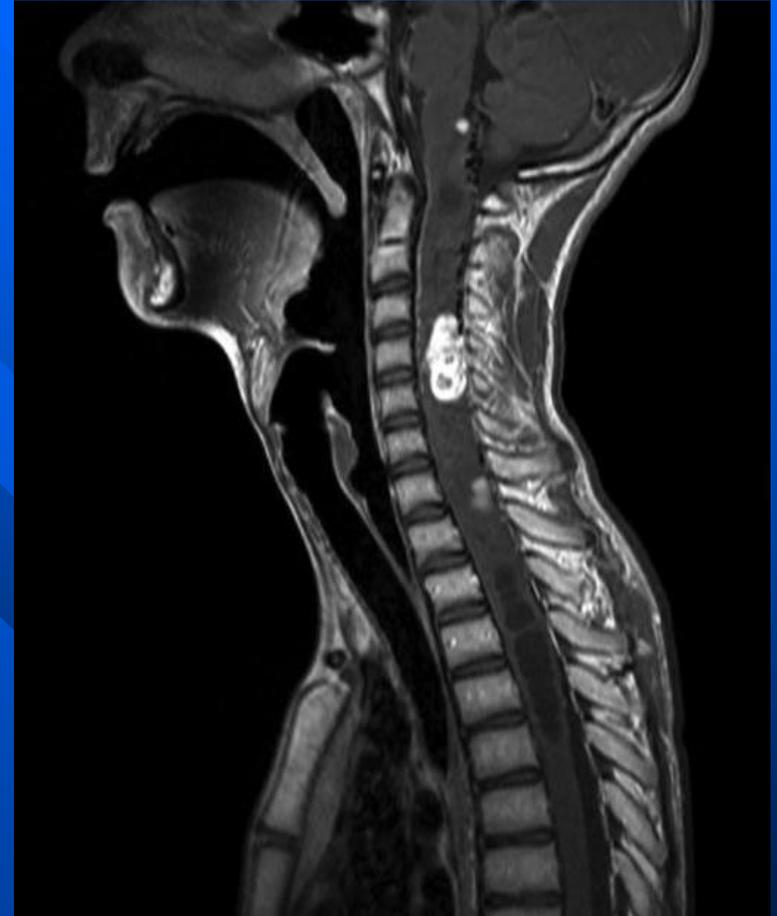


# Von Hippel-Lindau disease

- Multi-system disorder
- characterized by the development of numerous benign and malignant tumors (at least 40 types), as well as several non-malignant lesions.

# CNS manifestations

- CNS hemangioblastoma(s)
  - cerebellar (~75%)
  - spinal (~25%)
- Retinal hemangioblastoma(s)
- Choroid plexus papilloma (CPP)
- Endolymphatic sac tumors (ELST)



# Non CNS manifestations

- pheochromocytoma(s)
- numerous pancreatic lesions (may be the earliest manifestation <sup>3</sup>)
  - pancreatic cysts
  - pancreatic islet cell tumours
  - microcystic adenoma(s) of pancreas
  - pancreatic adenocarcinoma (rare)
- renal lesions
  - renal cell carcinoma(s): usually of the clear cell type <sup>7</sup>; can occur in up to 40% of cases and is one of the most feared complications; RCCs present at an earlier age in those with vHL
  - renal cysts: can occur in up to 75% of cases <sup>5</sup>; often tend to be bilateral and multiple
  - renal angiomyolipoma(s)
- liver cysts
- papillary cystadenoma(s) of the epididymis

# Von Hippel-Lindau Disease

- Autosomal dominant, gene VHL (3p25-p26)
- Incidence 1:36.000
- Retinal angiomas (70%)
- Hemangioblastomas of the brain (50%) and spinal cord
- Renal cell carcinoma (40%)
- Pheochromocytoma
- Endolymphatic sac tumors (10%)
- Cysts of the pancreas, kidney, liver and epididymis.

