



The complex endocrinology of Von Hippel Lindau

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Von Hippel Lindau

Endocrine manifestations

Pancreatic neuroendocrine tumors

Pheochromocytomas/ paragangliomas

Adrenal insufficiency after bilateral adrenalectomy

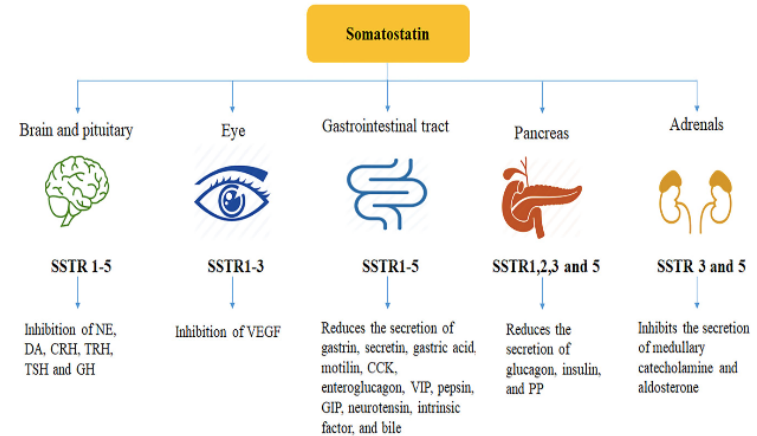
Hemangioblastomas in pituitary region

Endocrine system

- Illustrations follow
Glands
- Hormones

Somatostatin

- Carcinogenesis
- Tumorgrowth
- Tumorprogression
- Tumorspread



^{68}Ga -DOTATOC/DOTATE imaging

- High detection rates of pNETs
- Lower radiation exposure
- SR positive lesions in hemangioblastomas in cerebellum and spine

Shell et al. Eur J of Radiology 2011

Prasad et al. Eur J Nucl Med Mol Imaging 2016

Somatostatin receptor expression

- Gastro enteropancreatic neuroendocrine tumors
- Breast tumors
- Brain tumors: glial tumors and meningiomas
- Renal cell carcinomas
- Lymfomas

SSTR expression in the brain

- Dense network of SSTR receptors in several areas
- Hippocampus and cerebral cortex high affinity
- Spine and cerebellum intermediate affinity
- Meningiomas, astrocytomas, oligodendrogliomas have a high receptor expression

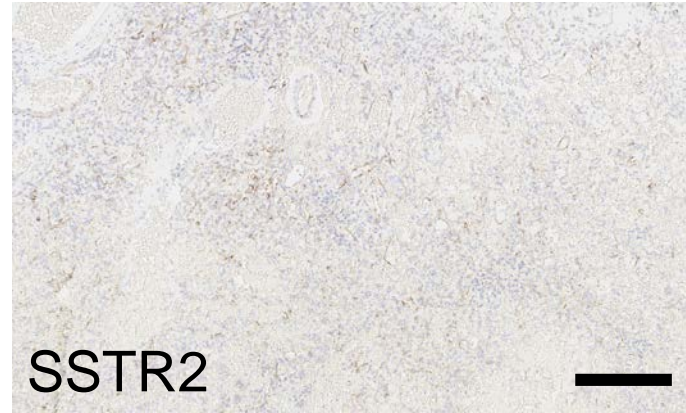
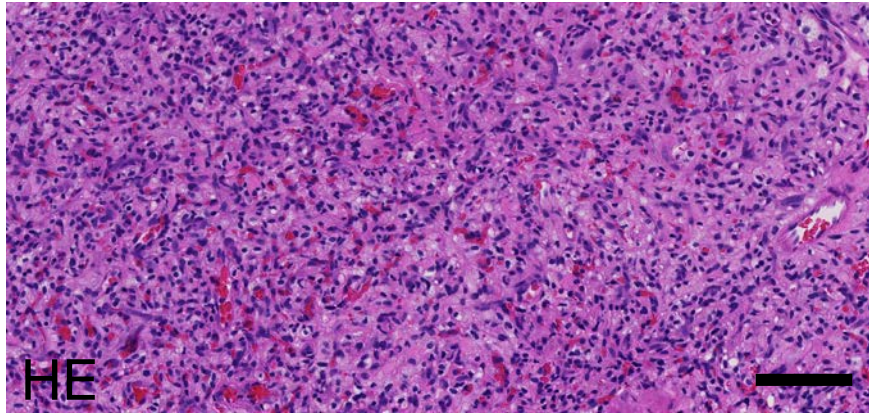
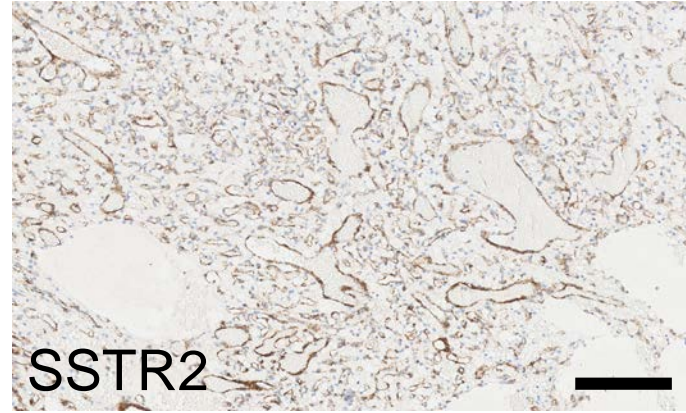
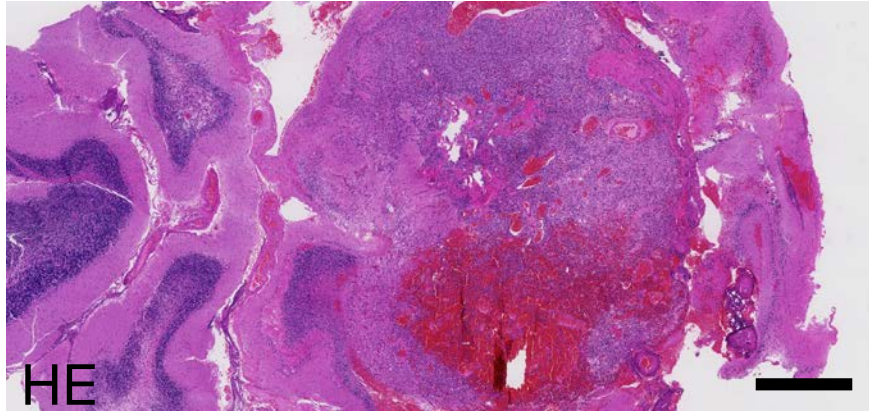
Reubi et al. Neuroscience. 1986
Reubi et al. Cancer Research 1987

Somatostatin expression in VHL

- SSTR receptor 1, 2a, 4 and 5 expression
- SSTR2 expression >> imaging & treatment
- 9 months of treatment with octreotide resulted in central photopenia on dotatate imaging

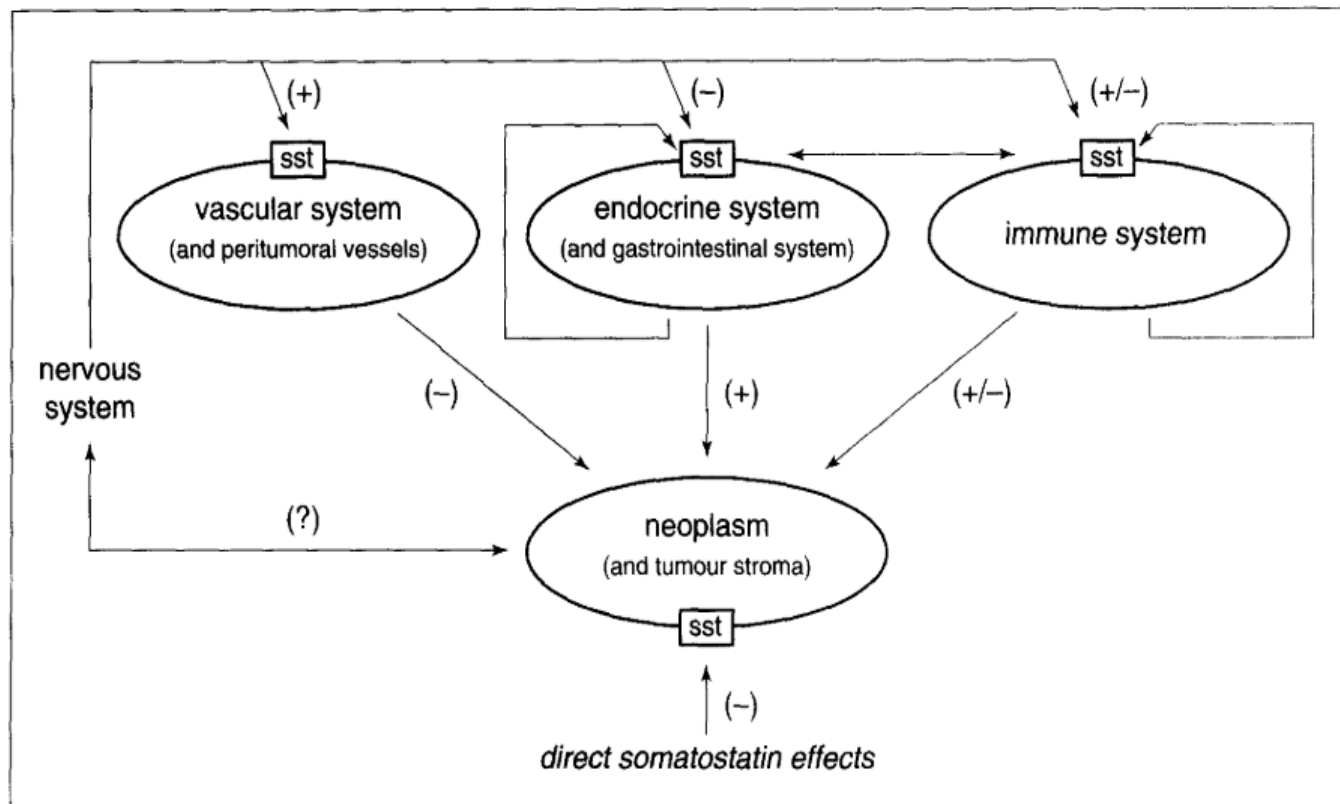
Somatostatin expression in VHL

- Plaatje hemangioblastoma van ons.



Peritumoral vascular system

- Colon carcinomas, renal cell carcinomas, breast cancer
- Independent of receptor in the tumor
- Vasoconstriction → hypoxia and necrosis of tumor;
hypoxia induces angiogenesis



Additional picture

Somatostatin related issues in VHL

Should we routinely perform Ga-68 scan?

- Large pNETs and contraindication for major surgery
- Detection of hemangioblastomas with high SSTR2 expression; eg when unresectable or many comorbidities?

Conclusion