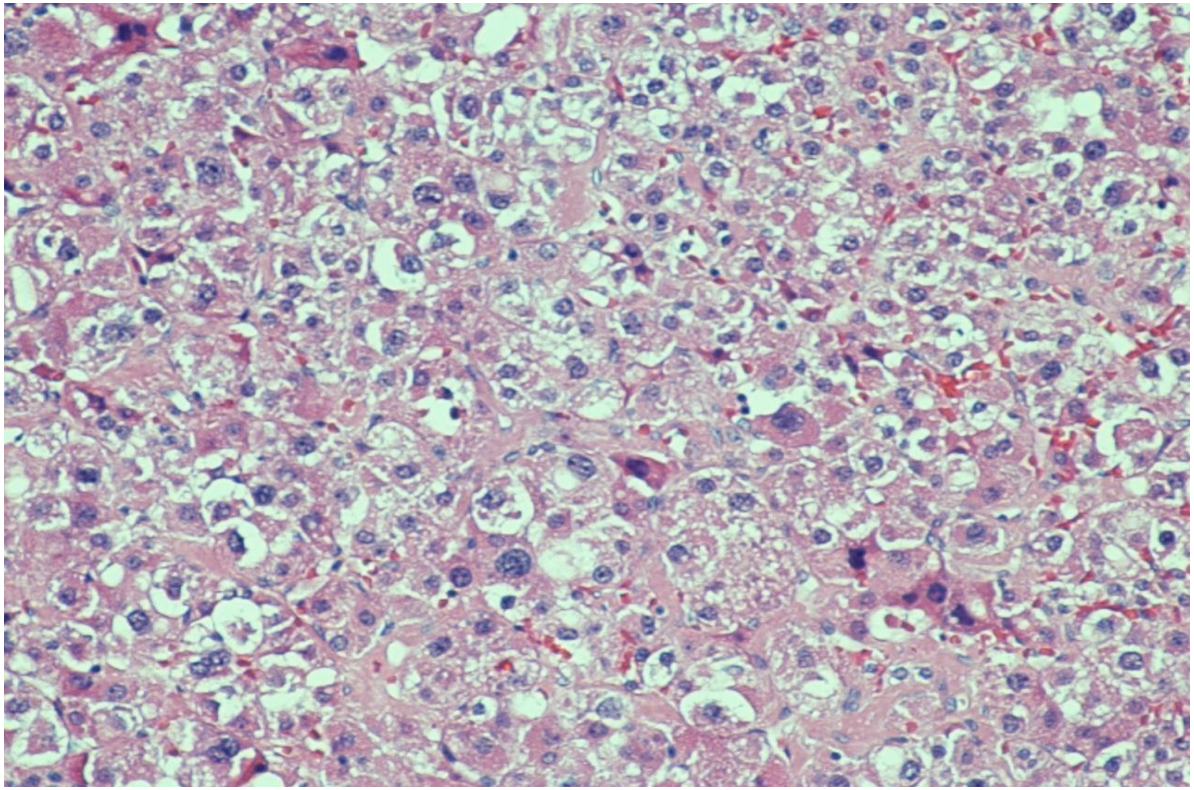
Cortical carcinoma



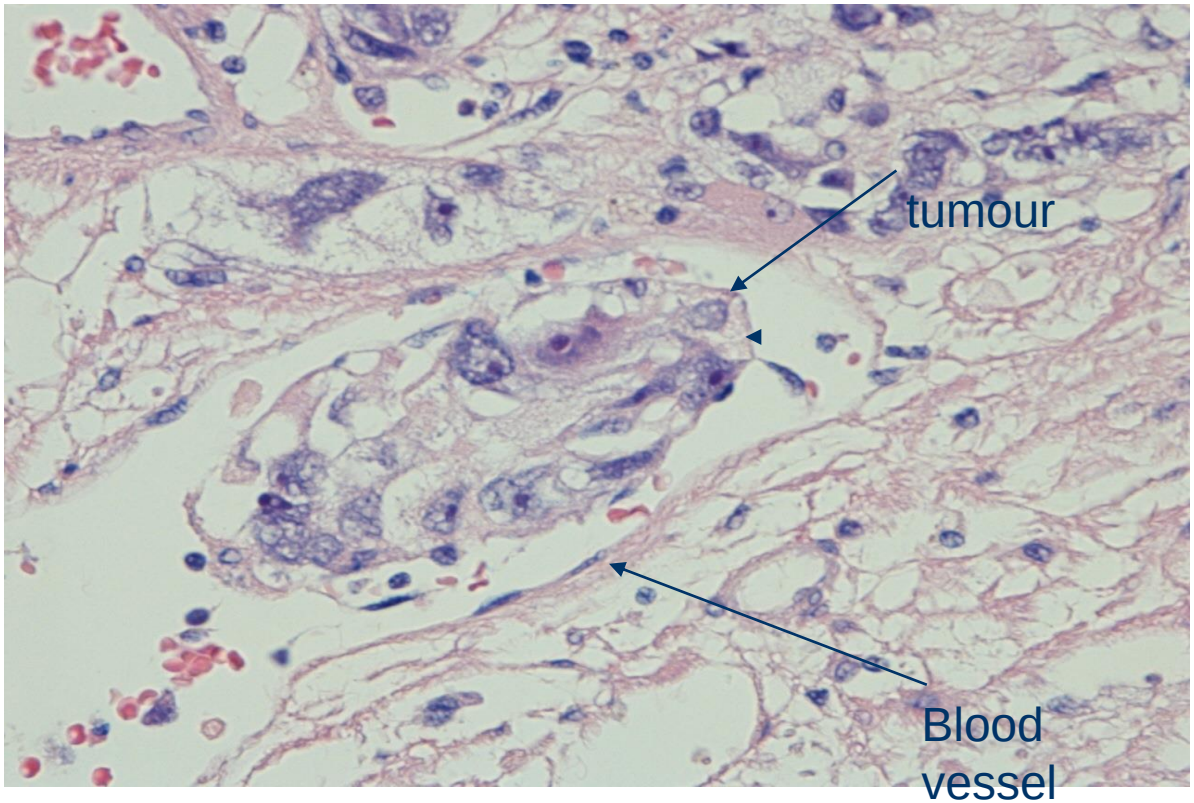
Cortical carcinoma



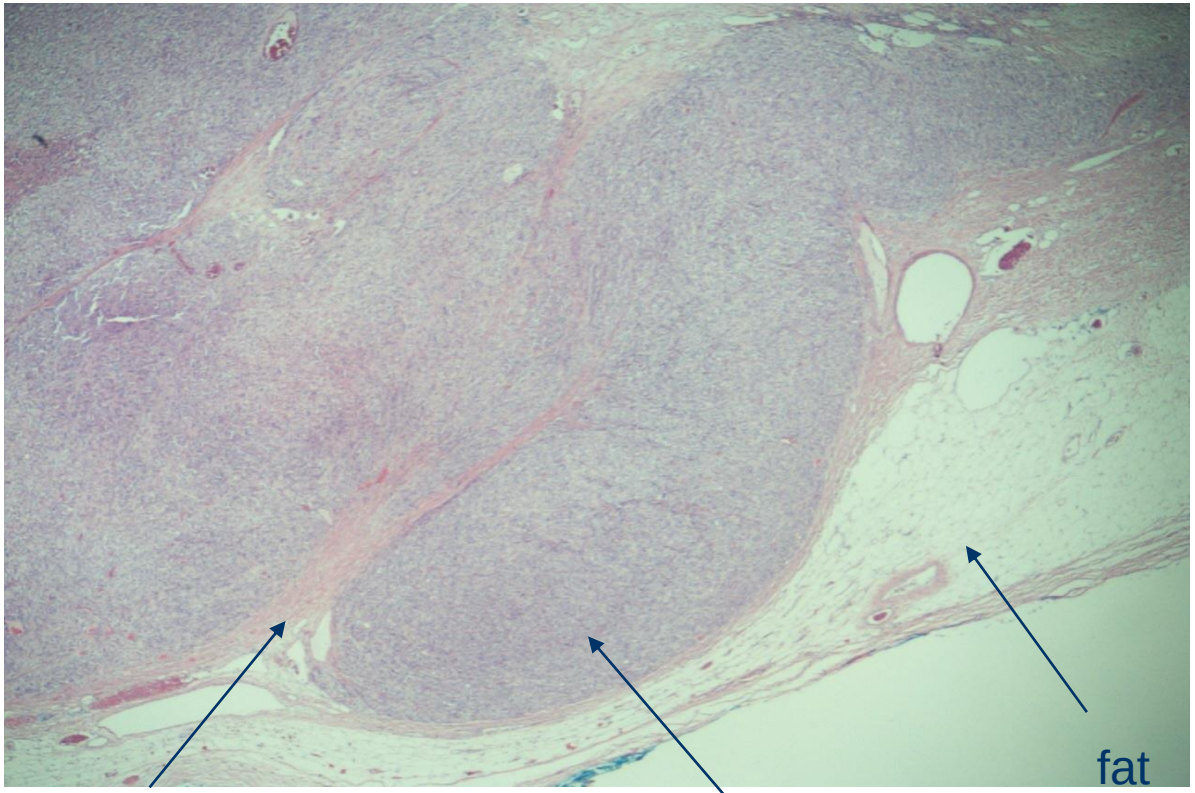
Cortical carcinoma



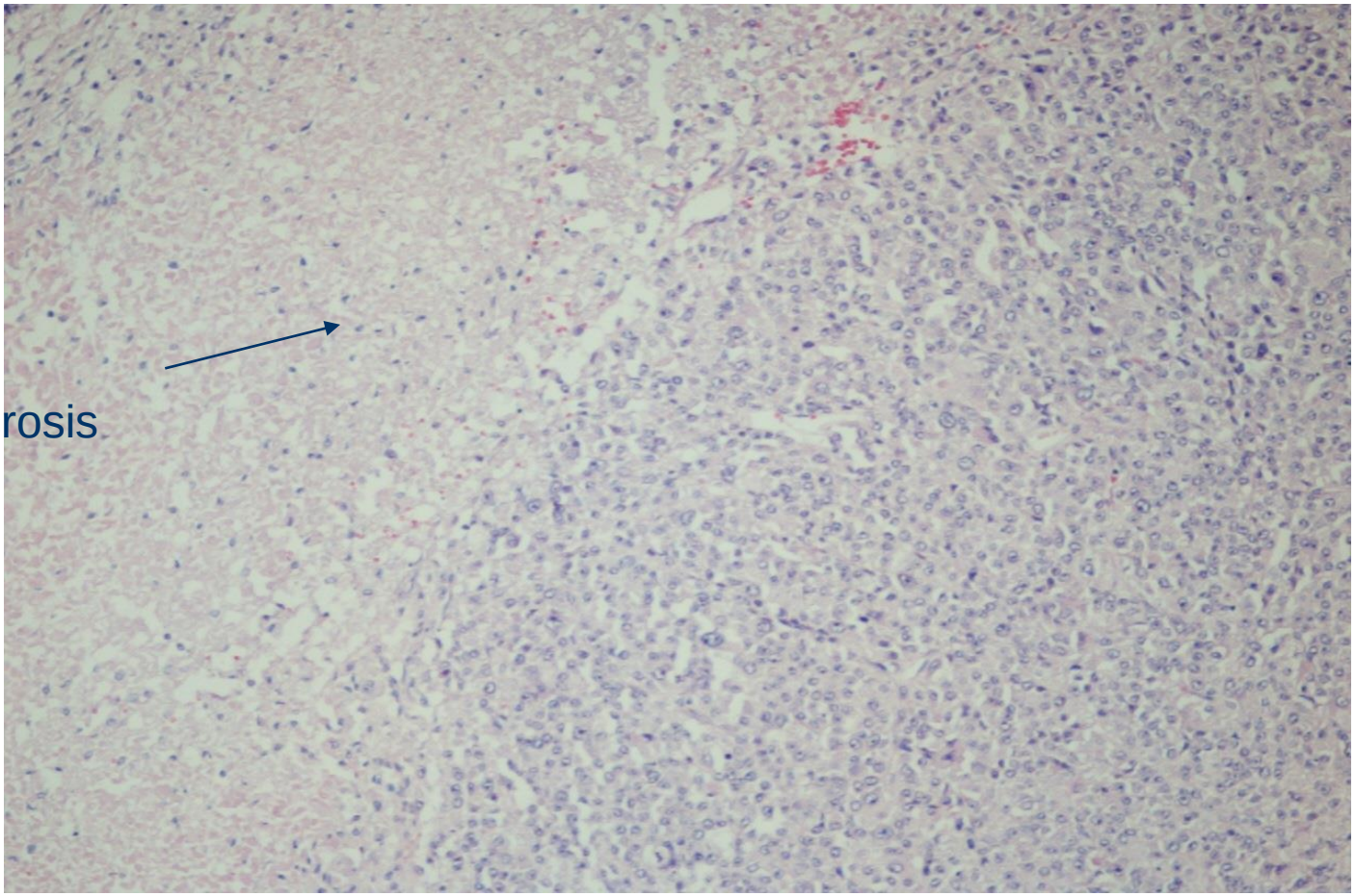
Vascular Invasion

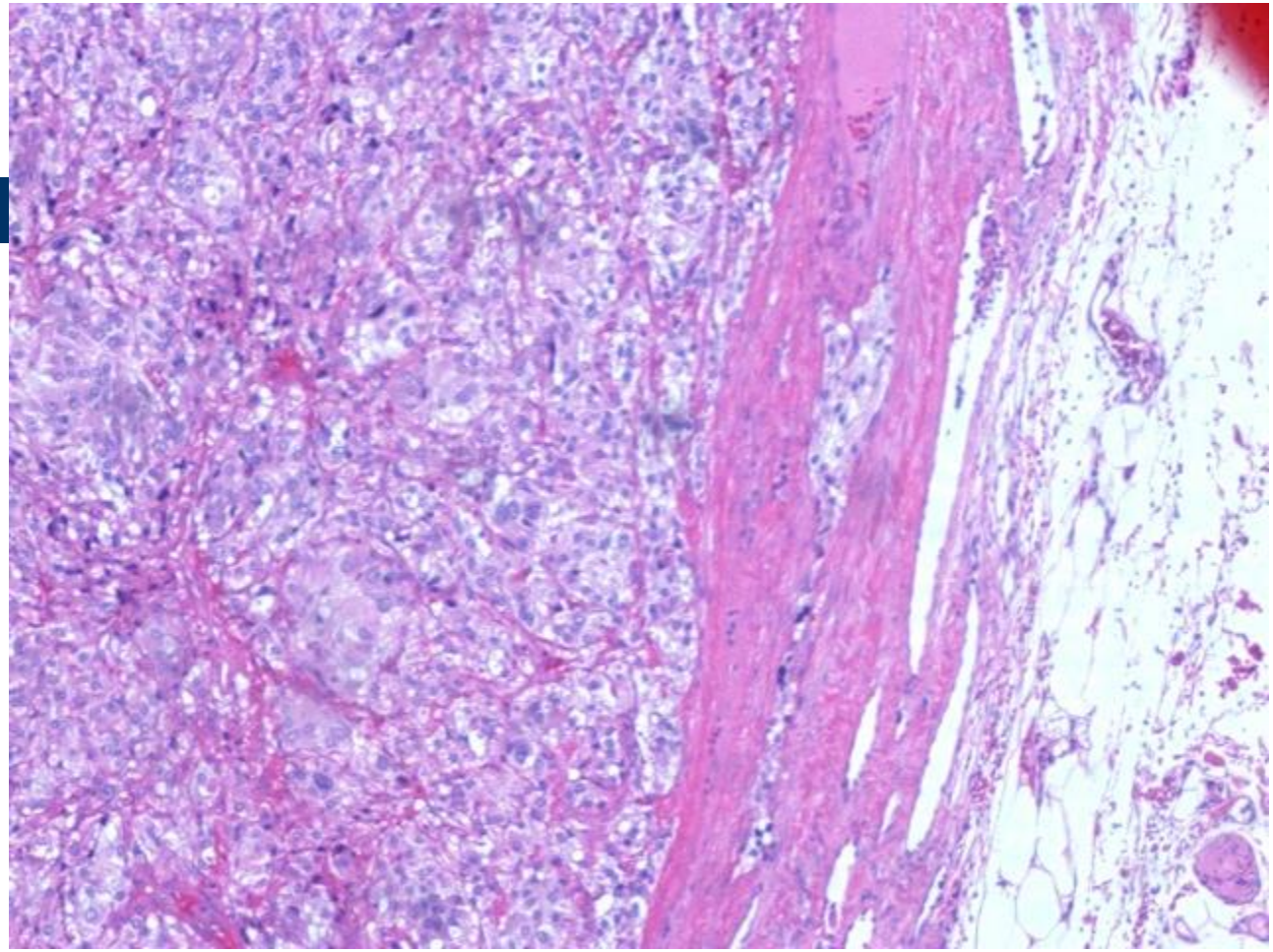


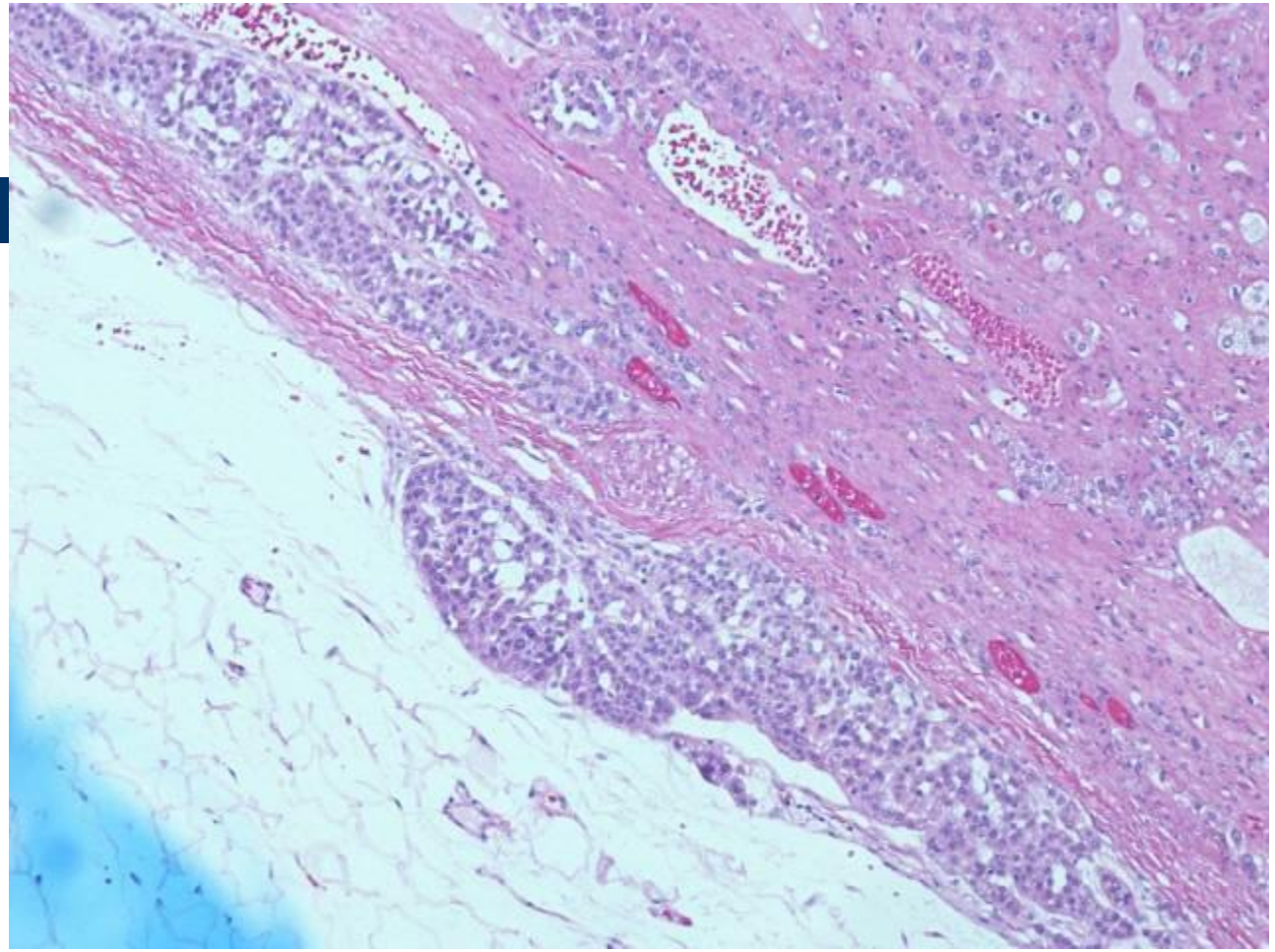
Capsular invasion

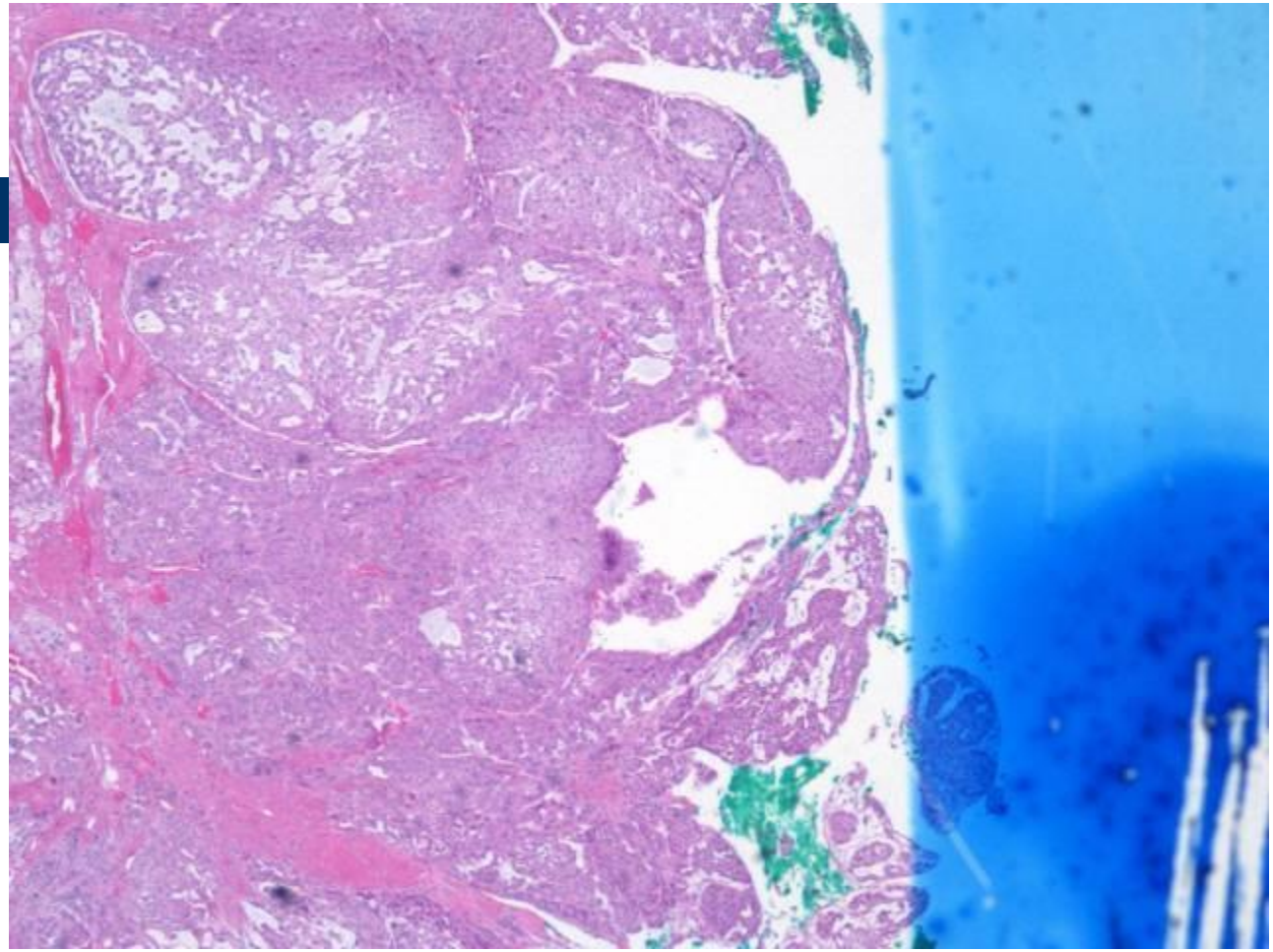


Cortical carcinoma









Cortical carcinoma - immuno

- inhibin and Melan A positivity is sensitive for adrenal cortical carcinomas
- Cytokeratins are usually negative or only very weakly positive
- MiB 1

Prognosis

- 5 year survival is 50 – 70% and improving due to better radiological techniques picking up smaller tumours
- The crucial factor may be whether neoplasms are resectable
- Most important pathological prognostic factors are mitotic rate, tumour size and Ki 67 index

Cortical carcinoma - spread

- To right atrium and lung via renal veins and in inferior vena cava
- Most commonly spread to liver, lung,retroperitoneal lymph nodes and bones
- 40% of cases present with distant metastases

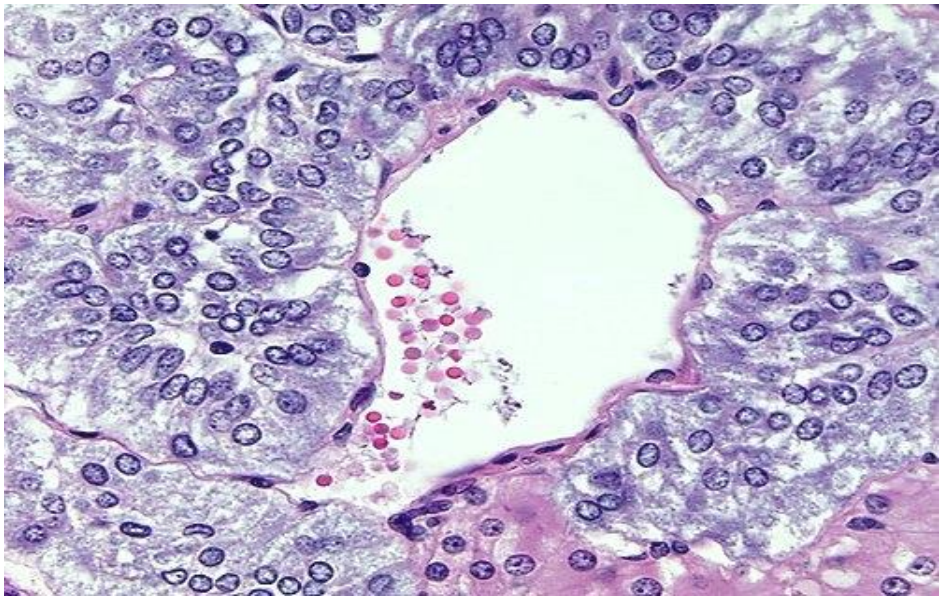
Genetic susceptibility

- Almost all cortical carcinomas are sporadic
- Very rare cases are associated with Li Fraumeni syndrome (sarcomas, leukaemia, carcinomas of breast, brain and adrenal) - mutation of p53 tumour suppressor gene

Genetic susceptibility

- Inherited syndromes that are associated with cortical adenomas include Carney complex (adrenal and pituitary tumours, spotty skin pigmentation, atrial and soft tissue tumours) and MEN 1(multiple endocrine neoplasia)

Adrenal medulla



Adrenal Medulla

The medulla is characterised by catecholamine (adrenaline and noradrenaline) producing cells. Secretion is stimulated directly by neurons in response to stress.

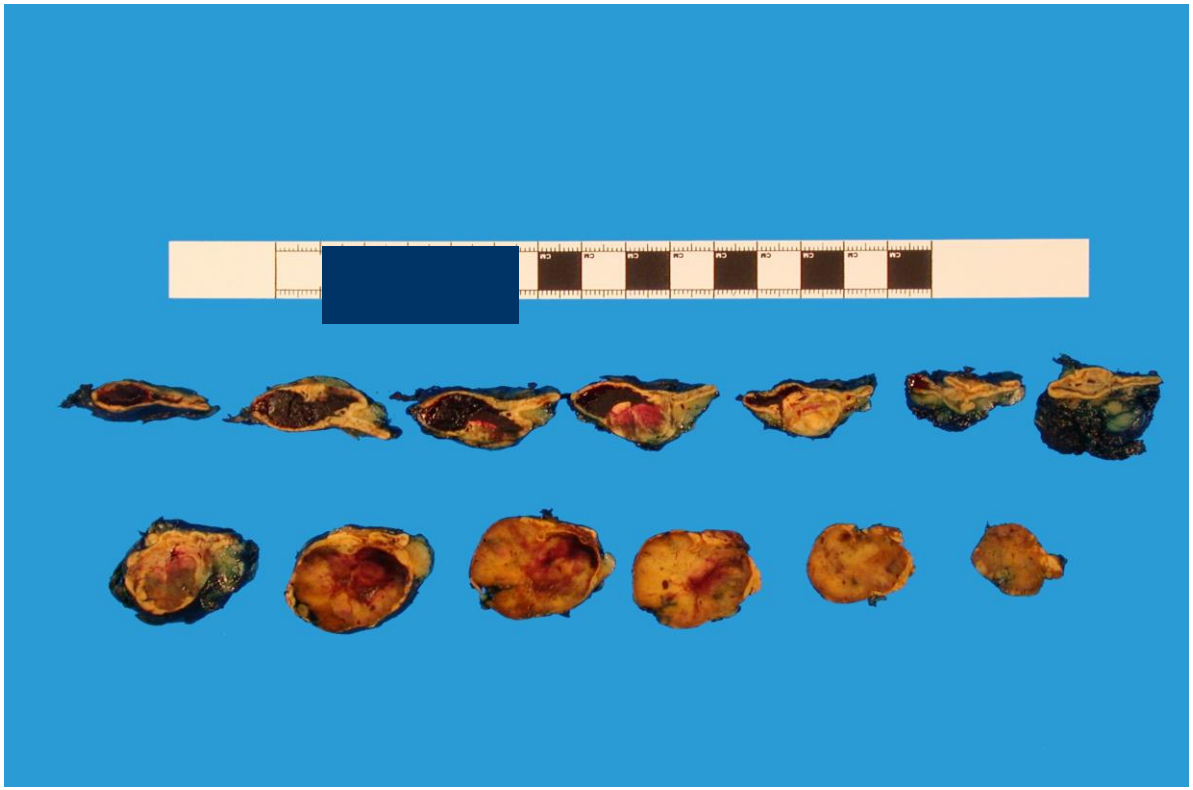
Phaeochromocytoma

- Tumour of adrenal medulla
- Most are sporadic
- 10% are malignant

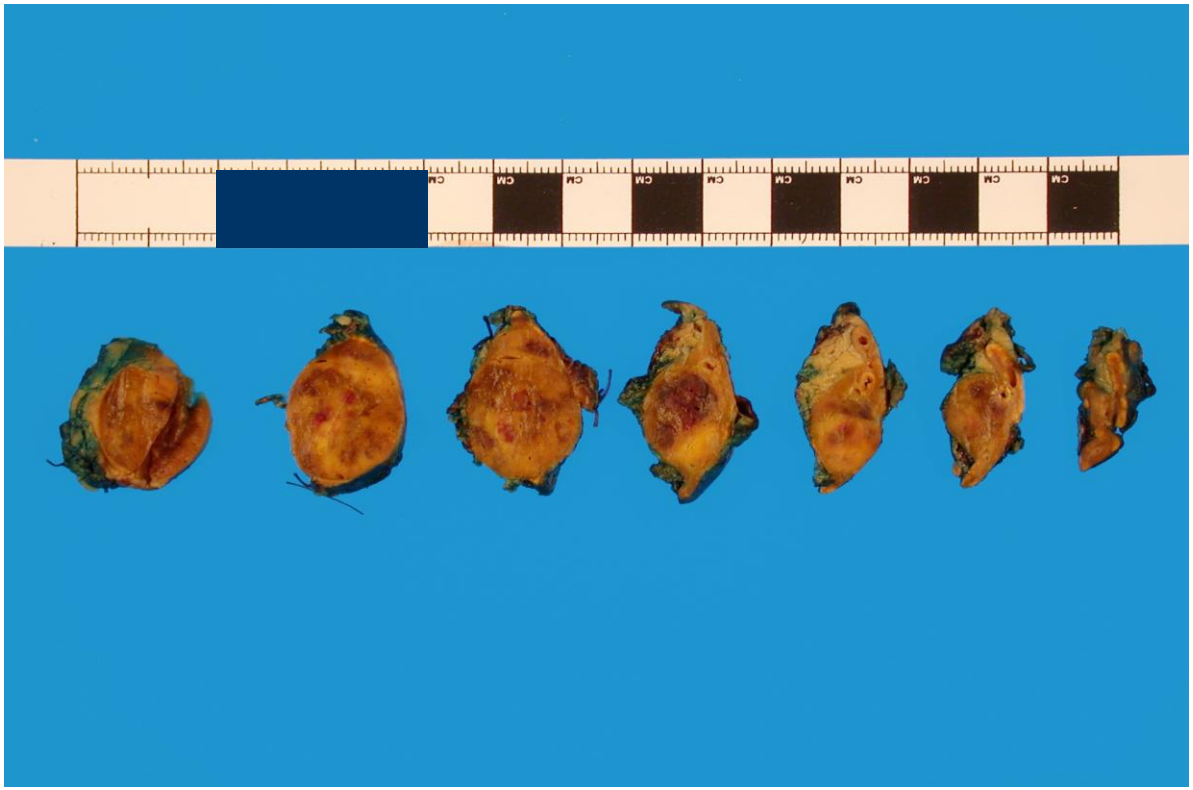
Phaeochromocytoma – clinical features

- Effects are due to excess circulating catecholamines
- Headaches, palpitations, anxiety, chest and abdominal pain.
- Phaeo “ spell” may last 10 to 60 minutes and occur daily to monthly

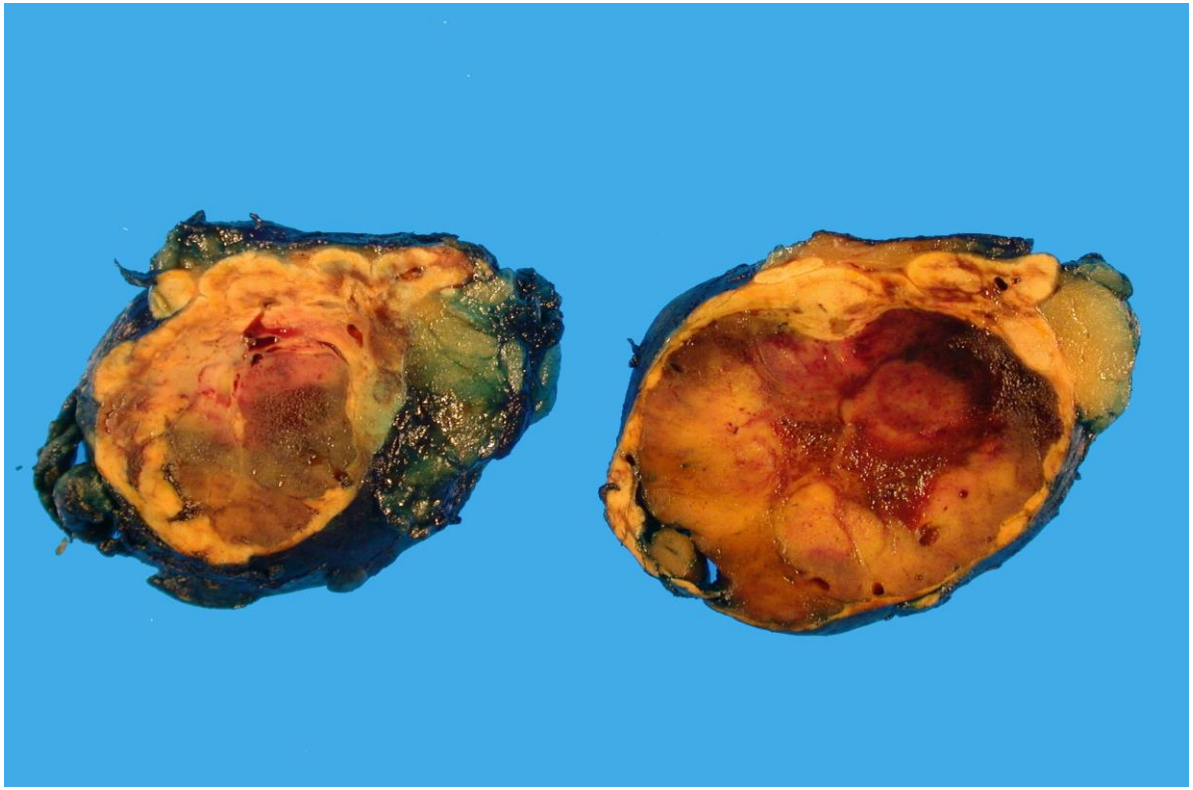
Phaeochromocytoma



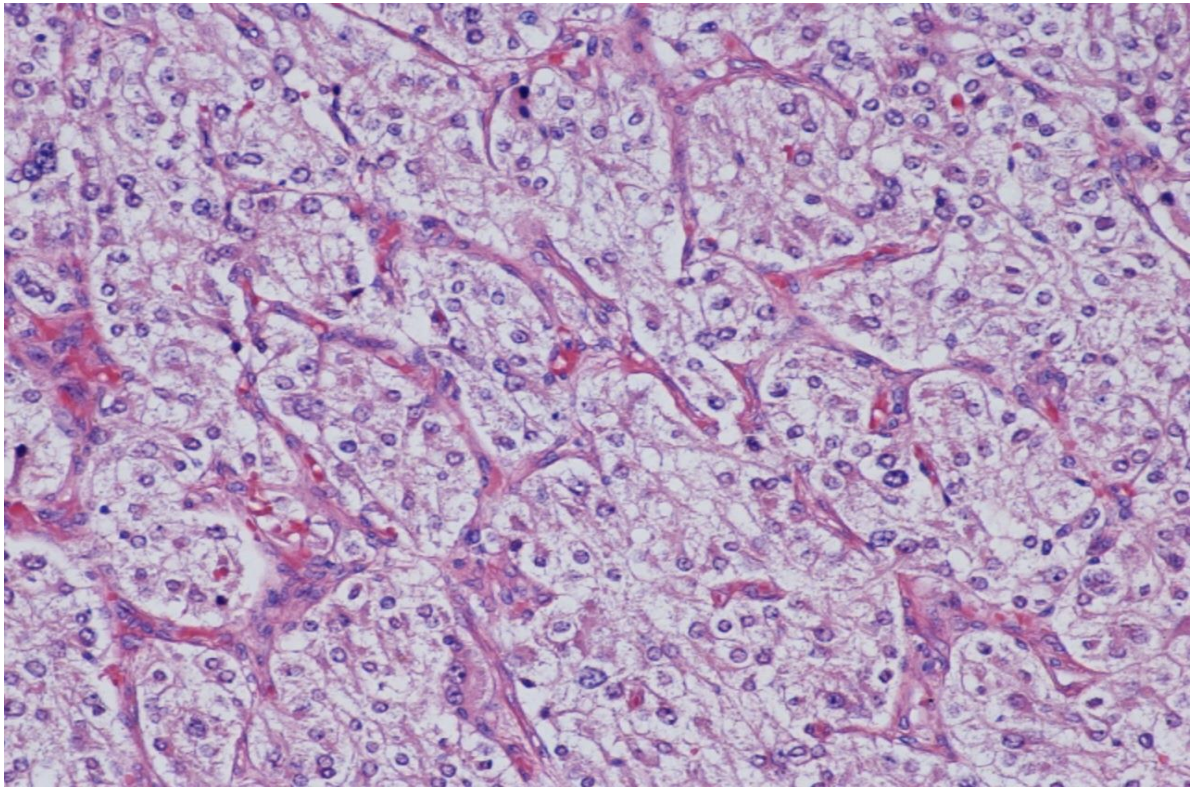
Phaeochromocytoma



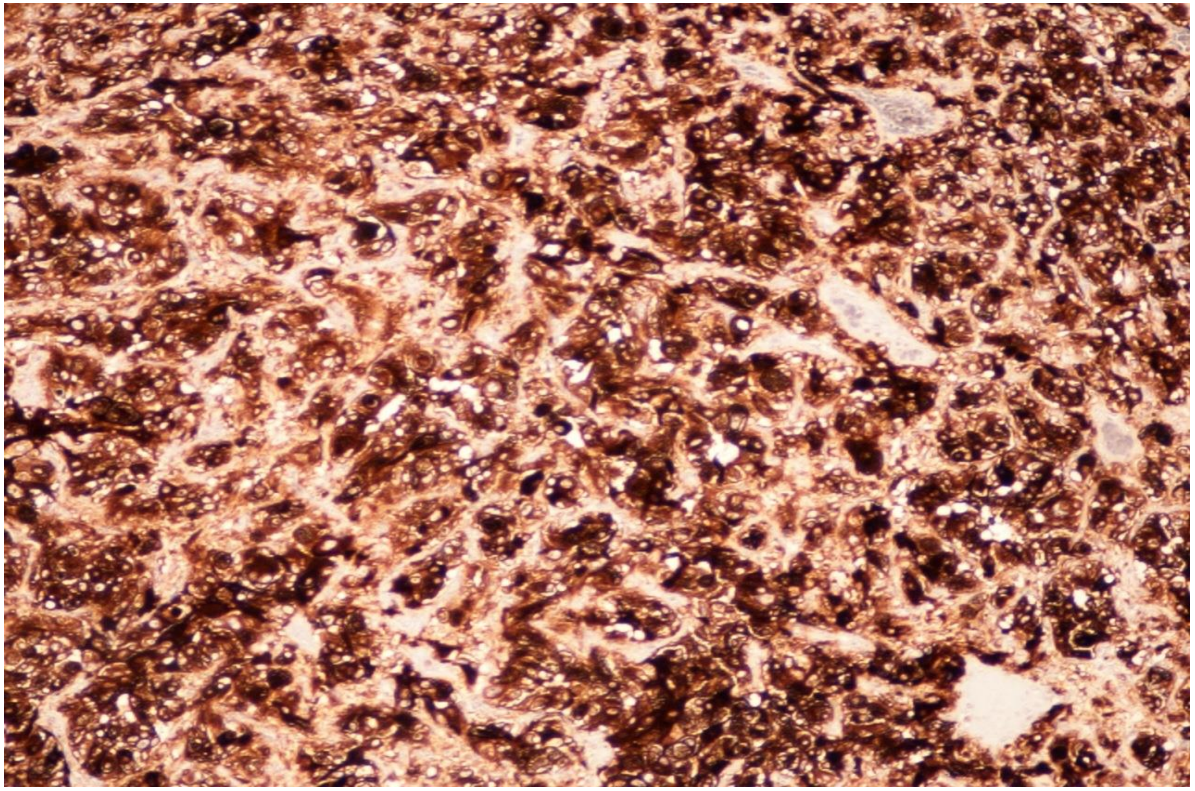
Phaechromocytoma



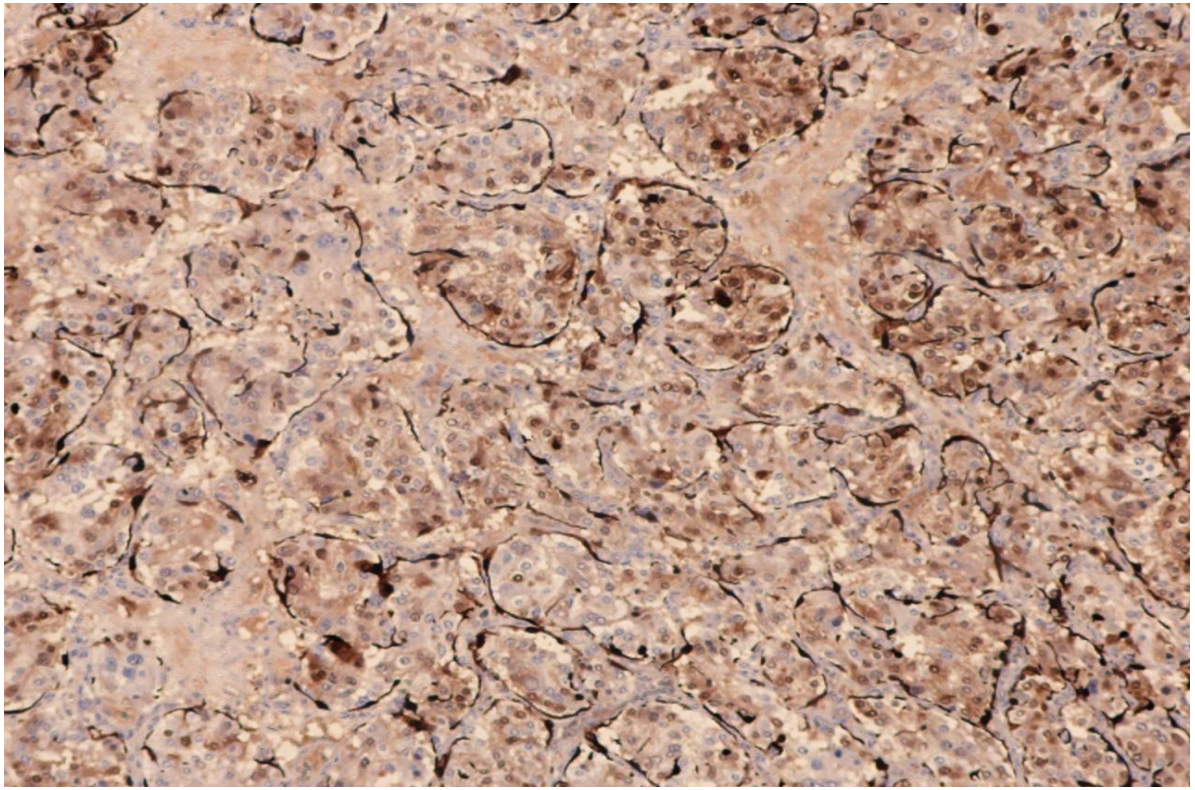
Phaeochromocytoma



Phaeochromocytoma- Chromogranin A



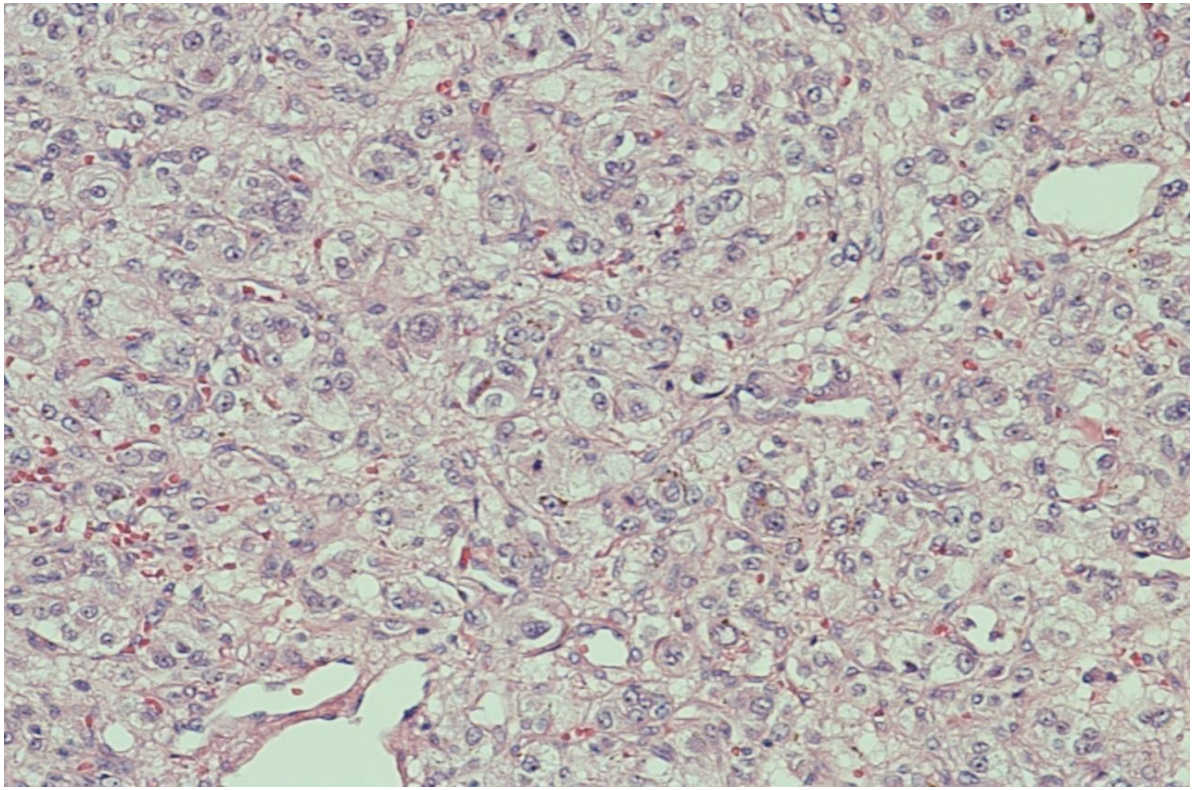
Phaeochromocytoma – S100



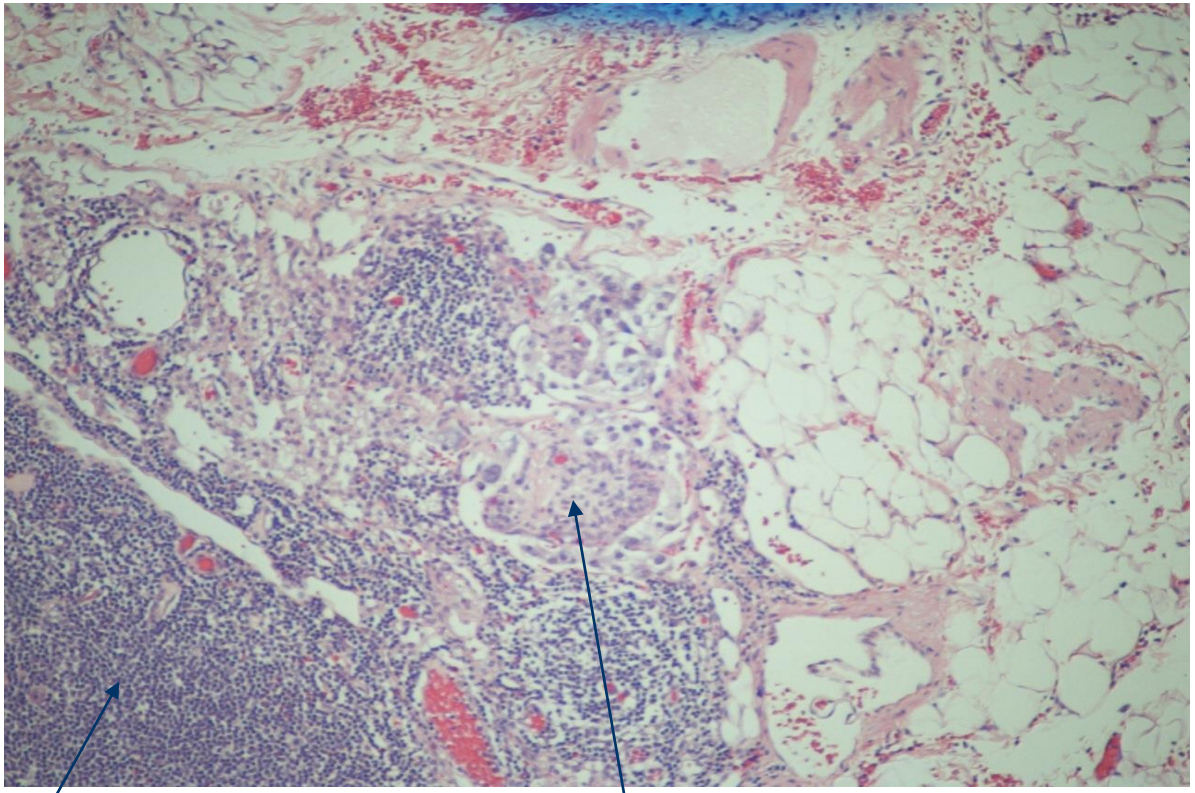
Features seen in malignant phaeochromocytoma

- Capsular invasion
- Vascular invasion
- Extension into fat
- Expanded nests
- Diffuse growth
- Necrosis
- Increased cellularity

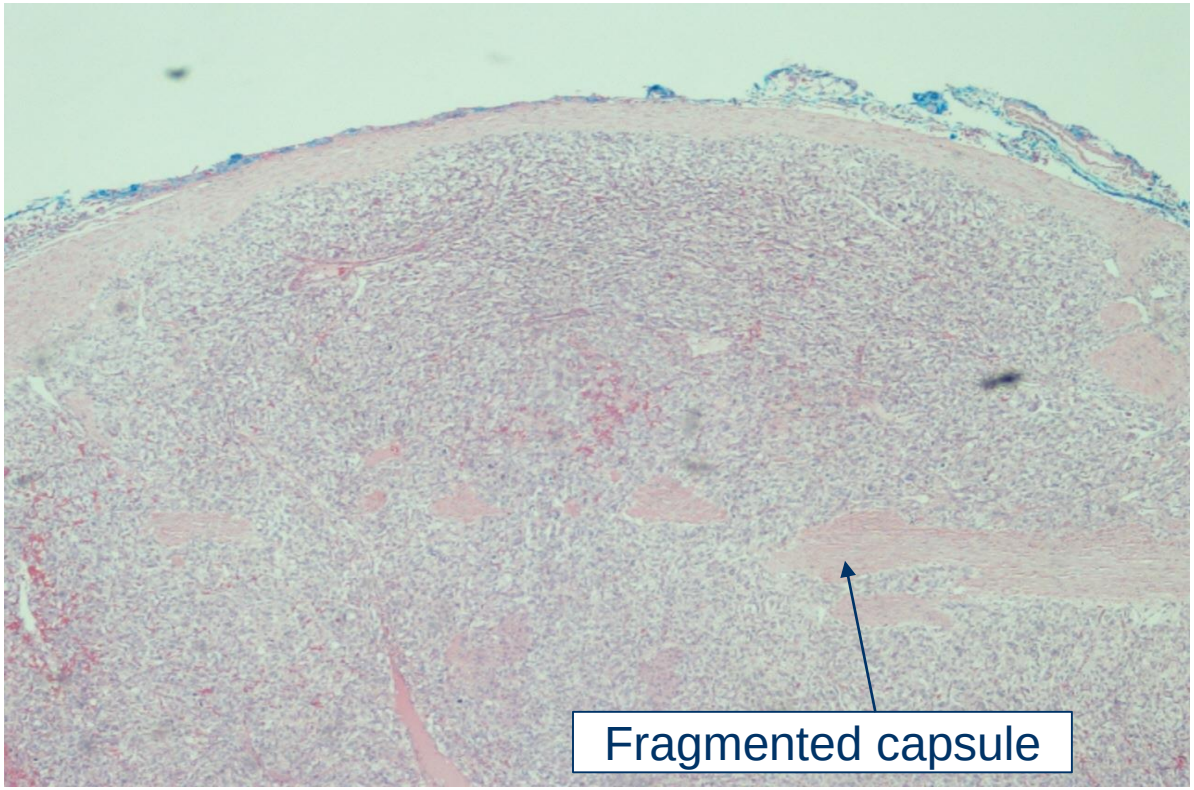
Malignant phaeochromocytoma



Malignant phaeochromocytoma



Malignant phaeochromocytoma



Phaeochromocytoma – genetic susceptibility

- MEN type 2 (marfanoid, mucosal neuromas)
- Von Hippel Lindau syndrome (Renal tumours, brain tumours or may be only feature)
- NF type 1(café au lait spots and subcutaneous neuromas)
- SDHB and SDHD (succinate dehydrogenase mutations)

Conclusion

- Although a very small number of specimens adrenal tumours are fascinating in their presentation and appearances
- MDT crucial involving lifelong follow up for some tumour types